Case Report / Olgu Sunumu

Extracranial Meningioma: A Case Report

Ekstrakraniyal Meningiom: Bir Olgu Sunumu

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Abstract

Extracranial meningiomas are rare tumors that occur in soft tissue or skin on the scalp and along the vertebral axis. The etiology of extracranial meningiomas is unknown, although four hypotheses have been postulated according to World Health Organization: (i) meningothelial cells carried along nerve sheaths as they exit the skull or vertebral column; (ii) ectopic arachnoidal cap cells; (iii) meningothelial cells displaced during trauma, and (iv) pluripotent mesenchymal cells capable of under going meningothelial differentiation or metaplasia. Extracranial meningiomas can be presented at all ages, with bimodal peaks occurring in the second decade of life and in the fifth to seventh decades. There is a slight female predominance.

Located swelling in the neck for almost a year, 46-year-old female patient was made mass excision. Macroscopically the mass was 1 cm in greatest dimension, nodular in appearance, cross-sectional face-cream-white color and solid. In microscopic examination, tumor, composed of large oval-round nucleated cells and hyaline stroma were observed. Tumor surrounding thin fibrous pseudocapsule and a plurality of peripheral nerve sections were observed around it. In the stroma psammom body like calcifications were observed. Tumor cells were positive for vimentin, S100, progesterone receptor, cytokeratin and focally epithelial membrane antigen; and negative for glial fibrillary acidic protein, and actin. Based on these findings the patient was diagnosed as extracranial meningioma. Slow growing and with very good prognosis of these tumors, surgical excision is the treatment of choice and do not require further treatment. In the differential diagnosis of soft tissue tumors, extracranial meningiomas should always be considered. It can be skipped because it is rare, so we have found the value to present this case.

Keywords
Extracranial meningioma, scalp, tumor

Anahtar Kelimeler
Ekstrakraniyal meningiom, skalp, tümör

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Öz

Ekstrakraniyal meningiomlar, vertebral aks boyunca, skalpte yumuşak doku ya da deride oluşan, nadir görülen tümörlerdir. Etiyolojisi çok iyi bilinmemesine rağmen patogenezlerinde Dünya Sağlık Örgütü'ne göre 4 hipotez öne sürülmektedir: (i) meningotelyal hücrelerin sinir kılıfları boyunca taşınarak kafatası ve vertebranın dışına çıkması; (ii) ektopik araknoid kap hücreleri; (iii) travma esnasında meningotelyal hücrelerin yer değiştirmesi; ve (iv) pluripotent mezenkimal hücrelerin meningotelyal farklılaşma ya da metaplazi göstermesi. Tüm yaşlarda görülebilmesine rağmen yaş dağılımı bimodaldir, 2. dekatta ve 5-7. dekatlar arasında daha sık görülür. Hafif bir kadın cinsiyet baskınlığı gösterir. Yaklaşık bir yıldır ense bölgesinde şişlik şikayeti bulunan 46 yaşındaki kadın hastaya kitle eksizyonu yapılmıştır. Kitlenin makroskopik değerlendirmesinde en büyük boyutu 1 cm olan, nodüler görünümde, kesit yüzü krem-beyaz renkli, solid, yumuşak doku materyali izlenmiştir. Mikroskopik incelemede ise arada lenfositik hücreler içeren hyalinize stromaya sahip, iri oval-yuvarlak nükleuslu hücrelerden oluşan tümör

izlenmiştir. Tümör çevresinde ince, fibröz bir psödokapsül ve bu kapsül çevresinde çok sayıda periferik sinir kesitleri gözlenmiştir. Tümöral alanlarda yer yer psammom cismi benzeri kalsifikasyonlar dikkat çekmiştir. Tümör hücreleri vimentin, S100, progesteron reseptörü, sitokeratin ve yer yer de epitel membran antijeni ile boyanmış, glial fibriler asidik protein ve aktin ile boyanmamıştır. Hastanın klinik olarak incelenmesinde intrakraniyal meningioma rastlanmamıştır. Bu bulgularla hastaya ekstrakraniyal meningiom tanısı konulmuştur. Yavaş büyüyen ve prognozları çok iyi olan bu tümörlerin tedavisinde cerrahi eksizyon tercih edilir ve ileri bir tedavi yöntemine ihtiyaç duyulmaz. Yumuşak doku tümörlerinde ayırıcı tanıya giderken ekstrakraniyal meningiom mutlaka düşünülmelidir. Ancak nadir görüldüğünden sıklıkla atlanır. Nadir görülen bir tümör olması sebebiyle biz bu olguyu sunulmaya değer bulduk.

Introduction

Meningiomas are second most common benign tumors of the central nervous system. 20% of intracranial meningiomas spread to extracranial areas (1). Primer extracranial meningiomas are rare tumors of scalp, soft tissue or skin along the vertebral axis. By definition, they are not associated with a meningioma of the underlying neuroaxons, and it should always be kept in mind that there may be an extracranial extension of an intracranial meningioma before it is accepted as a soft tissue or skin primary tumor. Primary extracranial meningiomas constitute less than 2% of all meningiomas (2,3). Two categories are examined: primary and secondary extracranial meningioma. Primary type is independent of intracranial meningioma. Secondary type is developed by direct release of the intracranial mass. The differential diagnosis includes various benign and malignant neoplasms, such as epithelial, neurogenic, vascular and mesenchymal tumors (4).

Case Report

A 46-year-old woman was admitted to the hospital for complaints of swelling in the nape region for approximately one year. Mass excision was applied. In the macroscopic evaluation of the mass, 1x0.6x0.4 cm nodular appearance, cross-section cream-white colored, solid, soft tissue material was observed. A well-defined nodular lesion was observed on microscopic examination. The cells forming the lesion were composed of solid islands and occasionally trabecularized in a hyalinized stroma. Lymphocytic cells were observed in the stroma between the islands. Psammom bodies were noticed in places (Figure 1). Tumor cells are large, oval-round nucleus and contain small nucleoli (Figure 2). A thin fibrous pseudocapsule around the lesion and numerous peripheral nerve sections around the capsule caught attention. Tumor cells were immunohistochemically stained by vimentin (Dako, Glostrup, Denmark), S100 (Dako, Glostrup, Denmark), progesterone receptor (Dako, Glostrup, Denmark), cytokeratin (Dako, Glostrup, Denmark), and occasionally epithelial membrane antigen (EMA) (Dako, Glostrup, Denmark). Glial fibrillary acidic protein (GFAP) (Dako, Glostrup, Denmark) and actin (Dako, Glostrup, Denmark) did not showed expression (Figures 3, 4). Ki-67 (Dako,

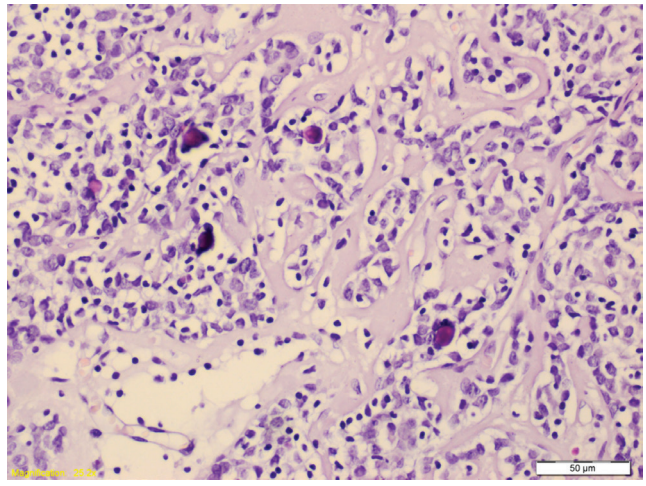

Figure 1. Psammom bodies among the tumoral islands (x400, hematoxylin and eosin)

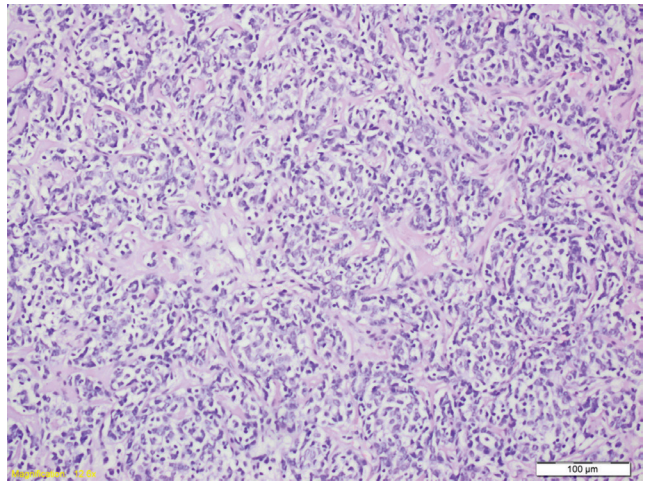

Figure 2. Tumor cells forming islands in the hyalinized stroma (x200, hematoxylin and eosin)

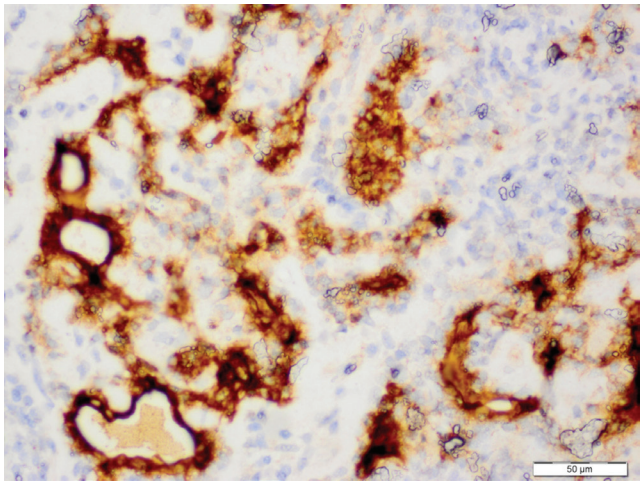

Figure 3. Tumor cells showing focal staining with EMA (x400, EMA)

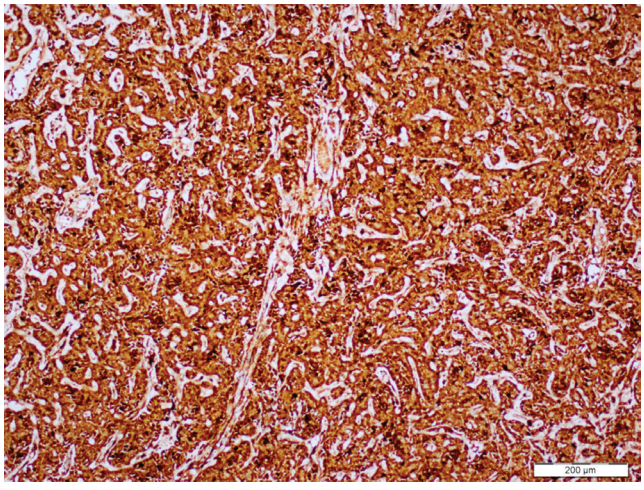

Figure 4. Tumor cells showing diffuse staining with S100 (x100, S100)

Glostrup, Denmark) was stained 2% of tumor cells. With these findings, the patient was diagnosed with extracranial meningioma.

Informed consent was not necessary because the case report was a retrospective study.

Discussion

Meningiomas are slow-growing, relatively common tumors arising from arachnoid cap cells of the meninges and constitute 13-18% of adult intracranial neoplasms. They are more frequent in women and appear more frequently in middle age (5). They are divided into 3 types according to their malignancy degree: benign [World Health Organization (WHO) grade 1], atypical (WHO grade 2), and anaplastic (WHO grade 3). Approximately 80% of meningiomas are benign. The most common histologic variants are meningothelial, fibrous, and transitional (1). Extracranial meningiomas are less common and constitute less than 2% of all meningiomas. Although it can be seen at all ages, the age distribution is bimodal, they are more common in 2th and 5th-7th decades. There is a slight female gender dominance. They occur more frequently in the vertebrae, neck, thorax, shoulder and peritoneum (3). In many cases, orbita is shown as the most common region, but tumors that have no connection with the optic nerve should be considered extracranial. Meningiomas with intracranial and intraspinal components should be excluded by definition. For example, en plaque meningiomas are usually invasive to the skull, and these tumors should not be evaluated as pure intraosseous. Although the etiology is not well known, 4 hypotheses are suggested in pathogenesis according to the WHO: (i) meningothelial cells moving out of the skull and vertebra along the nerve sheaths; (ii) ectopic arachnoid cap cells; (iii) displacement of meningothelial cells during trauma; and (iv) meningothelial differentiation or metaplasia of pluripotent mesenchymal cells. Tumor-related symptoms are associated with tumor size, location, and growth rate. They are usually seen as painless, slow growing masses.

Although macroscopically like intracranial meningiomas, the spread to the surrounding tissue is more common in these tumors. The color and consistency properties vary depending on the cellularity, collagen accumulation, and tumor grade (6).

Histologically, these tumors are indistinguishable from normal intracranial meningiomas (7). In the literature, extracranial meningiomas are divided into various histological types and grades, such as those with intracranial location (5,8,9). Solid meningiothelial cell nests are layered or coiled and occasionally contain psammom bodies. Generally, the tumors are irregular and composed of neoplastic, epithelioid cell lobules and folds. The nuclei are usually round-oval, contains thin nuclear chromatin and occasionally intranuclear pseudoinclusions. Although intranuclear inclusions are not always obvious and can not be easily detected, they are present in 71% of tumors (5). There are various hypotheses about the formation of psammom bodies. They are thought to be derived

from endothelial cells of obliterated vessels or may be formed by the secretion of arachnoid vessel cells and the calcification of necrotic cells (10).

Immunoprofiles of extracranial meningiomas are the same as meningiomas located in the central nervous system. They all express EMA and vimentin, while some express S100 protein (28%). Progesterone receptor positivity is also frequently seen (6). GFAP is negative. The Ki-67 index is below 5% and supports the slow progress of this clinical course (5,7-9).

Clinically, the differential diagnosis is extensive and includes nevi, fibroma, glioma, hemangioma, lipomas and many other benign or epithelial tumors and many malignant entities such as neurogenic tumors, melanoma, olfactory neuroblastoma, vascular tumors, mesenchymal tumors (4). Another entity that needs attention in differential diagnosis is psammomatous melanocytic schwannoma. This entity is seen as a finding of the Carney complex. For this reason, the patient needs to be investigated for myxoma, pigmentation or endocrine disorders that might suggest the Carney complex. These two tumors can be separated based on their histopathological characteristics. Extracranial meningioma does not have melanin pigment as well as areas of Antoni A and Antoni B expected to be seen in psammomatous melanocytic schwannoma. Careful microscopic examination and selection of appropriate immunohistochemical stain helps to confirm the diagnosis when cutaneous meningiomas is in the differential diagnosis. Nevertheless, more frequent tumors should be considered in differential diagnosis and the diagnosis should be based on histopathologic examination (7,8).

Depending on tumor grade and resection width, the prognosis of these slow-growing tumors is generally excellent (1,6). It is more likely to recur in intraosseous areas. Surgical excision provides complete cure and no further treatment is required. Distant metastases have been reported in only 6% of cases, frequently in anaplastic variants (6). It is important to note that these tumors have the potential to appear in places we have never expected. This, in turn, removes the potential difficulties associated with diagnosis and treatment management (5,8). As a result, extracranial meningiomas are tumors that cause diagnostic difficulties from time to time because of their abundant entity in their differential diagnosis. However, the prognosis is very good because of the slow growth and complete cure with complete surgical excision.

Ethics

Informed Consent: Informed consent was not necessary because the case report was a retrospective study.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: C.T., Ö.E.Ö., Concept: Ö.E.Ö., C.T., Design: Ö.E.Ö., Data Collection or Processing: Ö.E.Ö., C.T., Analysis or Interpretation: Ö.E.Ö., S.G., T.Ö., A.G.Ö., Literature Search: Ö.E.Ö. Writing: Ö.E.Ö., S.G., T.Ö., A.G.Ö.

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