Case Report (Olgu Sunumu)

Secretory Meningioma: A Rare Entity That Can Be Confused Radiologically and Pathologically with Malignant Tumors

Sekretuar Meningiom: Radyolojik ve Patolojik Olarak Malign Tümörlerle Karıştırılabilen Nadir Bir Antite

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Özet

Meningiomlar, araknoidin meningotelyal hücrelerinden kaynaklanan, yavaş büyüyen, ekstra aksiyal beyin tümörleridir. Meningiomlar en sık görülen beyin tümörleridir, ancak sekretuar meningiomlar son derece nadir bir varyant olup radyolojik ve patolojik olarak kötü huylu tümörlerle karıştırılabilir. Kırk sekiz yaşında erkek hasta sol gözde görme bozukluğu ve baş ağrısı şikayeti ile başvurdu. Manyetik rezonans görüntülemede sol pontoserebellar bölgede 57x48x30 mm boyutlarında ekstra aksiyal kitle lezyonu ve kitle çevresinde belirgin ödem görüldü. Mikroskobik olarak, oval-yuvarlak nükleuslu, ince veziküle kromatinli ve orta derecede eozinofilik sitoplazmalı uniform meningotelyal hücrelerden oluşan tümöral lezyon gözlendi. Tümör hücreleri arasında eozinofilik sekresyon içeren lümenlerin oluşturduğu psödopsammom yapılarıyla karakterize epitel farklılaşmasının varlığı belirlendi. Lümenlerdeki eozinofilik sekresyon histokimyasal periyodik asit schiff (PAS) boyası ile gösterildi. Karsinoembriyonik antijen (CEA), epitelyal membran antijen (EMA) ve pan sitokeratin (Pan-CK) ile psödopsammom yapılarında ve çevreleyen tümör hücrelerinde immün reaksiyon gözlendi. Tümör hücreleri, Vimentin ve progesteron reseptörü (PR) ile immünopozitifti. Ki67 proliferasyon indeksi <%1 olarak belirlendi. Nekroz veya mitoz saptanmadı. Tümör "sekretuar meningiom" olarak rapor edildi. Olguya ek tedavi uygulanmadı ve cerrahi rezeksiyon sonrası 32 aylık takipte nüks saptanmadı.

Sekretuar meningiomlar benign seyri nedeniyle klinik olarak önemli olmamakla birlikte radyolojik olarak belirgin beyin ödemi, serum CEA düzeylerinin yükselmesi ve epitel farklılaşması nedeniyle malign tümörlerle karıştırılabildiklerinden ayırıcı tanı önemlidir.

Anahtar kelimeler: Beyin, Beyin ödemi, Meningiom, Sekretuar meningiom

Abstract

Meningiomas are slow growing, extra axial brain tumors that originating from the meningothelial cells of the arachnoid. Meningiomas are the most common brain tumors, however secretory meningiomas are extremely rare variant and can be confused radiologically and pathologically with malignant tumors.

A 48 years old male was presented with a complaint of headache and visual impairment in the left eye. Magnetic resonance imaging revealed an extra axial mass lesion in the left pontocerebellar region, 57x48x30 mm in size, and marked edema around the mass. Microscopically, a tumoral lesion consisting of uniform meningothelial cells with oval-round nuclei, thin vesiculated chromatin, and moderately eosinophilic cytoplasm was observed. The presence of epithelial differentiation characterized by pseudopsammom structures formed by lumens containing eosinophilic secretion was appointed among the tumor cells. Eosinophilic secretion within the lumens were highlighted with histochemical periodic acid schiff (PAS) stain. Immunoreaction was observed in the pseudopsammom structures and the surrounding tumor cells with carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA) and pan cytokeratin (Pan-CK). Tumor cells were immunopositive with Vimentin and progesterone receptor (PR). Ki67 proliferation index was determined as <1%. No necrosis or mitosis were detected. The tumor was reported as ''secretory meningioma''. No additional treatment was applied to the case and no recurrence was detected in 32 month follow-up after surgical resection.

Although secretory meningiomas are not clinically important due to their benign course, differantial diagnosis is critical, since they can be confused with malign tumors because of marked brain edema radiologically, elevated serum CEA levels and epithelial differentiation.

Keywords: Brain, Brain edema, Meningioma, Secretory meningioma

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INTRODUCTION

Meningiomas are slow growing, extra axial brain tumors that originating from the meningothelial cells of the arachnoid. Most of them are grade 1 tumors with low risk of recurrence and benign course. Meningiomas are the most common brain tumors, and 36% of brain tumors are meningiomas while the probability of developing meningioma for whole life is 1% (1). However secretory meningiomas are extremely rare variant and its frequency among the meningiomas range 1.2-9.3% (2,3). They are more common in women and are most commonly reported in the frontal convexity and sphenoid ridge (4).

Secretory meningiomas have unique radiological, histopathological, and immunohistochemical features. Although they are grade 1 meningiomas, they can be exhibit marked brain edema radiologically, that is a common feature of aggressive high grade intraparenchymal brain tumors. Pathologically they are characterized by epithelial differentiation, which appears as intracellular lumens containing eosinophilic, PAS positive secretions that called pseudopsammom bodies (5).

CASE REPORT

A 48 years old male was presented with a complaint of headache and visual impairment in the left eye. Magnetic resonance imaging revealed an extra axial mass lesion in the left pontocerebellar region, 57x48x30 mm in size, and marked edema around the mass. The mass was excised.

On macroscopic examination of the excision material, fragmented pieces of 5x5x2.5 cm in size, with white in colour, and elastic consistency were seen. Microscopically, a tumoral lesion consisting of uniform meningothelial cells with oval-round nuclei, thin vesiculated chromatin, and moderately eosinophilic cytoplasm was observed. The presence of epithelial differentiation characterized by pseudopsammom structures formed by lumens containing eosinophilic secretion was appointed among the tumor cells that were forming whorls in some areas (**Figure 1**). Eosinophilic secretion within the lumens were highlighted with histochemical PAS stain (**Figure 2**). Immunoreaction was observed in the pseudopsammom structures and the surrounding tumor cells with CEA, EMA and Pan-CK (**Figure 3** and **4**). Tumor cells were immunopositive with Vimentin and PR (**Figure 5**). Ki67 proliferation index was determined as <1% (Figure 6). No increased cellularity, small cell changes, prominent nucleolus, sheeting, necrosis or mitosis were detected. Rare mast cells were also observed among the tumor cells. In addition, microcalcification was observed.

The tumor was reported as "secretory meningioma, grade 1". No additional treatment was applied to the case and no recurrence was detected in 32 month follow-up after surgical resection.

A written consent was obtained from the patient for the publication of this case report and accompanying images.

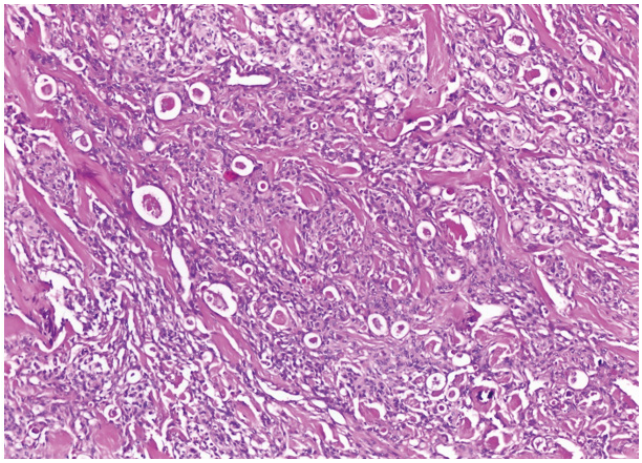

Figure 1. Microscopic view of secretory meningioma: Tumoral lesion consisting of uniform meningothelial cells with oval-round nuclei and thin chromatin, and epithelial differentiation characterized by pseudopsammom structures formed by lumens containing eosinophilic secretion. Haematoxylin Eosin (HE) X200

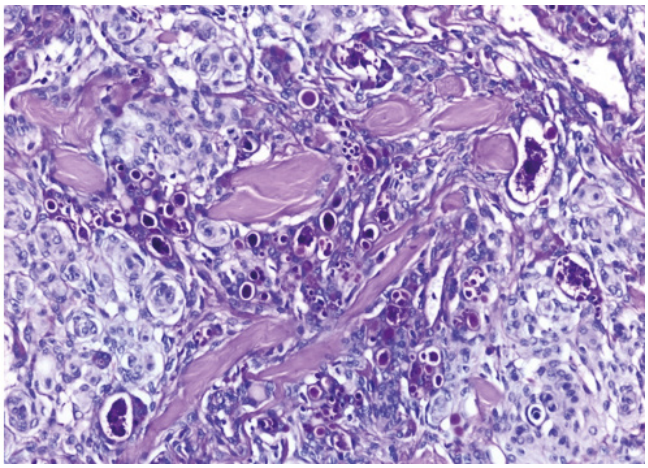

Figure 2. Histochemical PAS positivity in secretion within the lumens. PASX400

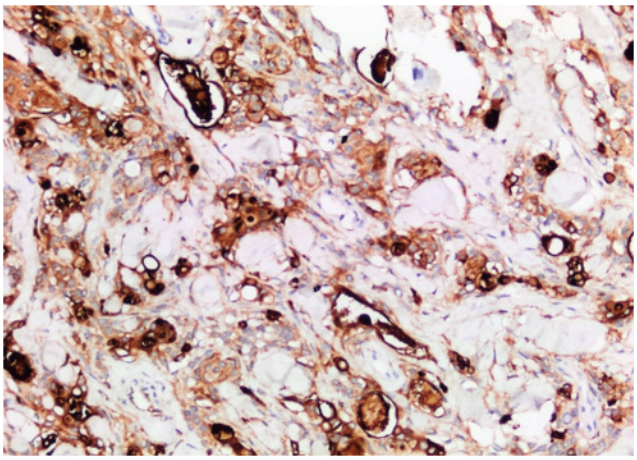

Figure 3. Immunohistochemical EMA positivity in tumor cells. EMAX400

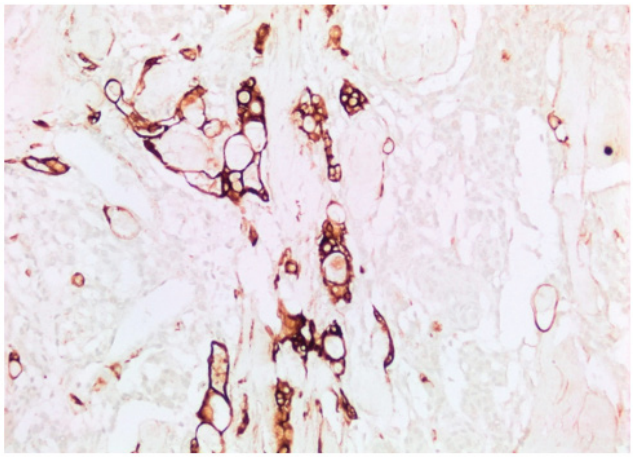

Figure 4. Immunohistochemical Pan-CK positivity in the pseudopsammoma structures and the surrounding tumor cells. Pan-CK X200

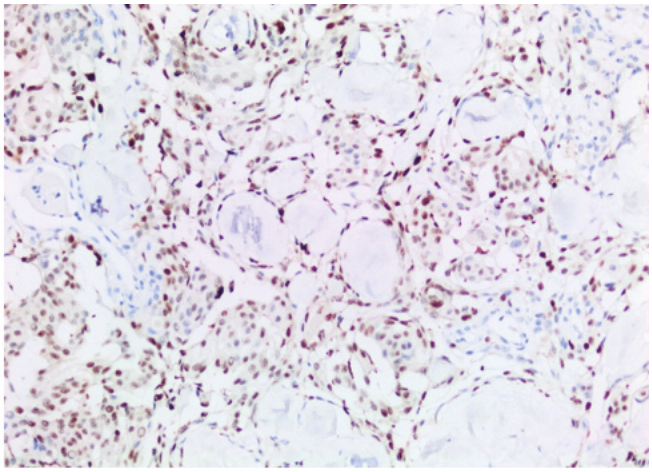

Figure 5. Immunohistochemical PR positivity in tumor cells. PRX400

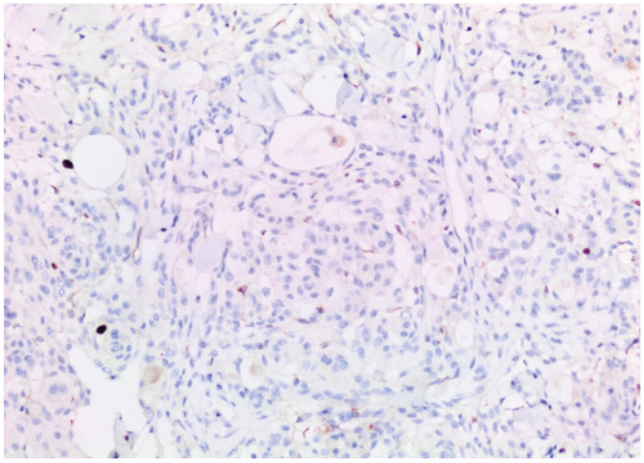

Figure 6. Nuclear staining in a small number of cells with immunohistochemical Ki67. Ki67x400

DISCUSSION

A combination of KLF4 and TRAF7 mutation is characteristic in secretory meningiomas. While TRAF7 mutation is observed also in other meningiomas, KLF4 mutation is not. This situation indicates that the KLF4 mutation plays a special role in the development of secretory menimgiomas, and also introduces that the KLF4 mutation can be used as a specific marker in the diagnosis of secretory meningiomas (6).

Secretory meningiomas consist mainly of meningothelial cells. Tumor cells have oval or round nuclei, eosinophilic cytoplasm, indistinct cell borders and they don't exhibit cellular atypia. Scattered whorl formations can be observed. Psammoma bodies which are round concentric calcium accumulations are absent or scarce. On the other hand the presence of pseudopsammoma structure is characteristic for secretory meningiomas. Pseudopammoma structures are intracytoplasmic inclusions that are round hyaline structures varying in size, and are homogeneous and bright eosinophilic or pale eosinophilic with hematoxylin-eosin stain (7). Characteristically, inclusions are PAS positive and diastase resistant. In addition, hyaline bodies and surrounding tumor cells exhibit immunohistochemical CEA, EMA, Alpha 1-antitrypsin, IgA, IgG and IgM expression (8). In these cases, serum CEA levels are also elevated and decrease rapidly after surgical resection of the tumor (7). In our case, pseudopsammom structures were noticeable and histochemical PAS staining was detected in these structures. Immunoreaction with Pan-CK, EMA and CEA was observed in pseudopsammom structures and surrounding tumor cells. In secretory meningiomas,

Ki67 proliferation index is not high and varies between 0 and 4% (9). Serum CEA level was not evaluated preoperatively in our case and Ki67 proliferation index was determined as <1%. As with other meningiomas, PR immunoreactivity is observed in secretory meningiomas and PR positivity has been reported in relation to good prognosis. In contrast, p53 positivity is thought to indicate the probability of recurrence (10).

The presence of varying amounts of mast cells in secretory meningiomas is remarkable. It has been suggested that mast cells are an important source of vascular endothelial growth factor and serotonin in secretory meningiomas and these mediators play a part in the causes of vasogenic brain edema in secretory meningioma (11). Edema observed in secretory meningiomas causes confusing the tumor radiologically with malignant tumors as well as increasing intracranial pressure and causing various neurological complications in the postoperative period (12).

Although secretory meningiomas are not clinically important due to their benign course, pathological differential diagnosis is critical. They can confuse with carcinoma metastases due to their epithelial differentiation. Since prognosis and management of these entities are so different, carcinoma metastasis must be excluded before the diagnosis of secretory meningioma. It should also be borne in mind that it can also be confused with microcystic meningiomas.

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Author Contribution Rate Statement Summary: The authors declare that, they have contributed equally to the manuscript.

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