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Calvarial bone cavernous hemangioma with intradural invasion: An unusual aggressive course—Case report and literature review

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ABSTRACT

INTRODUCTION: Cavernous hemangioma of the skull is a rare pathological diagnosis, accounting for 0.2% of bone tumors and 7% of skull tumors. Usually calvarial bone cavernous hemangioma are associated with a benign clinical course and, despite their enlargement and subsequent erosion of the surrounding bone, the inner table of the skull remains intact and the lesion is completely extracranial.
PRESENTATION OF A CASE: The authors present the unique case of a huge left frontal bone cavernous malformation with intradural extension and brain compression determining a right hemiparesis.
DISCUSSION: Calvarial cavernous hemangiomas are benign tumors. They arise from vessels in the diploic space and tend to involve the outer table of the skull with relative sparing of the inner table. More extensive involvement of the inner table and extradural space is very unusual and few cases are reported in literature. To the best of our knowledge, intradural invasion of calvarial hemangioma has not been previously reported.
CONCLUSION: Our case highlights the possibility of an aggressive course of this rare benign pathology.

1. Introduction

Cavernous hemangioma of the skull is a rare pathological diagnosis, accounting for 0.2% of bone tumors and 7% of skull tumors [1]. They can occur at any age, but are commonly found in females who are in the fourth or fifth decades of life [2]. These cranial lesions are usually solitary and most often found in frontal and parietal bones, but the lesion could occur in almost any skull region (occipital, sphenoidal, clival and ethmoidal regions) [3].

Calvarial cavernous hemangiomas arise from vessels in the diploic space and are supplied by the branches of the external carotid artery. The middle meningeal and superficial temporal arteries are the main sources of blood supply [4].

Calvarial hemangiomas tend to involve the outer table of the skull and the diploe, with relative sparing of the inner table [5]. More extensive involvement of the inner table and extradural space is very unusual [6].

The authors present the unusual case of a frontal bone cavernous malformation with intradural extension. To the best of our knowledge, intradural invasion of calvarial hemangioma has not been previously reported. This case highlights the possibility of an aggressive course of this rare benign pathology.

2. Presentation of a case

2.1. History and examination

A 60-year-old man with a left frontal mass was admitted to our Department after 2 week of increasing headaches, nausea, right arm weakness with poor coordination, and difficulty walking because of a “clumsy” right leg. A clinical history of uncontrolled hypertension, diabetes mellitus and prostatic hypertrophy is reported. The patient denied previous trauma. The mass had been present for at least 6 months and had shown gradual growth. Physical examination revealed an approximately four cm in diameter palpable and non-tender mass in the left frontal region. Neurologic examination revealed a patient with right hemiparesis. The patient had 3/5 strength in the right upper and lower extremities. The deep tendon reflexes were slightly increased on the right side. Magnetic resonance imaging (MRI) revealed a large extra-axial lesion with epicenter in left frontal bone, predominantly solid (Fig. 1A–C). It measured 4 cm in its largest dimension and approximately 3 cm in depth. There was discontinuity in the cortical surfaces of both the inner and outer tables of the skull with intracranial invasion. The intracranial component of the lesion compressed the left

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frontal lobe. The extracranial component was also seen in the left frontal scalp region. The lesion was predominantly isointense on T1-weighted images (Fig. 1A) and hyperintense on T2-weighted images (Fig. 1B). After the administration of Gadolinium, there was heterogeneous enhancement of the mass (Fig. 1C). A surgical plan was made under the assumption that this lesion was a meningioma.

2.2. Surgical treatment

During surgery, the patient was positioned in the supine decubitus position and head was fixed in 3-pin head holder and rotated to the right. A question mark-shaped incision was given just behind the posterior margin of the mass and the skin flap was raised subgaleally. Soft, reddish-brown, highly vascular lesion was seen in the left frontal region with erosion of the outer table. The whole extracranial portion was removed. Then we proceed to remove the intracranial portion of the tumor. After placing a burr hole behind the lesion, a circular craniotomy of about 6 cm was performed around the lesion. At this point, the tumor was found to be not completely extradural. In fact, the lesion eroded the dural mater with invasion of intradural compartment. The lesion is then coagulated and followed up to identify the opening of the dural mater. A defect of the dural plane caused by the tumor has been used to remove the residual intradural part of the lesion. At this stage, the lesion did not have arterious pial afferents anymore. The pia mater was thick and whitish. After total removal of the lesion, a polymethyl methacrylate cranioplasty was performed.

2.3. Postoperative course

Postoperatively, the patient presented immediate relief of symptoms, and results of his neurological examination were normal, with complete recovery from his previous motor weakness. Postoperative CT scan showed a good restoration of the cranial deformity (Fig. 1D).

He was discharged home on postoperative Day 3 after an uneventful course. The patient has been followed up for 48 months post-operatively and remains asymptomatic with an excellent cranial contour and no evidence of recurrence on serial MRI imaging (Fig. 1E and F).

2.4. Pathological findings

Macroscopically, the pathologic bone presented a huge purple-red blush mass that eroded completely the inner table. The dura appeared eroded throughout its thickness by a bluish red spongy lesion.

Histological examination of the bone revealed a capillary hemangioma consisting of thin-walled blood vessels, some of which were distended with blood (Fig. 2A). Microscopic examination of the dura showed pathological vessels perforating the dura mater (Fig. 2B). The vessels were separated by scarce connective tissue intensely blue stained (Fig. 2C). Immunohistochemical staining with CD34 highlights the contours of the malformed vessels (Fig. 2D).

The present work has been reported in line with the CARE criteria [7].

3. Discussion

Hemangiomas are benign tumors of blood vessels and are histologically classified as cavernous and capillary [8]. Most of the calvarial hemangiomas are of the cavernous type. The cavernous type is composed by a group of large, dilated blood vessels sepa-

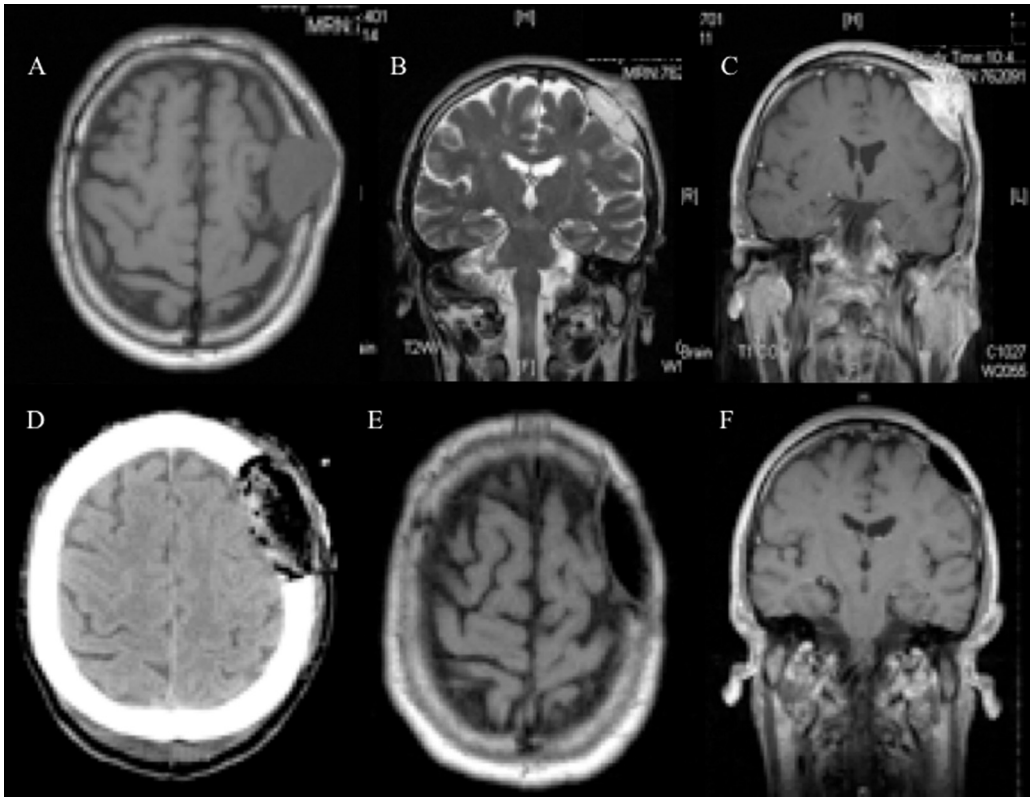

Fig. 1. (A–C) Pre-operative MRI imaging revealed a large extra-axial lesion with epicenter in left frontal bone and intradural invasion, predominantly isointense on axial T1-weighted image (A), hyperintense on coronal T2 weighted image (B) and with contrast enhancement (C). (D) Postoperative CT scan showed a good restoration of the cranial deformity. (E, F) Follow-up MRI axial and coronal T1 weighted images with Gadolinium demonstrated no evidence of recurrence.