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Case Report

Cavernoma of the Right Lateral Ventricle: A Rare Case Report

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Abstract

Intraventricular cavernoma (IVC) is a rare pathological entity constituting 2.5%–10.8% of cerebral cavernomas. The lateral ventricles are the most frequent site, followed by the third and fourth ventricles. IVCs usually attain a large size compared to parenchymal cavernomas and cause signs and symptoms mainly due to mass effect. IVCs lack specific clinical manifestations and radiological features. Microsurgical excision of IVCs is a safe and effective treatment option. We present a 71-year-old male patient with right lateral ventricle cavernous angioma. The patient underwent microsurgical resection of the vascular lesion with good neurological outcome.

Keywords: *Cavernoma, cavernous angioma, intraventricular, lateral ventricle*

Introduction

Cavernous angiomas are vascular malformations that may occur in any part of the central nervous system (CNS). Cavernomas constitute 10%–15% of vascular malformations of the CNS.[1] They most commonly occur between the 20 and 50 years of age group. They occur in equal frequency in both males and females.[2] The involvement of the ventricular system by cavernomas is a rare entity. The incidence of intraventricular cavernomas (IVCs) is 2.5%–10.8% of cerebral cavernomas.[3] The first report of an IVC was given by Finkelnburg in 1906.[4] The lateral ventricles are the most frequent site, followed by the third and fourth ventricles.[3] Cavernomas have a tendency for frequent intralesional hemorrhage that leads to hemosiderin deposition and reactive gliosis in the surrounding parenchymal tissues. Cerebral cavernomas usually present with seizures. IVCs present with features of raised intracranial tension due to hydrocephalus may present as an acute condition due to intraventricular hemorrhage (IVH). Magnetic resonance imaging (MRI) brain is the diagnostic modality of choice for cavernomas of CNS. Cavernoma appears as heterogeneous lesions with mixed-signal intensity in the center of the lesion in both T1- and T2-weighted images. Hyperintense signal in MRI corresponds to hemoglobin degradation products, namely methemoglobin, whereas gliosis gives hypointense signal. Peripheral rim of hypointensity is due to the presence of hemosiderin, which has a paramagnetic effect. Computed tomography (CT) angiography is not helpful in the diagnosis of cavernoma. Cavernoma is angiographically occult vascular lesion since there is no feeding vessel to the lesion.[5] Since very few cases have been discussed in the literature, we would like to report the present case. A brief review of pathogenesis, clinical manifestations, radiological features, and treatment was also discussed.

Case Report

A 71-year-old male patient presented with complaints of altered sensorium, vomiting since 1 day. He was having a history of on and off headache since 3 months. On examination, the patient was drowsy and arousable. Neurological examination showed bilateral papilledema. MRI brain of the patient revealed a well-defined heterogeneously hyperintense lesion on T2/fluid-attenuated inversion recovery images measuring 3.5 cm × 2.8 cm × 2.9 cm in the right lateral ventricle extending up to foramen of Monro. Areas of hemorrhage were seen on T1/susceptibility weighted images with poor contrast enhancement. The lesion is causing compression of foramen of Monro and causing mild obstructive

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hydrocephalus [Figure 1a and b]. The radiological differential diagnosis given was central neurocytoma and subependymal giant cell astrocytoma. The lesion was excised through interhemispheric transcallosal approach [Figure 2]. The patient was positioned in supine position with head slightly flexed. A horseshoe-shaped incision was made in the right frontoparietal region extending to midline. A parasagittal craniotomy was done by placing burr holes on either side of the superior sagittal sinus. Durotomy was done, and flap reflected medially up to the superior sagittal sinus. Through the interhemispheric transcallosal approach, the lesion was excised. The lesion was extending up to foramen of Monro. A ventricular drain was placed in view of the risk of postoperative hydrocephalus. There was mild IVH [Figure 3] in the immediate postoperative scan, which was managed with external ventricular drainage for 48 h. Follow-up scans at 6 weeks showed resolution of hydrocephalus [Figure 4]. The histopathological examination revealed the lesion as cavernoma [Figure 5].

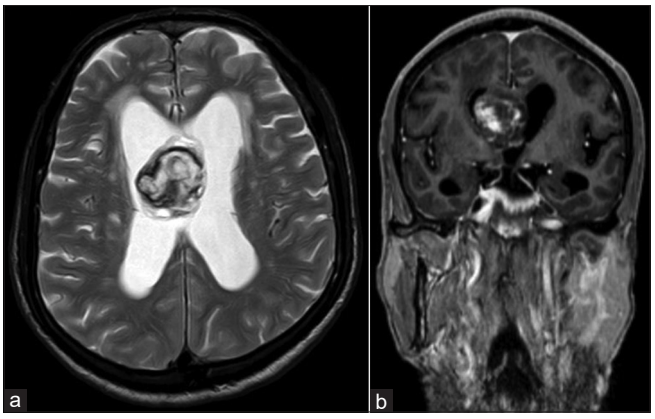

Figure 1: (a) Magnetic resonance imaging of brain axial section T2-weighted images showing a right ventricular mass of 3.5 cm × 2.8 cm. The lesion is well-defined, heterogeneously hyperintense with poor contrast enhancement. (b) Coronal T1-weighted magnetic resonance imaging showing areas of hemorrhage and obstruction of both foramina of Monro, causing biventricular hydrocephalus

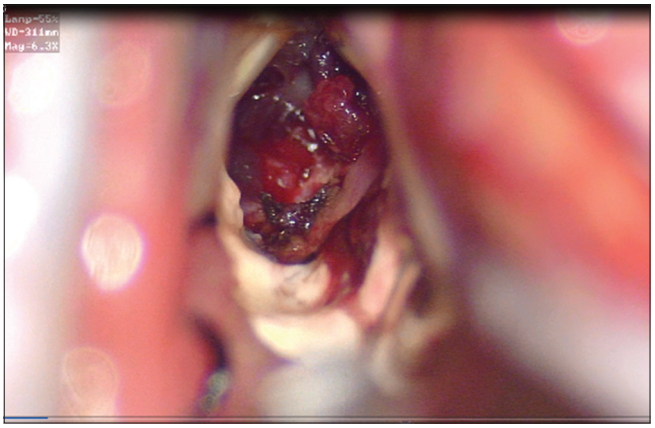

Figure 2: Intraoperative photograph showing interhemispheric transcallosal approach to the right ventricular cavernoma

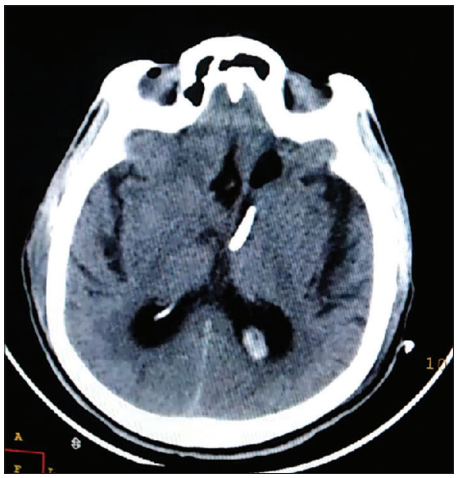

Figure 3: Immediate postoperative computed tomography scan brain showing minimal intraventricular hemorrhage

Discussion

Cerebral cavernous malformations are angiographically occult lesions. Histologically, they consist of dilated sinusoidal channels lined by a single layer of endothelium without any intervening normal tissue, smooth muscle cells, or elastin.[6] Cavernomas account for 10%–15% of all vascular malformations. Cavernous malformations occur in two forms: spontaneous and familial. The spontaneous form occurs as an isolated variant, with patients most commonly having a single lesion, whereas the familial form is characterized by multiple lesions and an autosomal dominant mode of inheritance. Three distinct gene foci on chromosomes 7q, 7p, and 3q have each been linked to familial cavernous malformations. The proteins encoded by these genes interact with the endothelial cytoskeleton during angiogenesis in the CNS and probably result in the development of these lesions.[7]

IVCs are a rare pathological entity constituting 2.5%–10.8% of cerebral cavernomas. The lateral ventricles are the most frequent site, followed by the third and fourth ventricles.[8]

IVCs have a relatively benign course. Patients mainly present with features of IVH, hydrocephalus, and seizures. The clinical features include nausea, vomiting, gait imbalance, and memory disturbances vision disturbances.[2]

Intraventricular bleeding from IVC may sometimes be potentially life-threatening requiring immediate surgical management. The incidence of rehemorrhage with IVCs is higher compared to cavernomas in other locations.[9] Patients with IVCs have a low incidence of seizures when compared to supratentorial cerebral cavernomas in whom the incidence of seizures is approximately 70%.[10] IVCs have low frequency of seizures because they do not cause perilesional intracerebral gliosis and hemosiderin deposition which have epileptogenic activity.[11]

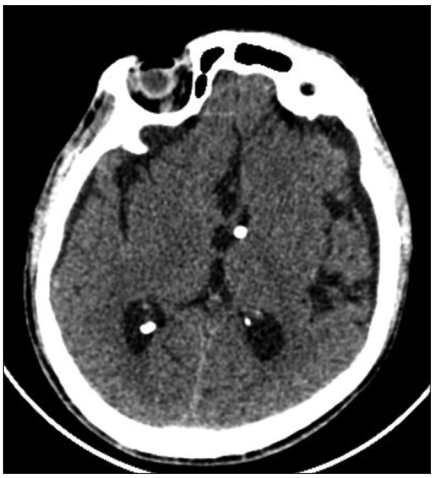

Figure 4: Follow-up computed tomography scan brain at 6 weeks

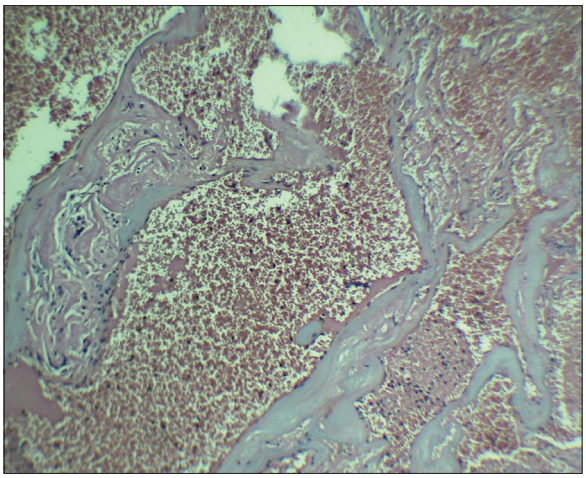

Figure 5: Photomicrograph of hematoxylin and eosin-stained preparation showing a lesion comprising of multiple large thin-walled lattice pattern of vessels, lined by a thin endothelium

MRI brain is the diagnostic modality of choice for cavernomas of CNS. Cavernoma appears as heterogeneous lesions in both T1- and T2-weighted images without enhancement after contrast administration. Cavernoma has a classical popcorn appearance on MRI. The MRI features correlate with different stages of degradation of blood products within the cavernoma. Hyperintense signal in MRI corresponds to hemoglobin degradation products, namely methemoglobin, whereas gliosis gives hypointense signal. Peripheral rim of hypointensity is due to the presence of hemosiderin, which has a paramagnetic effect. CT findings of cavernoma are often nonspecific. On CT imaging cavernoma appears as hyperdense lesion with occasional calcifications with mild or no contrast enhancement. CT angiography is not helpful in the diagnosis of cavernoma. Cavernoma is angiographically occult/cryptic vascular lesion since there is no feeding vessel to the lesion.[12] Typically, lateral ventricle cavernomas (LVCs) are associated with a medullary venous malformation which has a compensatory role in the venous drainage of the lesion.[13]

The radiological differential diagnosis of IVCs is choroid plexus papilloma, ependymoma, central neurocytoma, intraventricular meningioma, and subependymal giant cell astrocytoma.[2]

Cavernomas located in the lateral ventricles are approached according to their location, size, and expansion/dilatation of ventricular system. The shortest and safest trajectory is often chosen to provide safe dissection, minimize complications. Traditional approaches include transcallosal and transcortical approaches. Each approach has its own advantages and disadvantages.[14]

Surgical approaches

Interhemispheric transcallosal approach

Cavernoma arising within the body of the lateral ventricle are approached through the anterior transcallosal approach. The patient is placed in a supine position with the head slightly flexed. Neuronavigation is useful in the planning of the incision and craniotomy. A preoperative cerebral angiogram/MR venogram (MRV) is often obtained to determine the position of major draining cortical veins. The classic anterior interhemispheric craniotomy is placed two-thirds in front of the coronal suture and one-third behind.[15]

A bicoronal incision is made anterior to the coronal suture, or a horseshoe-shaped incision can also be made crossing the midline. The exposure in the midline is extended up to the superior sagittal sinus. The dura is opened and reflected medially up to the superior sagittal sinus. Cortical draining veins must be preserved. The dura covering the cortical veins draining into the superior sagittal sinus can be left intact to preserve the veins. The falx is identified in the midline and followed to the depth. Further arachnoid dissection is done to identify the two pericallosal arteries. The dissection is carried out between the two vessels. The callosotomy is done posterior to the genu of corpus callosum, and the lateral ventricle is entered. Malformations adjacent to the lateral ventricles can then be excised. Achieving meticulous hemostasis is of paramount importance. A ventricular drain is often left in place due to the risk of postoperative hydrocephalus even with small amount of IVH.[15,16]

Transcortical approaches

The transcortical approaches are commonly used for lateral ventricle cavernoma associated with hydrocephalus. This approach is preferred over the transcallosal approach for large midline draining cortical veins. Intraoperative ultrasound/neuronavigation helps in guiding direct ventricular access. Cavernoma in the anterior lateral ventricle is approached through superior frontal sulcus whereas cavernoma in the atrium of the lateral ventricle is approached through interparietal sulcus or superior parietal lobule. As discussed earlier, a preoperative cerebral

angiogram and MRV is often obtained to determine the position of major draining veins.[16]

Transtemporal approach

This approach is used for excision of cavernoma located in the temporal horn of the lateral ventricle. The patient is placed in supine position with the head turned 45° away from the affected side and slightly extended. A reverse question mark incision is made starting at the level of zygoma just anterior to the tragus of the ear then curving posteriorly over the ear and anteriorly toward the forehead. A temporal craniotomy is done. Temporal horn of the lateral ventricle is accessed through a cortical incision in the middle or inferior temporal gyrus.[16]

The transcortical approach provides better exposure and minimizes the risk of injury to bridging veins. The main complications of transcallosal approach are venous infarct, superior sagittal sinus thrombosis, disconnection syndrome, and forniceal injury.[17] Cavernoma located in the frontal horn is reached through either transcortical transventricular approach or through the interhemispheric transcallosal approaches. For excision of cavernomas situated in trigone and temporal horn of lateral ventricle transtemporal and superior parietal approach are employed. The transtemporal approach transcortical has a risk of injuring to visual fields, the vein of Labbe and speech.[5,16]

Another alternative approach that can be used for resection of trigonal cavernoma is the transsylvian transventricular approach which involves minimal disruption of the visual pathways.[13,17]

Conclusion

The lateral ventricle is an uncommon location for cavernoma of the brain. Preoperative diagnosis of IVC is difficult due to their low incidence, atypical location, lack of specific clinical manifestations, and neuroradiologic features. Cavernoma must be included in the differential diagnosis of intraventricular mass lesions. IVCs are associated with a high risk of IVH.

Progressive neurologic deterioration, mass effect causing hydrocephalus is indications for surgery. Microsurgical excision is the treatment of choice for IVCs. The approach depends on the location of the lesion and size of the ventricles.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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