CASE REPORT

Third Ventricular Cavernous Angioma

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Summary

Third ventricular cavernous angiomas are rare vascular malformations of the brain. We report an eight-year old boy with a rare third ventricular cavernous angioma that hemorrhaged presenting with symptoms of acute hydrocephalus. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) showed a heterogenous ill-defined, solid and cystic intraventricular mass in the third ventricle which was mildly enhanced with contrast and there was associated hydrocephalus. The mass was removed with success and follow up after two years revealed no neurological abnormalities.

Key Words: Cavernous angioma, Paediatric, Third ventricular tumor

Introduction

Cavernous angiomas (CA) or cavernoma of the central nervous system are vascular malformations that usually present in the cerebral hemispheres. Though considered rare before the common use of computerised tomography and magnetic resonance imaging, more cases of CA in the cerebrum, cerebellum,brain stem and spinal cord are being diagnosed in recent years in adults and less commonly in children[1,2]. However, they occur very rarely in the ventricles. Still rarer is CA involving both the third and the lateral ventricles[2]. The present case report describes a case of intraventricular mass with obstructive hydrocephalus which turned out to be a third ventricular cavernous angioma projecting into both lateral ventricles.

Case Report

An eight-year old Malay boy was admitted with reduced conscious level and headache for one week prior to admission. Prior to that, he had a history of high grade fever for two weeks followed by headache and vomiting, one week later. He had no history of recent seizures, blurring of vision or recent upper respiratory tract infections. He also had no cerebellar symptoms such as gait ataxia, frequent falls from disequilibrium or tremor. There were no symptoms to suggest endocrinopathy such as polyuria, delayed developmental milestones or growth retardation. On examination, he was stuporous with Glasgow Coma Scale (GCS) of 10/15. Neurological examination revealed a left lateral rectus palsy, papilloedema and left sided hypertonia with bilateral upgoing plantars. Non-contrasted CT showed an intraventricular tumour in the region of third ventricle with obstructive hydrocephalus associated with intratumoural bleed (Fig.1).

An urgent bilateral external ventricular drainage procedure was performed. MRI done five days later, revealed a multilobulated heterogenous midline mass with both solid and cystic components, in the region of the third ventricle. The mass was ill-defined, irregular superiorly and projected into both the lateral ventricles. Intratumoural bleed was confirmed. There was no calcification. Following intracavernous gadolinium,

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there was minimal enhancement. The adjacent structures such as optic chiasma, pituitary gland, suprasellar cistern and cavernous sinus were not involved. There was a huge extradural haematoma seen over both frontal regions (Fig.2) which was secondary to the post-intraventricular drainage procedure. Magnetic Resonance Angiography did not show any vascular abnormalities.

A semi-urgent bifrontal craniotomy, evacuation of the extradural haematoma and total tumour excision was performed via the transcorpus-callosal approach to the third ventricle. A yellowish 'mullberry' tumour was seen arising in the third ventricle at the junction of the Foramen of Monro. The tumour consisted of both a solid vesicular mass and cystic lesion containing altered blood. Post-operatively, he was extubated and subsequently discharged two weeks later with GCS of 15 without any neurological deficit. Repeat MRI one month later showed no evidence of residual tumour or recurrence. On follow up, two years later, he was doing well academically and functionally, without any neurological impairment.

Histopathological examination of the lesion revealed closely approximated thin and thick-walled vascular channels of varying sizes with scanty interposed brain parenchyma between the channels. The wall of the blood vessels were collagenized and devoid of muscle fibres or elastic laminae. The lumina of the blood vessels showed red blood cells and an occasional organized thrombus. The fragment of adjacent white matter showed gliosis with scattered haemosiderin laden macrophages and a few congested thin-walled blood vessels. The findings were consistent with cavernous angioma (Fig. 3).

Discussion

Cavernous angiomas of the central nervous system make up 8-15% of cerebrovascular malformations in postmortem studies. CA are outnumbered only by arteriovenous malformations in surgical pathology specimens[2,3]. Intraventricular cavernous angiomas (IVCA) constitute 2.5 to 10.8% of the cases of intracranial CA. The majority of the cases of IVCA occur in the average age group of 33.7 years and most of the cases occur in the third ventricle (44%). Paediatric cases of IVCA vary from 20% to 28.5% of all the cases of IVCA. It is very rare to have the involvement of both the third and the lateral ventricles

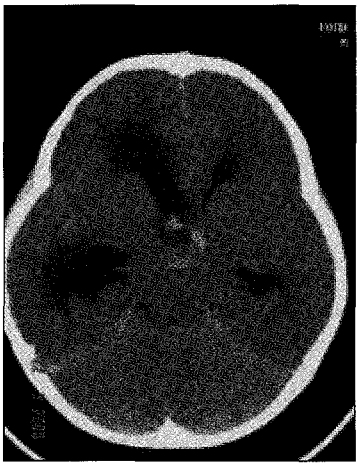

Fig. 1: Non-contrasted CT showed an intraventricular tumour at the region of third ventricle with obstructive hydrocephalus associated with intratumoural bleed.

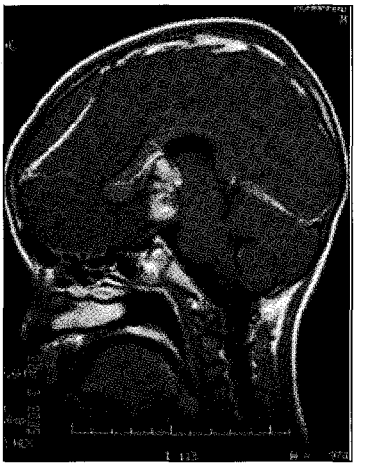

Fig. 2: Sagittal view of T1WI with gadolinium showed a multilobulated heterogenous midline mass with both solid and cystic components with minimal enhancement following intracavernous gadolinium. At the region of the third ventricle the tumour was projecting into the lateral ventricle. There was also an extradural haematoma over the frontal region.

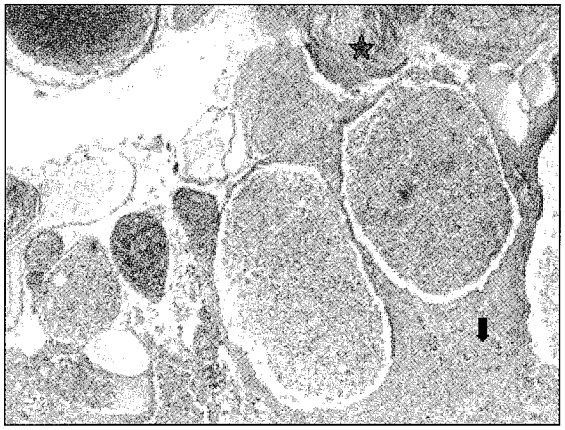

Fig. 3: Several closely packed blood vessels having collagenised walls devoid of muscle or elastic tissue are separated by fibrous tissue. The blood vessels are dilated and contain red blood cells. Two blood vessels show organized thrombui (red star). Haemosiderin-laden macrophages (black arrow) are also seen in the adjacent gliotic brain tissue. Elastic Van Gieson stain x 25.

as seen in our case[2]. IVCA present with symptoms of acute haemorrhage, seizures or progressive neurological deficit[2,3]. In this case report, headache and altered conscious level were due to the development of acute hydrocephalus secondary to the obstruction of the foramen of Monro by the IVCA, typical of any third ventricular mass lesion.

The important issue in IVCA is their potential to bleed. Angiographically, IVCA are occult with poor connections with the vascular system, and rarely show abnormalities other than a non-specific avascular mass, whereas others have described a blush supplied by an enlarged choroidal artery[2]. Though according to some, bleeding from IVCA is rare, others find that evidence of bleeding in IVCA is not uncommon[3]. Intratumoural bleed was confirmed in the present case. Rapid growth and mass effect of IVCA was attributed to repeated intralesional haemorrhage in surgical and autopsy cases[3]. The characteristic appearance of cavernomas on MRI of a central area of hypersignal correlated to the presence of methaemoglobin, associated with areas of hyposignal caused by calcifications and fibrosis within the lesion in T1- and T2-weighted images. Some authors noticed that contrast enhancement after gadolinium ranges from strong to moderate to none[2]. The angiographic appearance resembling caput medusae, a myriad of small tapering veins arranged in a radial fashion in the deep white matter and converging into a relatively large draining vein, classical of venous haemangioma was not observed in this case. Magnetic Resonance Imaging did not show any vascular abnormality. The solid and cystic components seen in the intraventricular mass enhanced minimally following intravenous gadolinium. The CT and the MRI findings in this case are probably due to recent haemorrhage. The cystic component was noted to contain altered blood intraoperatively. Some cases of IVCA have been thought preoperatively to be gliomas, meningiomas, teratomas, craniopharyngiomas and germinomas and preoperative streotactic biopsies have been done successfully to avoid erroneous diagnosis and mismanagement despite the danger of haemorrhage[2].

Histological examination of CA reveal thin and thick-walled veins closely approximated without the interposition of brain parenchyma. The wall of the blood vessels are hyalinised without any smooth muscle and elastic laminae. In contrast, in venous angioma and arteriovenous malformation, the blood vessels are separated by brain tissue and show smooth muscle in the walls[1,4]. The number or the size of the surrounding arteries are also not increased. A familial prevalence of CA has been described in some cases[1]. No history of familial involvement was obtained in this case.

Currently, the management of asymptomatic CA is conservative. Surgery is recommended if the lesion is safely assesible, is symptomatic either by mass effect and/or haemorrhage or seizure or shows evidence of having bled in the past. Patients who presented with haemorrhagic episode is dealt with in the same manner as any intracerebral haematoma of whatever cause. CA is not uncommon in the cerebral hemispheres and other parts of the brain, and are commonly asymptomatic. Although the more common presentations such as headache, seizure or the rare intracranial haemorrhage have been described, it is essential to look for another lesion which might produce these symptoms before attributing it to this benign pathology.

This case report has illustrated the rare location of IVCA in the third ventricle in a child and proves that if huge enough it can produce symptoms typical of any third ventricular mass lesion. Total surgical excision without any neurological deficit was achieved.

References

1. Scot RM, Barnes P, Kupsky W, Adelman LS. Cavernous angiomas of the central nervous system in children. J Neurosurg 1992; 76: 38-46.
2. Reyns N, Assaker R, Louis E. Intraventricular cavernomas: Three cases and review of the literature. Neurosurgery 1999; 44: 648-55.
3. Katayama Y, Tsubokawa T, Maeda T, Yamamoto T. Surgical management of cavernous malformations of the third ventricles. J Neurosurg 1994; 80: 64-72.
4. Burger PC, Scheithauer. Tumors of the central nervous system; Atlas of tumour pathology : AFIP, Washington DC, 1994; 287-92.