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Case Report / *Olgu Sunumu*

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Primary extracranial meningioma of the lower limb

Alt ekstremitede primer ekstrakraniyal menenjiyoma

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This paper is about a primary extracranial meningioma case of a 40-year-old male patient who presented with a complaint of a rapidly growing painless stiff mass located in his left thigh. The magnetic resonance imaging examination revealed that the lesion was well-circumscribed and with isosignal intensity to muscle on T1-weighted images and with a relatively hiperintense on T2-weighted images. The histopathological analysis of the specimens both from the incisional biopsy and the excisional surgery were typical of meningioma showing spindle cell proliferation with a whirling pattern. Although extracranial meningiomas are very rare, it should be considered in the differential diagnosis of a spindle cell neoplasm.

Key words: Meningioma; neoplasm's; unknown primary.

Bu makalede sol uylukta hızlı büyüyen, ağrısız, sert kitle yakınması ile başvuran 40 yaşında bir erkek hastada tespit edilen primer ekstrakraniyal menenjiyom olgusu sunuldu. Manyetik rezonans görüntüleme incelemesinde kitlenin iyi sınırlı olduğu, T1 ağırlıklı görüntülerde kas ile isosignal yoğunlukla, T2 ağırlıklı görüntülerde ise oldukça hiperintens olduğu görüldü. İnsizyonel biyopsi ve eksizyonel cerrahi sonrası yapılan histopatolojik analizlerde kitleye yumaklar oluşturan iğsi hücrelerden oluşan tipik menenjiyom tanısı konuldu. Ekstrakraniyal yerleşimli menenjiyomlar nadir görülmelerine rağmen, iğsi hücre neoplazilerinin ayırıcı tanısında düşünülmelidir.

Anahtar sözcükler: Meningioma; neoplasms; unknown primary.

Unusual tumors in unusual locations have been reported in the literature.[1-3] Meningioma adjacent to bone in the thigh hasn't been reported yet.

Meningiomas are one of the most common primary intracranial tumors and their locally aggressive nature is well known. Metastasis of meningiomas is rarely reported in the literature. Even more rarely are primary extracranial or ectopically-located cases seen in the limbs. Despite their unusual localizations, their morphology and immunoprofile are similar to their intracranial counterparts.[4] In this report, we present a rare case of primary extracranial submuscular meningioma located in the thigh.

CASE REPORT

A forty-year-old male patient presented with a left lower extremity mass, which had grown rapidly over a six-month period. Physical examination revealed a nontender, well demarcated, immobile, approximately 10x5 cm solid mass located in the anteromedial side of the distal 1/3 of the left thigh. The roentgenograms of the left femur showed no abnormality on anteroposterior (AP) and lateral views. However, magnetic resonance imaging (MRI) revealed an 8x6x4 cm well-circumscribed mass with isosignal intensity on T1-weighted imaging (Figure 1) and high signal intensity on T2-weighted imaging (Figure 2). The tumor was non-homogeneously enhanced by gadolinium. Both axial and sagittal MRI depicted the tumor had no invasion of bone and was surrounded by the muscle mass.

An incisional biopsy of the tumor was performed and the specimen was histopathologically diagnosed as meningioma. After that, the patient underwent marginal excision of the tumoral mass, which was located beneath the vastus medialis muscle and

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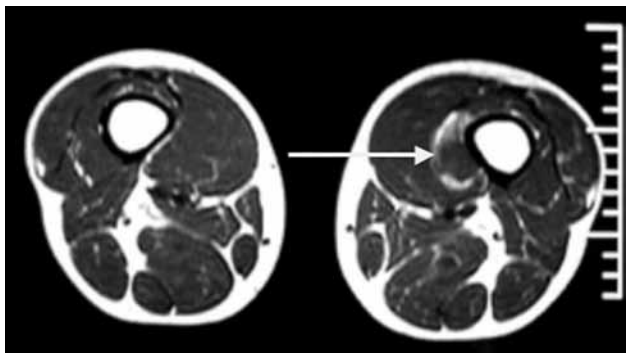

Figure 1. T_1 weighted magnetic resonance imaging of the tumoral mass (arrow).

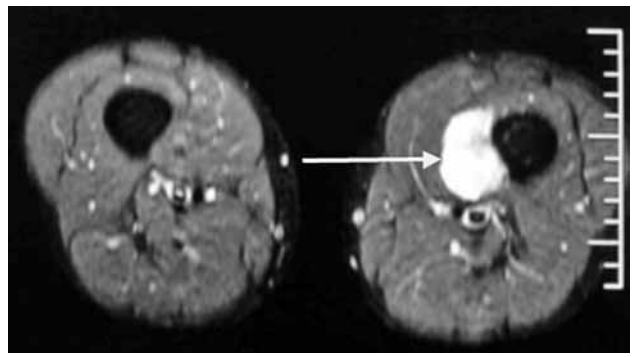

Figure 2. T_2 weighted magnetic resonance imaging of the tumoral mass (arrow).

adherent to the periosteum of the femur. The mass was surrounded by unclear and thin fibrous capsule. There was no association with peripheral nerves or blood vessels.

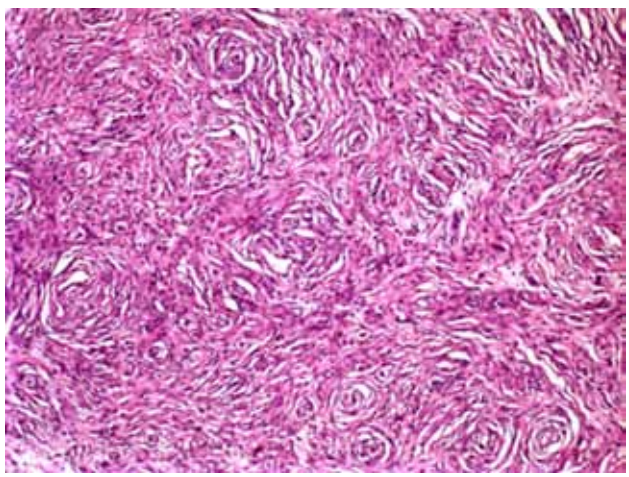

Figure 3. Tumor consist of whorls of plump epitheloid cells (H-E x 100).

Grossly, an encapsulated, nodular mass of 8x6x4 cm in diameter was observed. The cut surface of the tumor was solid, homogeneous dirty cream in color. Histologically, the tumor consisted of meningoepithelial and fibroblast like cells, which were arranged in sheets and whorls (Figure 3). There were no necrosis, mitotic figures and cytologic atypia. The tumor cells expressed antibodies for epithelial membrane antigen (EMA) (Figure 4) and vimentin (Figure 5), while no positivity was observed with the antibodies for CD34, CD117, desmin, actin, S100 and pan-cytokeratin. These findings were compatible with meningioma, World Health Organization (WHO) grade I.

After surgery and histopathological examination, full clinical work-up did not detect a primary or any other metastatic lesions including those of the central nervous system. The patient was discharged and mobilized with full weight-bearing. The patient has shown no signs of recurrence or metastasis during two-years of follow-up.

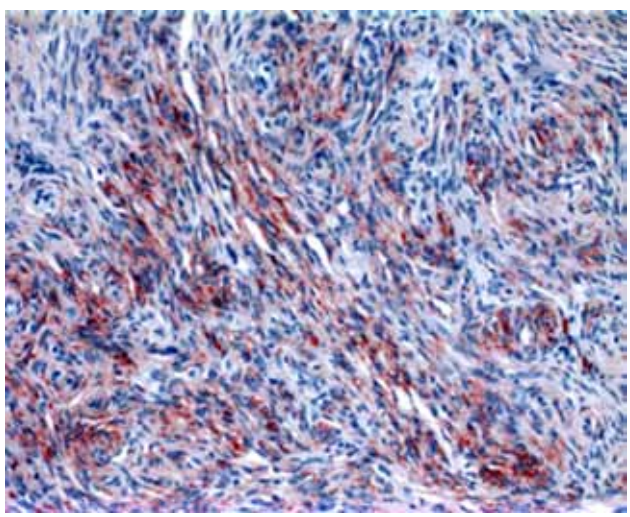

Figure 4. Immunohistochemically; EMA immunostaining of tumor cells (EMA x 200).

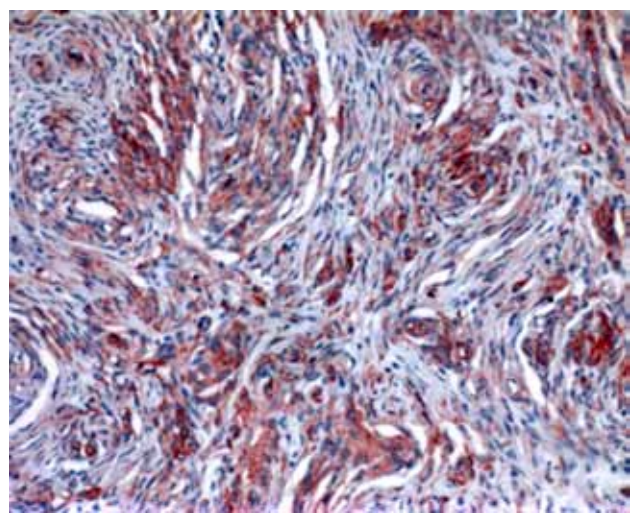

Figure 5. Immunohistochemically; vimentin immunostaining of tumor cells (vimentin x 200).

DISCUSSION

Meningiomas are well recognized and one of the most common tumors of the central nervous system. They arise from the arachnoid cells of the meninges. Most commonly, ectopic meningioma cases are seen as direct extensions from central nervous system meningiomas to head and neck areas via the optic nerve, but also rare cases arise in the lungs, liver, lymph nodes and bones by metastatic spread of malignant variants.[4]

In the literature only sporadic primary extracranial meningioma cases were reported. They were mostly at the head-neck region, with only a few cases being reported in limbs; such as in the little finger with calcification,[5] in the thigh adherent to hamstring muscle,[6] in the elbow joint,[7] in the forearm,[8] and medial plantar sole of the foot.[9] In all of these reports, the tumors were well-circumscribed and had no direct connection to neurovascular bundles, except in the elbow joint case presented by Anderson in which perineural tumor in the radial nerve showed aggressive behavior and recurred at the distal anatomic site after surgery.[7] Likewise, there were no connections between tumor and the neurovascular bundle, muscle or bone in our case. On the other hand, in our case; the diameter of the tumor which measured 8x6x4 cm was larger than the tumors which were reported previously in the literature.

Despite their unusual localizations, the immunoprofile and morphological characteristics of other reported ectopic meningiomas were the same as meningomas in the central nervous system. Immunohistochemical analysis demonstrated that intracranial meningiomas showed diffuse and strong positivity for EMA and vimentin[10] and positive reactivity for S-100 protein is only 28%.[11] On immunohistochemical profile of our case the spindled tumor cells showed diffuse and intense positivity for EMA and vimentin, and no reactivity for CD34, CD117, desmin, actin and S-100 antibodies.

When the anatomic localization and histological structure are envisaged in differential diagnosis it is essential to think of benign and low-grade malignant soft tissue tumors that have fusiform cell morphology such as schwannoma, perineurioma and monophasic fibrous synovial sarcoma.[9] Schwannoma has been distinguished from meningioma by diffuse and strong expressions with S-100 protein. Perineurioma is composed of only perineurial cells without residual axons and schwann cells and like meningioma, its tumor cells show positive staining for EMA and vimentin, and no reactivity for S-100 protein. However, tumor cells show more elongated bipolar cytoplasmic processes than meningothelial cells, arranged in intersecting fascicles or astoriform pattern. Monophasic fibrous sarcoma like meningioma, tumor cells show positive staining for EMA and keratin, but it is consisted by less spindle cells and non-distinguished cytoplasm.

There are many theories about the origin of primary extracranial meningiomas. It has been suggested that there might be some extradural trapping of arachnoid cells during embryogenesis or during peripheral nerve development. Recent studies have shown that the perineurium of the peripheral nerve indicates continuity with the arachnoid membrane, and perineural cells are functionally similar to arachnoid cells.[12] In the head and neck region these tumors are usually related to cranial nerves and might be derived from ectopic arachnoid tissue.[8] Nevertheless in spite of all these theories, the origin of primary meningiomas is controversial.

In conclusion, the primary ectopic meningioma is a very unusual neoplasm. It should be included in the differential diagnosis of soft-tissue spindle-cell tumors and identified by its characteristic morphology and immunohistologic profile.

Declaration of conflicting interests

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