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Primary Intraosseous Meningioma with Extracalvarial Extension into Subcutaneous Tissue

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ABSTRACT

Meningiomas are extraaxial tumors that arise from arachnoid cap cells and can develop wherever these cells reside. Therefore, meningiomas are typically attached to dural leaflets with potential extension and erosion into surrounding tissue, including bone. Meningiomas arising primarily in bone are rare. In this paper, we present the case of a 46-year-old female patient with a primary intraosseous meningioma, characterized by extensive hyperostosis of the calvarium and intracranial and extracranial soft tissue tumor extension.

Keywords: Meningiomas, Intraosseous meningioma, Extracalvarial meningioma, Arachnoid cells

INTRODUCTION

Meningiomas are typically benign lesions that arise from arachnoid cap cells. These lesions are usually found in the intracranial and intraspinal compartments. Intracranial meningiomas can be found in a variety of locations including convexity as well as skull base following the dural folds. They can occasionally be intraventricular as well specifically in the adult population. Other rare reported locations of meningiomas include primarily intraosseus meningiomas as well as outside the central nervous system such as lung, mediastinum, adrenal gland [1]. We present in this paper a case of a primary giant calvarial meningioma with local extension.

Case Report

The patient is a 46-year-old female who presented with a 15-year history of bi-parietal scalp swelling and a several month history of intermittent localized headaches. Her headaches were sharp in nature and typically in the right parietal region. The patient was reported to have a history of Paget disease. Additionally, she described frequent scalp "infections" which intermittently required drainage procedures.

Given the severity of these symptoms she was referred for an MRI of the brain (**Figure 1**) by her primary care physician, which revealed significant meningeal thickening and enhancement, marked irregular calvarial thickening, and enhancing soft tissue extending both within the widened diploïc space and extracranially into the subgaleal and subcutaneous scalp soft tissues.

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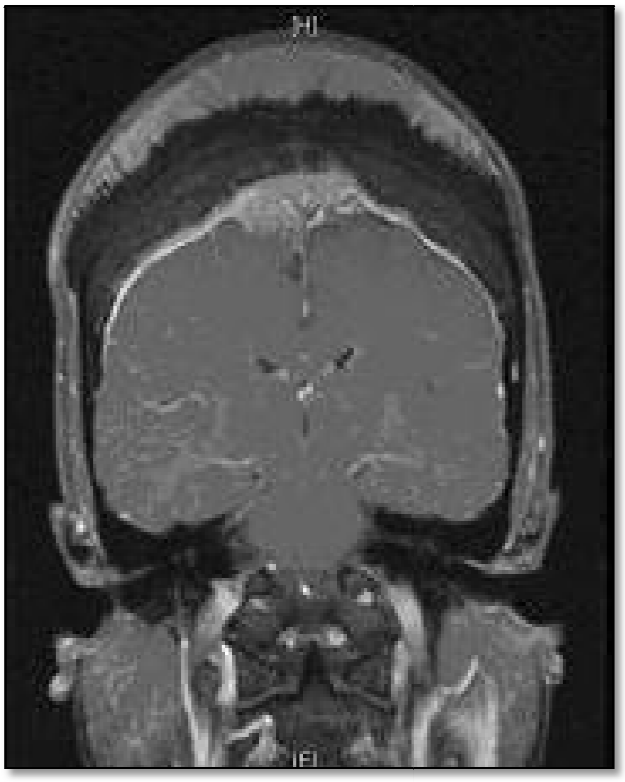

Figure 1. Post-gadolinium fat-saturated T1-weighted coronal plane spin echo MR images demonstrate marked calvarial thickening across the vertex, with linear low signal intensity structures extending into the enhancing scalp mass, and intra and extracranial soft tissue tumor extension. The intracranial component infiltrates the dura and exerts mass effect on the subjacent cortex and is largest where it appears to invade the superior sagittal sinus at midline.

The soft tissue mass extended 3.7 cm extracranially at one point, and 6 cm intracranially with signs of dural infiltration. There was as well concern for a local occlusion of the middle third of the superior sagittal sinus on MRV (**Figure 2**). A CT scan of the head (**Figure 3**) revealed spiculated reactive new bone that also radiated perpendicularly from the skull surface toward the scalp. Of interest, there was preservation of the bony structure along the sagittal suture as well as lack of focal lytic lesions. On physical examination, the patient was neurologically intact. She had marked fullness of the scalp in the bifrontal and parietal regions with scattered areas of superficial scalp nodularity and thickening. There were several areas with focal tenderness to pressure.

In the setting of a presumed paget's disease, the imaging and clinical findings were concerning for osteosarcoma. Therefore, the patient was taken for an en bloc biopsy of the right posterior frontal portion of the lesion (**Figure 4, Figure 5**) which ultimately revealed a WHO Grade I syncytial-type meningothelial neoplasm, which had infiltrated the calvarium extracranially and intracranially. Treatment strategies were discussed with the patient including surgical resection and adjuvant therapy. Patient refused any further intervention due to esthetic side effects and was therefore followed with serial imaging, initially with short follow-ups that were later spaced out. Patient is currently 7 years out from her biopsy with stable imaging and no new clinical complaints.

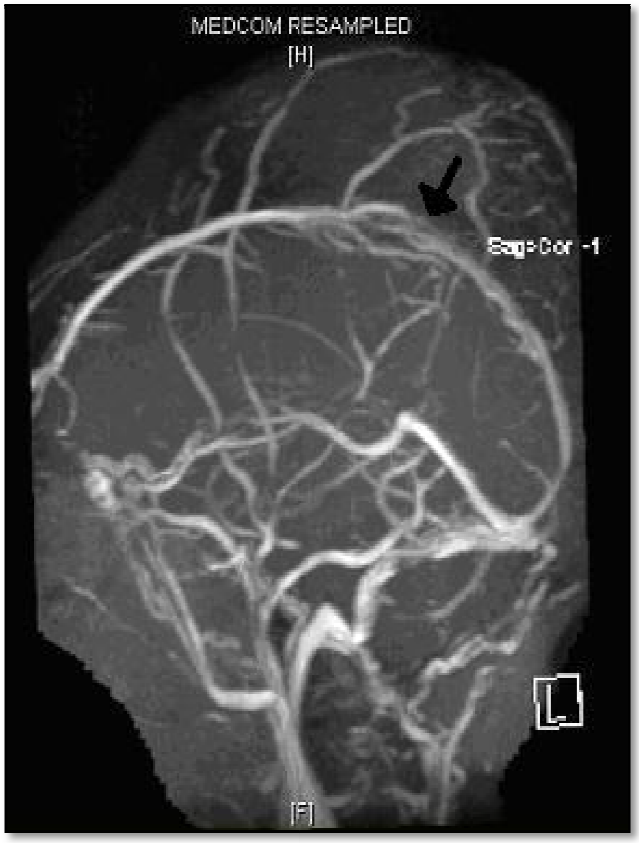

Figure 2. Sagittal MRV MIP reformat demonstrates attenuation of the posterior aspect of the superior sagittal sinus, with small associated venous collaterals (arrow).

DISCUSSION

The first case of primary extradural meningioma is thought to be reported in 1730 by Johan Salzmann [2]. The term "Primary Extradural Meningioma" (PEM) was first coined by Lang et al. as a designation of all meningiomas that arise away from the subdural space with no dural attachment [3]. These include locations such as calvarial, mediastinal, pulmonary, nasal and oral cavity, paranasal sinuses, neck, and abdomen [3-5]. PEM represent about 2% of all meningiomas the majority of which are located in the cranial vault [6].

Whilst the cell of origin of the meningiomas is the arachnoid cap cell in the meninges, the origin of PEM is not entirely clear. Multiple hypotheses have been advanced. Cranial

trauma with entrapment of dural/arachnoid cells in the fracture lines is one such explanation [7].

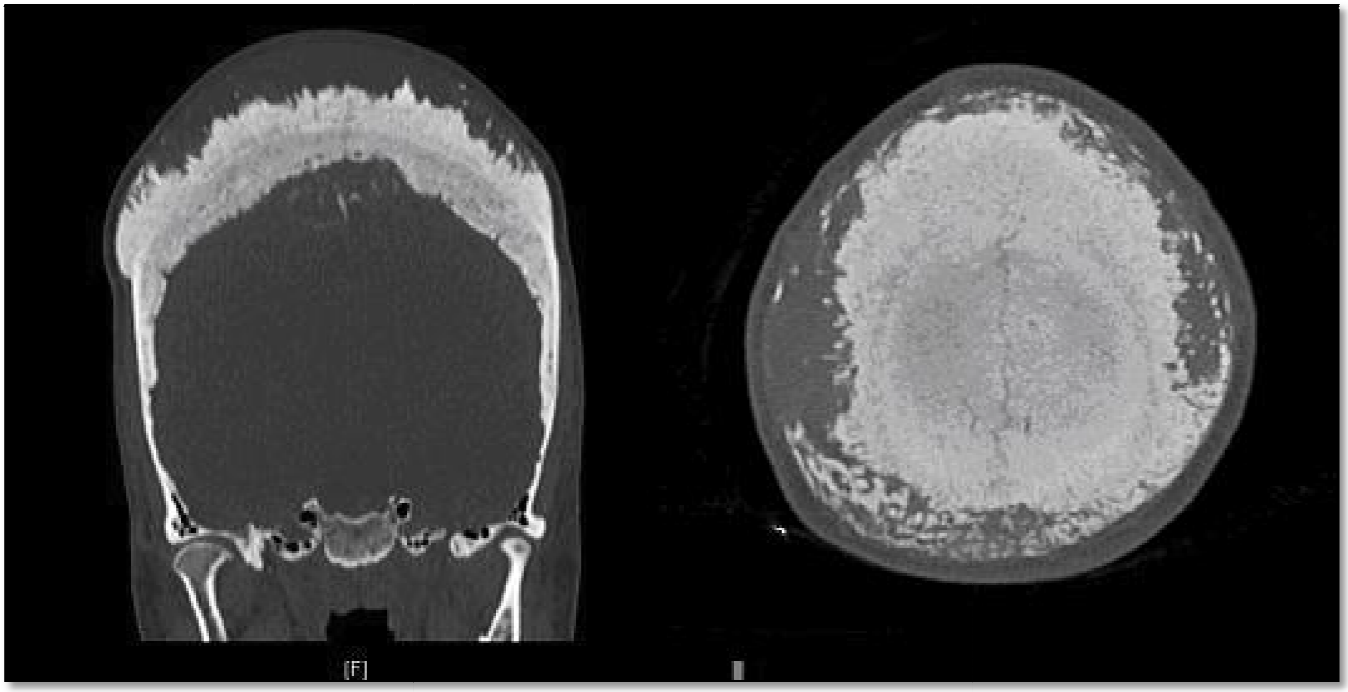

Figure 3. Coronal reformatted images from a noncontrast CT (bone window) nicely reveals the hyperostotic nature of this lesion, with perpendicular bony spicules extending into the scalp mass. The enduring sagittal suture and the lack of lytic bone destruction, mitigate against the diagnosis of a malignant tumor. There is no evidence of Pagetoid bone.

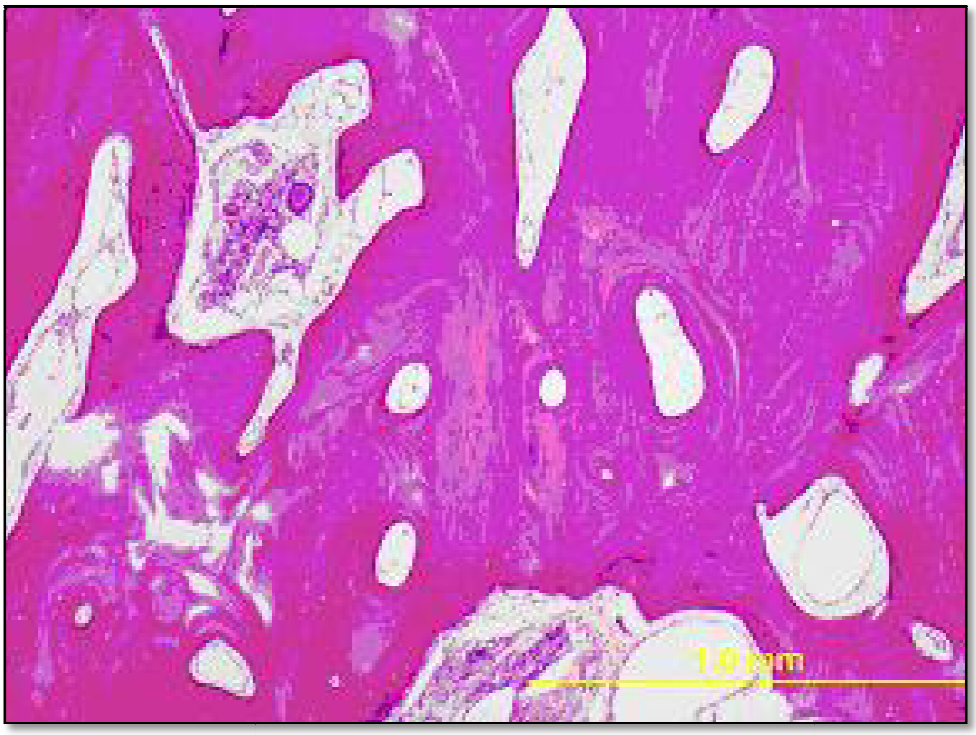

Figure 4. Wedge section of skull sample is fixed and decalcified. There is significant hyperostosis and intracanalicular tumor involvement.

Along the same lines, arachnoid cells are thought to become entrapped in the cranial sutures during the cranial molding as the baby passes through the birth canal [8,9]. However, only 0.2 to 4% of patient with PEM report a history of trauma [3,7]. Some authors have hypothesized that these PEM develop from pluripotent mesenchymal cells (Fibroblasts, schwann cells…) located extradurally [3,10,11]. Another hypothesis involve faulty arachnoid cell migration during the embryologic development to extracranial locations [3,12,13].

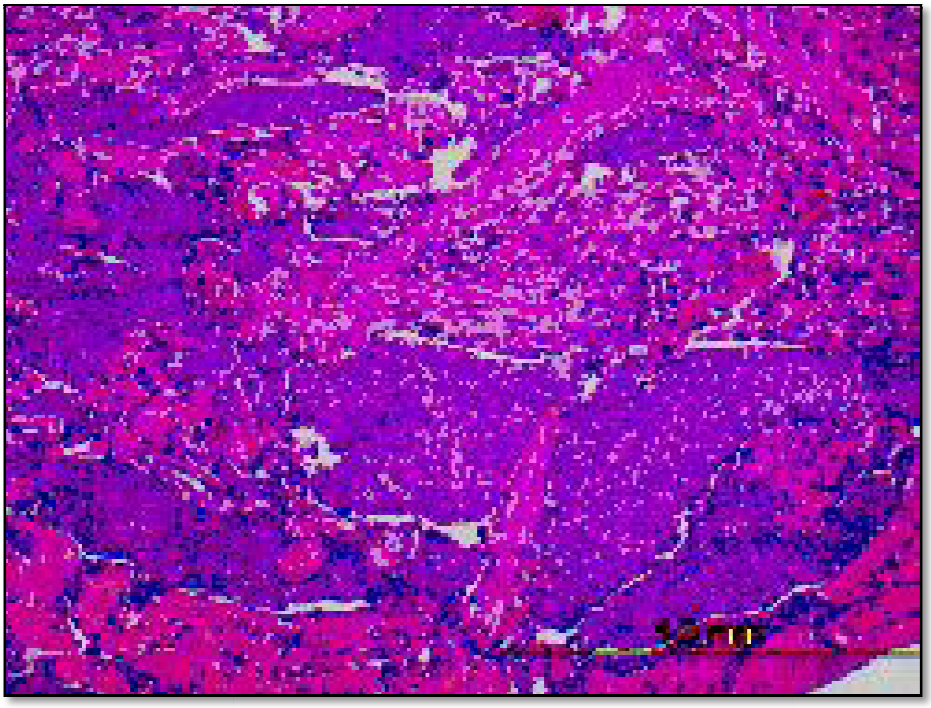

Figure 5. Higher power. The histoarchitecture is syncytial-type with no atypical features.

PEM have a bimodal distribution with the first peak in the second decade of life and a later peak between the fifth and the seventh decades [6]. They typically present as a progressively growing mass with rarely accompanying neurological symptoms. The clinical manifestations depend on the size and location of the tumor with varying symptoms such as deafness, painless expanding mass, proptosis [14,15]. In general, these slow growing meningiomas are usually asymptomatic and do not show neurologic signs unless the lesion extends through the dura to exert mass effect on intracranial structures. The most common radiologic finding for the calvarial meningiomas is hyperostosis [9], followed by osteolysis with the differential diagnosis therefore including bony metastasis, osteogenic sarcoma, epidermoid cyst, hemangioma, bone cyst, osteochondroma, multiple myeloma, eosinophilic granuloma, and fibrous dysplasia. In our case, the chronic history of scalp swelling and head enlargement led to a presumptive diagnosis of Paget disease before imaging was performed. It is likely that the episodic "scalp infections" requiring drainage were in fact areas of extracranial soft tissue tumor necrosis.

Lang et al. proposed a classification for cranial PEM according to their relative location vis-à-vis the skull. Purely extracranial tumors without bony attachment were classified as type I. Type II lesions had purely bony location and type III were primarily intraossesous tumors with extracalvarial extensions [3]. In 2006 Bassiouni et al. [12] proposed another classification taking into account the dural involvement as it has treatment and surgical implications. Type I tumors were epidural, type II calvarial, type III extracalvarial and type IV mixed with extension from the dura to the extracalvarial space.

The most common reported histopathological subtype is the meningothelial meningioma followed by transitional then psammomatous [6]. Atypical meningiomas constitute 3 to 5.6% of PEM while malignant meningiomas comprise 3.9 to 8% of all PEM [3,6,7].

The management strategy for a lesion of this nature is not all that dissimilar to that for other slow growing tumors. Wide marginal excision can be curative, however, given the diffuse infiltrative nature of the disease, a meaningful surgical excision can be difficult to achieve. In case of dural

involvement, resection of the dural area is advised. Other options include radiotherapy, and medical treatment options such as RU 486 [16,17] and hydroxyurea, [18,19] although some studies have shown these agents to be of marginal benefit.

CONCLUSION

Although rare, intraosseous meningiomas should be part of the differential diagnoses in patients with a skull-based lesion with associated soft-tissue components, although unlike the case we presented in this paper, the soft tissue component may be relatively minor. Plain x-ray and CT are superior to MRI in visualizing osteoblastic changes in the calvarium, a key finding in making the diagnosis.

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