Primary Intraosseous Meningioma with Extracalvarial Extension into Subcutaneous Tissue

Zoe Z. Zhang1, Rudy J. Rahme2, Eric J. Russell3, Alexander Nemeth3, Michelle Naidich3, Jin Chen Zhao4 and James P. Chandler5*

1Medical Resident, Neurosurgery Department, University of Minnesota Medical School, Chicago, USA
2Neurosurgery Resident, Northwestern University Feinberg School of Medicine and McGaw Medical Center, Chicago, USA
3Department of Radiology, Northwestern University, Feinberg School of Medicine, Chicago, USA
4Director, Surgical Simulation Laboratory, Division of Neurosurgery, North Shore University Health System, Evanston, USA
5Lavin/Fates Professor of Neurological Surgery, Professor in Neurological Surgery, Neurology - Ken and Ruth Davee Department and Otolaryngology - Head and Neck Surgery, Northwestern University, Feinberg School of Medicine, Chicago, USA

Received Feb 2, 2016; Accepted Mar 23, 2016; Published 30 Apr, 2016

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 intraosseous 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 soft tissue scalp structures.

Corresponding author: James P. Chandler, MD, Lavin/Fates Professor of Neurological Surgery, Professor in Neurological Surgery, Neurology - Ken and Ruth Davee Department and Otolaryngology - Head and Neck Surgery, Northwestern University, Feinberg School of Medicine, 676 N. St. Clair, Suite 2210, Chicago, IL, 60611, Tel: 312-695-1065; Fax: 312-695-0225; Email: jchandle@nm.org


Copyright: ©2016 Zhang Z, Rahme R, Russell E, Nemeth A, Naidich M, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
saturated T1

ts mass

d intracranially. Treatment
of interest, there was
-weighted

Figure ial
egions with

Figure 4, Figure
-type) revealed spiculated
hort follow-ups

2% of all
s well concern for a local occlusion of the

5]. PEM represent about
meningothelial neoplasm, which had infiltrated the
meningothelial neoplasm, which had infiltrated the
right posterior frontal portion of the lesion (Figure 3). Clinical findings were concerning for osteosarcoma.

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 syncytioid-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.

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].

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 psammomatosus [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

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

REFERENCES