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Hamartoma of the Scalp with Ectopic Meningothelial Elements in a Two-Year-Old Male

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Abstract

A two-year-old male presented with a 1.1 cm, firm, subcutaneous scalp mass. The lesion was discovered at birth, had grown with the child's head, and recently became painful. Magnetic resonance imaging and ultrasound demonstrated a soft tissue lesion associated with an anomalous connection between the deep venous drainage and the superior sagittal sinus, consistent with sinus pericranii. Surgical excision of the lesion was performed with obliteration of the sinus pericranii. Histologic examination showed subcutaneous tissue with fibrosis and a benign meningothelial proliferation. The cells exhibited a pseudo infiltrative pattern of growth dissecting along the connective tissue planes. The meningothelial cell population was immunopositive for epithelial membrane antigen. The diagnosis of hamartoma of the scalp with ectopic meningothelial elements was made. This is a rare diagnosis, and to our knowledge only nineteen cases have been reported in literature. The current case represents the second report of an intracranial communication, and the first reported case with an associated sinus pericranii. The potential for misdiagnosis as a malignant lesion necessitates knowledge of this entity.

Keywords: Hamartoma; Ectopic Meningothelial; lesion; Benign meningothelial proliferation; Scalp

Introduction

Hamartoma of the scalp with ectopic meningothelial elements was first described by Suster and Rosai in 1990, and there are approximately nineteen cases reported in the literature.[1-4] These lesions are benign and are described as being hamartoma to us due to the presence of both benign meningothelial cells and connective tissue stroma, instead of a neoplastic proliferation of pure meningothelial cells which would be indicative of a meningioma.[3, 4] A literature review of 17 cases by Curran-Melendez et. al. demonstrated that these lesions most often occur on the posterior scalp in children less than 5 years of age, with at least three arising in adults, the oldest of which occurred in a 46 year-old. There is no gender predilection.[1]Additionally, there has been one other reported case, which was associated with a nevus sebaceous on the vertex of the scalp.[4]These cases are summarized in Table 1. Clinically, patients present most often with a solitary painless mass, although some may become painful, especially upon palpation. Macroscopic examination generally demonstrates a fixed to mobile, solid to cystic nodule, which may appear in a variety of colors from amelanotic to red. [1](Table 1)

Histologically, they are characterized by a haphazard mixture of connective tissue elements, small to medium-sized blood vessels, and a minor component of meningothelial cells forming anatomizing, slit-like channels creating a pseudoinfiltrative growth pattern reminiscent of angiosarcoma.[3] Treatment typically consists of complete resection, and following complete resection there have been no reports of recurrence or metastasis. [1, 5]

Complete resection of hamartoma of the scalp with ectopic meningothelial elements is considered curative. A single case of recurrence was reported in an incompletely excised lesion, which was later re-excised without recurrence. [6]The risk of recurrence is theoretically increased if an intracranial connection exists, however, only one case prior to our study has been associated with an intracranial communication, and there was no recurrence 27 months after resection. [1,5]The current case had a Mib-1 index of approximately 3%, raising the potential concern for local recurrence.

Although the pathogenesis of hamartoma of the scalp with ectopic meningothelial elements is poorly understood, a few theories have been proposed. One theory states that these lesions form from ectopic arachnoid cell rests displaced during embryologic development. Others postulate that the delayed closure of an extracranial communication could cause the extrusion of intracranial meningothelial contents with eventual atresia or pinching off of the communicating stalk.[1, 5] A connection between the intracranial elements and subcutaneous tissue, similar to a traditional meningocele, or ectopic arachnoid cell rests, would explain the presence of ectopic meningothelial cells in the scalp.

Table 1: Clinical Characteristics of Hamartoma of the Scalp with Ectopic Meningothelial Elements Reported in the Literature.

Case [citation]	Age	Sex	Size (cm)	Location	Intracranial Extension	Follow-up
1[10]	17 days	Female	0.5	Midline posterior scalp	No	Not documented
2[10]	1 month	Female	1.1 x 1.5	Occipital	No	Not documented
3[3]	4 months	Male	1.5	Occipital	No	No recurrence at 1 year
4[10]	4 months	Male	1.3×1.3	Midline posterior scalp	No	Not documented
5[11]	4 months	Male	6 x 7.5	Parietooccipital	No	No recurrence at 10 years
6[5]	5 months	Male	0.8×0.9	Posterior scalp	No	No recurrence at 1year
7[6]	6 months	Female	2	Posterior Scalp	No	No recurrence at 9 months
8[6]	6 months	Female	2.2	Posterior scalp	Fibrous cord extending from mass to intracranial compartment though small osseous defect/suture	No recurrence at 27 months
9[1]	9 months	Female	3.0 x 2.5	Parietal	No	No recurrence at 2 months
10[5]	15 months	Female	2 lesions; (a) 0.75×0.75 (b) 1.0×1.0	(a) Anterior parietal (b) Posterior occipital	No	No recurrence at 6 months
11[4]	17 months	Female	0.6	Vertex	No	Not documented
12[3]	2 years	Male	3.2	Parietooccipital	No, extended to pericranium	No recurrence at 7 years
13 [current case]	2 years	Male	1.5 x 0.6 x 1.1	Vertex	Yes, anomalous vascular connection to superior sagittal sinus through 8 mm calvarial defect	No recurrence at 10 months
14[2]	3 years	Female	1.2	Vertex	No	No recurrence but follow-up interval not documented
15[3]	3 years	Male	2.5	Occipital	No	No recurrence at 1 year
16[6]	5 years	Male	1.6	Posterior Scalp	No	Recurrence after incomplete excision, No years recurrence at 9 post re-excision
17[6]	5 years	Male	2.7	Posterior Scalp	No	Not documented
18[3]	20 years	Male	3.5	Occipital	No	No recurrence at 1.5 years
19[8]	28 years	Male	1.5	Forehead	No	Not documented
20[3]	46 years	Female	3	Occipital	No	No recurrence at 3 years

Case Report

An otherwise healthy two-year old male presented with a 1.1 cm, firm, subcutaneous scalp mass posterior to the vertex. The lesion was initially discovered at birth, had grown with the child's head, and recently became painful. Nothing else of note was discovered on physical exam.

Magnetic resonance imaging demonstrated a 15 x 6 x 11 mm T1 and T2 dark enhancing midline subcutaneous soft tissue lesion of the parietal scalp (Figure 1). This lesion was associated with an anomalous connection between the deep venous drainage and the superior sagittal sinus through an 8 mm calvarial defect, consistent with sinus pericranii. In addition, a persistent falcine sinus was identified (Figure 2). No other abnormalities were identified.

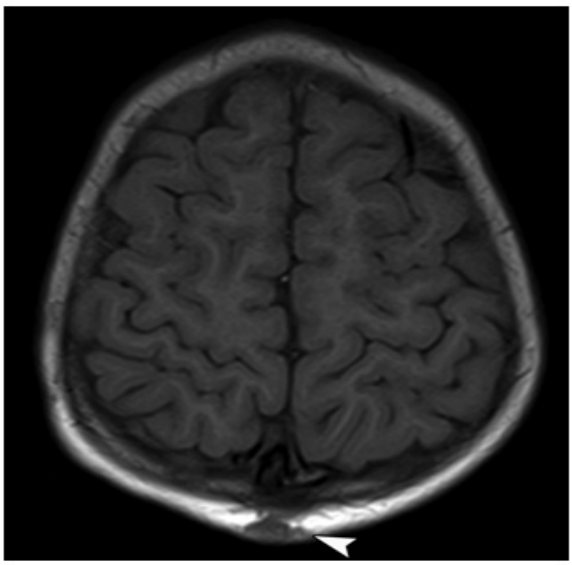

Figure 1: Axial T1 weighted MRI without contrast demonstrating a 15 x 6 x 11 mm midline subcutaneous soft tissue lesion (indicated by white arrowhead).

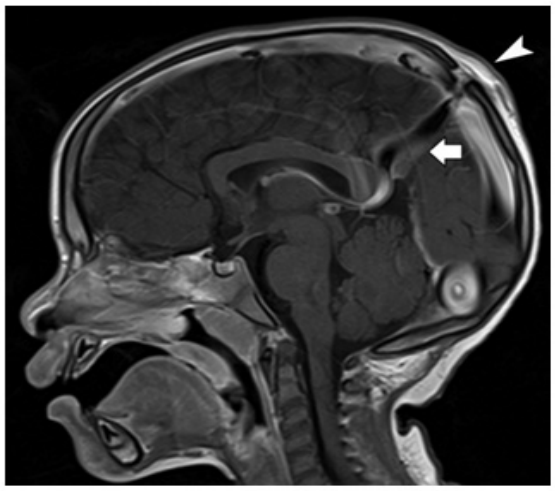

Figure 2: Sagittal T1 weighted MRI with contrast again demonstrating the 15 x 6 x 11 mm midline subcutaneous soft tissue lesion with an underlying calvarial parietal defect measuring 8 mm in diameter with a connection to the superior sagittal sinus (indicated by the white arrowhead). In addition, a persistent falcine sinus was identified (indicated by the white arrow).

The patient underwent surgery to remove the lesion and the sinus pericranii. The skin was sharply opened over the mass. Spreading and then sharp dissection was used to isolate the lesion transversing the skull. As fibrous bands were encountered, they were lysed. The lesion was isolated and the pulsatile large venous structure was visualized which was then tied off and the lesion removed. A pericranial graft was then hinged over the bony defect and sutured. There were no intraoperative complications. No decrease in cognitive abilities was noted postoperatively. Approximately one month post-surgery, he presented to the emergency department with acute swelling of the incision site with associated fever and was found to have a scalp abscess. He was then taken to the operating room for irrigation and debridement of the scalp abscess and cultures of the wound revealed group A Streptococcus pyogenes. He was treated with appropriate antibiotics and discharged home without further complications.

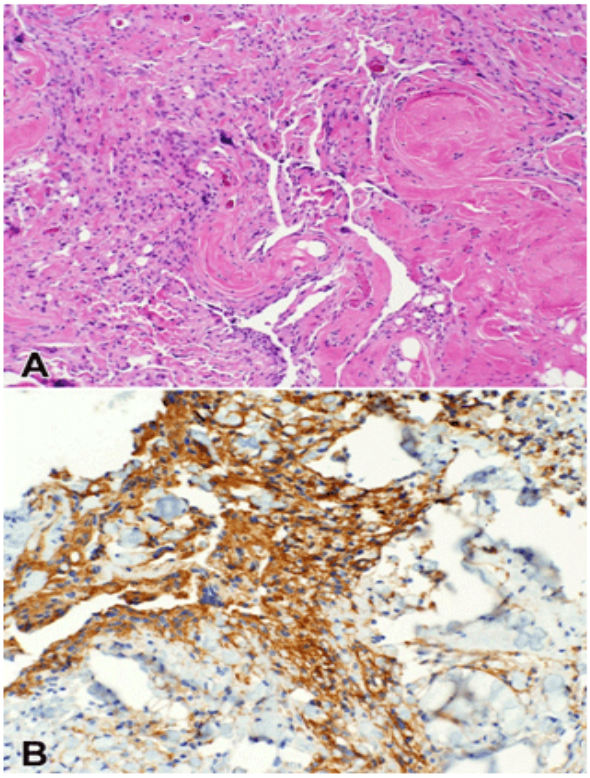

Figure 3: A. Hematoxylin and eosin (H&E) stained section of the lesion demonstrates dense collagen, vasculature, adipocytes, slit like spaces, and benign meningothelial cells (20x). **B.** An immunohistochemical stain for epithelial membrane antigen (EMA) highlights the meningothelial cells (20x).

Histologic examination showed subcutaneous tissue with fibrosis and a benign meningothelial proliferation. The cells exhibited a pseudoinfiltrative pattern of growth dissecting along the connective tissue planes in a pattern reminiscent of angiosarcoma (Figure 3A). Thermal artifact on the edge of the specimen was present and the vascular malformative appearance of sinus pericranii could not be confirmed histologically. The meningothelial cell population was immunopositive for EMA (Figure 3B). The Mib-1 proliferation-related index was approximately 3%. The diagnosis of hamartoma of the scalp with ectopic meningothelial elements was made. Ten months following the surgical procedure there is no evidence of recurrence.

Discussions

Hamartoma of the scalp with ectopic meningothelial elements are rare. Often these pediatric scalp lesions are clinically mistaken for dermoid/ epidermoid cysts or vascular lesions such as angiomas. [5] In a series of 65 pediatric patients with scalp lesions, dermoid/ epidermoid cysts were the second most common diagnosis and angiomas the sixth most common diagnosis. [7]

In the same case series, several malignant conditions including rhabdomyosarcoma, neuroblastoma, and angiosarcoma were also noted. [7] This is of particular importance in hamartoma of the scalp with ectopic meningothelial elements because the histologic appearance of pseudoinfiltrative growth of hyperchromatic, plump cells lining vascular-like channels overlaps with the appearance of angiosarcoma. However, angiosarcoma is rare in the pediatric population. In difficult cases these two entities may be separated by immunohistochemistry. The malignant cells of angiosarcoma are positive for Ulex europaeus and Factor VIII and the meningothelial cells of hamartoma of the scalp with ectopic meningothelial elements are positive for vimentin and epithelial membrane antigen (EMA). [3]

The current case not only represents a rare entity, but is also unique in that it is only the second reported case with an intracranial communication through an osseous calvarial defect. In the one other case, the lesion contained an intracranial connection via a fibrous cord through a small osseous defect. [1, 8]The current case is the only reported case associated with a sinus pericranii. Sinus pericranii is an anomalous vascular communication between the intracranial dural sinuses and extracranial vasculature which often presents as a soft tissue scalp mass in the pediatric population. [9] Prior to resection of the suspected lesion, it is imperative to image the mass and determine if a transosseous vascular communication is present. Although imaging can suggest a venous connection, surgical and histopathologic correlation is necessary for confirmation. [5, 7] In our case, the surgical finding of a large pulsatile venous connection to the lesion confirmed the imaging results. However, after surgical ablation of the suspected sinus pericranii, confirmation of its diagnostic histologic features was limited by thermal artifact.

Although rare, hamartoma of the scalp with ectopic meningothelial elements should be considered in the differential diagnosis of subcutaneous scalp lesions.

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References

1. Curran-Melendez SM, Dasher DA, Groben P, Stahr B, Burkhart CN, Morrell DS. Case report: Meningothelial hamartoma of the scalp in a 9-year-old child. Pediatr Dermatol. 2011; 28(6):677-680. DOI:10.1111/j.1525-1470.2011.01382.x
2. Cummings TJ, George TM, Fuchs HE, McLendon RE. The pathology of extracranial scalp and skull masses in young children. Clin Neuropathol. 2004; 23(1):34-43.
3. Suster S, Rosai J. Hamartoma of the scalp with ectopic meningothelial elements. A distinctive benign soft tissue lesion that may simulate angiosarcoma. Am J Surg Pathol. 1990; 14(1):1-11.
4. Crittenden SC, Sonnier GB. Meningothelial hamartoma associated with nevus sebaceus. Pediatr Dermatol. 2014; 31(2):208-211. DOI:10.1111/pde.12269
5. David LR, White WL, Jain AK, Argenta LC. Meningothelial hamartoma of the scalp: two case reports and literature review. European Journal of Plastic Surgery. 1996;19(3):156-9.
6. Marrogi AJ, Swanson PE, Kyriakos M, Wick MR. Rudimentary meningocele of the skin. Clinicopathologic features and differential diagnosis. J Cutan Pathol. 1991; 18(3):178-188.
7. Martinez-Lage JF, Capel A, Costa TR, Perez-Espejo MA, Poza M. The child with a mass on its head: diagnostic and surgical strategies. Childs Nerv Syst. 1992; 8(5):247-252.
8. Di Tommaso L, Fortunato C, Eusebi V. Meningothelial hamartoma located in the forehead. Virchows Arch. 2003;442(5):509-510. DOI:10.1007/s00428-003-0788-0
9. Bollar A, Allut AG, Prieto A, Gelabert M, Becerra E. Sinus pericranii: radiological and etiopathological considerations. Case report. J Neurosurg. 1992; 77(3):469-472. DOI:10.3171/jns.1992.77.3.0469
10. Herron MD, Coffin CM, Vanderhooft SL. Vascular stains and hair collar sign associated with congenital anomalies of the scalp. Pediatr Dermatol. 2005; 22(3):200-205. DOI:10.1111/j.1525-1470.2005.22304.x
11. Ferran M, Tribo MJ, Gonzalez-Rivero MA, Alameda F, Pujol RM. Congenital hamartoma of the scalp with meningothelial, sebaceus, muscular, and immature glandular components. Am J Dermatopathol. 2007; 29(6):568-572. DOI:10.1097/DAD.0b013e31815710ed