AN INTERESTING CASE OF BRAINSTEM CAVERNOMA

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Abstract:
A 45 yr old post menopausal lady admitted with insidious onset of focal neurological deficit in the form of right hemiparesis, right hemi hypoesthesia and failure of conjugate gaze to left side was diagnosed to have a pontine cavernoma with sub acute bleed. Surgical treatment was planned since the lesion reached a surgical surface and was causing significant neurological morbidity and was referred to higher centre for further management.

Keyword: Cavernoma, Magnetic Resonance Imaging, Brainstem, Haemorrhages

INTRODUCTION:
Cavernoma is a vascular hamartoma involving blood vessels of the central nervous system. It contributes to 5-10% of all cerebrovascular malformations. Cavernoma is also called as cerebral cavernous malformation, cavernous angioma or cavernous hemangioma. Although once considered as rare entity, with the development of the modern neuroimaging techniques especially MRI, understanding of these lesions were revolutionised as more number of cases were reported. The observation of large number of familial clustering of cases have stimulated research into genetic basis of the disease. Cavernous angiomas are typically <1 cm in diameter and are often associated with a venous anomaly. They may present in various ways and understanding the natural history of the lesion is important in deciding the medical or surgical treatment strategy. Our case is a middle aged lady with symptomatic infratentorial cavernoma who was planned for surgical treatment.

CASE REPORT:
A 45 yr old post menopausal lady came to the outpatient clinic with complaints of weakness of right side of body, diminished sensation of right side of body and double vision of 4 months duration. The weakness was insidious onset, spastic type and slowly improved over the period. It was associated with decreased sensation over the right half of the body in the form of difficulty in perceiving feeling of clothes and hot and cold water sensation. Diplopia was noticed 2 wks after these symptoms and was present when both eyes were open. Patient also had diffuse headache. It was not associated with any seizure.
loss of consciousness or head injury. There was no history suggestive of other cranial nerve involvement, no history of swaying while walking, no history of thinning of limbs or muscle twitching. No history of cough with expectoration or low grade fever or weight loss was present. No history of waxing and waning of symptoms were present. There was neither any history of any medical co-morbidities like hypertension, diabetes mellitus, tuberculosis, coronary artery disease nor history of similar illness. No family history of CVA could be elicited.

On examination at the time of admission patient was conscious oriented. There was no pallor, icterus, clubbing, cyanosis, generalised lymphadenopathy or edema. There was no neurocutaneous markers, no marker of TB or syphilis or atherosclerosis. Vital signs were within normal limits and stable. On neurological examination higher mental function was normal. There was failure of conjugate gaze to the left side. Also there was pyramidal type of weakness with grade 4 power and hypertonia on right upper limb & lower limb along with exaggerated reflexes with hemi hypoesthesia of the right side of body. Fundus was normal on both sides. Other systems examinations were within normal limits. Laboratory investigation showed a complete blood count of Hb-11.8g%, TC-8800cells/mm3, DC-P60% L38% E2%, RBC-3.4 million, Platelet: 1.9 lakh/mm3, PCV-30% and ESR- 30 mm/1st hr. Patient's blood sugar, renal function tests and liver function tests were normal. CT brain was done and showed a well circumscribed hyperdense lesion in the brainstem on the left side with heterogeneities and we proceeded with MRI brain with angiogram and venogram which showed evidence of focal hematoma with various stages of breakdown products with heterogenous signal intensity in the pons and brainstem predominantly on the left side measuring about 3.1*2.2*2.7cm suggestive of subacute hemorrhage within the left side of pons and brainstem with mass effect probably cavernoma. Thus a final diagnosis of brainstem cavernoma with subacute hemorrhage was made. Patient was treated with prophylactic antiepileptics, physiotherapy. Neurosurgeon opinion was sought and advised surgery. Patient was subsequently referred to Madras medical college, Chennai for surgical management. This case was presented 1) Due to the rarity of lesion. 2) Since this patient with brainstem cavernoma with hemorrhage presented with stable vitals.
multilobulated rounded areas of increased signal intensity

Figure 3: showing T2 image of cavernoma with central mixed intensity surrounded by peripheral hypointensity

Figure 4: Gradient-echo axial MRI showing cavernoma

Figure 5: MR Angiogram showing normal study

Figure 6: T2 image with central mixed intensity surrounded by peripheral hypointensity

Figure 7: T2 Picture showing cavernoma
DISCUSSION: Vascular malformations in brain have been divided into four types- AV malformations, cavernous malformations, capillary telangiectasia and venous angioma. Cavernomas are focal vascular abnormalities of blood vessels supplying brain although rarely can occur in spinal cord as intramedullary lesion. It contributes to 5-10% of all vascular anomalies. With introduction of MRI the incidence of cavernoma is increasing. They are low flow vascular anomalies consisting of grossly dilated ectatic blood vessel with single layer of endothelium that displaces normal neurological tissue. The wall of blood vessel does not have any smooth muscle or elastic tissue support which account for the slow perilesional or intralesional oozing. There is no intervening brain parenchymal tissue within the collagenous stroma that separate the vascular channel which is considered the histological hallmark of cavernoma. The blood vessels are filled with slow moving or stagnant blood that is usually clear or in a state of decomposition. Epidemiology: cavernoma occur at a frequency of 0.4-0.9% and contribute to 8-15% of all cerebral vascular anomalies. They can be of two types: spontaneous and familial. Familial cases express in autosomal dominant pattern and are related to three genes. KRIT1 located on Chromosome 7 and CCM2 located on chromosome 7 are genes involved in angiogenesis while PDCD 10 located in 3rd chromosome is an anti apoptotic gene. Hormonal factors, prior radiation exposure are the various hypotheses behind spontaneous cavernomas. Cavernomas occurs commonly in 2nd to 5th decade and shows no sex predilection although male patients present earlier in life while females have more predisposition for hemorrhage. Two third of total cases occur in supratentorial compartment while one third is in infratentorial region. Frontal and Temporal lobe are the commonest site for supratentorial cavernoma while Pons and Cerebellum are the commonest infratentorial sites. Most of the lesions are solitary. But of the multiple lesions two third are familial type. Clinical features: Many lesions are clinically silent and are incidental finding on MRI. Seizures are the most common form of presentation and are due to the deposition of iron in hemosiderin produced by the haemorrhages. Another major symptom is sudden onset of focal neurological deficit and is more seen in brainstem lesions. Usually symptoms are maximal at onset and improve as the hemorrhage is organised and absorbed although recurrent episodes of hemorrhage may lead to progressively worsening neurological deficits and permanent impairment. Hemorrhages contribute to 10-30% of symptomatic cases. It is characterised by more subcortical location, smaller hematoma, and insidious onset of symptoms unlike hypertensive hemorrhage. There is a female predilection for hemorrhage and aggravation during pregnancy supposed to be due to the growth of lesions under hormonal influence. Patient also can have transient neurological deficits due to subclinical bleeds. The risk of bleeding from cavernoma is around 0.7-1.5% per year although after one episode of hemorrhage it increases for a short period and comes around 5%. Familial cases have relatively more aggressive since incidence of hemorrhage or formation of newer lesions are higher. Differential diagnosis for cavernoma include primary hemorrhagic or metastatic tumours, infectious or granulomatous diseases, cryptic or partially thrombosed AV malformations and multiple
sclerosis Investigations: CT brain: CT findings are non specific. It commonly appears either hyperdense lesion or isodense lesion. Faint contrast enhancement may be there. Calcifications are common. MRI Brain: is the best modality to diagnose cavernoma and T2 images are more sensitive than T1. It shows characteristic T2 picture with central core of mixed signal intensity surrounded by a rim of decreased intensity produced by hemosiderin which indicates old hemorrhages from leaking vessels. This characteristic picture of hemorrhage in various stages of evolution is produced by the leakage of blood from the thin walled weak vessels. An improvised T2 imaging called as Gradient Echo increases the sensitivity of MRI in detecting cavernoma and is the technique of choice and is particularly useful in cases of multiple lesions and small lesions. In T1 images it is seen as multilobulated rounded areas of increased signal intensity and T1 images helps in more anatomical resolution and help for planning surgery. The more the surrounding rim of hypointensity more active the lesion. Angiogram: usually does not reveal the lesion and hence they are called as angiographically obscure malformations (AOM). Treatment: is decided on the basis of site of lesion and symptomatology of the patient and age of the patient. In asymptomatic patients with accidental detection of cavernoma conservative treatment with observation and periodic MRI is preferred. In such patients if lesion enlarges in repeat MRI or symptom arises, surgery is advised. If seizures are the presenting symptom and seizures are well controlled with no overt hemorrhage medical treatment can be continued. In case of refractory seizures lesionectomy will lead to control of seizures. But Surgical removal is very likely to result in a cure and eventual cessation of the need for antiepileptic medication hence surgery is considered for well accessible lesions. In clinically overt hemorrhage involving supratentorial & cerebellar cavernoma surgical excision is preferred. In case of brainstem cavernoma with clinically significant hemorrhage once they become symptomatic, almost always from the mass effect of a new hemorrhage, they tend to progress with repeated “intralational” hemorrhages and thus progressive growth of the mass occur and hence surgery is advised once the lesion reaches a surgical surface(e.g. floor of the fourth ventricle, lateral surface of the Pons or Medulla, anterolateral surface of the Pons or mesencephalon, or quadrigeminal plate). In deeper lesions that does not come to surface surgery is done only when if repeated hemorrhage cause significant morbidity. Radiosurgery appears to be ineffective for these lesions, although some uncontrolled series have suggested a slight improvement in the natural history of future hemorrhage. Currently most neurosurgeons do not recommend this form of treatment for angiographically occult malformations.

CONCLUSION:
Neurological deficits of insidious onset should arise suspicion of space occupying lesions. Conventional angiogram has little role in diagnosis of cavernoma and MRI Brain has revolutionised the understanding of cavernoma.

ABBREVIATIONS: MRI- Magnetic resonance imaging, CT scan- Computed tomographic scan, CVA-Cerebro vascular accident, TB- Tuberculosis, TC- total WBC count, DC-Differential count, PCV-Packed cell volume, ESR-Erythrocyte sedimentation rate, AV Malformation-Arteriovenous malformation

Explanations for Reviewers Suggestions: 1 Facial nerve was not involved in our case. Although the lesion was in pons, it was not involving the facial nerve nucleus.
was discussed with professor of Radiology and according to him- The lesion was an expansive hematoma which displaced the surrounding structures rather than destroying it and hence facial nerve nuclei was not involved. 2 The total MRI pictures, nine in number have been submitted as image proof for cavernoma. 3 1- YOUMAN’S Neurological Surgery- 6th Edition 12 “MRI characteristics of cavernous malformations are sufficiently unique enough to allow diagnosis of majority of the lesions on the basis of MRI findings alone” 2- Cavernous malformations Of Brain and spinal cord by Giuseppe Lanzino, Robert F Spetzler 2 “The introduction of MRI allowed cavernous malformations to be diagnosed without the need of pathological confirmation” 3- Magnetic Resonance Imaging Of Brain And Spinal cord BY Scott. W. Alas 13 “MRI features of cavernous malformations are characteristic and considered diagnostic of these lesions” According to above cited references MRI itself can diagnose Cavernoma without need of Pathological confirmation 2,12, 13. The patient was referred to higher centre (Madras Medical College, Chennai) for surgical management and has lost follow up.

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