16 Hemangioblastoma

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BACKGROUND

Hemangioblastoma is a vascular tumor of the central nervous system (CNS). It most commonly occurs within or on the surface of the cerebellum and represents the most common primary tumor of the posterior fossa in adults. Hemangioblastomas may also occur within the spine. Single tumors may be sporadic, but multiple tumors are almost always associated with von Hippel-Lindau (VHL) disease. Sporadic tumors appear in the fifth and sixth decades of life, whereas VHL-associated tumors are detected earlier, in the third and fourth decades. One-third of patients with cerebellar hemangioblastoma have VHL disease. Two-thirds of VHL patients develop hemangioblastomas; thus screening and surveillance programs are required for this population.

Progressive evolution of hemangioblastomas from solid tumor to cyst with mural nodule has been proposed in the literature. Patients typically present with symptoms due to mass effect, most often from an enlarging peritumoral cyst. An understanding of this progression can assist in clinical decisions regarding follow-up intervals and the timing of surgical intervention.

TYPES OF HEMANGIOBLASTOMA

Hemangioblastomas are traditionally categorized as one of four types by either histology or imaging (Fig. 16.1). Type 1 (5% of posterior fossa hemangioblastomas) is a simple cyst without a macroscopic nodule. Type 2 is a cyst with a mural nodule (60%). Type 3 is a solid tumor without cyst (26%), and type 4 is a solid tumor with small internal cysts (9%). Types 3 and 4 lesions predominate in the spinal cord. Of note, many authors have disputed the existence of type 1 (purely cystic tumors), questioning the quality of presurgical imaging (contrast not given or slice thickness limitations) or detail of histologic sectioning.

IMAGING APPEARANCE

Hemangioblastomas are vascular tumors; thus the solid tumor components demonstrate intense enhancement following contrast administration (Fig. 16.2). It should be noted that when a cyst

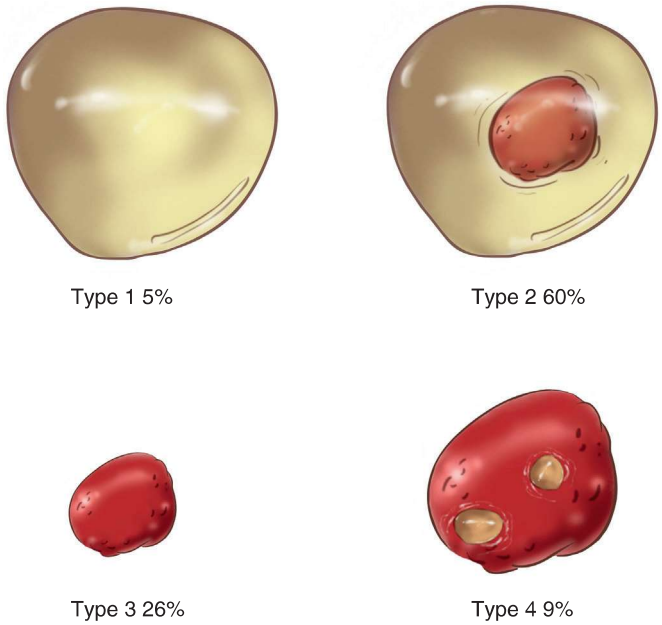

Figure 16.1. Types of hemangioblastomas with percent prevalence in the posterior fossa. Type 1 tumors likely represent type 2 tumors with solid components that escape imaging or histologic detection.

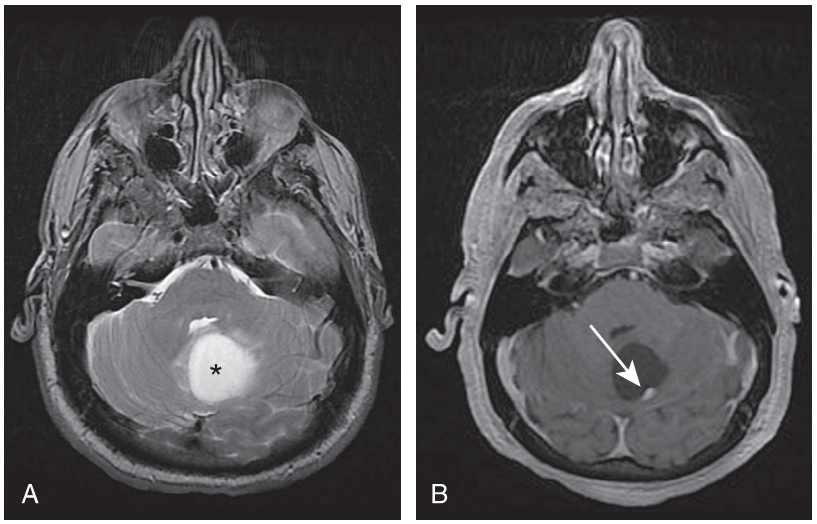

Figure 16.2. Type 2 hemangioblastoma. Large cyst with small enhancing mural nodule. (A) Axial T2-weighted image with cyst *(*)* partially effacing the fourth ventricle. (B) Postcontrast axial T1-weighted image demonstrates a small enhancing mural nodule *(arrow)* along the posterolateral wall of the cyst. Only the enhancing nodule represents tumor. The wall of the peritumoral cyst does not enhance and does not contain tumor (see section titled "Stages of Evolution").

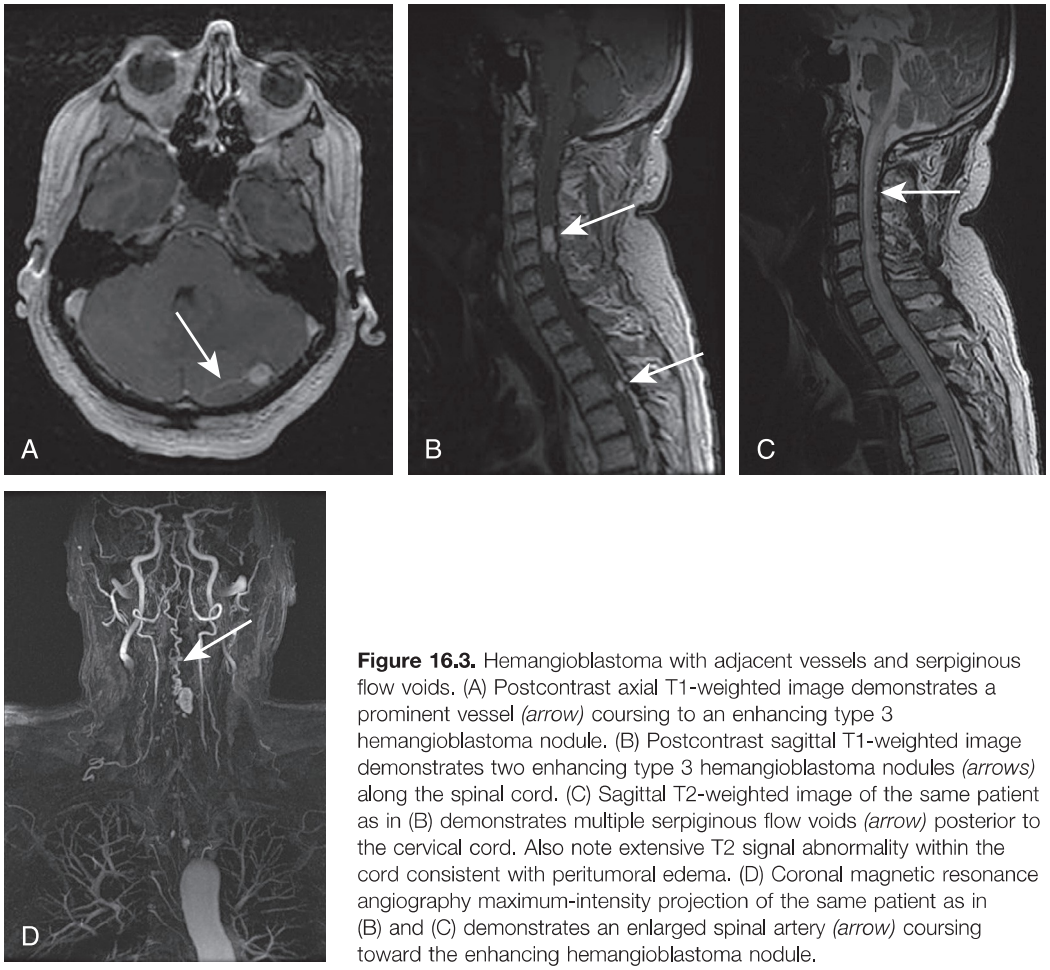

Figure 16.3. Hemangioblastoma with adjacent vessels and serpiginous flow voids. (A) Postcontrast axial T1-weighted image demonstrates a prominent vessel *(arrow)* coursing to an enhancing type 3 hemangioblastoma nodule. (B) Postcontrast sagittal T1-weighted image demonstrates two enhancing type 3 hemangioblastoma nodules *(arrows)* along the spinal cord. (C) Sagittal T2-weighted image of the same patient as in (B) demonstrates multiple serpiginous flow voids *(arrow)* posterior to the cervical cord. Also note extensive T2 signal abnormality within the cord consistent with peritumoral edema. (D) Coronal magnetic resonance angiography maximum-intensity projection of the same patient as in (B) and (C) demonstrates an enlarged spinal artery *(arrow)* coursing toward the enhancing hemangioblastoma nodule.

is associated with this tumor, it is a true "peritumoral cyst"; the wall does not enhance and the wall does not contain tumor (see section titled "Stages of Evolution," further on). Hemangioblastomas often have enlarged feeding vessels that may enhance or manifest as serpiginous hypointense flow voids on T2-weighted images (Fig. 16.3).

Multiple lesions would suggest underlying VHL. A less common VHL-associated tumor that may be detected on CNS screening examinations is the endolymphatic sac tumor, which typically causes permeated destruction of the posterior surface of the temporal bone.

STAGES OF EVOLUTION IN HEMANGIOBLASTOMA

Patients with sporadic tumors often present when the lesion has grown large enough to cause marked mass effect, resulting in symptoms referable to the area of the lesion. In the case of type 2 hemangioblastomas (cyst with a mural nodule), the cystic component is typically the predominant feature and largely responsible for the degree of mass effect.

Screening of VHL patients will often detect small solid hemangioblastomas, which are asymptomatic. Surgical resection at this stage is associated with unnecessary risk of neurologic injury. However, progression from solid tumor to cyst with mural nodule has been described in the VHL population, with surgery required for decompression of mass effect related to the enlarging peritumoral cyst.

A study by Lonser et al. describes the development of peritumoral edema prior to the development of a peritumoral cyst. The mean time required for peritumoral edema to evolve into a cyst was 27 ± 19 months (range, 8–67 months) in the cerebellum and 47 ± 22 months (range, 9–72 months) in the spinal cord. Cases of sporadic (not VHL-associated) hemangioblastomas progressing from solid tumor to cyst with mural nodule requiring surgery have also been reported.

Peritumoral cysts develop as leakage of ultrafiltrate from the tumor into the surrounding normal brain parenchyma exceeds the parenchymal reabsorption rate. The resultant increase in interstitial pressure causes a cyst with a rim of gliosis to develop. This evolution is supported by reports of simple cyst drainage being insufficient for definitive management due to prompt return of the cyst and associated mass effect.

The proposed evolution of hemangioblastoma is outlined schematically in Fig. 16.4 with a case example in Fig. 16.5.

MIMICS AND DIFFERENTIAL DIAGNOSIS

Primary neoplasms such as pilocytic astrocytoma (Fig. 16.6) are more common in the pediatric and young adult population. Metastatic tumors (Figs. 16.7 and 16.8) should always be considered in the differential diagnosis of adult patients. When a cystic component is present in a pilocytic astrocytoma or metastasis, the wall typically enhances due to tumor involvement of the cyst wall. By contrast, the gliotic wall of the hemangioblastoma cyst does not enhance.

CONCLUSION

The notion that hemangioblastomas follow a specific pattern of progression suggests that anticipation of this progression is

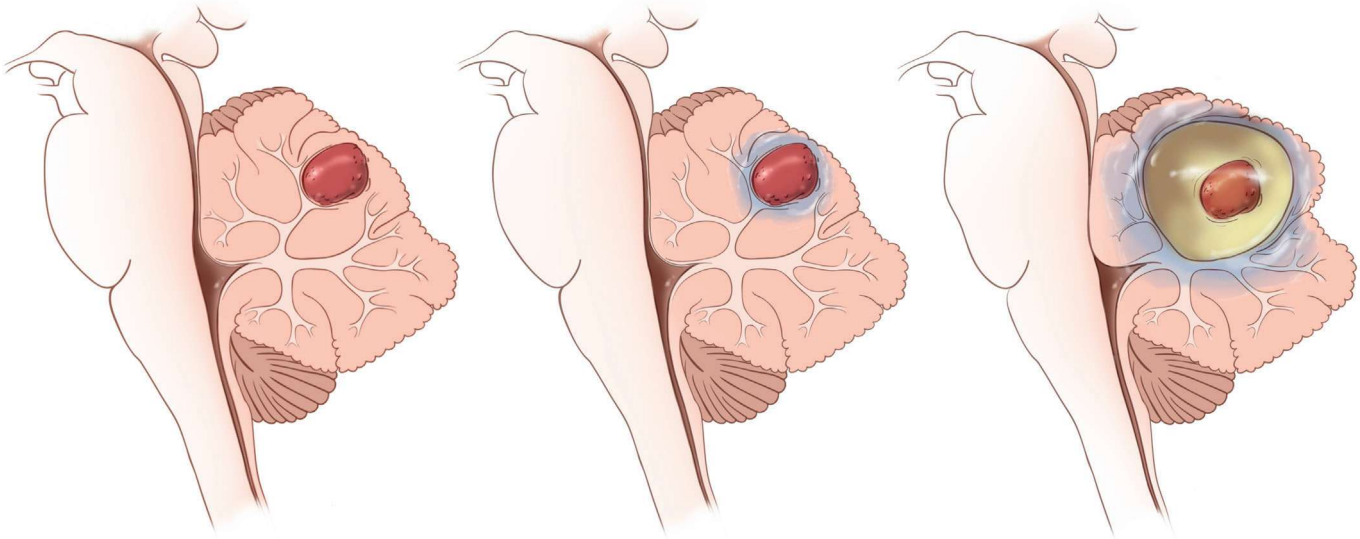

Figure 16.4. Evolution of hemangioblastoma. Stage 1 *(far left)*: solid enhancing tumor without peritumoral edema; stage 2 *(middle)*: solid enhancing tumor with peritumoral edema; stage 3 *(far right)*: solid tumor with peritumoral cyst.

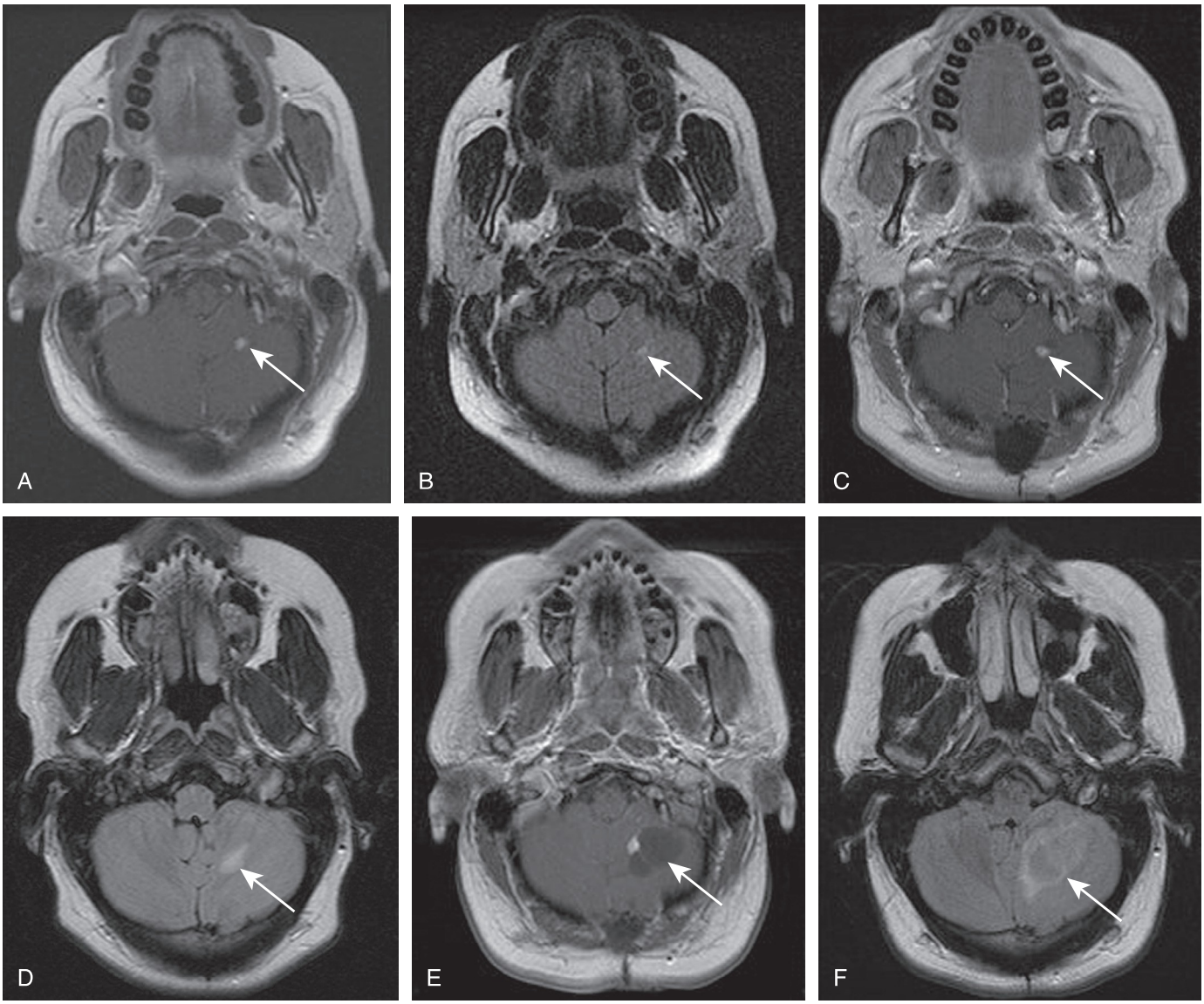

Figure 16.5. Evolution of hemangioblastoma. Axial T1 postcontrast (A) and fluid-attenuated inversion recovery (FLAIR)-weighted (B) images demonstrate an enhancing nodule (*arrow* in A) without associated edema (*arrow* in B). Axial T1 postcontrast (C) and FLAIR-weighted (D) images at 1 year follow-up demonstrate a stable enhancing nodule (*arrow* in C) with interval development of surrounding edema (*arrow* in D). Axial T1 postcontrast (E) and FLAIR-weighted (F) images at 3-year follow-up demonstrate a slight interval increase in the nodule size, interval development of a cyst (*arrow* in E), and increased surrounding edema (*arrow* in F).

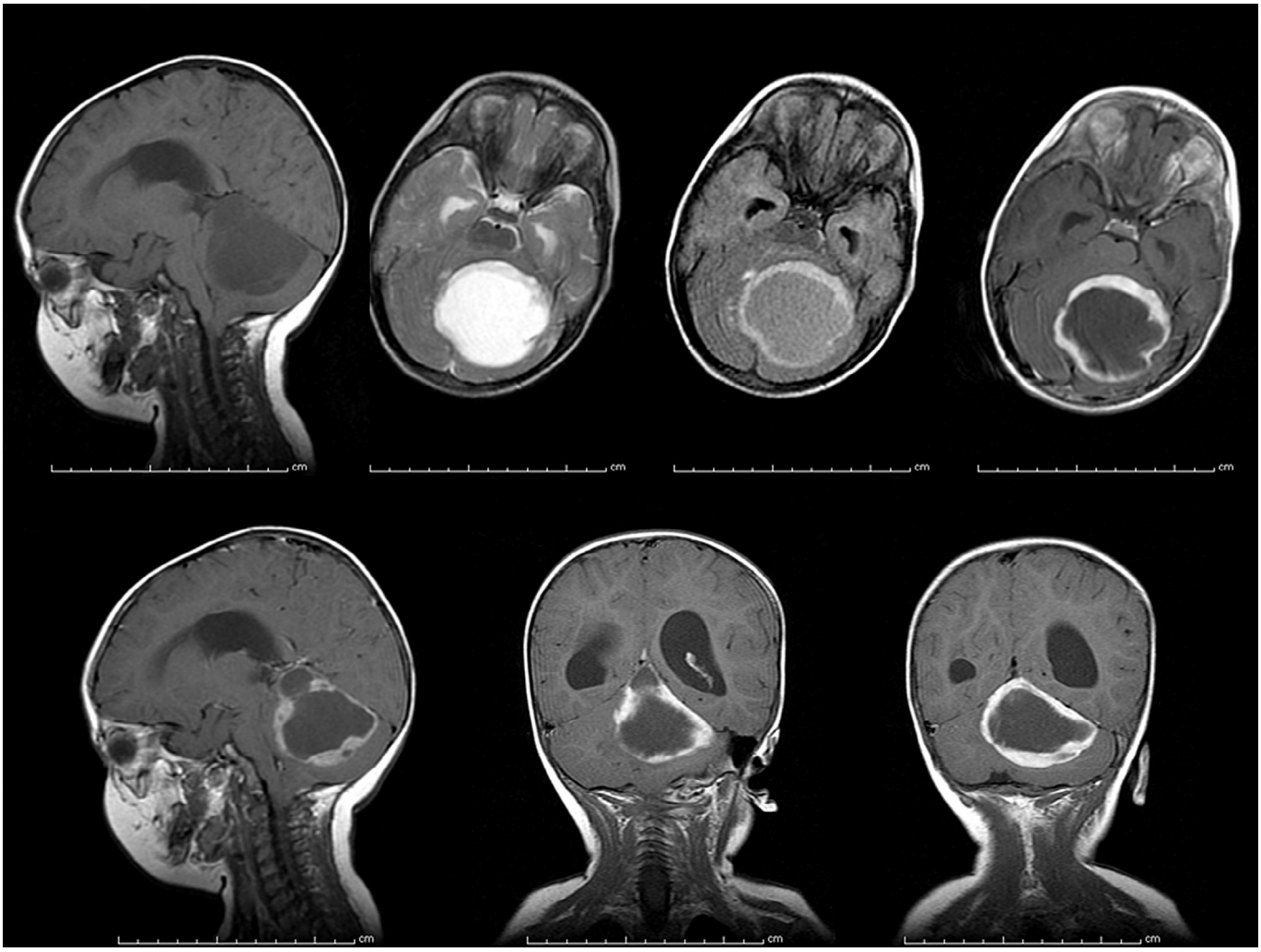

Figure 16.6. Pilocytic astrocytoma in a child. *Upper panel* (*left* to *right*): Sagittal T1, axial T2, axial fluid-attenuated inversion recovery, and postcontrast axial T1-weighted images. *Lower panel:* Postcontrast sagittal and coronal T1-weighted images. A T1 hypointense and T2 hyperintense peripherally enhancing cystic posterior fossa cerebellar mass lesion effaces the fourth ventricle, resulting in hydrocephalus.

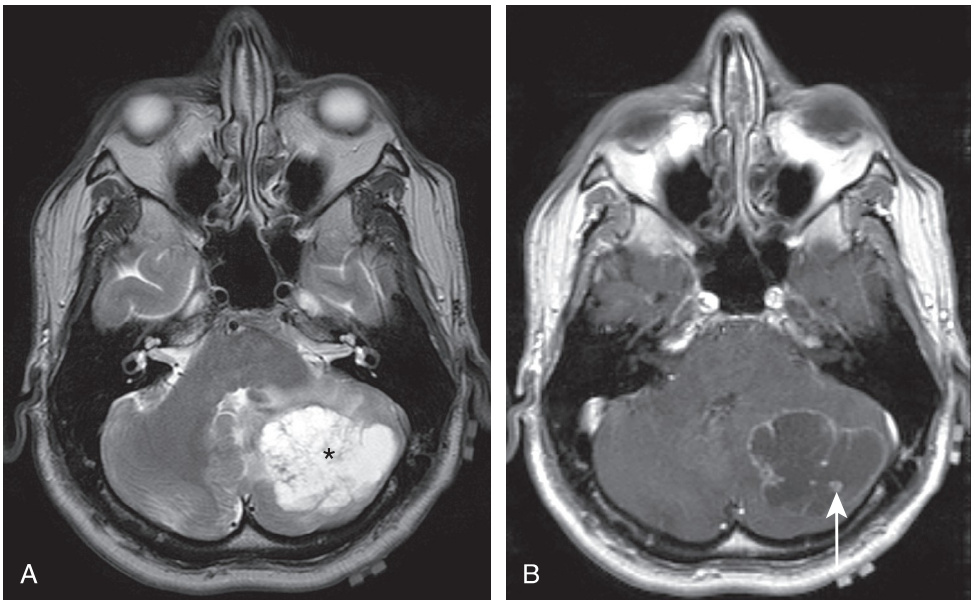

Figure 16.7. Necrotic metastatic tumor in a patient with a history of stage 1 non–small cell lung carcinoma treated 1 year prior. (A) Axial T2-weighted image demonstrates a cystic cerebellar lesion *(*)* with internal septations and perilesional edema. (B) Postcontrast axial T1-weighted image with contrast demonstrates a cystic cerebellar mass with predominantly peripheral smooth and nodular enhancement. There are also three internal enhancing nodules *(arrow)*.

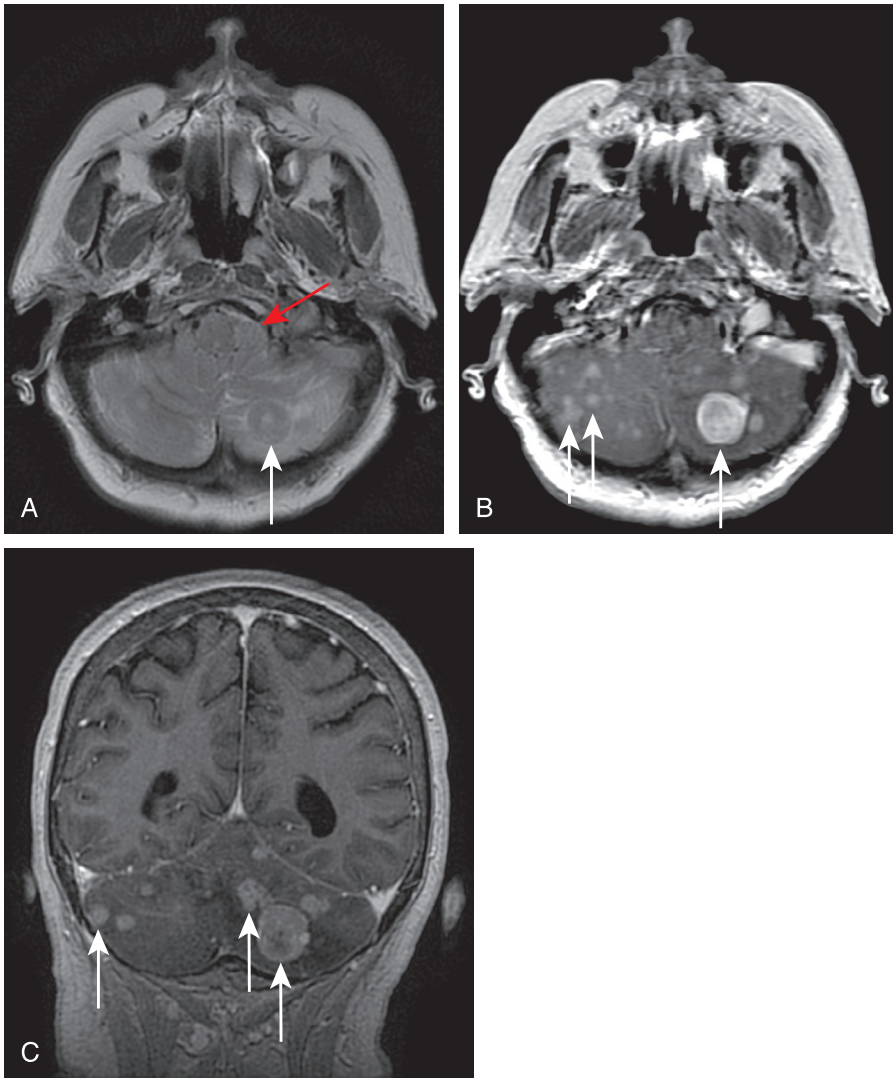

Figure 16.8. Multiple cerebellar metastases in a patient with breast carcinoma mimicking the appearance of the multiple type 3 hemangioblastomas seen in von Hippel-Lindau disease. (A) Axial T2-weighted image demonstrates a dominant isointense left cerebellar lesion *(white arrow)* with surrounding edema. Tonsilar herniation is also seen *(red arrow)*. (B) Postcontrast axial T1-weighted image demonstrates multiple enhancing tumors *(arrows)*. (C) Postcontrast coronal T1-weighted image demonstrates multiple enhancing tumors *(arrows)* exclusively involving the cerebellum.

important for the recommendation of follow-up imaging or timing of surgical resection.

The importance of this evolution in the cerebellum has been the focus of this discussion. However, spinal lesions often present at earlier stages (solid tumor stages types 3 and 4) because of the smaller space of the spinal column; they often require earlier intervention due to mass effect.

SUGGESTED READING

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