Sclerosing stromal tumour of the ovary

Abstract
Sclerosing stromal tumours (SSTs) are rare, benign, sex cord stromal tumours of the ovary. They affect women, who complain mainly of menstrual irregularity, in their second and third decades. We report a histologically confirmed case of a 23-year-old woman who presented with complaints of abdominal pain and irregular menstruation. She later underwent surgery when a pre-pathology workup raised the suspicion of a malignant tumour. It is imperative to consider the differential diagnosis of an SST of the ovary in a young woman with an ovarian tumour.

Introduction
Sclerosing stromal tumours (SSTs) are rare, benign, sex cord stromal tumours of the ovary that affect mainly young women. Chalvardjian and Scully first described these tumours as having a heterogeneous pattern, often characterised by pseudo-lobulation of cellular areas, a prominent tendency to sclerosis and marked vascularity, which distinguished them from fibromas, thecomas and other more common types of ovarian stromal tumours.

Case report
A 23-year-old woman was referred to us with complaints of vague abdominal pain, fullness and irregular menstruation which she had been experiencing for two years. On clinical examination, a firm, cystic, ballotable hypogastric mass was palpable, arising from the pelvis. The rest of the examination was unremarkable except for goiterous enlargement of the thyroid. There were no features of hirsutism.

Contrast-enhanced computed tomography (CT) of the abdomen and pelvis revealed a heterogeneously enhancing solid mass of 4.7 x 4.4 x 3.8 cm, with areas of necrosis, involving the right adnexa (Figure 1). The mass was well defined with maintained fat planes. However, there was a suspicious extension into the right Fallopian tube. There were subcentimetre lymph nodes along the iliac vessels bilaterally and minimal fluid in the pouch of Douglas.

Figure 1: Contrast-enhanced CT of the pelvis showing a well-defined, heterogeneously enhancing solid mass in the right adnexa, with areas of necrosis.
A Doppler study revealed a complex heterogeneous mass with solid and cystic components, with marked intraläsional and perilesional vascularity and a speck of calcification.

Thyroid sonography revealed multinodular goitre with a biochemical euthyroid state.

In light view of a raised serum CA 125 and peritoneal fluid adenosine deaminase, the patient underwent an exploratory laparotomy with right ovarian cystectomy and biopsies of the left ovary, omentum and peritoneum. The right ovary was found to be 5 x 5 cm, with a smooth surface covered in tubercles, an intact capsule and no adhesions or vascularity. Tubercles were present on the uterus, Fallopian tubes and left ovary. The intestinal loops cut-section was solid, greyish-white, with no areas of haemorrhage or necrosis. Approximately 200 ml of straw-coloured fluid was drained from the peritoneal cavity. The rest of the findings were unremarkable. There was no obvious lymphadenopathy.

The patient was started on antitubercular treatment in view of the operative findings. This was later discontinued because of lack of evidence of tuberculosis.

Gross pathological examination revealed a well-demarcated, grey-white to brown soft tissue mass with a smooth surface. On cut-section, it was solid, grey-white with yellow flecks and lobulated with focal mucinous and cystic change. Normal ovarian tissue was not identified.

Histological studies revealed ill-defined, cellular pseudo-lobules separated by hyalinised and oedematous stroma (Figure 2). Two cell types were present within the lobules: spindle cells producing collagen, and polygonal cells with round to oval nuclei, fine chromatin and prominent nucleoli. Mitotic figures were not seen and cytoplasm was vacuolated.

Figure 3: Vimentin-positive immunohistochemistry (x 10)

Figure 4: Inhibin-positive immunohistochemistry (x 10)

Figure 5: Desmin-negative immunohistochemistry (x 10)
Prominent thin-walled blood vessels were seen within some nodules.

Sections from omental tissue, the parametrium and the left ovary showed the presence of noncaseating epitheloid cell granulomas surrounded by mononuclear cells.

The specimen was diagnosed as an SST in the right ovary, with granulomatous inflammation on the surface of the left ovary and omentum.

Immunohistochemical analysis on the tissue blocks revealed positivity for vimentin and inhibin, and negativity for desmin (Figures 3-5).

CA 125 levels were raised (170 U/ml) post-surgery, and returned to normal levels six weeks later.

Peritoneal fluid was negative on repeated cultures for acid-fast bacilli.

Chromosomal analysis revealed a normal female karyotype in the cells that were analysed.

Discussion

An SST of the ovary is a rare tumour and falls within the classification of sex cord stromal tumours. Usually, the presenting symptoms are menstrual disturbances, abnormal vaginal bleeding and abdominal or pelvic pain or discomfort. A few patients also present with infertility, hirsutism or no symptoms. Generally, the tumour has been reported to be unilateral. The common naked-eye appearance is mostly solid and cystic in various proportions.3

An contrast-enhanced CT scan usually shows a well-defined round mass with strong peripheral enhancement anterior to the uterus,4 although in our case, it was a complex-enhancing mass. On colour Doppler study, a high degree of peripheral and slight central vascularisation may strongly suggest the diagnosis of SST of the ovary,5 which in this case had a marked intralesional and perilesional vascularity.

On immunohistochemistry, SSTs are usually vimentin positive, sm-actin positive, inhibin positive or negative, calretinin positive or negative, desmin positive or negative, and pancytokeratin negative.6 In this case, the tumour demonstrated positivity for vimentin and inhibin and was negative for desmin. Cyto genetic analysis commonly reveals a trisomy of twelfth chromosome,7 but in our case, divulged a normal female karyotype. Clinically, there were no features that were suggestive of hirsutism, which has been reported in a few cases.3

In view of the characteristic histopathology and imaging (including Doppler and immunohistochemical studies), these tumours are increasingly being diagnosed. It is important to consider the diagnosis of SST in a young female with an ovarian tumour.

References

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