

Case 2798

Steroid cell tumour of the ovary -- a case report

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HCIS

Section: Genital (Female) Imaging

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Patient: 19 year(s), female

Clinical Summary

A 19-year-old female patient presented with hirsutism and primary amenorrhoea.

Clinical History and Imaging Procedures

A 19-year-old patient presented with hirsutism and primary amenorrhoea. She had been treated for precocious puberty since the age of seven. The results of therapeutic trials and computed tomography of the adrenal glands were found to be negative. A pelvic transabdominal ultrasonography revealed the presence of a hypoechoic lesion measuring 20 x 13 mm, with a 10 mm hyperechoic image with distal acoustic shadowing on the left adnexal region (Fig. 1). These aspects are suggestive of a teratoma. An MRI was proposed for the characterization of the lesion. When obtained, the MRI showed the presence of a solid mass in the left ovary, with a higher signal intensity than the ovarian stroma in the T1-weighted image and a low signal intensity in the T2-weighted image. After contrast-enhancement with Gd-DTPA, the fat-saturated T1-weighted image demonstrated the enhancement of the ovarian stroma and a lower intensity of the lesion (Fig. 2). On the basis of MRI findings, a diagnosis of a left ovarian tumour associated with androgenic production was suggested. The left ovary was resected at surgery. A pathological examination revealed the presence of a well-circumscribed tumour, measuring 20 mm along its longest axis, and brown in colour (Fig. 3). Histologically, the presence of clusters of polygonal large cells, uniform nuclei, eosinophilic and granular cytoplasm without mitosis, and cellular atypia or crystals of Reinke were identified (Fig. 4).

Discussion

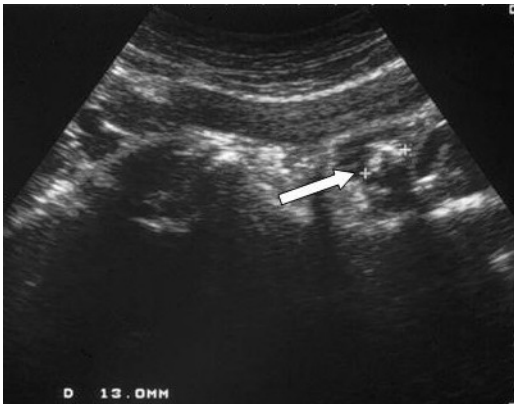
The term "steroid cell tumour" has been accepted by the World Health Organization (WHO) to describe an ovarian neoplasm which arises from the connective cells which are specialized in secreting steroid hormones. These tumours account for only 0.1%- 2% of all ovarian tumors, and have been subdivided into two subtypes of known origin, the stromal luteoma and the Leydig cell tumour, and a third subtype whose cell lineage is uncertain, known as a steroid cell tumour not otherwise specified (NOS) (1). The NOS tumours represent about 60% of steroid cell tumours, and are large stromal luteomas or Leydig cell tumours, but because they lack crystals of Reinke and because of their large size, they obscure their topographic features and cannot be identified as any other type of tumour. NOS tumours occur at any age, typically at a younger age (mean 43 years) and occasionally before puberty. These tumours are associated with androgenic changes in half the cases, which may occur over a duration of many years; oestrogenic changes in 10% of the cases and occasionally progestagenic changes. In the medical literature, five cases were reported as being associated with cortisol secretion, causing Cushing's syndrome, some without clinical manifestations but with elevated serum cortisol levels; and one case associated with aldosterone secretion. Rare examples have been associated with hypercalcaemia, erythrocytosis and ascites (2). The treatment of steroid cell tumours NOS is primarily surgical. Unilateral oophorectomy is indicated in stage 1A cases in young patients because of the low frequency of bilaterality (only about 5%). Chemotherapy has not been effective for high stage tumors or those that have recurred. Extra-ovarian spread is present at the time of operation in 20% of the cases. 25%-40% of these tumours are malignant with clinical manifestations, but no malignant tumours have been reported in the first two decades of life. Ultrasonography is the investigative modality of choice in women when a pelvic mass is suspected. In most cases, ultrasonography can characterize the mass and suggest its malignant potential on the basis of the morphological features of the mass and the presence of ascites or visceral metastases (3). The MRI is restricted when ultrasound findings are equivocal or when the diagnosis of an adnexal abnormality is not adequately characterized by ultrasonography, and for staging the malignancies. Gadolinium-enhanced MR imaging is highly accurate in the detection and characterization of complex adnexal masses, as reported above (4). This case reports a rare entity that could be of many years duration, can present with a variety of clinical manifestations and radiological appearances.

Final Diagnosis

Steroid cell tumour NOS.

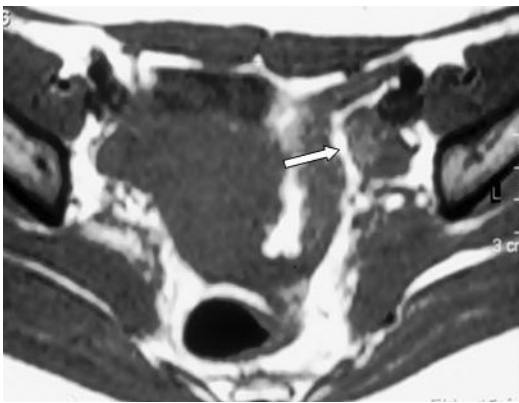
Figures

Figure 1 Pelvic ultrasonography



The transabdominal sonogram of the left ovary revealing a central region of calcification (arrow).

Figure 2 Pelvic MRI



The axial T1-weighted image showing a solid mass in the left ovary with a higher signal intensity than the ovarian stroma (arrow).



The axial T2-weighted image showing that the lesion has a low signal intensity (arrow).



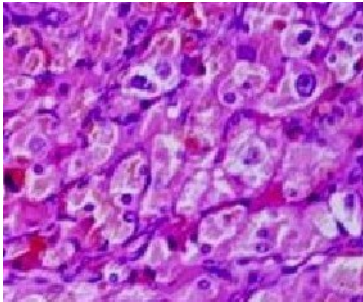
After contrast-enhancement with Gd-DTPA, the fat-saturated axial T1-weighted image, showing an enhancement of the ovarian stroma and a lower intensity of the lesion. A low signal intensity mass is well-delineated against the enhancing stroma (arrow).

Figure 3 Tumourectomy specimen



A well-circumscribed tumour, measuring 20 mm along the longest axis, brown in colour.

Figure 4 Microscopy findings



Large tumour cells with granular and eosinophilic cytoplasm and uniform nuclei (H&E).

MeSH

Ovary [A05.360.319.114.630]

The reproductive organ (GONADS) in female animals. In vertebrates, the ovary contains two functional parts: the OVARIAN FOLLICLE for the production of female germ cells (OOGENESIS); and the endocrine cells (GRANULOSA CELLS, THECA CELLS, and LUTEAL CELLS) for the production of ESTROGENS and PROGESTERONE.

Pelvic Neoplasms [C04.588.699]

Tumors or cancer of the pelvic region.

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Citation

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