

Meningothelial hamartoma of the scalp

Taeki Kim¹,
Junhyung Kim¹,
Jaehoon Choi¹,
Sangho Oh¹,
Sunyoung Kwon²,
Woonhyeok Jeong¹

Departments of ¹Plastic and
Reconstructive Surgery and ²Pathology,
Dongsan Medical Center, Keimyung
University School of Medicine, Daegu,
Korea

Meningothelial hamartoma is a benign tumor composed of ectopic meningotheial elements in the dermis and subcutaneous tissue. It mainly occurs in the scalp; however, the incidence is extremely low. The origin of meningotheial hamartoma has not been elucidated; nevertheless, it has been theorized that it derives from ectopic meningotheial rests displaced during embryologic development. It can be diagnosed histologically as proliferation of connective tissue elements and cells arranged in solid nests, resembling vascular tumors. On immunohistochemistry, it stains positively for epithelial membrane antigen and vimentin. At least 17 cases have been reported, verifying the rarity of the lesion. We present the case of a 16-year-old male patient with a soft scalp mass which was thought to be a lipoma, but turned out to be a meningotheial hamartoma on histology.

Keywords: Hamartoma / Meningocele / Scalp / Skin neoplasm

INTRODUCTION

Soft tissue masses of the scalp are commonly encountered in clinical practice. Among these, lipoma is the most common benign tumor; only 13% of them are seen in the head and neck [1]. Nevertheless, the incidence of lipoma on the scalp comprises 30%, followed by that of trichilemmal cysts [2]. Meningotheial hamartoma is a rare benign tumor composed of ectopic meningotheial elements in the dermis and subcutaneous tissue. Since its first description by Suster and Rosai in 1990 [3], it has been reported in at least 17 cases [4,5]. It occurs mainly on the posterior scalp [4]. Although the incidence of a tumor arising on the scalp is increased compared to those occurring elsewhere on the skin, most are benign tumors [2].

We present a case of soft scalp mass that was thought to be a lipoma until diagnosis of meningotheial hamartoma on histology.

Correspondence: Woonhyeok Jeong

Department of Plastic and Reconstructive Surgery, Keimyung University School of Medicine, 1035 Dalgubeol-daero, Dalseo-gu, Daegu 42601, Korea
E-mail: psjeong0918@gmail.com

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CASE REPORT

A 16-year-old male presented to our department for evaluation of a soft, nontender, well-defined, 4-cm mass on the posterior scalp (Fig. 1). He did not have any mass-related symptoms except for sparse hair at the lesion. A computed tomography scan revealed a 4.7 × 3.6 cm fatty mass in the right parietal scalp without any abnormal high- or low-density lesion in the brain (Fig. 2). There was no intracranial connection and the cranium was intact. Surgical excision was planned because the clinical impression was a lipoma (Fig. 3).

Total excision was performed. A pale tan soft tissue mass was totally removed from the patient's scalp. There was no cranial involvement on visual inspection. Low-power field histologic examination (Fig. 4A) showed haphazardly arranged infiltrative epithelioid cell nests admixed with connective tissue components, adipose tissues and pseudovascular pattern within deep dermis. A high-power field observation revealed that these epithelioid cells had plump, large, oval, and vesicular nuclei (Fig. 4B). To further differentiate the diagnosis from malignant tumors such as angiosarcoma or melanoma, immunohistochemistry (IHC) was performed.



Fig. 1. Preoperative photograph of a mass on the posterior scalp.



Fig. 2. Preoperative computed tomography revealing bulging at the posterior scalp without cranial involvement.

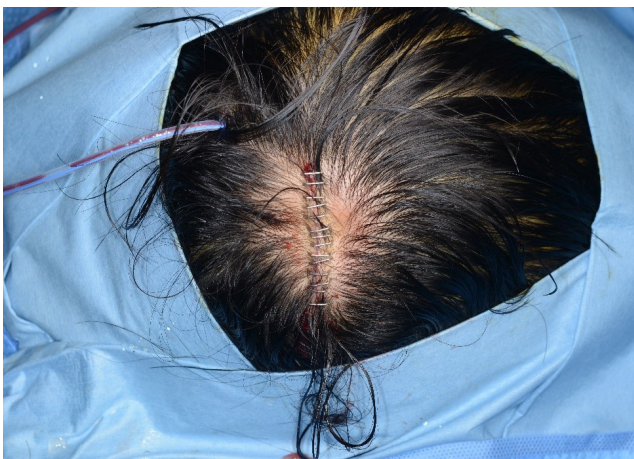


Fig. 3. Immediate postoperative photograph after excision.

IHC for HMB-45 and CK was negative and very low Ki-67 proliferation was noted so malignant melanocytic lesions could be excluded. Moreover, further stained IHC for factor VIII (to

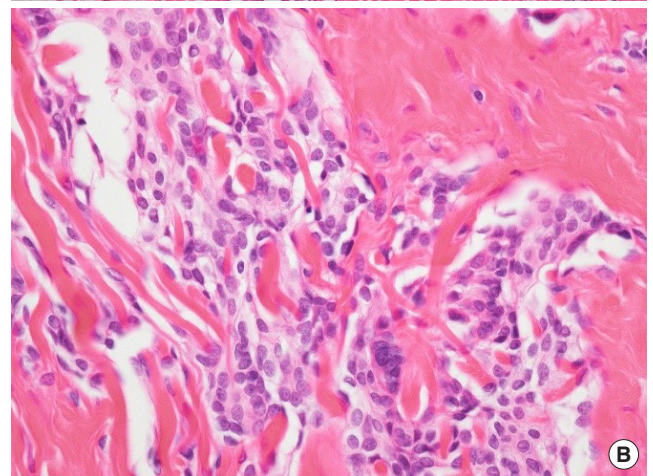
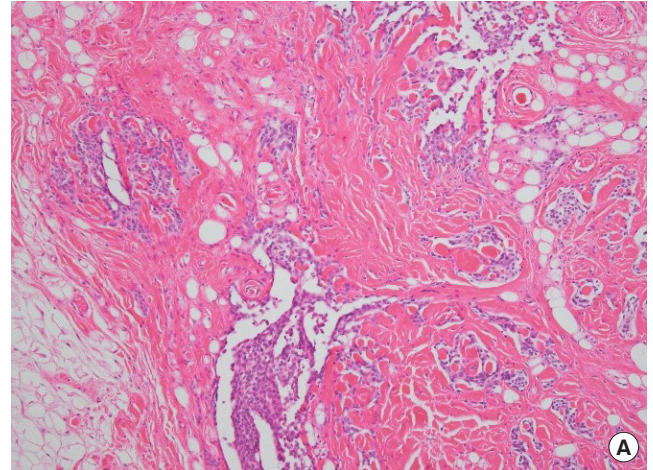


Fig. 4. Histologic examination. (A) Low-power magnification showing haphazardly arranged infiltrative epithelioid cell nests admixed with connective tissue components, adipose tissue, and a pseudovascular pattern within the deep dermis (H&E, $\times 100$). (B) High-power magnification showing plump, large, oval, and vesicular nuclei (H&E, $\times 400$).

screen for angiosarcoma) revealed negative. IHC for epithelial membrane antigen (EMA) (Fig. 5A) and vimentin (Fig. 5B) showed strong positivity in epithelioid cells and these histological findings were compatible with the diagnosis of meningothe-lial hamartoma. Three months after surgical excision, the patient showed no evidence of postoperative complications or recurrence.

DISCUSSION

We described a meningothe-lial hamartoma in a 16-year-old male patient. In the scalp, a remarkably broad range of tumor types may arise, including over a hundred types of neoplasm, hamartoma, malformation, and cysts, both benign and malignant. About 98%–99% of scalp tumors are benign; diagnosis and treatment of these lesions may pose significant challenges

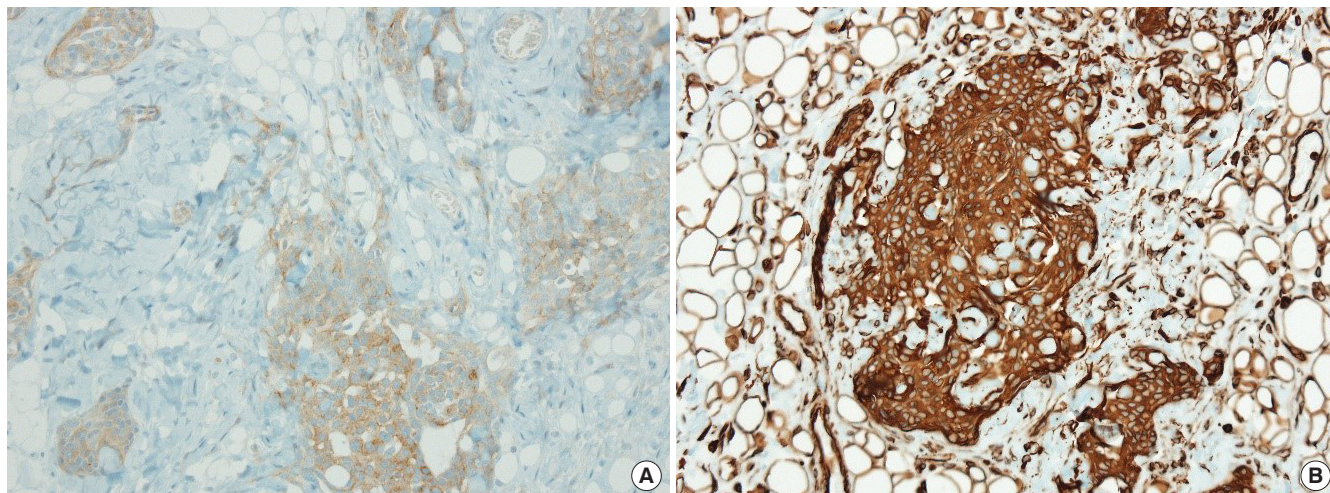


Fig. 5. Immunohistochemistry revealing strong positivity for (A) epithelial membrane antigen and (B) vimentin in meningothelial cell nests ($\times 200$).

because of anatomical circumstances and distinct histological features. Of all the benign tumors located on the scalp, lipoma accounts for 30% [2]. In the present case, the mass was a well-circumscribed, solitary lesion that resembled a lipoma. Furthermore, the results of initial imaging were suggestive of lipoma; it was difficult to suspect the meningothelial hamartoma before the histological study.

Meningothelial hamartoma was first described by Suster and Rosai in 1990 [3]. It is characterized by the admixture of abnormally-arranged various types of mature connective tissue elements, especially meningothelial elements in the dermis and tissue. Because of these characteristics, it is classified as a hamartoma. It is given various names, including meningothelial hamartoma, rudimentary meningocele, sequestered meningocele [6-11], and hamartoma of the scalp with ectopic meningothelial elements [3]. Most are solitary lesions, except for one reported case of multiple lesions in the scalp [12]. The most common location is in the occipital midline and posterior portion of the scalp. There has been one reported case in the parieto-occipital area and one in the vertex of skull [4]. The lesion has been reported once on the forehead [6]. It may have associated with alopecia as in our case [9,11]. In general, it does not extend to below the subcutaneous tissue; however, there were two reported lesions, one extending through the galea down to pericranium and another extending to intracranial compartment with a small osseous defect [3,6]. The treatment of lesions without extension is simple surgical excision.

Suster and Rosai [3] suggested that meningothelial hamartomas come from ectopic meningothelial rests, arachnoid cells displaced in the scalp during embryologic development. Some proposed that meningothelial hamartomas are part of a spec-

trum that encompasses primary cutaneous meningiomas, suggesting the abortive migration of cells from the neural crest [10]. Others suggested that it is form of meningocele with an obliterated intracranial communication [6]. There is controversy as to whether it is due to proliferation of meningothelial cells along the routes of the cutaneous nerve or whether it is classified as a meningocele under the name of rudimentary meningocele because of the identification of connections extending from the lesion to the dura [13]. There have been several theories such as these; nevertheless, the cause of meningothelial hamartoma has yet to be elucidated.

In the preoperative phase, meningothelial hamartoma is mostly indistinguishable from other common tumors of scalp such as lipoma. The patient merely complains of a nontender and soft mass on the scalp. The final diagnosis can be made after surgical removal and histopathological examination. Histologically, meningothelial hamartomas usually arise in the subcutaneous tissue and sometimes the dermis can be involved; however, the epidermis is histologically normal [5]. It is characterized by the proliferation of connective tissue elements and epithelioid cells with pseudoinfiltrative growth pattern. Occasionally, adipose tissues or smooth muscle fibers, or apocrine and eccrine glands can be found. Plump and cohesive epithelioid cells can test positive in IHC for EMA and vimentin and these findings indicate meningothelial origin. Differential diagnosis of meningothelial hamartoma was cutaneous angiosarcoma which shows thin-walled infiltrative vascular patterns; however, the pseudovascular pattern does not express IHC of endothelial markers such as factor VIII-related antigen. This histologic finding was noted in our case that revealed negativity of IHC for factor VIII-related antigen. Additionally, malignant

melanocytic tumor should be excluded using melanocytic markers such as S-100 and HMB-45.

In this case, the lesion was initially thought to be a lipoma, and was treated by surgical simple excision. It turned out to be a meningotheial hamartoma on histological examination. There are no differences between the two in the sense that complete excision is the best option for treatment. Nevertheless, if a surgeon encounters a meningotheial hamartoma in the clinic, understanding its characteristics and histological similarity to vascular neoplasm should enable them to make a differential diagnosis.

NOTES

Conflict of interest

No potential conflict of interest relevant to this article was reported.

Ethical approval

The study was approved by the Institutional Review Board of Keimyung University Dongsan Medical Center (IRB No. 2019-11-025) and performed in accordance with the principles of the Declaration of Helsinki. Written informed consent was obtained.

Patient consent

The patient provided written informed consent for the publication and the use of his images.

ORCID

Taeki Kim	https://orcid.org/0000-0002-8760-3546
Junhyung Kim	https://orcid.org/0000-0001-9077-2853
Jaehoon Choi	https://orcid.org/0000-0001-6180-1619
Sangho Oh	https://orcid.org/0000-0002-8739-908X
Sunyoung Kwon	https://orcid.org/0000-0002-8410-0185
Woonhyeok Jeong	https://orcid.org/0000-0002-0221-3347

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