

Tumors and Tumor-like Lesions of Blood Vessels

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16.1 Introduction

Tumors and tumor-like conditions of the vascular system are divided into three categories according to their degree of malignancy: benign vascular lesions, lesions of intermediate malignancy, and malignant vascular tumors. The vast majority of the lesions belong to the benign group. These are found predominantly in younger children and adolescents. They may involve either the skin and subcutis or the deep soft tissues. Classification of these lesions is still the source of much controversy and is based on clinical appearance, pathology, embryology, and endothelial growth characteristics [12, 41,

42]. There are two major classification schemes for vascular tumors. That of Enzinger et al. [12] relies on pathological criteria and includes clinical and radiological features when appropriate. On the other hand, the classification of Mulliken and Glowacki [42] is based on endothelial growth characteristics and distinguishes hemangiomas from vascular malformations. The latter classification shows good correlation with the clinical picture and imaging findings.

Hemangiomas are characterized by a phase of proliferation and a stationary period, followed by involution. Vascular malformations are no real tumors and can be divided into low- or high-flow lesions [65].

Cutaneous and subcutaneous lesions are usually easily diagnosed and present no significant diagnostic problems. On the other hand, hemangiomas or vascular malformations that arise in deep soft tissue must be differentiated from malignant neoplasms. Detailed assessment by medical imaging is necessary for adequate planning of surgery.

On magnetic resonance imaging (MRI) benign vascular lesions have a characteristic configuration, generally allowing a correct diagnosis. MRI is superior to other imaging techniques in defining the extent of these lesions, which is important since some types may involve large segments of the body.

Generally the classification of soft tissue vascular anomalies based on endothelial growth characteristics [66] shows good correlation with MRI appearance of these lesions [37]. Since this classification has been useful clinically [54], recognition of these characteristic MRI features is essential for improving therapeutic outcome in these patients.

Vascular lesions of intermediate malignancy and malignant vascular tumors are far more rare. Hemangioendothelioma is a neoplasm of endothelial cells that can be benign or malignant. Angiosarcoma is an aggressive tumor with high local recurrence rate and risk of distant metastases. Imaging findings have only been sparse, probably due to the tendency of these lesions to involve skin and superficial tissue, in contrast to other soft tissue sarcomas. MRI is used for staging rather than for characterization of these lesions.

16.2 Definition and Classification

16.2.1 Benign Vascular Tumors

In the past it was debated whether vascular tumors are developmental malformations or true tumors. In the nineteenth century vascular lesions were thought to be 'produced by the longing of the mother, for particular things, or her aversion to them'. Expressions such as *nexus materneus* or *stigma metrocelis* were in reference to the mother. Because vascular tumors sometimes closely resemble normal vessels, it is difficult to distinguish clearly between neoplasm and malformation on histological examination [41].

The term hemangioma has frequently been used incorrectly, although accurate nomenclature is of utmost importance for correct diagnosis and treatment of these lesions [66].

16.2.1.1 Classification of Mulliken

Some authors have suggested that the clinical presentation provides the necessary perspective for classifying vascular lesions. Mulliken et al. presented a useful scheme for separating cutaneous vascular lesions based on endothelial growth characteristics [42]. Their studies reveal two major types of vascular lesions. One exhibits a rapid growth phase followed by a period of stabilization and finally involution; these show a female

Table 16.1. Differentiating features of hemangiomas and vascular malformations (modified from [65])

Hemangiomas	Vascular malformations
Exhibit cellular proliferation	Comprised of dysplastic vessels
Small or absent at birth	Present at birth
Rapid growth during infancy	Growth proportional to child
Involution during childhood	No regression

predominance and are usually not present at birth. Because of cellular proliferation and the mass effect, these are called (infantile) hemangiomas (Table 16.1).

The majority of hemangiomas do not need treatment.

On the other hand, many vascular lesions show no cellular proliferation; they usually are not present at birth but grow with the child and have no involution phase. These lesions are called vascular malformations, and they are divided into capillary, venous, arterial and lymphatic types depending on the predominant vessel type [42].

The classification scheme was updated during the 1992 meeting of the International Society for the Study of Vascular Anomalies (ISSVA) [75, 82] (Table 16.2).

The endothelial lining of the vascular malformation group is not proliferative. Although they are stable from a cellular point of view, they can be clinically devastating, specially when there is arteriovenous shunting.

16.2.1.2 Classification of Enzinger

Enzinger et al., however, do not rely on clinical presentation for classification of benign vascular lesions [12]. Some congenital lesions do not become apparent until adult life, depending on their location and growth. In view of this limitation no attempt is made to separate malformations from benign neoplasms. All lesions are called hemangiomas, and hemangioma is defined as 'a benign but nonreactive process in which there is an increase in the number of normal or abnormal-appearing vessels [12]. Hemangiomas may be either of two types: those localized in one area and those involving large segments of the body. The histological classification of vascular tumors proposed by Enzinger et al. [12] is as follows (Table 16.3).

Localized hemangiomas are the more common. They have been classified according to clinical, embryologic, or pathological criteria but no system is entirely satisfactory.

Table 16.2. ISSVA classification of vascular anomalies (modified from [76, 83])

Vascular malformations		
Vascular tumor	Simple	Combined
Hemangioma		
Proliferative phase	Capillary malformation	Arteriovenous fistula, arteriovenous malformation, capillary-venous malformation, capillary-lymphatic-venous malformation (Klippel Trénaunay syndrome)
Invulsive phase	Lymphatic malformation	Lymphatic-venous malformation, capillary-arteriovenous malformation (Parkes-Weber syndrome), capillary-lymphatic-arteriovenous malformation
Other tumors	Venous malformation	

A more pertinent issue considering imaging and therapy is classifying vascular malformations as either low-flow or high-flow lesions [84]

Table 16.3. Histological classification of vascular tumors (modified from [12])

Benign vascular tumors
Localized Hemangioma
Capillary Hemangioma
Cavernous Hemangioma
Venous hemangioma
Arteriovenous hemangioma
Epithelioid hemangioma
Hemangioma of the granulation tissue type
Deep soft tissue hemangioma
Angiomatosis
Vascular tumors of intermediate malignancy
Epithelioid hemangioendothelioma
Spindle cell hemangioendothelioma
Malignant endovascular papillary hemangioendothelioma
Malignant vascular tumors
Angiosarcoma
Kaposi's sarcoma

Table 16.4. WHO classification of vascular tumors (modified from [68])

Benign
Hemangiomas of
Subcutis/deep soft tissue
Capillary
Cavernous
Arteriovenous
Venous
Intramuscular
Synovial
Epithelioid hemangioma
Angiomatosis
Lymphangioma
Intermediate (locally aggressive)
Kaposiform hemangioendothelioma
Intermediate (rarely metastasizing)
Retiform hemangioendothelioma
Papillary intralymphatic angioendothelioma
Composite hemangioendothelioma
Kaposi sarcoma
Malignant
Epithelioid hemangioendothelioma
Angiosarcoma of soft tissue

They are usually located superficially but may involve deep structures, such as skeletal muscle. Depending on the predominant vessel type, the former are grouped as capillary, cavernous, venous or arteriovenous. Other forms are more rare.

Hemangiomas involving the deep soft tissues are grouped as intramuscular, synovial or intraneuronal types. Intramuscular hemangioma usually shows an overgrowth of adipose tissue, giving the impression of angiomyoma. Unlike their cutaneous variants, it is not always possible to make a clear distinction between different types of intramuscular hemangiomas. A classification into small-vessel, large-vessel and mixed types with different clinical behavior was proposed by Allen to avoid misinterpretation since some of the small-vessel type lesions show alarming histological features resembling those of a sarcoma [1].

Angiomatosis is a rare condition in which large segments of the body are involved by proliferating vessels. Mostly the extremities are affected. A characteristic feature of this condition is the large amount of mature fat that accompanies the proliferating vessels [12].

The distinction between intramuscular hemangioma and angiomatosis is made better by clinical than pathological criteria.

16.2.1.3 WHO Classification

The classification of the WHO (Table 16.4) closely resembles that of Enzinger. In this classification, no distinction is made between benign vascular neoplasms or vascular malformations. Similarly, it is impossible to

reliably distinguish vascular from lymphatic endothelioma. Therefore, they classify lymphangioma as a vascular tumor.

16.2.2 Vascular Tumors of Borderline or Intermediate Malignancy

The term hemangioendothelioma is currently used for lesions which are histologically intermediate in appearance between hemangiomas and angiosarcomas.

According to Enzinger, the three types are epithelioid hemangioendothelioma, spindle cell hemangioendothelioma and malignant endovascular papillary hemangioendothelioma. Only the epithelioid subgroup occurs in deep soft tissues, most commonly of the extremities. The other types develop preferentially in the dermis or subcutaneous tissues [13].

The WHO classification differentiates locally aggressive tumors of intermediate malignancy and rarely metastasizing types. Kaposiform hemangioendothelioma is characterized by a 'Kaposi sarcoma like' fascicular spindle cell growth pattern. It most commonly occurs in the retroperitoneum or the skin.

16.2.3 Malignant Vascular Tumors

Angiosarcomas are tumors that can vary from highly differentiated, resembling hemangioma, to those whose anaplasia makes it difficult to distinguish from carcinomas. They are characterized by irregular anastomosing vascular channels lined by atypical endothelial cells.

The terms hemangiosarcoma and lymphangiosarcoma are no longer appropriate [14, 45]. The lesions occur preferentially in the skin or soft tissues of the scalp and face. Kaposi's sarcomas have spindle cell areas containing vascular channels.

The disease is associated with the Human Herpes Virus (HHV-8) infection. It presents as cutaneous lesions in the form of multiple patches. According to the WHO classification Kaposi sarcoma is of intermediate malignancy and epithelioid hemangioendothelioma is malignant.

16.2.4 Glomus Tumor

The glomus tumor is a neoplasm consisting of cells which closely resemble smooth muscle cells of the normal glomus body. The glomus body is an arteriovenous anastomosis that has an important role in thermoregulation. It is located in the subungual region, digits, and palms. The lesion shows both muscle fibers and epithelial-appearing glomus cells [15].

16.2.5 Hemangiopericytoma

Hemangiopericytoma is an uncommon tumor, first described by Stout and Murray, and is composed mainly of pericytes [8, 15, 26, 28, 33, 44, 52, 55, 63]. According to the most recent WHO classification [67], hemangiopericytoma will be discussed in Chap. 13 (Tumors of Fibrous Tissue).

16.3 Incidence and Clinical Behavior

16.3.1 Benign Vascular Tumors

To avoid confusion we follow here a combination of the classification of Enzinger and the WHO [12, 67].

The differentiation of Mulliken and Glowacki between hemangiomas and vascular malformations is only considered when specifically mentioned.

Although hemangiomas are uncommon tumors, they make up about 7% of all benign soft tissue tumors. Most of the lesions, however, are located in the skin or subcutaneous tissues, while only a minority is deeply seated [12].

Capillary hemangiomas form the largest group. Except for cherry angioma they occur predominantly in childhood. Juvenile hemangioma is an immature form of capillary hemangioma with a characteristic clinical evolution. They occur mostly in superficial tissue of head and neck region and arise a few weeks after birth. They grow slowly to reach a maximal size at six months of age and then regress. Some lesions may pose cosmet-

ic problems or threaten vital structures. They are treated by systemic steroid therapy and subsequent surgery. Acquired tufted angioma, verrucous hemangioma, and senile angioma are clinical variants of the same subtype of hemangioma.

Cavernous hemangiomas are less frequent but share age and anatomical distribution with the capillary hemangiomas. However, they are usually larger and less circumscribed and may be locally destructive. The majority require surgical resection.

Superficial arteriovenous and venous types are less common. They are distinguished from the capillary and cavernous counterparts by the presence of thick-walled vessels. Superficially located arteriovenous hemangiomas cause no clinical problems. On the other hand, shunting of blood causing cardiac overload, pain and hypertrophy of the involved extremity are problems associated with deep lesions and require an adequate therapeutic approach.

Epithelioid hemangioma and granulation type hemangioma are unusual vascular tumors.

Epithelioid hemangioma consists of well formed but immature vessels and a prominent inflammatory component. They occur predominantly at the forehead and the digits, and present as a nodular mass which gives the impression of an epidermal cyst.

Compared to superficial hemangiomas, deep-seated lesions are quite uncommon, with a reported frequency of 0.8% of all lesions [44]. However, they deserve specific mention because of their different clinical presentation. Because they present with pain or swelling, it is difficult to make a diagnosis based on physical examination. Some 80–90% of intramuscular hemangiomas occur in the first three decades of life. There is no female predilection, as for superficial hemangiomas. The majority of lesions are located in the lower extremity, especially in the muscles of the thigh. Pain is reported to be more common in tumors involving long, narrow muscles, probably through stretching of the muscle fibers.

Lesions have often been present for many years and it is therefore likely that many examples are congenital.

Physical deformity, cardiac decompensation, or destruction of adjacent vital structures may require surgical intervention. Therapy is aimed at complete excision, with or without prior embolization [10, 63]. Intramuscular hemangiomas are benign, but with a high incidence of local recurrence.

Synovial hemangioma and hemangioma of peripheral nerve are rare. The synovial type almost always involves the knee joint, presenting with pain, swelling and joint effusion. The most common site is the suprapatellar pouch. Because of repetitive bleeding into the joint, a radiographic appearance identical to that of hemophilic arthropathy is seen [12, 28]. Only a small proportion of lesions are correctly diagnosed before surgery (Fig. 16.1).

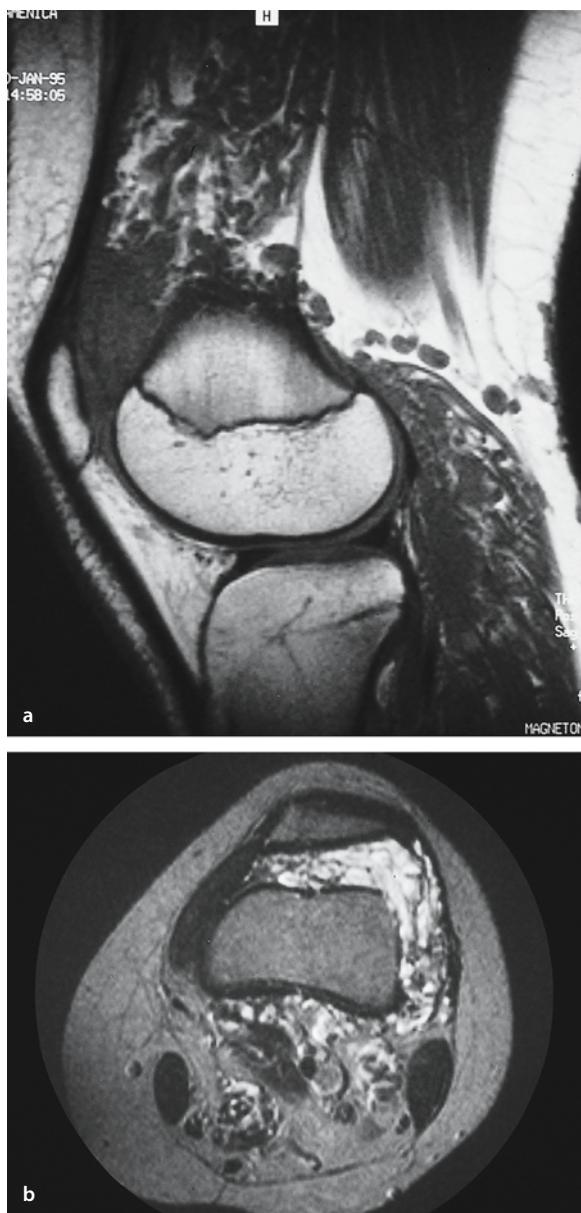


Fig. 16.1 a, b. Synovial hemangioma of the knee. **a** Sagittal T1-weighted MR image. **b** Axial T2-weighted MR image. A multilobular inhomogeneous lesion involving both the suprapatellar bursa and the extra-articular fatty tissue and muscles is seen. The lesion is isointense to muscle on T1-weighted images and a large draining vessel is seen in the popliteal fossa. Axial T2-weighted images show a lesion of high signal intensity. There is no joint effusion

16.3.2 Angiomatous Syndromes

Angiomatous syndromes include Kasabach-Merritt syndrome, Maffucci's syndrome, Klippel-Trénaunay-Weber syndrome, Osler-Weber-Rendu disease, Gorham disease, Proteus syndrome and angiomyomatosis.

Kasabach-Merritt syndrome was first reported in 1954 [25]. The cardinal features of this syndrome include an enlarging hemangioma, thrombocytopenia and microangiopathic hemolytic anemia with acute or chronic consumptive coagulopathy. The etiology of the coagulopathy is still unknown. Mortality is estimated at approximately 21%. The major cause of death is bleeding. Many therapeutic modalities have been applied, aiming at a twofold objective: to control the bleeding and to reduce the size of the lesion [11, 25, 32, 38]. Maffucci's syndrome is a rare dysplasia characterized by multiple hemangiomas and enchondromas. The vascular tumors are usually noted at birth and are of the cavernous type. According to Silverman et al. [54], the lesions that are termed hemangiomas are in fact vascular malformations. The cartilaginous lesions typically develop after the vascular tumors. The bones are shortened and have multiple exostoses and enchondromas [6, 12] (Fig. 16.2). Malignant sarcomatous degeneration is noted in about 21% of patients.

Klippel-Trénaunay-Weber syndrome consists of cutaneous hemangioma, bone and soft tissue hypertrophy and varicose veins. The syndrome is usually unilateral and involves the lower extremity [68].

Proteus syndrome is a complex hamartomatous disorder defined by local overgrowth (macrodactyly), subcutaneous tumors and various bone, cutaneous and/or vascular anomalies. Vascular anomalies are common and are distributed at random sites of the body. The clinical presentation is highly variable [69, 70].

Angiomatosis is a rare condition in which large segments of the body are involved by proliferating vessels [71]. Principally the extremities are affected. A characteristic feature of this condition is the large amount of mature fat that accompanies the proliferating vessels [12]. The disorder probably starts during intrauterine life when the limb buds form.

Approximately two-thirds of cases develop within the two first decades of life and nearly all are apparent by age 40 years.

It usually presents as a swelling, induration, or discoloration of the affected area with or without limb hypertrophy. Mortality is high because of space occupying effects and consumption coagulopathy [12, 28].

16.3.3 Hemangioendothelioma

Epithelioid hemangioendothelioma of the soft tissues does not occur preferentially in children. All age groups are affected, and there is no sex predilection. Most lesions are solitary. Overall prognosis of this tumor is quite favorable. Only a small proportion does metastasize and cause death. Therapy includes wide local excision without adjuvant chemotherapy or radiotherapy. Regional lymph nodes should be evaluated [30, 41].



Fig. 16.2 a, b. An 18-year-old woman with Maffucci's syndrome. **a** Plain radiograph of the left hand. **b** Plain radiograph (lateral view) of the left wrist. Multiple, expansile, well-defined and predominantly lytic lesions within the phalanges and metacarpals of the left hand are shown. The second digit has previously been am-

putated. Phleboliths are seen within a soft tissue swelling at the volar aspect of the wrist. The combination of multiple enchondromas and soft tissue hemangiomas is characteristic for Maffucci's syndrome

16.3.4 Angiosarcomas

Angiosarcomas of the deep tissue are one of the rarest forms of soft tissue neoplasms, accounting for less than 1% of all sarcomas. These tumors are evenly distributed throughout all decades and show predilection for the lower extremity and the abdominal cavity, in contrast to the cutaneous forms which are found mostly in head and neck. Chronic lymphedema is the most widely recognized predisposing factor in angiosarcomas. Angiosarcomas are often characterized as enlarging, painful masses lasting for several weeks and are occasionally associated with hemorrhage, anemia or coagulopathy. Epithelioid angiosarcoma is the most frequently observed pattern, but the morphological spectrum is wide [36]. Malignant degeneration in a preexisting benign lesion is probably an unusual event [15].

The paucity of data precludes statements concerning the optimal mode of therapy [5, 45].

16.3.5 Glomus Tumor

Glomus tumors are uncommon, with an equal frequency of occurrence in both sexes. The most common location is the subungual tissue at the tip of the finger. For

this location however, there is a striking female predominance. Glomus tumors are also seen at the palm, wrist, forearm, and foot. Multiple lesions may be present, especially during childhood. Glomus tumors cause a radiating pain which is elicited by a change in temperature. Clinical examination usually reveals a characteristic blue-red nodule. Therapy aims at complete excision, still leaving a recurrence rate of 10% [15].

16.3.6 Hemangiopericytoma

As earlier mentioned, hemangiopericytoma (HPC) will be discussed in Chap. 13, which is in line with the most recent WHO classification (2002).

16.4 Imaging

16.4.1 Imaging Studies Other than MRI

Although findings on plain radiography are frequently normal, they are sometimes helpful in diagnosing soft tissue hemangiomas. Plain radiographs reveal the presence of a lesion by displacement or loss of the normal tissue planes. The presence of phleboliths is a specific sign, but unfortunately of low sensitivity.



Fig. 16.3a, b. A 35-year-old man with painless soft tissue mass at the left hand. **a** Radiograph of the left hand. **b** Coronal spin echo T1-weighted MR image. Plain radiography shows an increased density of the soft tissues at the left thenar. Multiple rounded calcifications of varying size are present, corresponding to phleboliths. This finding is characteristic for intramuscular hemangiomas. There is an erosion of the ulnar sided cortex at the base of metacarpal I (**a**). On the MR image the muscles of the thenar are slightly hyperintense due to infiltration by the hemangioma. The rounded spots of low signal intensity correspond to the phleboliths. Pressure erosion of metacarpal I is shown without change in the adjacent bone marrow (**b**)

Phleboliths are found in venous malformations and correspond to calcifications in thrombosis [72].

Lesions that are in close proximity to bone may cause bone erosion or periosteal reaction (Fig. 16.3). Twenty percent of hemangiomas of deep soft tissues cause adjacent bony changes.

The osseous changes can be categorized as periosteal, cortical or medullary. All three categories correlate with

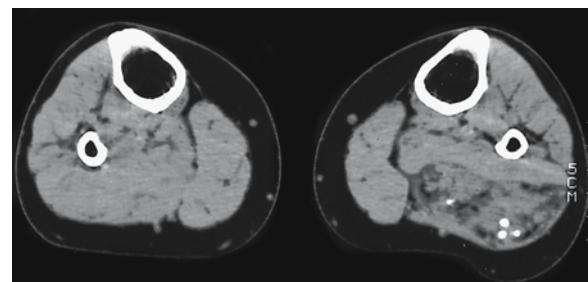


Fig. 16.4. A 47-year-old woman with a swelling of the left calf. Unenhanced CT slice shows an inhomogeneous aspect of the lateral head of the gastrocnemius muscle. The muscle is enlarged and has dispersed internal areas of decreased attenuation. A few rounded phleboliths are seen. There is no involvement of the underlying soleus muscle. Histological examination revealed the presence of a hemangioma. Areas of decreased attenuation correspond to intralesional fat

the proximity of the lesion to bone, whereas medullary changes correspond to size of the hemangioma [73, 74].

Plain film studies are superior to MRI in demonstrating the type and extent of reaction [36]. On the other hand, information about size and extent of the lesion is limited [2, 29, 43, 44, 50, 57].

On computed tomography (CT) benign angiomatic lesions show a mottled low density pattern, partially due to the mixture of fatty, fibrous, and vascular tissue elements. The slow flow and pooling of blood sometimes cause the presence of punctate and curvilinear structures. CT has a greater sensitivity in detecting associated phleboliths (Figs. 16.3 and 16.4) and also clearly shows the relation to adjacent structures in the axial plane. CT may also be useful for exclusion of other soft tissue lesions such as lipomas, which are characterized by a homogeneous low-density attenuation [2, 21, 22, 51].

On angiography the appearance of benign vascular tumors is variable, ranging from small poolings of contrast material over coarse and fine hypervascularity to the presence of large tortuous blood vessels. Angiography has long been the method of choice for defining the lesions extent, vascularity and feeding vessels, allowing distinction from other well-vascularized masses [39, 40].

Angiography of venous malformations, although not indicated, shows contrast pooling in dilated vessels. In arteriovenous malformation, feeding and draining vessels as well as large arteriovenous shunts can be demonstrated [65].

However, underestimation of the extent, false-negative results and inability to define the relationship to adjacent neural structures or fascial planes, have been reported [31, 32]. Moreover, differentiation between hemangiomas and other soft tissue tumors is often not possible. Angiography is, however, still helpful in identi-

fication of feeding and draining vessels for planning embolic therapy or preoperative embolization [44, 50].

Excellent palliation has been reported with embolotherapy of symptomatic arteriovenous malformations [11, 64]. The pain is thought to be secondary to perineural congestion.

Subselective embolization using ethanol, coils and particles usually relieves pain [65].

Ultrasound is a very sensitive technique for detecting soft tissue abnormalities, as well as for demonstrating hemangiomas. If phleboliths are present, they are seen as an echogenic focus with acoustic shadowing. Hemangiomas usually present as oval masses with smooth margins. They are heterogeneous and generally hyperechoic. However, there is no ultrasound appearance specific to muscular hemangiomas [7, 58].

Reports on the use of color Doppler are sparse and usually do not deal with vascular lesions in particular. In some hemangiomas no Doppler signal is noted because of low flow. On the other hand, a characteristic low resistance flow pattern can be noted in high flow vascular malformations [75].

Doppler sonography can also be used for follow-up of AVM after therapy [65].

High vessel density and high peak arterial Doppler shift can be used to distinguish hemangiomas from other soft tissue masses [9].

However, literature findings are contradictory [75]. Paltiel et al. have even tried to distinguish hemangioma from vascular malformation on the basis of mean venous peak velocity and mean resistive index [9, 37, 47, 59].

Only one report in the literature mentions the role of positron emission tomography in the evaluation of hemangioma. PET is promising for differentiating benign hemangiomas from other soft tissue tumors [76].

On plain radiographs a glomus tumor is seen as a soft tissue mass usually at the dorsal surface of the finger. A characteristic bone erosion, possibly with sclerotic margin is sometimes noticed [15, 44]. Ultrasound reveals a hypoechoic lesion but is not sensitive [17].

It is not possible to differentiate between hemangiobendothelioma and angiosarcoma on the basis of plain radiography [16].

Reports on CT and ultrasound findings in malignant vascular tumors are sparse and usually non-specific [16, 18]. CT show a heterogeneous mass which becomes sharply delineated owing to marked contrast enhancement. Angiosarcoma following chronic lymphedema is characterized by fibrous thickening, increased fat attenuation, and fluid collections surrounding the muscles [27, 50].

16.4.2 Imaging Findings on MRI

MRI is considered the modality of choice in evaluating soft tissue masses, and hemangioma is no exception to this rule [2, 4, 21, 24, 44, 46, 48, 50, 53, 60, 73, 74].

Deeply located and intramuscular hemangiomas usually have no specific clinical signs, unlike their cutaneous variants, and detection is usually delayed [12, 44]. Medical imaging is indispensable for preoperative assessment and is carried out preferentially by MRI.

The descriptions in the literature can be divided into two groups: those that use the term hemangioma and do not distinguish between different subtypes [4, 24, 46, 56]. On the other hand we share the opinion of some authors who believe that MRI can be used to differentiate slow and high flow vascular malformations and hemangiomas, which is of major therapeutic importance [3, 37, 49].

If benign angiomatic lesions are grouped under the term hemangiomas, high signal intensity on T2-weighted images is the most characteristic MRI finding of these lesions [56]. The signal intensity is indeed higher than that of subcutaneous fat. Increased T2-weighting demonstrates increased brightening of the hemangioma and usually improved delineation of the edge and extent of the lesion [46] (Fig. 16.5). On T1-weighted images the signal intensity of the lesions is usually intermediate between that of muscle and fat. Histological comparison shows that slow flow within dilated venous channels, pooling of blood in cavernous spaces, thrombosis, and the presence of fatty elements are responsible for the higher signal intensity on T1- and T2-weighted images (Fig. 16.6). Capillary hemangiomas and small hemangiomas may not present with high signal intensity on T1-weighted images [2].

Except for small lesions (diameter less than 2 cm) and some located in the extremities [4, 73, 77] all hemangiomas are inhomogeneous on both T1- and T2-weighted images. On T2-weighted images punctate or reticular low signal intensity areas may be present corresponding to fibrous tissue, fast flow within blood vessels, or calcified or ossified foci. The presence of those areas is an important feature in the differential diagnosis between hemangiomas and lipomas or intramuscular hematomas [2, 49, 54, 56, 73, 74].

High flow in vessels or fibrous tissue or calcifications can be differentiated using gradient echo sequences with gradient-moment nulling [37].

Hemangiomas are generally composed of multiple lobules of high signal intensity with a 'bunch of grapes' appearance (Fig. 16.7).

Fig. 16.5 a,b. High signal intensity of hemangioma on T2 weighted images: **a** subcutaneous hemangioma of the wrist. Coronal T2-weighted image with fat suppression shows a multilobular mass which involves the subcutaneous fat. The lesion is hyperintense and inhomogeneous due to internal septations; **b** capillary hemangioma of the neck in a two-years old boy. Axial T2-weighted image shows an inhomogeneous septate mass which is sharply delineated and is hyperintense to subcutaneous fat. There is no invasion of the underlying muscle

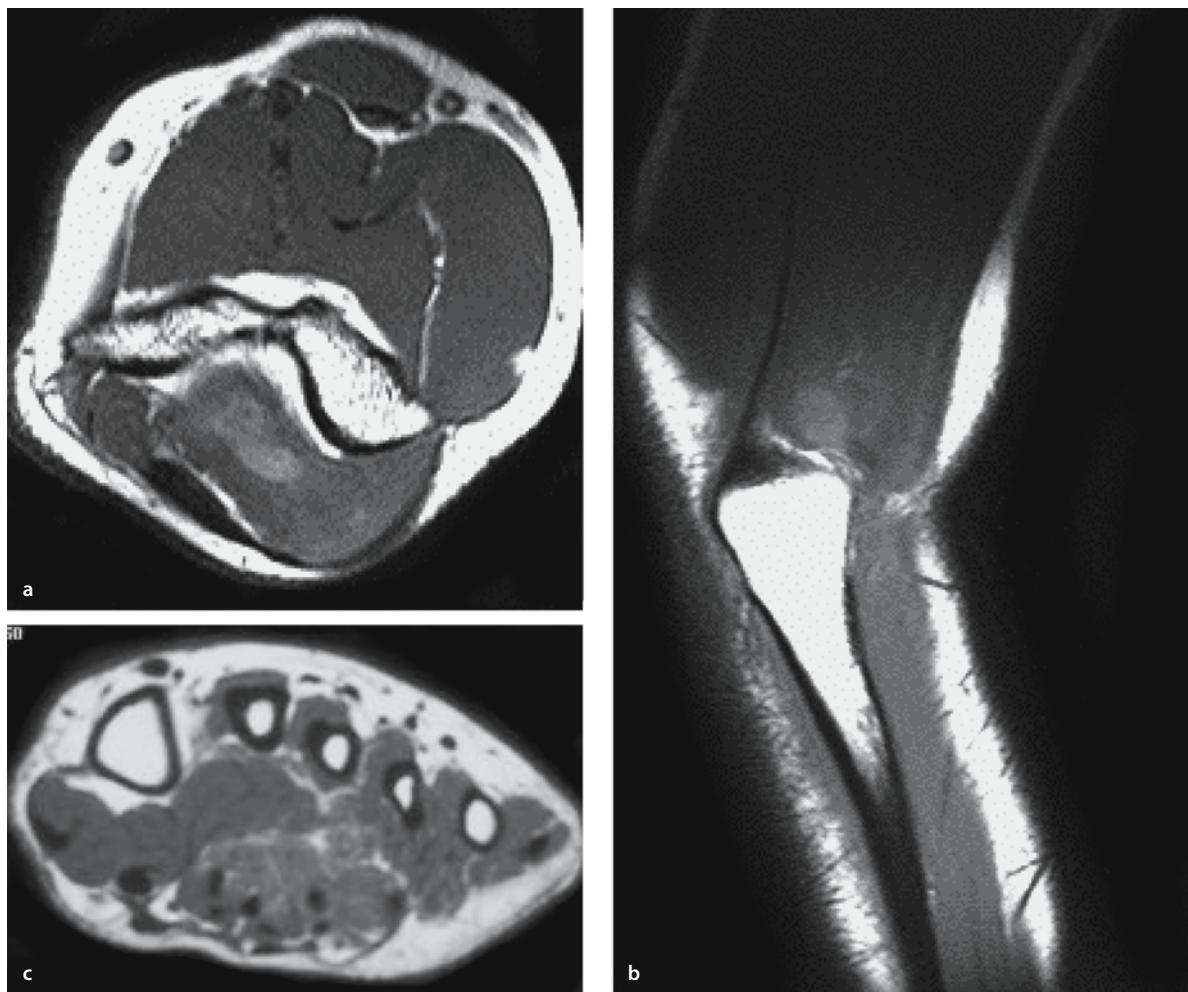
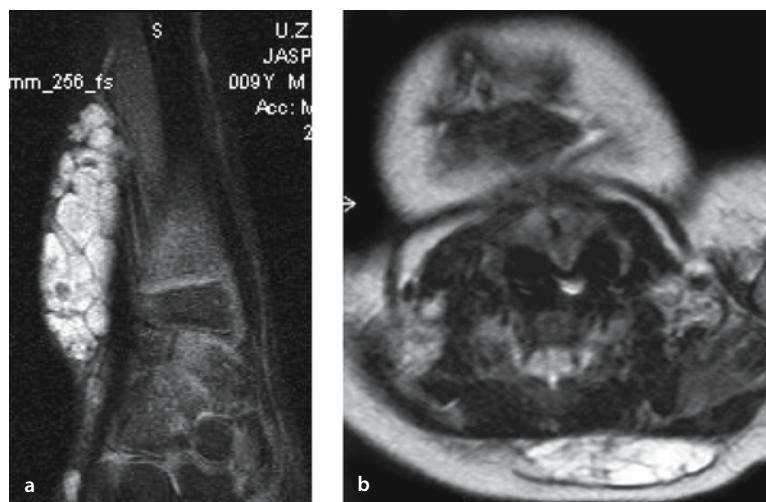


Fig. 16.6 a–c. Signal intensity of hemangioma on T1-weighted images: **a,b** synovial hemangioma of the left elbow. The lesion is located in the posterior recess of the joint and is hyperintense compared to the triceps muscle. High signal intensity is probably

caused by stagnant blood; **c** the signal intensity of the flexor digitorum muscles is minimally increased due to the presence of intra-muscular hemangioma. Note also the presence of peripheral fatty components

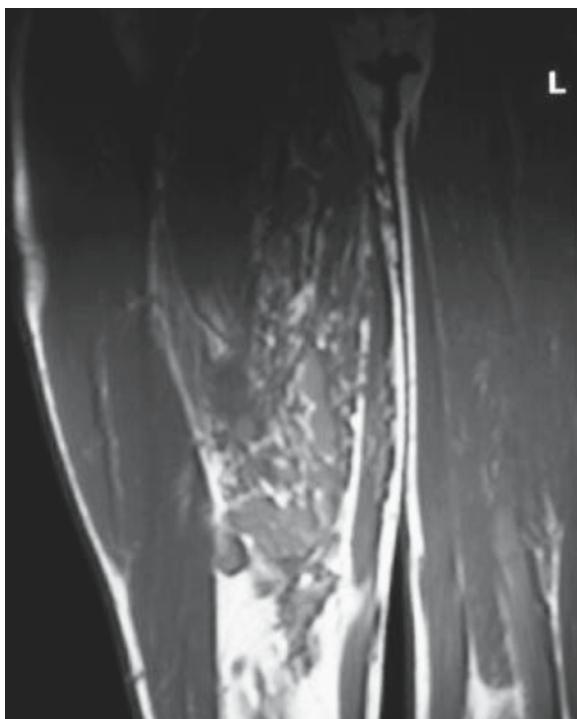


Fig. 16.7. Coronal T1-weighted image of the right thigh shows an intramuscular mass which involves the hamstring muscles. The lesion is composed of multiple nodular components which make it resemble 'a bunch of grapes'

This is probably due to cavernous or cystic vascular spaces containing stagnant blood [4]. It is also within these spaces that fluid-fluid levels can be noted. These fluid-fluid levels are mostly seen in cavernous hemangiomas and are caused by hemorrhage. On T2-weighted images a high signal intensity 'supernatant' corresponding to serous fluid overlies a low signal intensity lower layer. Fluid-fluid levels are not appreciated as easily on T1-weighted images, but they are nevertheless present. On T1-weighted images the signal intensity of the higher layer is low, and that of the lower layer is high (Fig. 16.8).

As noted above, hemangiomas are generally inhomogeneous on T1-weighted image. The lesions are predominantly isointense with muscle, with internal serpiginous high signal intensity strands. These strands correspond to enlarged vessels and generally are orient-

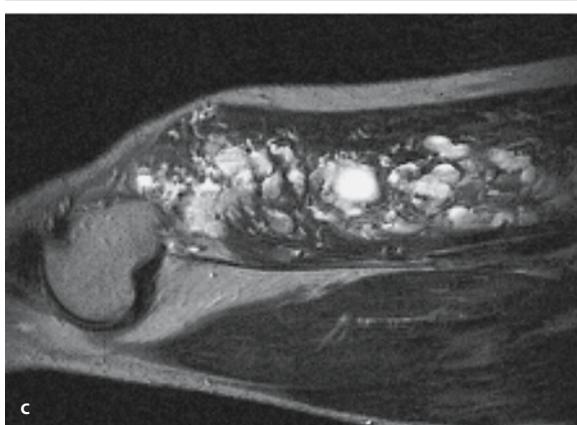
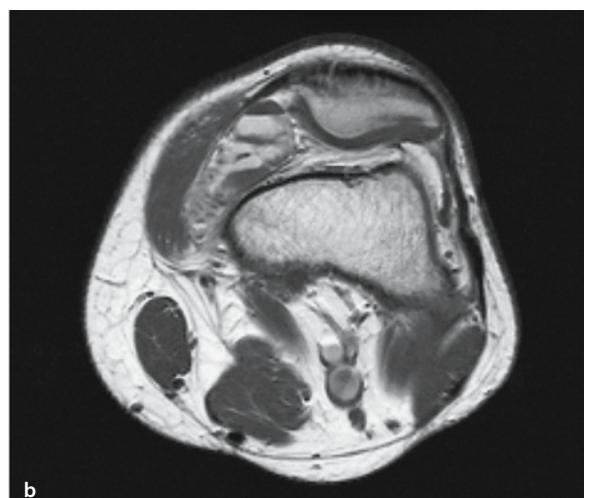
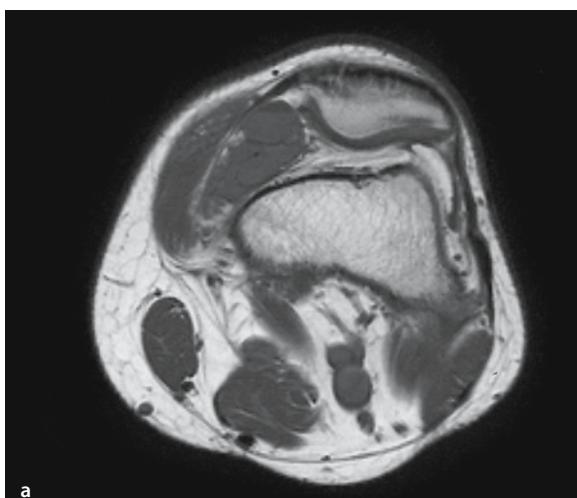


Fig. 16.8. **a** Axial T1-weighted MR image. **b** Axial T1-weighted MR image after gadolinium contrast injection. **c** Sagittal T2-weighted MR image. A fluid–fluid level is seen on axial T1-weighted images. The supernate is slightly hypointense compared with the lower layer due to its serous composition (**a**). The fluid–fluid level becomes clearer after administration of gadolinium due to the sedimentation of gadolinium to the lower part of the lesion, the same phenomenon as is noted within the bladder after intravenous administration of gadolinium chelates (**b**). On T2-weighted images there is increased contrast between the hyperintense serous upper layer and the lower layer, which is hypointense due to the sedimentation of erythrocytes (**c**)

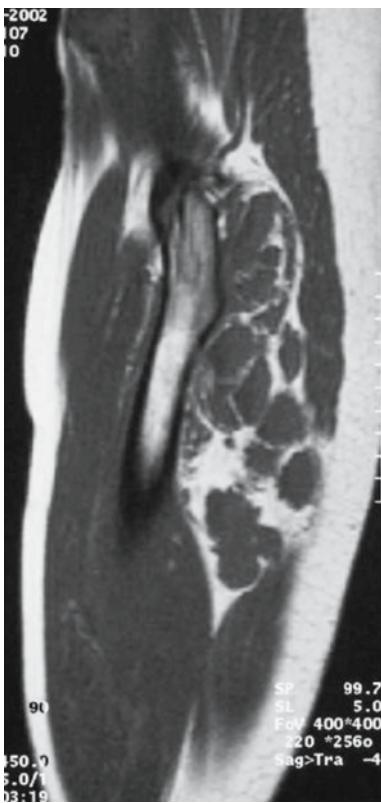


Fig. 16.9. Intramuscular hemangioma in a 40-years-old man. On sagittal T1-weighted images, the signal intensity of the lesion is inhomogeneous. There are central nodular components that are isointense to muscle. The peripheral parts are mainly composed of fatty tissue and are hyperintense to muscle

ed parallel to the muscle fibers [21, 56]. In some hemangiomas peripheral high signal intensity areas are noted on T1-weighted images, corresponding to fat within the lesions (Fig. 16.9).

A frequent finding is hypertrophy or atrophy of the muscle or the subcutaneous fat involved by the hemangiomas (Fig. 16.10). Hypertrophy of the limb in angiomyomatosis is also known.

The classification of Mulliken differentiates hemangiomas, venous malformations and arteriovenous malformations.

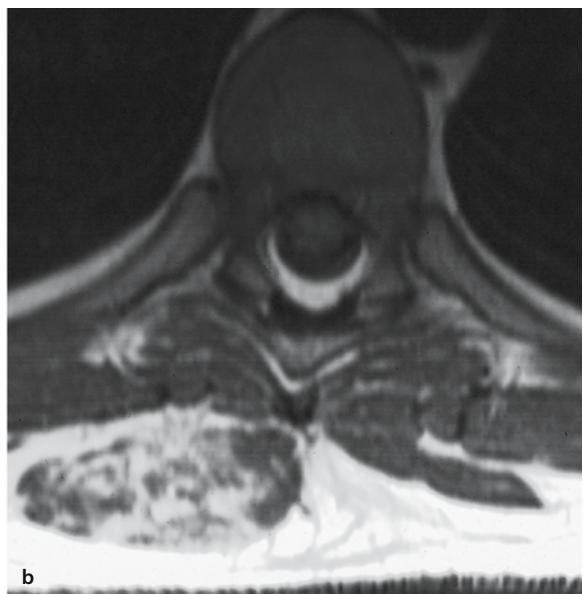
Slow flow vascular malformations include venous, capillary, cavernous and mixed types. The appearance on MRI depends on the composition. Venous portions show large spaces with internal linear or serpentine structures of low or high signal intensity depending on the pulse sequence and blood flow velocity [78, 79].

In addition they are oriented along the long axis of the limb, follow neurovascular distributions, and are sometimes multifocal (Fig. 16.11). Usually there is an enlargement of the neighboring subcutaneous fat. The combination of all findings suggests a congenital tissue dysplasia.

On the other hand, high flow arteriovenous malformations have signal voids on all pulse sequences. Although the lesions can be associated with surrounding edema or fibrofatty stroma, no focal soft tissue mass is found [37, 49] (Fig. 16.12).



Fig. 16.10 a, b. A 55-year-old woman complaining of interscapular pain. **a** CT after iodinated contrast injection. **b** Axial spin echo T1-weighted MR image. There is a marked hypertrophy of the right trapezoid muscle compared to the contralateral side. Central and peripheral enhancing areas are noted, corresponding to enlarged



vessels (**a**). In addition to the hypertrophy, signal intensity of the right trapezoid muscle is inhomogeneous and increased, nearly isointense to subcutaneous fat. The increase is caused by slow flow in enlarged veins (**b**). The presence of enlarged veins and the hypertrophy of the muscle are characteristic for hemangioma

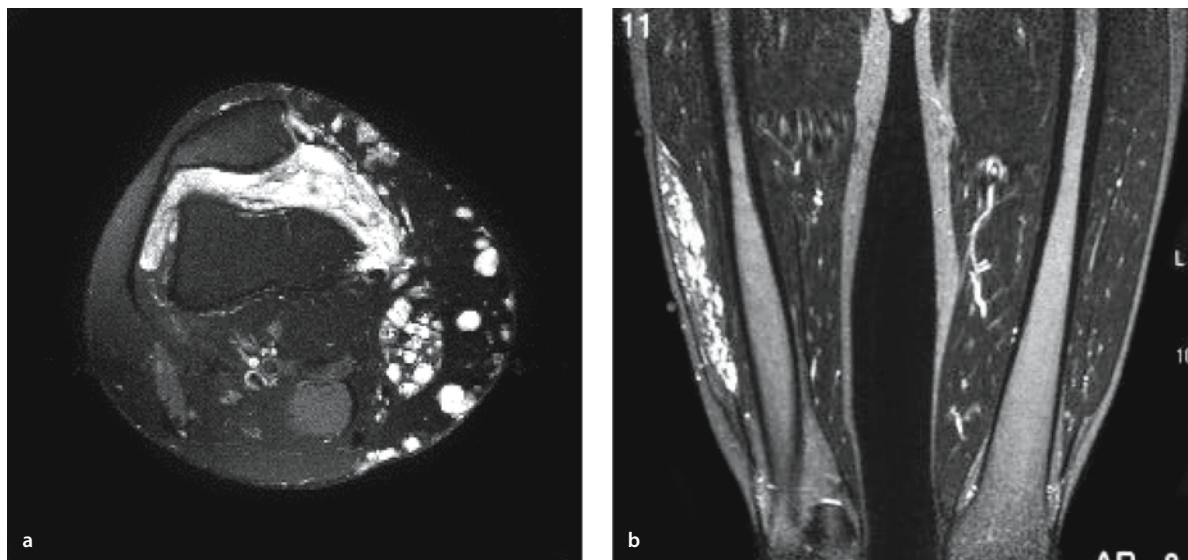


Fig. 16.11 a, b. Venous hemangiomas: **a** axial T2-weighted image with fat saturation shows the involvement of multiple compartments of the knee region by the lesion. There are hyperintense nodular and serpentine areas in the subcutis, the semimembranous muscle and within the joint, in the anterior fat pad. High signal intensity is caused by slow flowing blood in dilated venous channels; **b** coronal inversion recovery T2-weighted image in another patient shows pooled blood within saccular dilated blood vessels. There is extension along the long axis of the involved leg

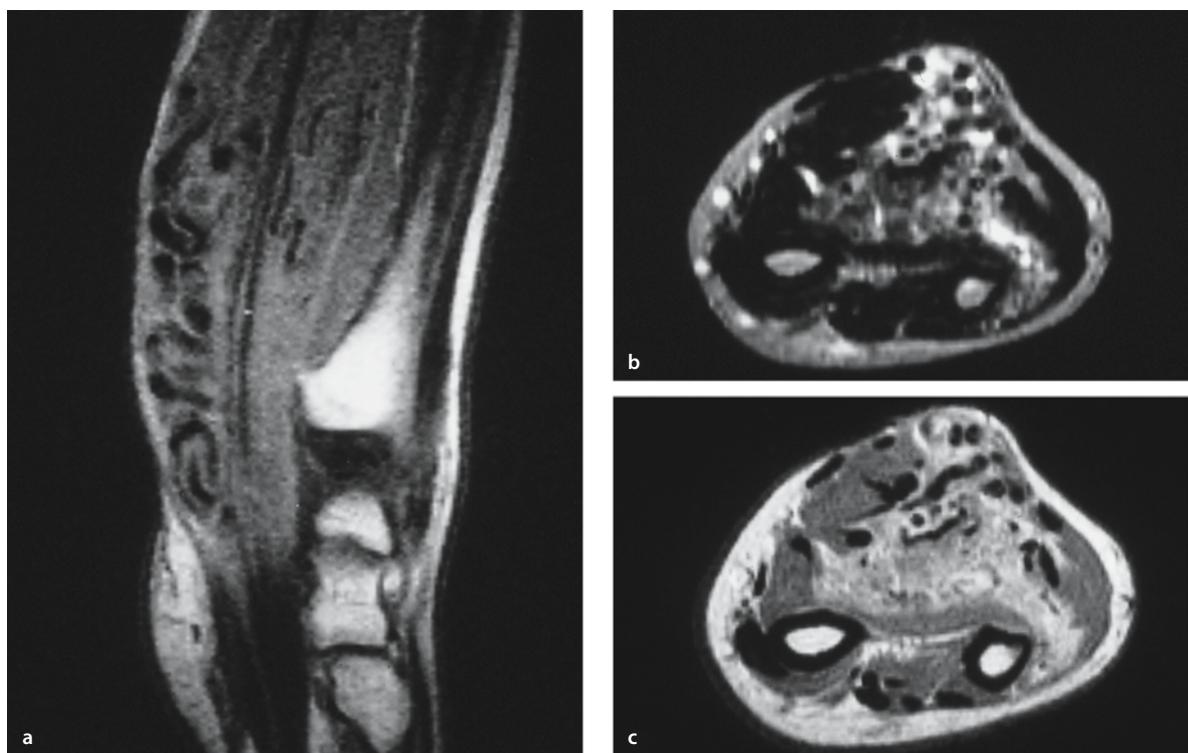


Fig. 16.12 a–c. High-flow arteriovenous malformation in the flexor muscles of the forearm. **a** Sagittal T1-weighted MR image. **b** Axial T2-weighted MR image. **c** Axial T1-weighted MR image after gadolinium contrast injection. Multiple serpiginous areas of signal void are present within the flexor muscles of the fingers. These muscles are nearly isointense on T1-weighted images (**a**). On T2-

weighted images the involved muscles are slightly hyperintense, with internal areas of signal void. High signal intensity areas correspond to the matrix of vascular malformation, while signal void is caused by high flow in dilated vessels (**b**). After administration of Gd contrast, there is only enhancement of the solid part of the mass (**c**). (From H. Van Moer, with permission)

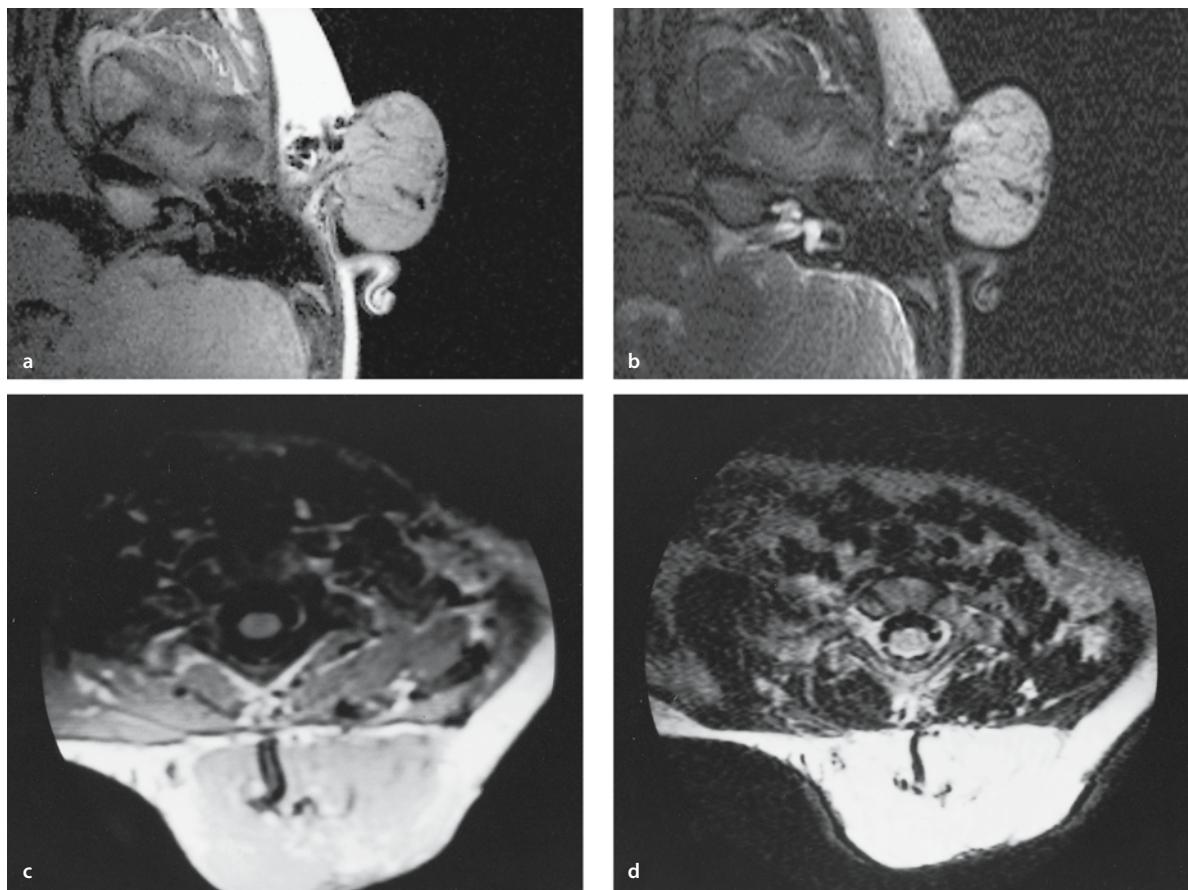


Fig. 16.13a–d. Hemangiomas of infancy. **a, b** Axial T1- and T2-weighted MR images in a 2-year-old girl. **c, d** Axial T1- and T2-weighted MR images in a 17-month-old girl. A preauricular mass that is isointense on the T1-weighted image and hyperintense on the T2-weighted image is seen (**a, b**). Enlarged feeding vessels are seen within the subcutis anterior to the lesion and peripherally

within the lesion. The same findings are noted on axial T1- and T2-weighted images of the neck in the other patient (**c, d**). A horizontal snakelike area of signal void is seen on both images. The presence of enlarged feeding or draining vessels is typical for hemangiomas of infancy and is not seen in venous malformations

Infantile hemangiomas display the features described above: a lobulated mass that is hyperintense on T2-WI and isointense to muscle on T1-WI. They have prominent feeding or draining vessels which are identified as central or peripheral high flow channels (Fig. 16.13).

T1-weighted images after the administration of gadolinium show a moderate to strong enhancement of the vascular lesions. The pattern of enhancement varies with the interval between imaging and the time of injection, depending on the rate of inflow of contrast into the blood-filled spaces [56]. Degree of enhancement varies with flow velocities of blood in the vessels of the hemangioma. Therefore more pronounced enhancement occurs in the low signal intensity parts on native scans, and the lesions become more homogeneous. The contrast-to-noise ratio between hemangiomas and surrounding tissues, however, remains lower than on T2-weighted images (Fig. 16.14).

Conventional MR imaging generally does not allow to making a distinction between different types of venous malformations.

However, Verstraete et al. maintain that dynamic contrast-enhanced MRI and ‘first pass’ images enable the differentiation between cavernous and capillary hemangiomas and between low flow and high flow vascular malformations [62] (Fig. 16.15; see Chap. 6).

Low slope values are usually found in cavernous types due to low perfusion, while capillary types show high perfusion and high slope values. The first-pass images show good correlation with angiographic findings.

More recent studies confirm that dynamic contrast enhanced MRI provides images that are comparable to conventional angiography. In addition time-intensity curves allow to distinguishing venous malformations from mixed capillary-venous and arteriovenous malformations [80, 81].

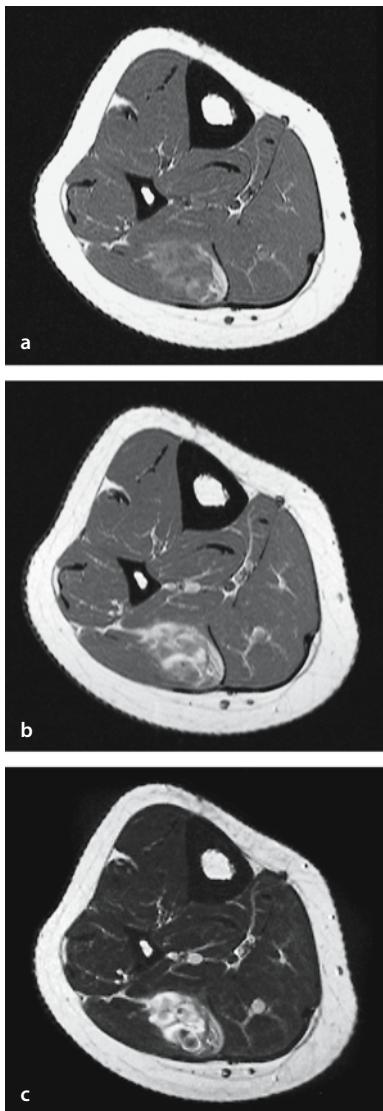


Fig. 16.14 a–c. A 27-year-old man presenting with a painful swelling of the right calf. **a** Axial T1-weighted MR image. **b** Axial T1-weighted MR image after gadolinium contrast injection. **c** Axial T2-weighted MR image. An ill-defined, inhomogeneous mass within the lateral head of the gastrocnemius muscle is seen. The mass has a hyperintense medial border. The lateral part is nearly isointense to muscle (**a**). After contrast administration homogenization of signal intensity takes place due to a higher degree of enhancement of the low signal intensity parts on native T1-weighted images (**b**). The exact extent of the lesion, however, is still better appreciated on T2-weighted images (**c**)

Teo et al. retrospectively reviewed the records of patients with soft tissue hemangiomas and compared them with those having malignant masses. No single MR feature was diagnostic, but analysis of morphology, and signal intensity on native and enhanced images allowed differentiation. The combination of high signal intensity on T2-weighted images, high contrast enhancement, and lobulated, septate morphology was typical for hemangioma [60].

Synovial hemangiomas present with similar features as intramuscular types, except for the high frequency of pressure erosions of adjacent cortical bone and the low signal intensity synovial lining [20, 33, 56] (Figs. 16.2 and 16.16). The appearance of angiomyomatosis on MRI is identical to that of solitary angiomyomatous lesions. MRI is ideally suited for defining the extent of soft tissue involvement [40] (Fig. 16.17).

On MRI a glomus tumor is seen as a homogeneous hyperintense lesion on T2-weighted images. Definition of extent is superior to that on other techniques [59].

MRI characteristics of hemangioendothelioma and angiosarcoma may be non specific (Fig. 16.18). Some authors mention the presence of prominent serpentine vessels suggestive for the diagnosis.

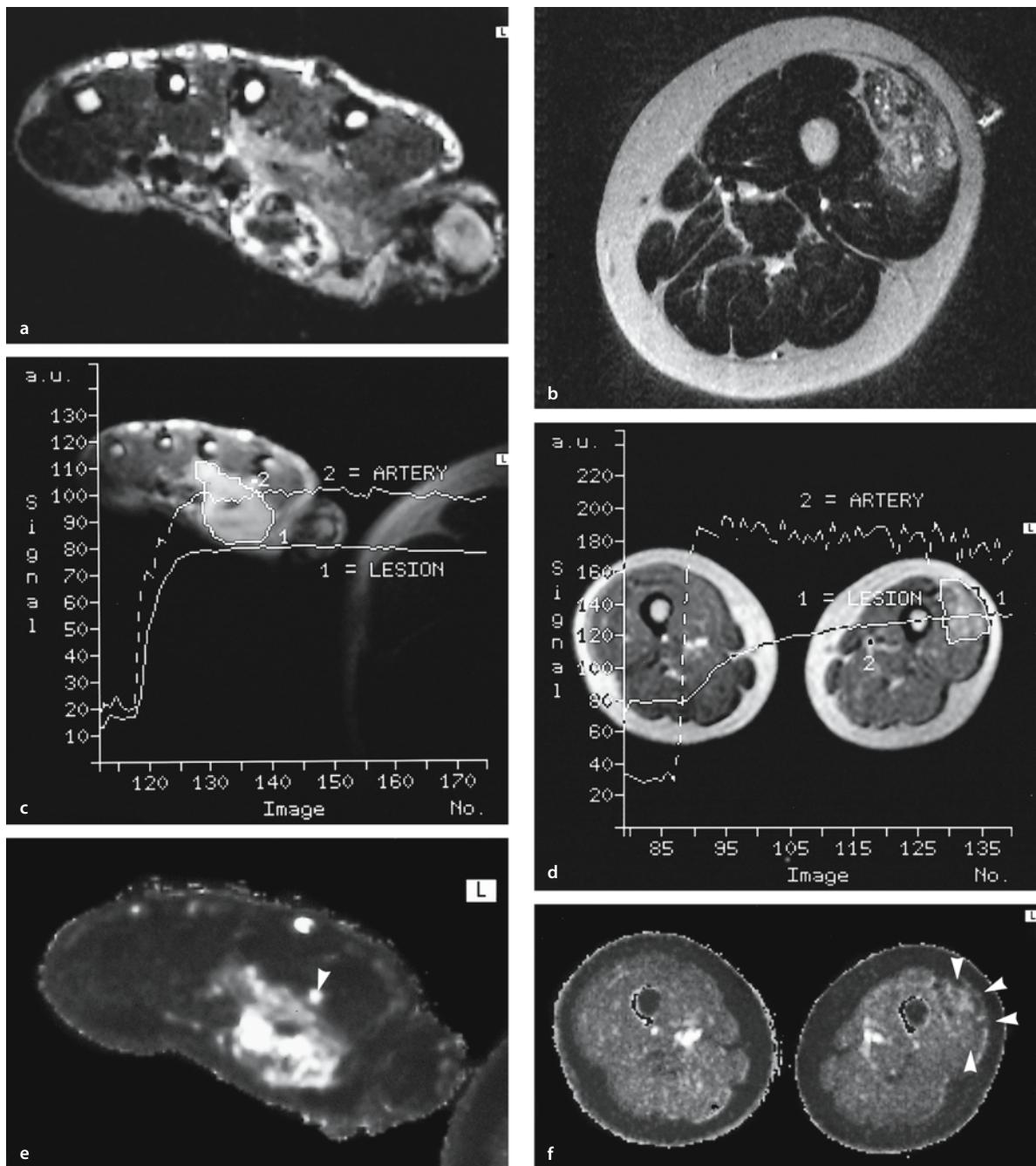


Fig. 13.15 a-f. Differentiation of high-flow from low-flow hemangioma with dynamic MRI. **a** Axial T2-weighted image in a 34-year-old man. **b** Axial T2-weighted MR image in a 19-year-old girl. **c, d** Time-intensity curves. **e, f** First-pass MR images (turboFLASH; TR/TE/TI/flip angle: 9ms/4ms/200ms/8°). Axial T2-weighted MR images show a soft tissue lesion of the right hand with low, intermediate, or high signal intensity, corresponding to an arteriovenous hemangioma (**a**) and to a cavernous heman-

gioma within the vastus lateralis muscle which displays intermediate to high signal intensity (**b**). On the time-intensity curve, the arteriovenous hemangioma has a high first-pass enhancement that parallels the arterial curve, indicating high perfusion, whereas the cavernous hemangioma has a slow perfusion (**c, d**). On first-pass images the arteriovenous hemangioma appears as bright as the feeding artery (**e, arrowhead**), whereas the cavernous hemangioma appears dark, due to slow perfusion (**f, arrowheads**)

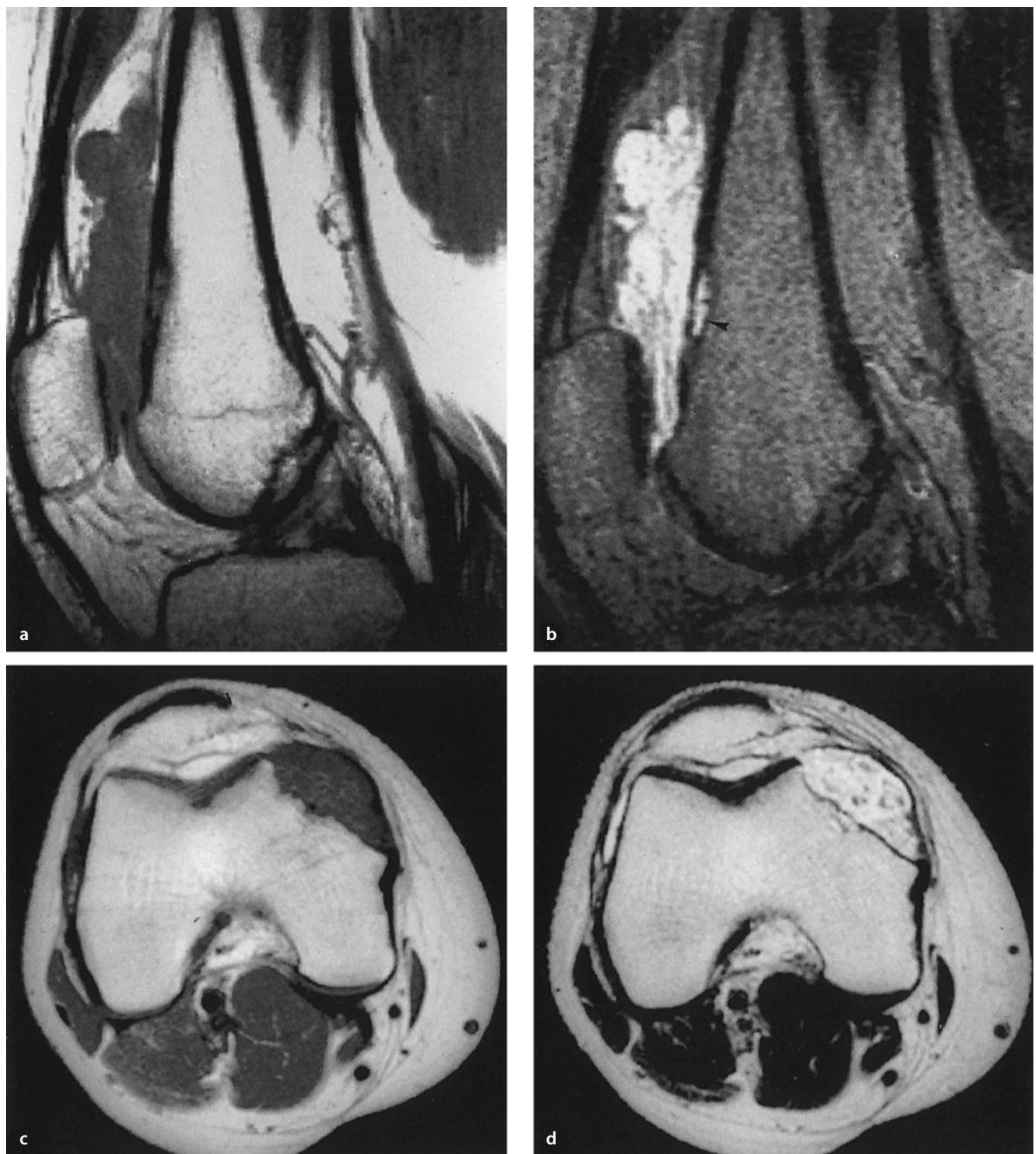


Fig. 16.16a–d. Synovial hemangioma. **a** Sagittal T1-weighted MR image. **b** Sagittal T2-weighted MR image. **c** Axial T1-weighted MR image. **d** Axial T2-weighted MR image. A well-defined homogenous lesion with lobulated contours is seen within the suprapatellar bursa. The lesion is isointense to muscle on T1-weighted images (**a, c**) and hyperintense to fat on T2-weighted images (**b, d**) with

small linear and punctate areas of low signal intensity. The axial slices reveal erosion of the anterior cortex of the distal femur with adjacent changes within the medullary fat, probably corresponding to bone marrow edema. (Reprinted from [33], with permission)

Fig. 16.17. Angiomatosis of the right lower limb. Axial T1-weighted image after gadolinium contrast injection shows the extension of the angiomatic lesion involving the subcutaneous tissues and all muscles with exception of the peroneal muscles, which display normal signal intensity. Note also the obvious hypertrophy of the right lower limb

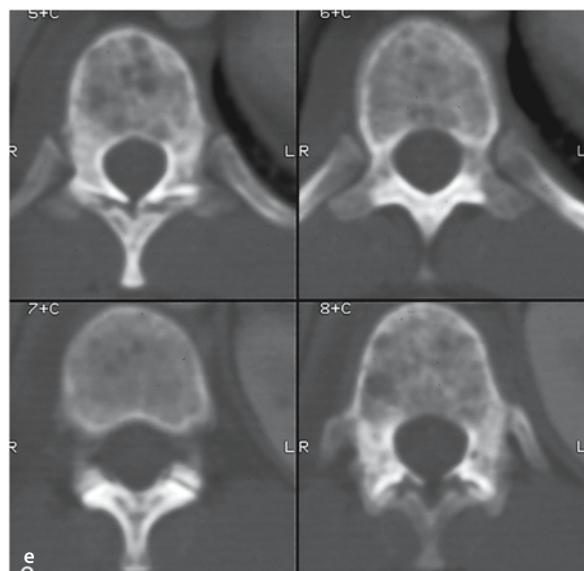
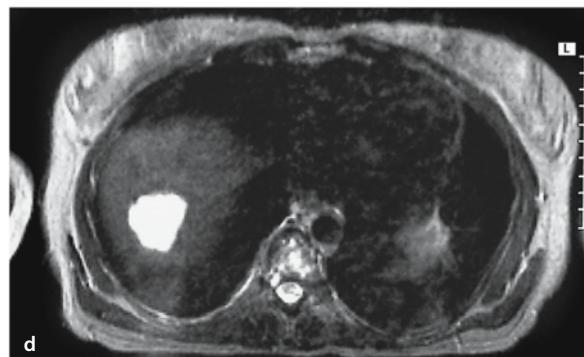
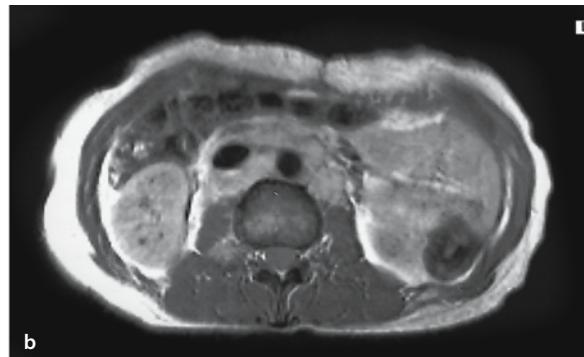
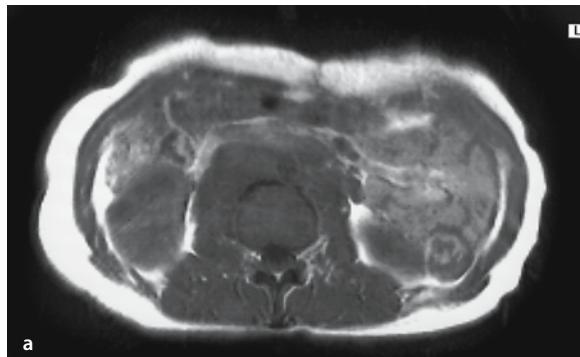
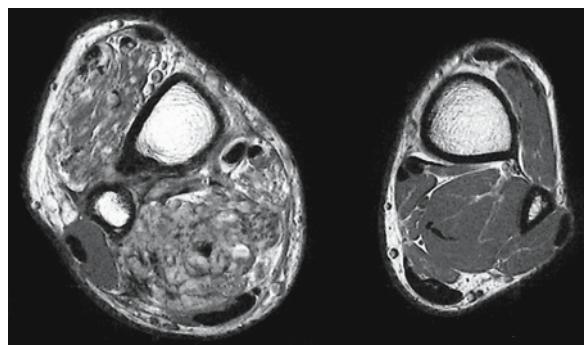


Fig. 13.18a–e. A 35-year-old woman with retroperitoneal bleeding. **a** Axial T1-weighted MR image. **b** Axial proton density weighted MR image. **c** Sagittal T1-weighted MR image after gadolinium contrast injection. **d** Axial T2-weighted MR image. **e** CT. Axial MR images demonstrate the presence of a retroperitoneal mass which encircles both the aorta and the inferior caval vein. The lesion is hypointense on the T1-weighted images (**a**) and has an intermediate signal intensity on the proton density weighted image (**b**). After administration of gadolinium there is a minimal enhancement of the lesion (**c**). The diagnosis of angiosarcoma is made after open biopsy. On the sagittal images inhomogeneous signal intensity of the intravertebral bone marrow is noticed (**c**). On the T2-weighted image multiple high signal intensity spots are seen within the vertebral bodies (**d**), which correspond to lytic areas on CT (**e**). Finally, a hyperintense liver lesion is seen on the T2-weighted image, which proved to be a hemangioma. MRI findings in angiosarcoma are only sparsely reported and are nonspecific. In this patient there is the coincidence of angiosarcoma, liver hemangioma, and diffuse skeletal hemangiomatosis

16.4.3 Imaging Strategy

Cutaneous and subcutaneous vascular tumors usually present with characteristic clinical features. For this reason diagnosis usually poses no problems. Surgical excision can be performed without preoperative radiological assessment [44]. However, if extension to or involvement of the underlying tissues is suspected, MRI has a definite role in the evaluation.

Because patients with deep-seated vascular tumors present with a mass or pain, it is difficult to make a diagnosis based on clinical examination. Nonspecific complaints lead to delayed detection and misdiagnosis. A majority of patients undergo surgery without a definite evaluation of the extent of the lesion or preoperative tissue diagnosis. A high recurrence rate due to incomplete resection is reported.

The choice of imaging techniques is dictated by the availability of equipment and patient-related considerations. When a deep hemangioma is suspected, imaging should start with plain radiography. Characteristic phleboliths and bone erosion can be depicted. Plain radiographs are ideally followed by MRI. MRI can provide characteristic features such as high signal intensity on T2-weighted images and curvilinear or serpentine inhomogeneities on all pulse sequences. Patterns of signal intensity and morphology may allow a presumptive diagnosis of hemangioma even in patients in whom vascular lesions are not suspected. The use of T1-weighted sequences is essential in evaluating hemangiomas since most other lesions have low signal intensity on T1-weighted images.

However, one must be aware of certain pitfalls. Myxoid tumors can show the same reticular high and low signal intensity and the infiltrative appearance of angiomyomatosis.

Other intramuscular tumors complicated by intralensional hemorrhage or intramuscular hematoma have the same signal intensity as hemangiomas but lack the presence of enlarged vessels. Tumors that infiltrate the subcutaneous fat or are primarily fatty may also resemble hemangiomas [19].

Differentiation of hemangiomas and low flow or high flow vascular malformations is also preferentially done by MRI. Donnelly et al. use a combination of axial T1-weighted, fat sat T2-weighted and gradient echo MR images with the addition of coronal or sagittal spin echo T2 weighted images. Differentiation of the lesions is essential for providing appropriate monitoring and therapy. Other authors believe that the combination of conventional MR characteristics with dynamic contrast enhanced features can be used for this purpose [78, 80, 81].

Several reports have shown the superiority of MRI over CT and angiography in delineating the extent of vascular lesions. The multiplanar format permits a complete assessment of size, location, extent and topo-

graphic relationship. Since about one third of all lesions, especially venous malformations, are multifocal, this is best achieved by slices oriented parallel to the limb axis. The precision of CT in defining the craniocaudal extent or the extent of lesions without a fatty margin is markedly inferior, due to similar attenuation of hemangioma and muscle. The better differentiation between vascular lesions and the surrounding tissues on T2-weighted images allows a precise definition of the size and the extent [23, 35].

MRI is also superior to evaluate the involvement of vital structures such as neurovascular bundles. Such information is vital to planning surgery or imaging-guided procedures [65].

The extent of venous angiomas is best demonstrated on short inversion time inversion recovery images. Slow-flowing blood is responsible for marked hyperintensity on these short inversion time inversion recovery images, permitting excellent depiction of topography [53]. T1-weighted images may not be useful in cases of subcutaneous hemangiomas because it is difficult to differentiate normal subcutaneous fat from that belonging to the tumor. The only major limitation of MRI compared with plain radiography and CT is the lower sensitivity to identify phleboliths. Large phleboliths can be seen as areas of low signal intensity on T1- and T2- weighted images. However, this is a non-specific finding, since this cannot be differentiated from fibrous tissue.

Concerning vascular tumors of intermediate malignancy and malignant vascular tumors, no specific literature findings concerning imaging strategy have been published. However, we recommend that imaging starts with plain radiography to demonstrate the presence of calcifications or ossifications. Plain radiographs are followed by MRI because of its superiority in staging of soft tumors [5, 64]. The administration of gadolinium contrast is indispensable since it is essential for the demonstration of intratumoral necrosis, which is a very specific sign indicating malignancy [61]. It is also essential for postoperative assessment in differentiation of postoperative fibrosis and tumor recurrence.

Things to remember:

1. The majority of vascular tumors are benign and are located in the skin or subcutis. They are classified as hemangiomas, which show cellular proliferation, and vascular malformations, which represent a dysplasia rather than a tumor.
2. Hemangiomas can involve large segments of the body, when they are a part of angiomatic syndromes or angiomyomatosis. However, they are usually small and clinically insignificant.

3. When a deep hemangioma is suspected, plain radiography is performed to demonstrate phleboliths, followed by MRI; T1-weighted images show characteristic high signal intensity, and T2-weighted images show high signal intensity with curvilinear or serpentine inhomogeneities.
4. Diagnosis of vascular malformations is done preferentially by MRI. MRI can be used to differentiate slow and high flow vascular malformations and hemangiomas using both conventional and dynamic contrast enhanced sequences.

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