Lumbosacral Meningial Hamartoma And Hypertrichosis: A Case Report

Hipertrikozisli Lumbosakral Meningial Hamartoma: Bir Olgu Sunumu

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Abstract: ***Objective:*** Ectopic meningiomas are rare tumors. In this paper, we describe a patient with lumbosacral meningeal hamartoma and hypertrichosis.
Methods: An 18-year-old female with a red-violet skin lesion on her lower back was determined spina bifida at L5-S1 by x-ray. Magnetic resonance imaging sequences showed a fibrous band arising from a posterior fusion defect at L5-S1. Light microscopy examination of the specimen were seen islets of meningothelial cells and calcified psammoma bodies spread among the vessels, neuronal structures, and dense collagenous tissue.
Result: We present a case lumbosacral ectopic hamartoma with hypertrichosis and a review of the nomenclature, classification and pathogenesis of this disorder.
Conclusion: Ectopic meningiomas are rare tumors in which etiopathogenesis is not clear. They are usually independent of intracranial meningiomas. These lesions characterize with clinical and pathological diagnostic difficulties. In the literature, these lesions have been given a lot of names like ectopic meningioma, meningial hamartoma, cutenous meningioma. Because this lesion did not consist solely of meningial elements, we suggested that the cases should be more accurately labeled as meningial hamartoma.

Key Words: Meningial hamartoma, hypertrichosis, lumbosacral region

Özet: ***Amaç:*** Ektopik meningiomalar nadir görülen tümörlerdir. Bu çalışmada hipertrikozisli, lumbosakral yerleşimli bir ektopik hamartoma olgusu sunulmuştur.
Metod: Sırtında kırmızı-mor cilt lezyonu olan 18 yaşında bayan hastada direct grafide L5-S1 de spina bifida saptandı. Magnetik rezorans incelemesinde L5-S1 deki posterior füzyon defektinden kaynaklanan fibröz bir bant görüldü. Lezyonun ışık mikroskopik incelemesinde damarlar, sinirler ve yoğun kollojen doku boyunca uzanan meningotelyal hücre adaları ve kalsifiye psammoma cisimleri görüldü.
Sonuç: Biz hipertrikozisli, lumbosakral yerleşimli bir ektopik meningeal hamartoma olgusu sunduk ve bu lezyonların isimlendirme, sınıflama ve patogenezini gözden geçirdik
Yorum: Ektopik meningiomalar etyopatogenezi tam olarak bilinmeyen, nadir görülen tümörlerdir. Genellikle meningiomalarla ilişkisiz olarak bulunurlar. Bu lezyonlar klinik ve patolojik tanı zorlukları içerirler. Kaynaklarda bu lezyonlara ektopik meningioma, meningial hamartoma, kutenöz meningioma gibi pekçok isim verilmektedir. Bu lezyonlar sadece meningial elementleri içermediği için biz meningial hamartoma olarak isimlendirmenin daha doğru olduğunu tavsiye etmekteyiz.

Anahtar kelimeler: Meningial hamartoma, hipertrikozis, lumbosakral bölge

INTRODUCTION

Meningioma is the most common neoplasm of the central nervous system (CNS). It is believed that these tumors arise from arachnoid cap cells (1-3). They are rarely encountered in regions other than the CNS. Ectopic meningiomas, which account for 1-2% of all meningiomas, most often develop in the scalp, the skin of other parts of the body, the subcutaneous tissue, the lungs, and the mediastinum. Those that arise in the skin or subcutaneous tissue have also been labeled cutaneous meningioma (1,3,6-9,16-18).

CASE REPORT

An 18-year-old female with a congenital red-violet skin lesion on her lower back that included a tuft of hair was admitted to the neurosurgery clinic at Adana Hospital. The patient's personal and family medical histories were unremarkable, and her neurological examination and routine laboratory tests were all normal. Physical examination revealed a 6x10 cm elliptical, circumscribed, red wine-colored lesion with localized hypertrichosis. The lesion was located on the midline of the lower lumbar region. There was no evidence of meningomyelocele. A lumbosacral x-ray demonstrated spina bifida at L5-S1. Magnetic resonance imaging sequences showed a fibrous band, arising from a posterior fusion defect at L5-S1. The band extended through the subcutaneous fat tissue in this area, and appeared to be connected to a nodular subcutaneous structure (figure 1). The subcutaneous nodular lesion was totally excised, with the surrounding skin and the associated fibrous "band," which extended as a tract that communicated with the spinal canal. The patient was discharged on the 3 rd postoperative day, and follow-up examination revealed no recurrence.

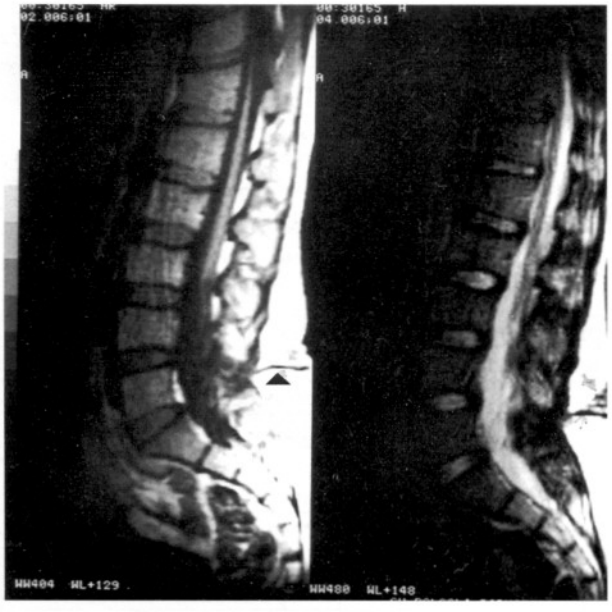

Figure 1: Magnetic resonance imaging shows the fibrous band arising from the posterior fusion defect at L5-S1.

The surgical specimen was 6x6x4 cm, and was composed of soft-tissue material covered by intact hairy skin. A firm mass of tissue with irregular borders was localized deep inside the specimen. Light microscopy examination showed areas of normal epidermis and dermis in the superficial parts of the specimen. The deeper areas contained islets of meningothelial cells and calcified psammoma bodies that spread among the vessels, neuronal structures, and dense collagenous tissue (figure 2 a-b). No pleomorphism, mitoses, or atypical cells were seen. On immunohistochemical study, the meningothelial cells

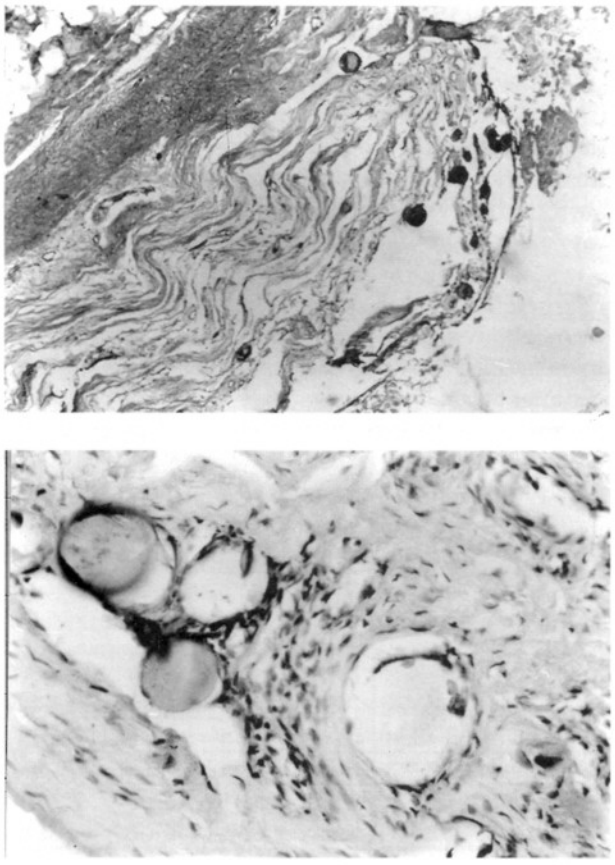

Figure 2 a: Islets of meningothelial cells and calcified psammoma bodies between vessel and neuronal structures in the subcutaneous tissue. (HEX40) 2 b: Group of meningothelial cells and psammoma bodies in higher magnification (HEX400)

stained positive for epithelial membrane antigen (EMA).

DISCUSSION

Ectopic meningiomas differ from intracranial meningiomas regarding their associated clinical and histopathological findings, and prognosis. In 1974, Lopez et al. reported a series of 25 cases that involved mostly the scalp, neck, and back, but only one patient had a lumbosacral cutaneous meningioma(7). These authors have developed a system for classifying cutaneous meningiomas in three different types (Table I). Type I is a congenital non-neoplastic lesion, and the other two types are forms of acquired neoplastic proliferation of meningothelial cells. Type I lesions, also called primary cutaneous meningioma (PCM), are typically misdiagnosed as alopecia, fibroma, or nevus in the clinical setting(7,14,18) PCMs have also been found in association with von Recklinghausen's disease, and a familial pattern has been reported(1,7,12,18)

It is suggested that PCMs arise from arachnoid cap cells in the subcutis or cutis as a result of a developmental defect. Light microscopy study of these tumors reveals masses of meningial cells that either surround the vessels or spread throughout a dense or myxoid stroma. One PCM subtype that Lopez and co-workers labeled as "rudimentary meningocele" is characterized with a cystic cavity obliterated by fibrocollagenous proliferation and arachnoid cell hyperplasia (7,11,13,14,16,18). Forty-seven rudimentary meningiocele cases had been reported in the English literature until 1994. Listed from highest to lowest frequency of occurrence, these lesions were noted in the scalp, vertebrae, forehead, and neck. In nine cases, the tumor was associated with a fibrous tract that communicated with the CNS. Six patients exhibited abnormalities of the bone underlying the lesion. The most common bone abnormality was spina bifida (1-18). Our patient's extracranial meningioma was located in the lumbosacral region, which is a rare site of occurrence. There was no cystic cavity, but the young woman had spina bifida and there was a fibrous tract extending from the tumor to the spinal canal.

Another variant of PCM, the "acoelic meningial hamartoma," has no cystic cavity, but hyperplastic meningial cells and psammoma bodies are observed within the surrounding tissue as untidy foci (5,7,11,16). Suster and Rosai claimed that the cases that as acoelic meningial hamartomas or rudimentary meningoceles in Lopez's classification should be labeled as "ectopic meningial hamartomas"(15). Their rationale for including the word hamartoma was the presence of a combination of meningial cells, vessels, adipose tissue, and other mesangial cells(5,13,15). In contrast to ectopic meningial hamartoma, the cellular composition of PCM is limited to meningial cells alone, but these tumors also contain many psammoma bodies. Theaker and colleagues reported six cases of ectopic meningioma characterized by meningothelial cells in the vicinity of small nerves, and called these lesions "cutaneous heterotropic meningial nodules" (17). Hirakawa et al. suggested that meningial hamartomas should be classified somewhere between meningocele and PCM (5).

The clinical and histopathological findings in our case were compatible with type I PCM; however, we suggest that, since this lesion did not consist solely of meningial elements, the label of meningial hamartoma would be more appropriate.

Regarding the localized hypertrichosis in our patient's lesion, we were able to find only one similar case in the literature. In this case reported by Penas

Table I: Classification of cutaneous meningiomas.

	Type I: Primary cutaneous meningioma	Type II: Ectopic meningioma of the soft tissue extending to the skin	Type III: CNS meningioma extending to the skin
Origin	congenital	acquired	acquired
Age of onset	childhood	adult	adult
Location	scalp, face, neck, paravertebral region	periorbital, perinasal, periauricular	
CNS meningioma	none	none	present
Behavior	benign	neoplastic	neoplastic
Histopathological features	meningothelial cell proliferation similar to meningioma	meningioma	meningioma

and associates, the tumor was located in the paravertebral region, but the patient did not have spina bifida and there was no communicating tract(13).

To establish the definitive diagnosis in ectopic meningioma, PCM must be histopathologically distinguished from perineuroma, glomus, schwannoma, and Spitz nevus (2,3,6,10,15). The immunohistochemical and ultrastructural features of ectopic meningiomas are different from these lesions. Like their intracranial counterparts, they also stain positive for EMA and vimentin and stain negative for cytokeratin, S-100 protein, desmin, neuron- specific enclose (NSE), chromogranin A. Also, this diagnostic staining property supports the theory that these tumors originate from arachnoidal cells (3,4,11,15). Glomus tumors tend to be more vascular in nature and they are immunohistochemically positive for chromogranin (6). Nevomelanocyctic and nerve sheat tumors (schwannoma, perineuroma) which are positive for S-100 protein can be distinguished by routine light microscopy and immunohistochemical methods (2). In our case in which a hair component was observed, Becker's nevus, familial hypertrichosis, and congenital spinal cord hypertrichosis must also be included in the differential diagnosis (13). Congenital localized hypertrichosis may be located away fom the spine and may be associated with other cutaneous abnormalities (13). Our patient has a single cutaneous lesion on her back and her familial history was negative for familial hypertrichosis.

The prognosis is good for both forms of PCM, and Lopez et al. reported no deaths in their ectopic meningioma type I cases during 20 years of follow-up (7). There is virtually no recurrence after appropriate surgical resection. In our patient, postoperative recovery was uneventful, and a follow-up examination at 24 months revealed no recurrence or metastases. This case is unusual because of the lumbosacral localization of the lesion, and the fact that the lesion exhibited hypertrichosis, and had a fibrous tract that communicated with the spinal canal.

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