Signet Ring Stromal Tumor of Ovary

Overin Taşlı Yüzük Hücreli Stromal Tümörü

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ABSTRACT

Signet-ring stromal tumor (SRST) is a rare benign ovarian neoplasm with only 10 reported cases in the literature. We report a case of signet-ring stromal tumor of the right ovary in a 43- year-old woman who presented with abdominal discomfort. Histologically, the tumor was composed of an admixture of spindle and round cells containing a large cytoplasmic vacuole which displaced the nucleus, creating a signed-ring appearance. Immunohistochemically, the tumor cells showed positivity for wimentin and negativity for PANCK, CK7, CK20. *(Gazi Med J 2012; 23: 167-9)*

Key Words: Signet-ring ovarian tumor, Krukenberg tumor, vimentin

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ÖZET

Overin taşlı yüzük hücreli tümörü, literatürde 10 kadar yayınlanmış vaka bulunan, oldukça nadir görülen benign bir tümördür. Biz abdominal ağrı şikayeti ile başvuran 43 yaşındaki hastanın sağ overinde tespit edilen overin taşlı yüzük hücreli tümörü olgusunu raporladık. Histolojik olarak tümör büyük sitoplazmik vakuol ile nükleusu kanara iten iğsi veya yuvarlak atipik epitelyal hücrelerden oluşmaktaydı. Tümör hücreleri immünhistokimyasal olarak vimentin ile pozitif, PANCK, CK7 ve CK20 ile negatif bulundu. *(Gazi Med J 2012; 23: 167-9)*

Anahtar Sözcükler: Taşlı yüzük hücreli overyan tümör, Krukenberg tümörü, vimentin

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INTRODUCTION

Signet-ring stromal tumor (SRST) is a very rare benign ovarian tumor. Only 10 cases have been reported in the literature (1). Microscopically, this ovarian neoplasm is characterized by a proliferation of stromal spindle-shaped cells merged with rounded cells containing eccentric nuclei and single vacuoles, resembling signet-ring cells. In this article, we report a case of a signet-ring stromal tumor of the right ovary in a 43 year-old woman and emphasize the histopathologic, histochemical and immumhistochemical characteristics that differentiate it from a Krukenberg tumor.

This case was presented at 20th National Pathology Congress, 29th September-3rd October, 2010

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CASE REPORT

A 43 year-old, nulligravida woman presented with a 4-month history of low abdominal discomfort. There was no relevant medical or family history. In the pelvic examination a palpable mass was located in the right hemiabdomen. Pelvic ultrasound examination confirmed a normal-sized uterus and a semisolid right ovarian mass measuring about 5 cm at its largest dimension. Serum tumor markers including CA-125, CA 15.3, CA 19.9 and CEA were all within the normal range. The patient underwent laparotomy, and a 5 cm sized solid tumor in the right ovary was observed.

Pathologic Findings

The ovarian tumor was solid and measured 5x3.5x3 cm. Its outer surface was smooth. Its cut surface was yellowish soft-to-firm solid. Cystic, hemorrhagic or necrotic areas were not seen. Microscopic examination revealed a circumscribed but unencapsulated tumor surrounded by a rim of normal ovarian stroma of varying thickness (Figure 1). The tumor revealed an intermixed arrangement of rounded cells elongated in fascicles containing eccentric nuclei and single large vacuoles resembling signet-ring cells (Figure 2). Very rare mitotic figures were seen. Clear vacuoles were negative for PAS with diastase digestion. Reticulin stain showed a delicate network of fibers investing signet-ring cells as well as the surrounding fibrous stromal cells. Necrosis and desmoplasia were absent. None of the tumors formed epithelial structures, such as glands , nests or cords. There was no intra and extracellular hyaline globules.

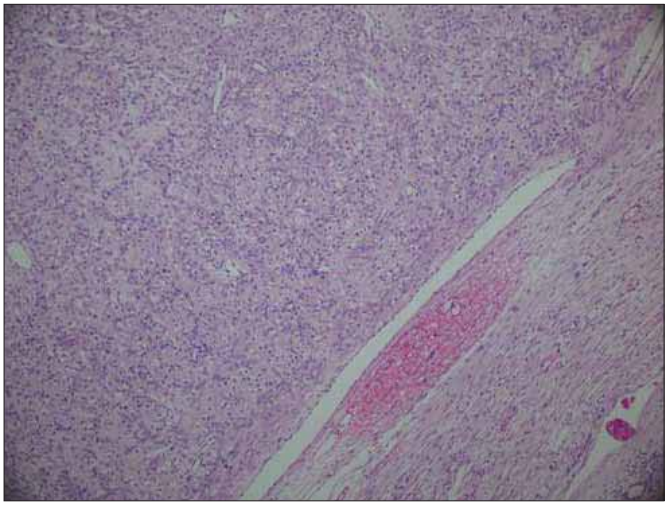

Figure 1. Circumscribed but unencapsulated tumor surrounded by a rim of normal ovarian stroma of varying thicknes

Immunhistochemical Analysis

Immunhistochemically, the tumor cells showed diffuse immunreactivity for vimentin (Figure 3) and focal staining for calretinin (Figure 4) except in their vacuoles . The signet-ring cells were negative

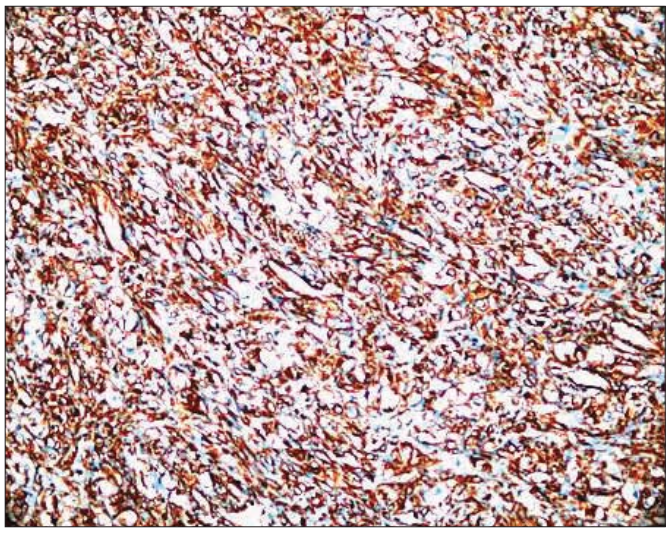

Figure 3. Tumor cells showed diffuse immunreactivity for vimentin

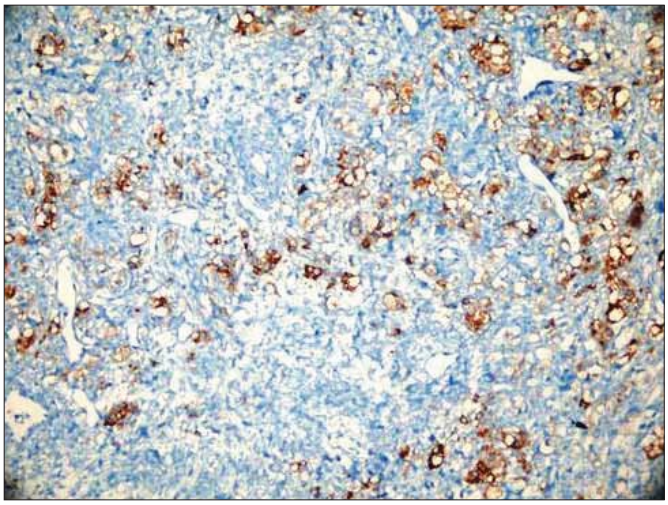

Figure 4. Tumor cells showed focal staining for calretinin

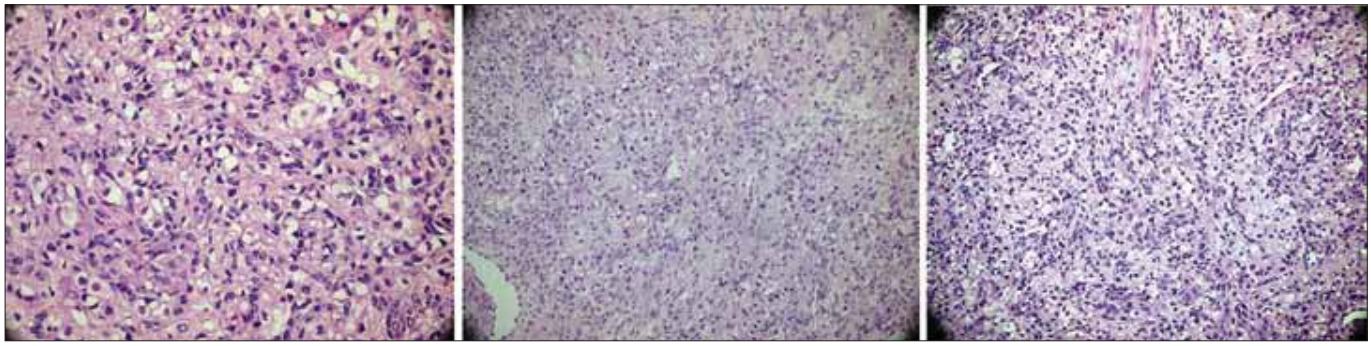

Figure 2. Tumor cells containing eccentric nuclei and single large vacuoles resembling signet-ring cells

for pan-cytokeratins, CK7, CK20. The nuclei of approximately 2% of cells were positive for Ki-67 (Figure 5). With these results we decided that it was a signet-ring stromal tumor of the ovary.

DISCUSSION

Signet-ring stromal tumor of the ovary is a distinct type of stromal neoplasm characterized by the presence of signet-ring cells that can mimic those of a Krukenberg tumor. It is very important to distinguish between these two tumors, because a SRST is a benign tumor, whereas a Krukenberg tumor is a metastatic cancer with poor prognosis (2). Ramzy first described this unique signet-ring type of ovarian tumor in 1976 and concluded that the cell type was of stromal origin. In all the previously reported SRSTs as well as in our case, the tumors were unilateral, confined to the ovary and measured 12 cm or less in greatest dimension. In the literature, bilaterality and extra-ovarian involvement were not seen in any of the SRSTs . Bilaterality has been observed in up to 93% of Krukenberg tumors (6). All the tumors reported so far occurred in adults at a mean age of 49.8 years and a range between 21 and 83 years of age. Abdominal pain was the most frequent presenting symptom (7). Ancillary techniques such as special stains for mucin are very helpful in discriminating SRST from Krukenberg tumors; all the SRSTs are negative for PAS-D and mucicarmen, whereas the signet-ring cells component is positive in all cases of a Krukenberg tumor (2, 8).

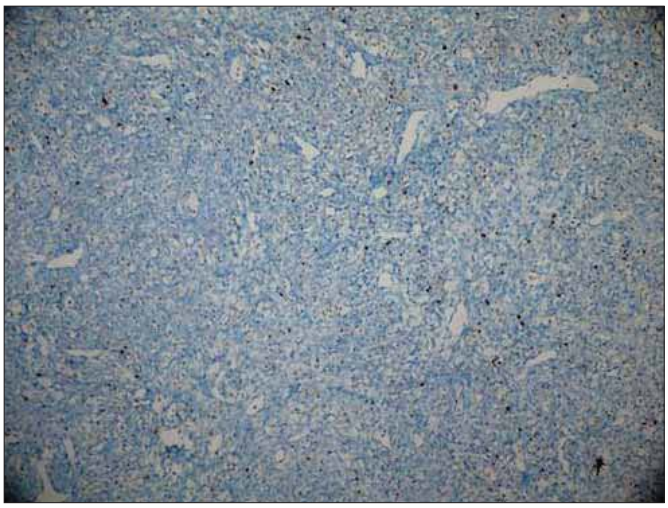

Figure 5. Ki 67 staining showing proliferation

Immunohistochemical stains, with a panel of epithelial and sex cord stromal markers, are also valuable in separating SRST from a Krukenberg tumor. In previous studies pancytoceratin was a very useful marker, with no expression in the SRSTs but with consistent staining of Krukenberg tumors (2, 8). The signet-ring cells in the case presented herein positive with pancytokeratin but were negative for CK-7 and CK-20. It's strongly recommended that immunohistochemical stains and mucin stains are used to differentiate between SRST and Krukenberg tumor.

CONCLUSION

Signet-ring stromal tumor should be considered a morphologically distinctive type of benign ovarian stromal tumor that may be mistaken for Krukenberg tumors. Although factors associated with SRSTs have not been fully elucidated, the signet-ring ovarian stromal tumor appears to have a good prognosis in view of the operative findings, histological features and follow up data. Therefore, in these cases we must be aware of that ıt will be a benign lesion although it has signet- ring cells and a malignant histologic appearance.

Conflict of Interest

The authors have no conflict of interest to declare.

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