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Dural-based posterior fossa giant cavernous hemangioma masquerading as hemangiopericytoma

Gigantický kavernózní hemangiom zadní jámy nasedající na duru napodobující hemangiopericytom

Dear editor,

Intracranial cavernous hemangiomas are one of the rarest among all vascular malformations, with a necropsy incidence between 0.39 and 0.53% [1] accounting for 10–20% [2] of all vascular malformations. They are usually found in a supratentorial location with 20.7% incidence in the infratentorial region but rarely in the region of cerebellum (6.1%) [3]. The rarity of the cavernous hemangioma in the region of the cerebellum may result in a frequent misdiagnosis thus making the treatment quite challenging. Giant cavernous hemangiomas (GCH) are exceedingly rare, imaging appearance is quite variable and because of slow perfusion, contrast enhancement is not regarded as a characteristic imaging finding of the cavernous hemangiomas, so straightforward diagnosis may again pose a significant challenge [4]. Some of the GCHs may have features similar to those of typical cavernous hemangiomas, while some may be purely cystic, and some may present with significant contrast enhancement along with a mass effect, mimicking neoplasms [5]. We present our experience with a case of GCH that mimicked the appearance of a hemangiopericytoma and discuss its radiological and histopathological findings.

A 25-year-old female patient was admitted to our hospital with a history of persistent occipital headache with intermittent vomiting and unsteadiness of gait during the past 3 months. On examination, she was conscious and oriented. Cerebellar signs were positive on the right side. There were no cranial nerve palsies and ophthalmoscopic examination was normal. Contrast-enhanced computed tomography (CECT) of the brain was suggestive of heterogeneously enhancing extra-axial mass lesion at the right posterior fossa with destruction of the overlying bone causing compression of the 4th ventricle and hydrocephalus. Magnetic resonance imaging (MRI) of the brain revealed a large, well-defined extra-axial mass lesion in the posterior fossa causing compression of the right cerebellum which was T1-isointense, T2/FLAIR-hyperintense with intensive post-contrast enhancement with necrotic areas and flow voids. There was dural enhancement and destruction of the right occipital bone along with cerebellar tonsillar herniation and obstructive hydrocephalus. There was non-visualization of the right transverse sinus suggestive of compression (Fig. 1). In view of the possible diagnosis of hemangiopericytoma, a retromastoid retrosigmoid approach was used to operate the patient in the sitting position. Intraoperatively, there was a lesion sized 5 × 5.5 cm in the right posterior fossa which was reddish-brown in color, firm and highly vascular with a well-defined plane of cleavage. The dura was hypertrophied, and the overlying bone was eroded by the tumor. Total excision of the lesion was done.

Histopathological examination of the specimen revealed a vascular lesion composed of a variable sized thin-walled vascular channel with dilated and blood filled lumina. Intervening stroma showed delicate collagenization. The lining cells were negative for EMA and S-100 (Fig. 2), was consistent with the diagnosis of cavernous hemangioma.

Postoperatively, the patient did well and was discharged on the 8th post-operative day. A repeat CECT of the brain showed that the lesion was completely removed. At the 3-month follow-up, she continued to do well with a complete resolution of pre-operative symptoms and cerebellar signs.

Cavernous hemangiomas are benign vascular lesions comprised of enlarged, clus-

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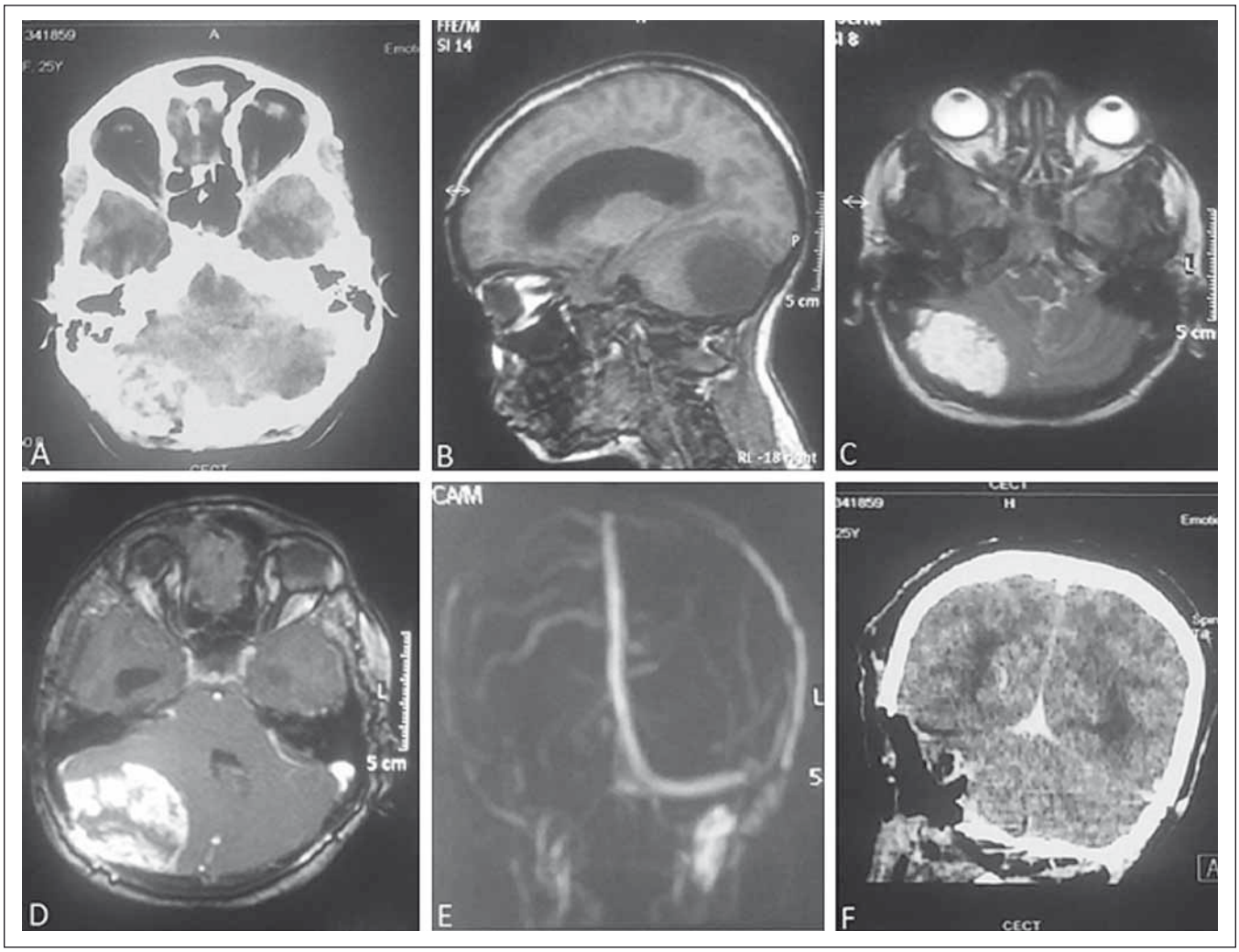

Fig. 1. Contrast-enhanced computed tomography (CECT) of the brain showing heterogeneously contrast-enhancing lesion at the right posterior fossa with erosion of the overlying bone (A). On MRI, the lesion is isointense on T1 (B) and hyperintense on T2-weighted image (C) with intensive post-contrast enhancement (D). There is non-visualization of the right transverse sinus (E) on MRA. Figure F is post-operative CECT of the brain showing complete removal of the lesion.

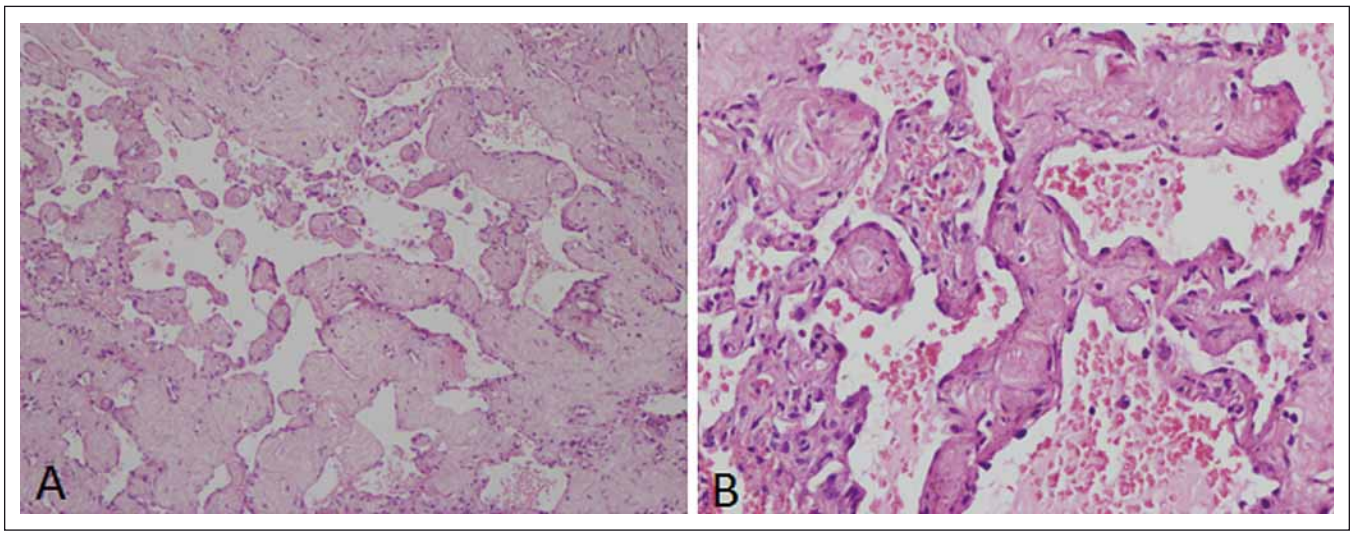

Fig. 2. H & E × 100 section showing irregular varying size vascular channel lined by endothelial cells and separated by thick collagenized tissue (A). H & E × 400 showing blood cells filled with collagenized intervascular spaces (B).

tered, sinusoidal vessels without intervening parenchymal tissue lined with epithelium. They are most frequently found in the brain parenchyma but can also arise in the spinal cord or in an extra-axial location [6]. The most common presenting symptoms are seizures (37%), hemorrhage (36%), headache (23%), and focal neurological deficit (22%); however, unlike parenchymal lesions, dural-based cavernous hemangiomas commonly present with headache, rather than seizures or hemorrhage [7]. Histologically, they appear as a cluster of hyalinized sinusoidal spaces without neural tissue which differentiates them from telangiectasias [6]. The mean size of cavernous hemangiomas reported in several large series varies from 15 to 19 mm in diameter and according to some authors, the larger the lesion, the higher the chances of symptom recurrence and GCHs were rarely seen [8].

Dural-based cavernous malformations mimicking meningiomas or other neoplastic processes have been reported in the literature and differential diagnosis of such lesions is wide, including hemangiopericytomas, hemangioblastomas, solitary fibrous tumors, dural-based metastases, and angiomatous meningiomas. These lesions tend to be iso- to hypointense on T1-weighted images, mixed to hyperintense on T2-weighted images, contrast enhancing, and hyperdense on CT [9]. However, these findings are not typical for cavernomas and can also be seen in hemangiopericytomas or meningiomas, thereby clouding the differential diagnosis based on history and radiologic analysis. Several authors have reported that lytic destruction of the adjacent skull on plain X-rays or CT suggests an hemangiopericytoma while hyperostosis suggests a meningioma [10]. Based on this, our pre-operative diagnosis was a hemangiopericytoma; however, the case turned out to be a GCH thus proving the fact that dural-base cavernous hemangiomas may mimic the clinical and radiological features of a hemangiopericytoma. This unique case strengthens the fact that lesions at the posterior fossa may cause diagnostic dilemmas and etiology of such tumors may not always be straightforward. The case also urges to keep cavernous hemangioma as a differential diagnosis of extra-axial lesions at the posterior fossa.

The lack of definitive radiologic and clinical entities differentiating cavernous hemangiomas from other neoplastic lesions can lead to unexpected findings in the operating room and may alter treatment plans especially in cases of hemangiopericytoma, as they are considered to be aggressive lesions. Given the lack of differentiable factors, it is very important for the neurosurgeon to keep in mind the possibility of dural-based cavernous hemangiomas when formulating a differential diagnosis and choosing the appropriate excision technique and postoperative treatment plan. These lesions do not tend to recur in contrast to hemangiopericytomas and can safely be monitored without adjunct therapy.

The unique nature of our case of GCH stems from its size, location in the posterior fossa, dural attachment and appearance similar to hemangiopericytoma. To the best of our knowledge, this has not been described in the literature. Although hemangiomas are benign entities, our patient's lesion was in the posterior fossa causing compression, hydrocephalus and cerebellar symptoms that necessitated resection. We would like to encourage others to consider the possibility of hemangioma in the differential diagnosis of dural-based posterior fossa lesions.

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