

CAVERNOUS ANGIOMA OF BRAIN: REVIEW OF 5 CASES

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Abstract:

There are four types of intracranial vascular malformations: AVMs, cavernous angiomas, capillary telangiectasias, and venous angiomas. Cerebral cavernous malformations (CCMs) are angiographically occult, mulberry like assembly of thin-walled vascular sinusoids lined by endothelium with no intervening brain parenchyma. CM (Cavernous Malformation) changes in size and number over time by progression, regression and denovoformation. Cavernous angiomas have a classic popcorn-like appearance on CT and MRI, indicating hemorrhage of multiple ages and calcification. They often have a classical hemosiderin ring (hypointense ring on T2-weighted images).

Here we have analysed 5 cases of cavernous malformation treated by surgery. All the patients had excellent operative outcome with no new neurological deficit.

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Introduction :

Cerebral cavernous malformations (CCMs) are a mulberrylike assembly of thin-walled vascular sinusoids lined by a thin endothelium lacking smooth muscle, elastin, and intervening parenchyma, surrounded by hemosiderin deposits and gliosis, which may or may not be thrombosed¹. Cavernous malformations can be found throughout the the brain and the brainstem in a volume distribution, and also the spinal cord, the cranial nerves, and the ventricles^{1,2}.

There are four types of intracranial vascular malformations: AVMs, cavernous angiomas, capillary telangiectasias, and venous angiomas^{1,2,3}. The incidence in the general population is roughly 0.5%, and clinical symptoms typically appear between 20 to 30 years of age. Once thought to be strictly congenital, these vascular lesions have been found to occur *de nov*)^{1,2,3}.

Most vascular malformations are present at birth (congenital) and are suspected to arise between three and eight weeks of gestation, but the specific defect in embryogenesis has not yet been identified for each type of malformation^{3,4}.

Cavernous malformations/angiomas are composed of cystic vascular sinusoids lined with a vascular endothelium monolayer and no intervening neural tissue^{3,4}. These are slow-flow lesions and hemorrhage at approximately 0.5% per year. Like AVMs they can present with either hemorrhage or seizure⁵. Approximately 70% of these lesions occur in the supratentorial region of the brain; the remaining 30% occur in the infratentorial region. the brainstem is the most common site of involvement in this compartment (9–35% of all cases⁵.

The incidence of CMs ranges from 0.4 to 0.9% of the general population³. They constitute 8 to 15% of all cerebrovascular malformations, and they occur in the supratentorial compartment in 63 to 90% of cases. Posterior fossa CMs represent 7.8 to 35.8% of all cases and the brainstem is the most common site of involvement in this compartment (9–35% of all cases.^{5,6}.

CM are angiogenically immature lesions with endothelial proliferation increased neoangiogenesis⁶. Cerebral cavernous malformations exhibit broad range of are dynamic behaviors, changing in size and number over time, progression, regression and denovoformation. They can range from 0.1 to 9 cm and usually reach a larger size in children than adults (who usually have cavernous malformations only 2-3 cm in size)⁶.

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Methods :

All the patients underwent clinical evaluation, routine and specific investigation mainly MRI of Brain with contrast was done. Proper craniotomy and upon the basis of neuroanatomy proper cortical incision were applied. Angioma was removed in pieces by sucker, microrounger under the illumination of high power microscope. Histopathological result were collected properly and postoperative care were monitored regularly.

Results :

In table -I results of all 5 cases were reviewed.

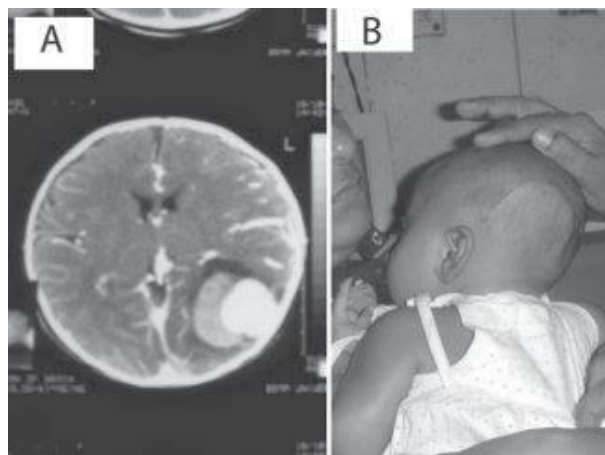


Fig.-1: (Case 1) *Partially thrombosed cavernous angioma in CT scan (A) and postop scar (B)*

Table-I

Summary of cases presenting symptoms, radiography findings operative outcomes.

Case no	Age/ Sex	Presentation	Location of angioma	Name of Operation	Complication	Follow-up
1.	2yrs /F	Headache, seizure	Parieto-occipital lobe	Craniotomy and total excision	Nil	3 yrs
2	28/M	Partial seizure	Left parietal lobe	Left Superior parietal lobule approach	Still occational partial seizure	3 yrs
3	32/M	Sudden severe headache, Rt sided weakness, leg>hand	Lt Fronto-parietal lobe	Left Superior Parietal lobule approach	Nil, Single attack of partial seizure in post op period.	3 months
4	18/M	Headache , Ataxia, visual blurring	Rt cerebello-tonsillar region	Midline suboccipital craniectomy	Nil	2 months
5	38/M	Sudden dysphasia, dysphonia, ataxia, quadreparesis H/o cerebral cavernoma surgery 8 years back	Lt Ponto medullary junction	Midline suboccipital craniectomy and cerebello-medullary fissure approach	Nil	2 months

Discussion :

Case –1: A 2 years old girl presented with excessive crying and occasional vomiting and irritability . Her visual problem was noted by her mother . Her CT revealed ruptured cavernous angioma with mass effect. She underwent craniotomy and gross total removal of the cavernous angioma without any further neurological deficit.

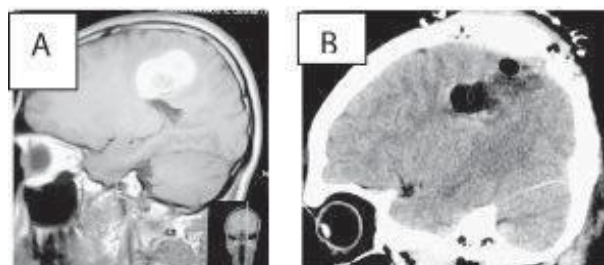


Fig. 2 (Case 2): Preoperative MRI(A) and immediate postoperative CT scan(B) of same patient showing no residual cavernous angioma.

Case –2: 28 years old man presented with focal motor seizure mainly in the right leg with occasional headache. He underwent MRI investigation and revealed cavernous angioma of left parietal lobe. Parietal craniotomy and superior parietal lobule approach done and total cavernous angioma was removed uneventfully.

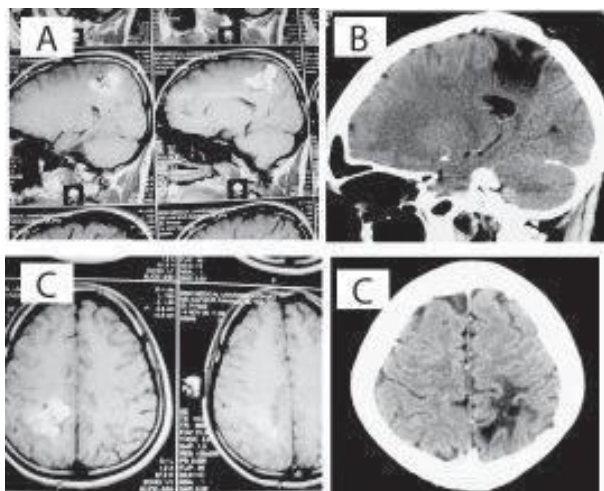


Fig. 3 (case 2): Preoperative MRI(A,C) show in typical popcorn appearance of cavernous angioma and delayed postoperative CT scan(B,D) of same patient showing no recurrence of cavernous angioma.

Case –3: 32 years old man presented with sudden severe headache, vomiting, and progressive right sided hemiparesis. Initially he was treated conservatively for 2 weeks. His motor weakness improved. He underwent parietal craniotomy and superior parietal lobule approach done and total cavernous angioma was removed uneventfully.

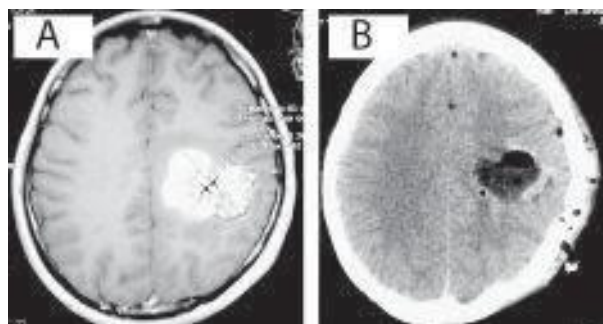


Fig. 4 (Case 3): Preoperative MRI(A) and immediate postoperative CT scan(B) of same patient showing no residual cavernous angioma.

Case –4: 18 years old boy presented with sudden headache, vomiting, ataxia and visual dimness . His CT scan revealed cerebellar ruptured cavernous angioma with fourth ventricular compression and moderate obstructive hydrocephalus. He underwent midline suboccipital craniectomy and with C1 laminectomy and gross total removal of angioma. His postoperative course was uneventful.

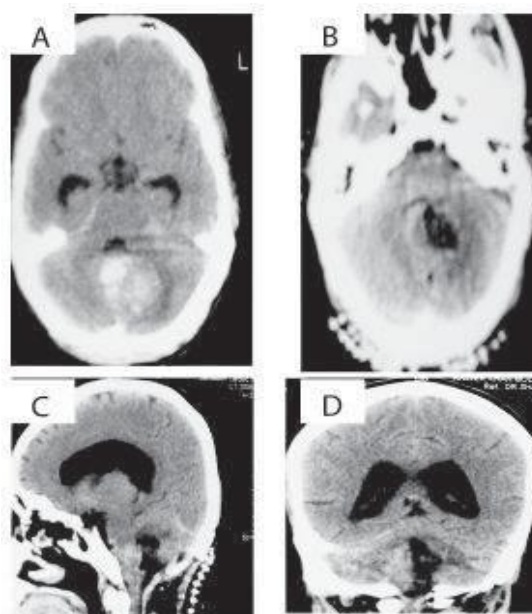


Fig.-5 (Case 4): Preoperative CT Scan(A,C) and immediate postoperative CT scan(B,D) of same patient showing no residual cavernous angioma.

Case –5: 38 years old man presented with sudden onset of dysphagia, dysphonia, dysarthria, and gross quadreplegia and respiratory distress. He underwent emergency tracheostomy to enhance trachio-bronchial toileting. He had a history of supratentorial craniotomy 10 years back where histopathology revealed cavernous angioma. His latest MRI revealed recent ruptured cavernous angioma of ponto-medullary region along with evidence of previous surgery and some discrete cavernous angioma in different location. He underwent midline suboccipital craniectomy and near total removal of cavernous angioma and his postoperative course was uneventful. His motor power in all limbs improved and tracheostomy wound closed. His dysphagia still persist.

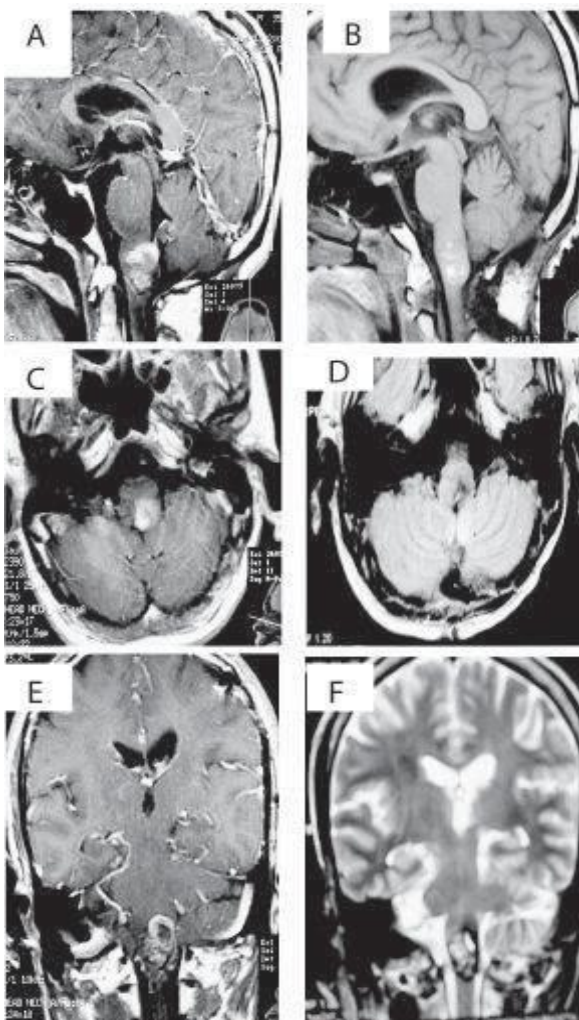


Fig. 6 (Case 5): Preoperative MRI(A,C,E) and early postoperative CT scan(B,D,F) of same patient showing some residual cavernous angioma in Medulla oblongata.

Magnetic resonance imaging:

Cerebral cavernous malformations are characterized by small, nonsymptomatic hemorrhages typically confined to the location of the lesion, only occasionally resulting in clinically significant hemorrhaging⁶. Hemoglobin degradation products such as methemoglobin, hemosiderin, and ferritin present at the site of the lesion alter the local magnetic environment allowing for magnetic resonance imaging (MRI) detection^{6,7}.

Cavernous angiomas have a classic popcorn-like appearance on CT and MRI, indicating hemorrhage of multiple ages and calcification. They often have a classical hemosiderin ring (hypointense ring on T2-weighted images). (Osborn). They show minimal to no enhancement on contrast CT or MRI and are not detectable by angiography⁷.

Best imaging tool is MRI T2 sequence, standard T1, T2WI may be negative in Type 4 lesions⁷.

DSA usually normal hence known as Angiographically occult vascular malformation. (osborn)⁸.

AVMs are direct artery to vein fistulae that hemorrhage at a rate of 4% per year. They usually have tortuous feeding arteries, a dense nidus, and large draining veins that may be seen on CT. AVMs may have associated feeding artery aneurysm secondary to the high flow state. AVMs most commonly present as an ICH or seizure and less commonly as focal neurologic deficit from vascular steal or mass effect⁸.

Approximately 70% of these lesions occur in the supratentorial region of the brain; the remaining 30% occur in the infratentorial region. Cavernous angiomas can also occur in the spinal cord⁸.

In up to 30% there is a coincidence of CCM with a venous angioma, also known as a developmental venous anomaly (DVA). These lesions appear either as enhancing linear blood vessels or caput medusae, a radial orientation of small vessels that resemble the hair of Medusa from Greek mythology⁹.

Causes : Cerebral cavernous malformations can be present at birth (congenital) or develop after birth (acquired). Most CCMs are caused by a mutation in one of three particular genes¹. The gene mutation may occur for the first time in the affected individual (sporadic) or be inherited from a parent (familial). Acquired CCMs can be caused by an injury to the brain or spinal cord^{1,9}.

Risk factors for developing clinically significant hemorrhage

The most widely cited risk factor for clinically significant hemorrhage. Rao et al¹⁰ found a more dramatic increase risk of haemorrhage (0.39% to 23% per annum). Another important risk factor is found in young women wishing to become pregnant. The hormonal state of pregnant women is such that endothelial cell proliferation may increase the risk for hemorrhage substantially¹. Other controversial risk factors include age and location¹⁰.

Clinical presentation:

The clinical presentation of these lesions is highly variable, ranging from incidental finding at neuroimaging to discovery in autopsy after fatal hemorrhage. The most common symptom of cavernous malformation is seizure followed by focal neurological deficits, acute hemorrhage, and headache¹⁰. The onset of symptoms occurs most commonly in the third and fifth decade of life but can occur at any point in life, from children to the very elderly. All seizure types, including simple seizures, complex partial, and generalized seizures, have been known to present in patients with supratentorial CCMs^{1,10}. The pathogenesis of seizure is related to the presence of iron products after red blood cell breakdown secondary to multiple micro-hemorrhages¹⁰.

Related disorder :

- Arteriovenous Malformations of the Brain
- Moyamoya Disease
- Blue Rubber Bleb Nevus
- Von Hippel-Lindau Disease¹⁰

Staging , Grading or classification criteria : Zabramski classification of Cavernous angioma:

Type 1= Subacute haemorrhage (hyperintense on T1WI, hyper or hypointense on T2WI.)

Type 2=Mixed signal intensity on T1. T2WI with degrading haemorrhage of various ages(classic popcorn ball lesion)

Type 3=Chronic haemorrhage (hypo to iso on T1-T2WI.)

Type 4=Punctate microhaemorrhage (black dots)⁴.

Role of Surgery :

The removal of CCMs in adults is usually fairly straightforward. Hemorrhage can occur, but blood flows slowly in a CCM so the risks of surgery are not great. During surgery, the lesion is removed and the bleeding around it is coagulated. Removal of a CCM in the

brainstem can sometimes prove to be more complicated than in other areas of the brain (i.e. cerebellum)¹⁰.

Role of Radiosurgery: It is suggested that surgical excision provides immediate protection from the risks of recurrent haemorrhage, establishes a tissue diagnosis, allows complete removal at the primary intervention, avoids complications of radiation-induced damage and is performed more easily in these vascular anomalies due to the presence of a capsule with surrounding gliotic tissue¹⁰.

Conclusion:

The management of patients with cavernous angioma continues to evolve. Our current recommendations for management are as surgery to the patients who are symptomatic with acute severe or progressive neurological deficits, seizure and a single hemorrhage in the cerebrum, cerebellum. When the hemorrhage is in the brainstem, thalamus, or basal ganglia, operation have to be done in subacute stage.

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