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CASE REPORT

Extra-axial Cavernoma Mimicking Meningioma: A Case Report

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Abstract

Background— Extra-axial cavernous malformations (ECMs) are rare benign vascular malformations that may mimic meningioma. ECMs may have different radiologic features but some specific radiologic characteristics may help to differentiate them from meningioma. Herein, we describe a rare case of ECM with radiologic features similar to meningioma.

Case Description— The patient is a 62-year-old woman who presented with 3 months of headache. Initial head computed tomography (CT) showed an extra-axial hyper-dense lesion in the middle cranial fossa. A brain MRI confirmed the presence of an extra-axial lesion in her left middle cranial fossa which was hypo-intense on T1-weighted image and iso-intense on T2-weighted image with fine heterogenous contrast enhancement on contrast-enhanced T1-weighted image. Based on her imaging findings, pre-operative diagnosis of cavernoma was made, and when the lesion was resected, biopsy confirmed the diagnosis.

Conclusions— ECMs are rare lesions which should be considered in the differential diagnosis of the extra-axial masses, particularly if neuroradiological features suggest a meningioma. Given histopathological confirmation is always essential, total resection of the lesion should be the treatment of choice.

Key words— Cavernoma, Cavernous malformation, Cavernous hemangioma, Extra-axial cavernoma, Meningioma, Surgery, Management, Outcome.

INTRODUCTION

Cavernous malformations, also known as cavernous hemangiomas or cavernomas are accounting for 5-13% of central nervous system (CNS) vascular malformations. They can occur sporadically or congenitally (1). Extra-axial cavernous malformations (ECMs) are rare and are mostly located in the middle cranial fossa associated with the cavernous sinus, although they could also be found in other locations such as cerebellopontine angle , tentorium, convexity, anterior cranial fossa, Meckel's cave, Sella turcica and internal auditory meatus with much less frequency (0.2-0.5%) (2).

ECMs are mostly asymptomatic but can present with headache, cranial nerve palsy, seizure and infrequently intracranial hemorrhage. The most important complications of ECMs are cranial nerve compression and endocrine dysfunction (3). ECMs have the tendency to mimic and sometimes even coexist with other CNS tumors (e.g. meningioma, neurinoma and pituitary adenoma), other vascular malformations (e.g. developmental venous anomalies) or inflammatory processes (3). Imaging modalities, particularly brain Magnetic resonance imaging , play a key role in the diagnosis of cavernomas. The treatment options depend on the accessibility and clinical presentation of the lesion and varies from conservative management to surgical resection. The most important clinical aspect of cavernomas, which also poses a surgical challenge, is their innate potential risk for hemorrhage and expansion (1). In contrast, meningiomas are the most frequent primary tumors of CNS that usually originate from dura and rarely can be found at the extradural sites (4, 5). Similarities in the originating location and radiological features of ECMs and

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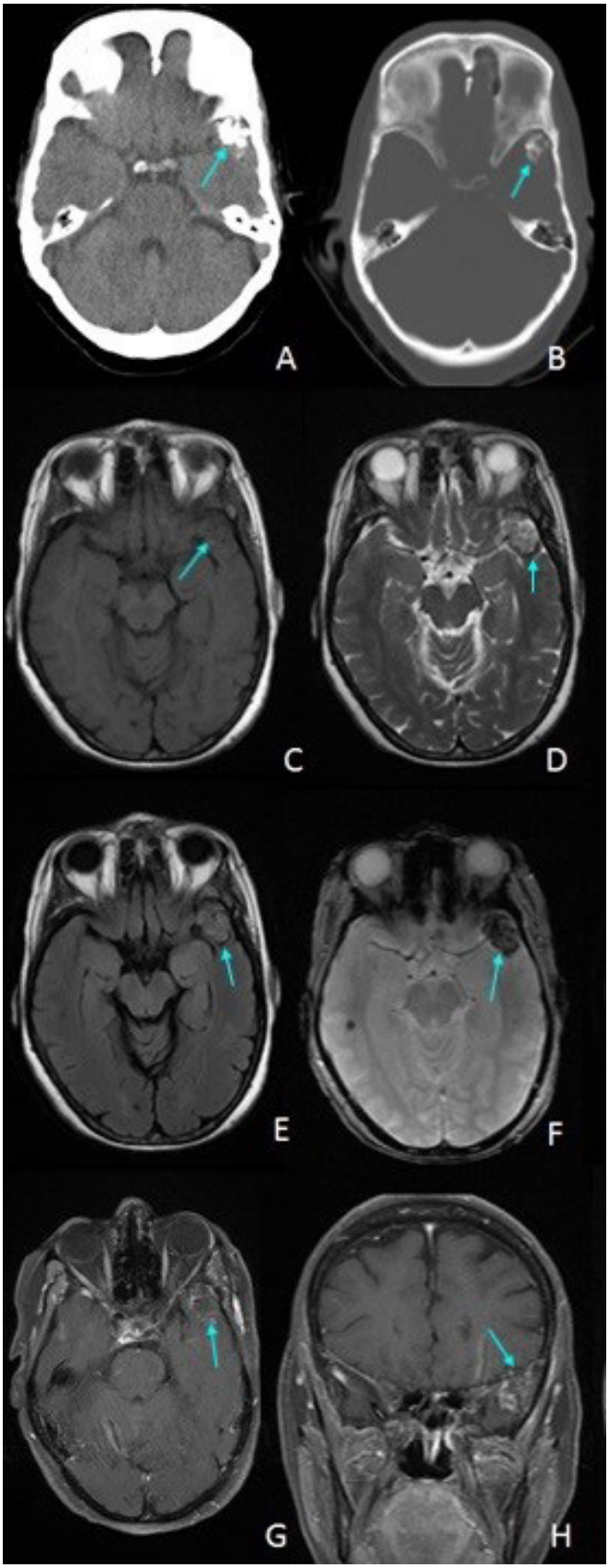

FIGURE 1: A and B: Parenchymal & bone windows of head CT without contrast show a hyper-dens extra-axial lesion in the anterior left middle fossa, posterior to the sphenoid lesser wing. C and D: Brain MRI T1 and T2-weighted sequences shows the hypo and hyper signal intensity of the lesion, respectively. E: Iso-intensity signal shown at FLAIR sequence. F: Hypo-intensity signal shown on Gradient echo sequence. G and H: Contrast-enhanced T1 image show fine heterogeneous enhancement of the lesion in axial and coronal sections, respectively.

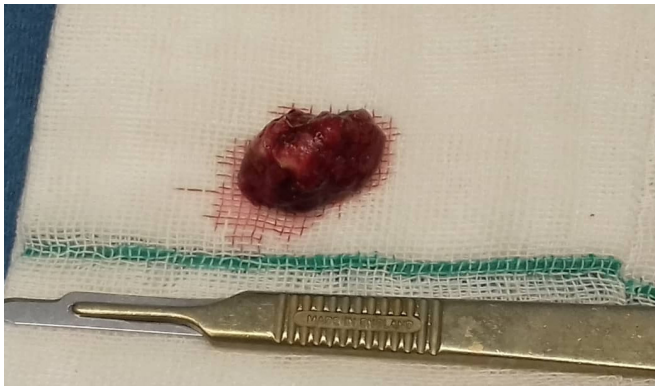

FIGURE 2: Macroscopic view of the lesion which was totally resected via left pterional craniotomy.

meningiomas can make the diagnosis and differentiation of these two challenging at times.

CASE PRESENTATION

A 62-year old woman presented with 3 months history of left temporal headache. Her past medical history and social history were unremarkable. She had a normal state of consciousness and orientation. Neurological examinations did not show any focal neurological deficit.

In her work up, a head CT without contrast revealed an extra-axial hyper-dense lesion in the left middle cranial fossa at the posterior aspect of the left sphenoid lesser wing (Figure 1, A and B). Subsequently a brain Magnetic resonance imaging with and without contrast performed. Gradient echo sequence confirmed the presence of an extra-axial hypo-intense lesion within the left middle cranial fossa. The hypo-intensity of extra-axial lesion on Gradient echo sequence was thought to be due to calcification or hemosiderin deposition. On T2-weighted image, the lesion was iso-hyper intense and a hypo-intense hemosiderin rim was noted and on T1-weighted image, the lesion was hypo-intense. The same pattern of intensity as T2-weighted image was seen on fluid attenuated inversion recovery (FLAIR) sequence. Fine heterogeneous contrast enhancement of the lesion was shown on contrast enhanced T1-weighted image (Figure 1). Based on these findings, cavernoma was considered as the pre-operative diagnosis.

Management

This lesion was considered responsible for patient's symptoms. The patient underwent a left pterional craniotomy and surgical resection of the lesion in the left middle cranial fossa. After opening of the cranium and dura, the lesion was noted to be calcified and hyper vascular with blueberry appearance (Figure 2). The lesion was completely resected and bone flap was replaced. Post-operative course was uneventful.

Histological studies

Histological specimens were stained with hematoxylin and eosin. Multilobulated sinusoidal spaces including proliferative vascular structures with thin and hyalinized wall, lined with flat endothelial cells with ectatic pattern were identfied. Multiple focal fine calcifications were also observed. These vascular spaces were separated by sections of cortical degen-

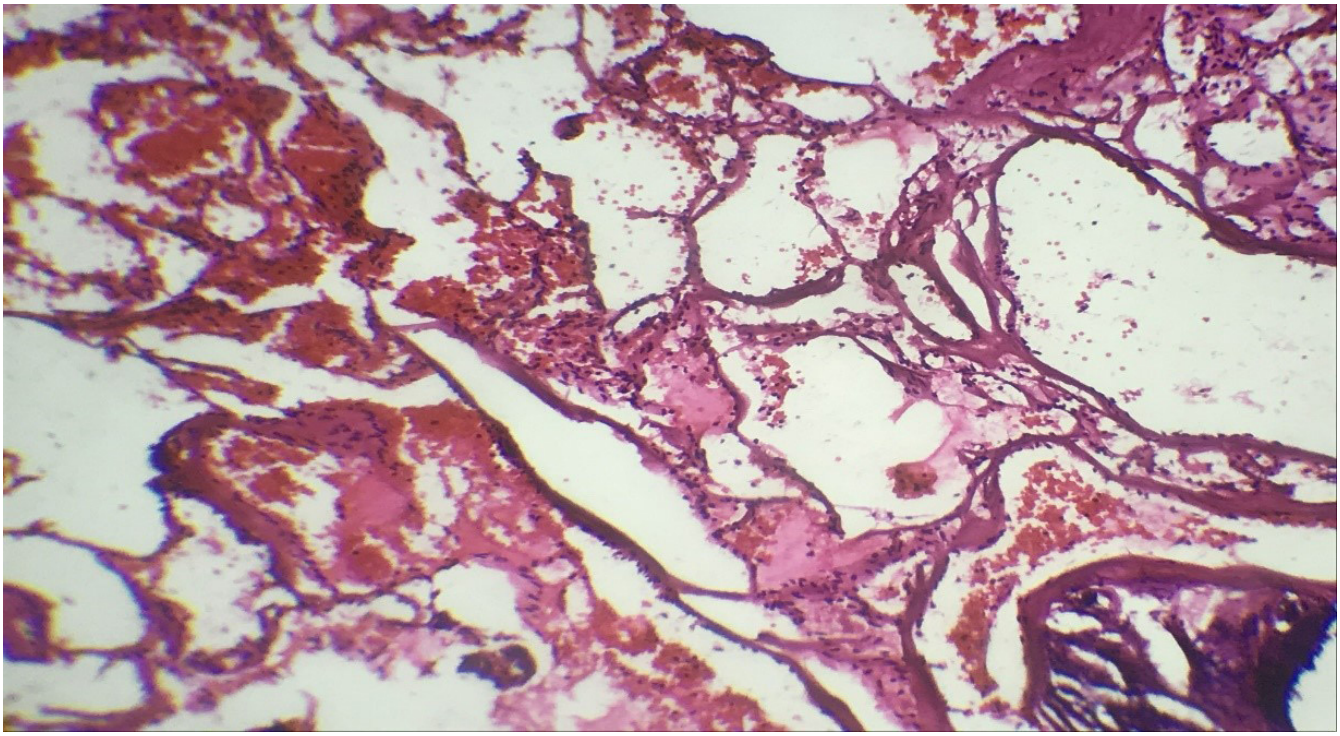

FIGURE 2: Photomicrograph of the extra-axial cavernoma specimen of the left middle fossa. High-power (X100) view of the cavernoma showing a sinusoidal space composing of thin walled vessels lined with one layer of endothelial cells. These spaces are separated by interstitial fibrotic tissue and a mixture of other tissues.

erated bony trabecula and fibrotic tissue (Figure 3). There was no evidence for neoplastic tissue necrosis or heterogenous differentiation. ECM associated with gliosis was the final definite diagnosis.

DISCUSSION

Dural based lesions have limited differential diagnosis including meningioma, hemangiopericytoma, sarcoma, granulomatous lesions, tuberculosis , metastatic masses and cavernoma with meningioma being the most prevalent one (6). In this study, we described the key diagnostic features of an ECM which can help differentiate the lesion from meningioma. In contrast to the intra-axial cavernomas, headache and cranial nerve involvements are more prevalent than intraparenchymal hemorrhage and seizures in ECMs (7-9).

On histological examination, ECMs are generally benign vascular malformations in which smooth muscles and elastic fibers of the vessel walls without interposing neural tissues. Additional features include inflammation, calcification, and ossification, mostly in larger lesions. Characteristically, they have a raspberry appearance with purple like clusters and usually follow a honeycomb pattern (10). Absence of proper junctions between the endothelial cells as well as the lack of the smooth muscles and elastic fibers in the vessel walls cause ECMs to bleed. . Furthermore, the growth rate of these lesions is very slow. Meningioma is generally a neoplastic lesion which has neoplastic features and grow slowly (10).

On head CT scan, cavernomas usually present as iso-hyper dense lesions and often demonstrate mild contrast enhancement (2, 7-10). In some cases , punctate calcification might be observed whereas calcification is quite rare in meningiomas (7). Brain Magnetic resonance imaging is more helpful in differentiating these entities. While on T1-weighted imaging, ECMs usually appear iso-intense or hypo-intense lesions, on T2-weighted imaging they often present as hyper-intense lesions. Meningioma has the same features but there are specific characteristics which can help to distinguish between the two. For intra-axial cavernoma, there is usually a hypo-intense hemosiderin rim surrounding the lesion on the T2-weighted image which gives the lesion a popcorn appearance and is diagnostic for intra-axial cavernoma (10). Notably, this popcorn pattern is usually absent in the cavernous sinus and dural based cavernomas but was noted in our case (8, 11). Another differentiating feature is that meningiomas demonstrate homogeneous post-contrast enhancement. In contrast, cavernomas usually have fine heterogenous enhancement and are usually multilobulated. The dural tail sign is a classic finding in meningioma which could be sometimes mimicked by cavernomas. Gradient echo sequence or susceptibility weighted imaging on MRI could be useful for detecting smaller cavenomas. ECMs could demonstrate variable and sometimes atypical radiological patterns such as homogeneity, hyperostosis or dural tail sign which can be misleading.

Total surgical resection of the ECMs is generally the treatment of choice for these lesions. However, due to the cranial nerve or other critical structures adherence and vascular nature of the mass, complete surgical resection may be challenging. Intra-operative hemorrhage, particularly when using ultrasonic guided aspiration, and cranial nerve injuries are among the main complications of the surgical approach (9). Stereotactic radiosurgery represents the other therapeutic option, which leads to reduction in the lesion volume. However, this option is only recommended in cases of partial surgical resection after histological confirmation (3).

CONCLUSION

In conclusion, cavernomas are rare extra-axial lesions which should be considered in the differential diagnosis of extra-axial masses, particularly if neuroradiological features are suspicious for a meningioma. Histopathological confirmation is necessary and total resection of the lesion should be the goal of treatment.

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