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Case Report

Ruptured Tentorium Originating Masson Tumor

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Abstract

Intravascular papillary endothelial hyperplasia (IPEH) also known as Masson's tumor, is a benign, slow growing, vascular lesion which is seen very rarely and only a few cases have been reported intracranially in the literature. It has been reported at many sites, but the posterior fossa involvement is very rare. The preoperative diagnosis is very difficult, as there is no enough cases to achieve a clear understanding about the details of its radiological findings. Differential diagnosis have to be made especially from angiosarcoma and meningioma. It is curable by total surgical removal. In this article we presented the characteristic clinical, radiological, perioperative and pathological findings in a case of IPEH in an unusual location, origin and behavior. To best of our knowledge, we presented the first case of IPEH originating from tentorium.

Keywords: *Dural attachment, extraaxial lesion, intravascular papillary endothelial hyperplasia, posterior fossa tumor*

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) (Masson's tumor) was first described by Pierre Masson in 1923, who named it "hemangioendotheliome vegetant intravasculaire."[1] IPEH is known as an unusual benign tumor defined as a form of nonneoplastic endothelial proliferation found in organizing intravascular thrombus and less common, in extravascular hematomas.[2] The deep vascular inner layers of skin, of the neck and the head regions, are the most frequent sites for IPEH.[3] In this article, we present the first case of IPEH that originates from the tentorium. Management of IPEH should be complete excision if it is possible.

Case Report

We report a case of 56-year-old male who presented with a sudden onset of headache, dizziness, and gait imbalance that gradually increased in 15 days. Romberg's sign, dysdiadochokinesis, and ataxia were positive. Investigations including magnetic resonance imaging (MRI) demonstrated a right-sided lesion attached to the tentorium. It did not have edema but there was evidence of a hematoma beginning from the lesion, and extending to the left cerebellum through the tentorium. Postcontrast T1-weighted images showed mostly peripheral contrast-enhancing lesion which was partly attached to the tentorium [Figure 1]. The patient underwent a right paramedian suboccipital craniectomy. Infratentorial supracerebellar approach was performed. Following the relaxation of the cerebellum and opening the supracerebellar corridor, the lesion was seen 3 cm in depth in a close anatomical relationship with the tentorium, which is not expected in such intraaxial lesion [Figure 2]. The lesion was well-circumscribed and originating from the tentorium which is not usual. The lesion had a tight relationship with the tentorium and attached to the tentorium firmly. As the first step, it is planned to disconnect the lesion from the tentorium in order to mobilize and free the lesion from its superior surface. There were large dilated drainage veins around the lesion which were easily identified. Our attention was focused on preserving these vessels by exposing the tumor circumferentially until the total arterial devascularization of the tumor. Most of the arterial feeders of the tumor were originating from the

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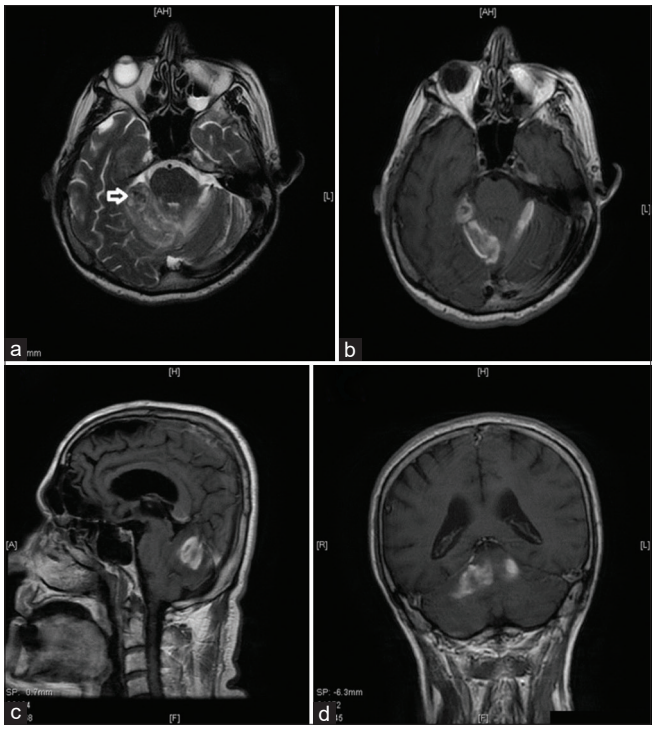

Figure 1: (a-d) Preoperative magnetic resonance images (a) axial unenhanced T2 weighted magnetic resonance imaging showing right-sided lesion (white arrow) and hyperdense areas which extends to the left cerebellum (hemorrhage) and there is no edema round the lesion. (b) Axial postcontrast T1 image, showing bilateral vermian lesions in which the right-sided mass is enhancing gadolinium peripherally. (c) Postcontrast sagittal image, demonstrating partly attachment of the mass to the tentorium. (d) Postcontrast coronal image demonstrates bilateral lesions close to the tentorium in mixt intensity

dura. After the coagulation of these arterial feeders, entire venous structures were also coagulated and dissected. Thus, the disconnection of the origin of the tumor which was the tentorium was achieved and the devascularization was carried out. The initial part of this surgery can be assessed as the crucial surgical step in terms of providing safe and controlled surgery since the tumor was avascularized. There was a thiny epiarachnoid space between tumor and cerebellar parenchyma which is one of the main features of extraaxial lesions. Due to the effective cerebellar relaxation, peritumoral dissection was performed easily, remaining feeders were coagulated, and tumor was totally removed en bloc [Figure 3]. Intraoperatively, hematoma extending along the tentorium was seen. The mass contained vascular structures.

Standard histopathological slides were performed, and additional immunohistochemistry was added. On Hematoxylin and Eosin staining slides, the mass was composed of fibrovascular tissue with numerous large, anomalous blood vessels. In some areas, larger blood vessels with thickened walls were also detected. The delicate papillary structures are covered by a single layer of swollen or plump endothelial cells around a core of fibrous connective tissue. An associated thrombus was also seen,

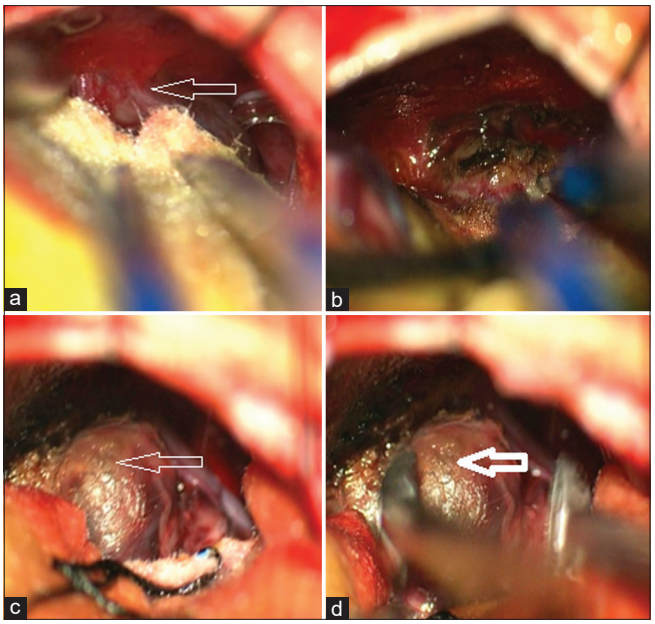

Figure 2: (a-d) Intraoperative images. (a) Initial surgical view of the tumor, originating from the tentorium (white arrow) after retraction of cerebellum inferiorly. (b) Bipolar coagulation of the tumor feeders and detachment of the tumor from the tentorium. (c) Surgical view of the tumor (white arrow) after total detachment. (d) Removal of the tumor in a single piece (white arrow) fashion

but no significant atypia, mitotic activity, or necrosis were present [Figure 4a and b]. Staining for CD34 and CD31 highlighted endothelial cells lining papillary structures and vascular channels [Figure 4c]. Stains for Epithelial membrane antigen (EMA), cytokeratin, and S-100 were negative. These findings are compatible with the diagnosis of the IPEH within a preexisting cavernoma.

The postoperative course was uneventful. The patient progressed well and was discharged in a good condition without any complaints before the surgery. The patient was followed-up for 5 years and no recurrence occurred. He died of lung cancer. Postoperative MRI showed no residual mass [Figure 3a-c].

Discussion

The involvement of Masson's tumor in the central nervous system is very rare. To the best of our knowledge, intracranially only 24 cases are reported so far in the literature, in which posterior fossa location is very rare.[2,4,5] In addition, there is no reported article before that shows the origination of a masson tumor from the tentorium, as described in this article. Meninges, cavernous sinus, skull base, subarachnoid space, and superior orbital fissure are the other locations intracranially reported for IPEH.[2,4,5]

On radiological findings, these pathologies tend to mimic high-grade tumors, necessary biopsy for diagnosis. Most cases of IPEH show contrast enhancement on MRI and computed tomography (CT).[2] The lesion often presents as

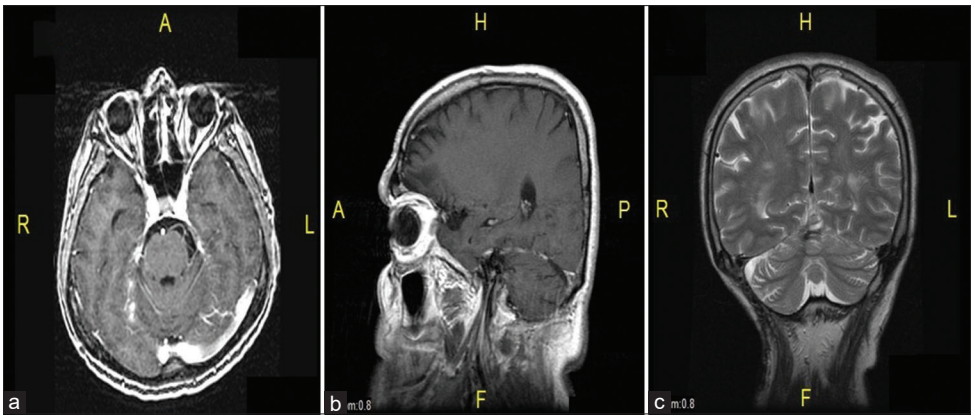

Figure 3: (a-c) Postoperative magnetic resonance images demonstrating no residual tumor. (a) Axial enhanced T1 imaging. (b) Sagittal enhanced T1 imaging. (c) Coronal T2 imaging

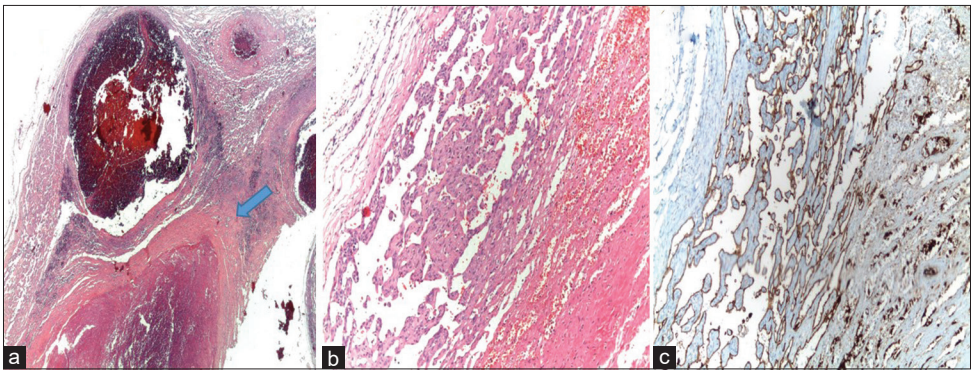

Figure 4: (a-c) Histopathological images. (a) Anomalous vascular channels, some of them are associated thrombus and fibrosis. Well circumscribed dilated large blood vessel containing papillary structures in the lesion is marked with an arrow, (H and E, ×40) (b) High magnification view of the (arrow) marked area. The delicate papillary structures are covered by a thin endothelial lining. No significant atypia, mitotic activity, or necrosis are seen, (H and E, ×100). (c) Strong immunopositivity for CD-31 in most of the endothelial cells lining small vessels and papillary structures CD31 ×100

a circumscribed vascular mass with mild hyperdensity on CT, hyperintense on T2 weighted MRI , and iso-to mildly hyperintense on T1-weighted imaging.[5]

Intracranial IPEH might be presented with symptoms secondary to the mass effect such as increased intracranial pressure, cranial nerve palsy, hemorrhage, and compression of adjacent neurovascular structures. Because of its vascular nature, hemorrhage is usually seen, as we experienced in this case.

Histopathologically differential diagnosis has to be made, especially from angiosarcoma. Our lesion is well circumscribed and located in a vessel. However, angiosarcomas are rarely intravascular and tend to invade surrounding tissues. Formation of multiple papillae and predominant hypertrophy of endothelial cells are common features in angiosarcoma and IPEH. However, necrosis, solid areas, mitotic figures, and moderate and severe nuclear atypia are only seen in angiosarcoma. The papillary structures in our case are covered by a single layer of swollen or plump endothelial cells around a core of fibrous connective tissue and associated thrombus is also seen, but no significant atypia, mitotic activity, or necrosis are present. For further confirmation of IPEH diagnosis CD31 and CD34 are used to highlight the endothelial lining around the papillary tufts.[1,2]

Despite the pathogenesis of IPEH remains unclear, intracranial form is associated with thrombosis in preexisting vascular malformation.[2] IPEH proliferation occurs as a response to combined arterial and venous reorganization. Angiogenic factors and consequent chronic vascular inflammation leads to IPEH proliferation. In our case, pathology is compatible with the diagnosis of the IPEH within a preexisting cavernoma. In such cases, it should be reminded that surgical aim has to be focused on total removal for curative treatment and long-term follow is needed due to slow-growing pattern of this pathology. In this case, the lesion was totally removed which is very important in the recurrence rate and in the necessity of adjuvant therapy. Because theoretically in total removal, there is no place for radiosurgery, radiotherapy, and chemotherapy.[2]

IPEH has to be considered as a benign tumor. Therefore the best prognosis is achieved by total tumor resection. To the best of our knowledge, we presented the first case of IPEH originating from tentorium.

Conclusion

Intracranial masson tumor is seen very rarely. Differential diagnosis has to be made very detailed. Intracranial IPEH has to be considered as a benign tumor, therefore the best prognosis is achieved by total tumor resection. When this is not possible the patient can get a second (adjuvant) therapy. Because of its slow-growing pattern, long-term follow-up is recommended.

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.

References

1. Masson M. L'Hemangioendotheliome intravasculaire. Ann Anat Pathol 1923;93:517.
2. Shih C, Burgett R, Bonnin J, Boaz J, Ho CY. Intracranial Masson tumor: Case report and literature review. J Neurooncol 2012;108:211-7.
3. Wang ZH, Hsin CH, Chen SY, Lo CY, Cheng PW. Sinonasal intravascular papillary endothelial hyperplasia successfully treated by endoscopic excision: A case report and review of the literature. Auris Nasus Larynx 2009;36:363-6.
4. Lee W, Hui Y, Sitoh Y. Intravascular papillary endothelial hyperplasia in an intracranial thrombosed aneurysm 3T magnetic resonance imaging and angiographic features. Singapore Med J 2004;45:330-3.
5. Miller TR, Mohan S, Tondon R, Montone KT, Palmer JN, Zager EL, *et al*. Intravascular papillary endothelial hyperplasia of the skull base and intracranial compartment. Clin Neurol Neurosurg 2013;115:2264-7.