Sclerosing stromal tumor of the ovary

A 14-year-old girl without relevant clinical history presented with pyelonephritis. On the renal and pelvic US performed for the evaluation of pyelonephritis, a solid mass in the left adnexal area was found. The patient had normal levels of CA-125, CEA, ß-FP, and ß-hCG.

Imaging Findings:

A pelvic MRI was performed and documented a well-defined solid ovoid mass with 8cm larger dimension, on the left adnexal area. This mass had homogeneous low-signal intensity on T1-weighted images (T1WI). The T2-weighted images (T2WI) showed central high-signal intensity areas with a peripheral rim of low-signal intensity. Contrast-enhanced MRI revealed early avid contrast enhancement of the periphery. There was a small amount of ascites on the Douglas recess. The uterus and the right ovary didn't show any abnormalities. The patient underwent surgical excision of the left adnexa. The histologic examination of the surgical specimen described a solid mass weighting 176gr and measuring 8 x 7 x 5 cm, of the left ovary, soft in consistency and slightly lobulated, compatible with a sclerosing stromal tumour of the ovary.

Discussion:

Sclerosing stromal tumours (SST) are an extremely rare sub-type of sex-cord stromal ovarian tumours, and were described for the first time by Chalvardjian and Scully in 1973 [1].

This tumour occurs predominantly in women younger than 30 years, different from other types of stromal tumours [1, 2].

The most common presenting symptom is pelvic pain. It is often hormonally inactive although there are few reports of irregular menses, genital bleeding or virilisation, more common during pregnancy.

Histologically, this tumour is characterised by cellular pseudolobules, interlobular fibrosis, marked vascularity and a dual cell population (collagen-producing spindle cells and lipid containing round or ovoid cells). The heterogeneity due to the variation in cellular size and shape are helpful features in the differential diagnosis of SST, and contrasts
with the relative homogeneity of thecoma and fibromas [3-6]. Sonographically SSTs are described as unilateral, multilocular cystic or solid masses with multiple small cysts or clefts. Colour Doppler imaging shows increased vascularity in the periphery and between the cysts. A small amount of ascites may be present [3, 4].

On T2WI shows hyperintense cystic areas and hypointense nodules located inside the hyperintense stroma, corresponding to the pseudolobulation that characterises histologically the SST, and a thin rim of low-signal intensity in the periphery. T1WI may show the same thin low-signal intensity outer rim that corresponds to the compressed ovarian cortex and a low-signal intensity central area. At dynamic contrast-enhanced imaging, early and strong peripheral enhancement with centripetal progression is characteristic. Lack of enhancement of the central area, even on delayed images, correspond to collagenous acellular areas [2, 5, 6].

The differential diagnosis should include in the paediatric and young adult population: Sertoli-Leydig cell tumour, granulosa cell tumour, cystadenofibroma; and in adults: thecoma/fibroma, malignant epithelial ovarian tumours and metastases.

Sertoli-Leydig cell tumour presents as a solid, well-defined, enhancing mass that can have some cystic/necrotic components, in a young woman with virilisation. Granulosa cell tumour most commonly present as a multilocular cystic/solid mass with cystic components, low-signal on T2WI, in a patient with a thickened endometrial stripe due to tumour oestogens secretion.

Cystadenofibroma can present as a complex unilocular/multilocular cystic mass with papillary solid projections.

Fibromas/thecomas usually appear with low-signal intensity on T2WI, and have a progressive enhancement. Ovarian metastases and malignant epithelial tumours are more common in older patients and usually don’t show the progressive centripetal enhancement [6].

All the SSTs described in the literature were benign and were treated successfully by tumour excision.

**Differential Diagnosis List:** Sclerosing stromal tumour of the ovary., Sertoli-Leydig cell tumour, Ovarian fibroma / thecoma, Granulosa cell tumour, Dysgerminoma, Ovarian carcinoma, Krukenberg tumour, Metastases to the ovary, Massive ovarian oedema

**Final Diagnosis:** Sclerosing stromal tumour of the ovary.

**References:**


Description: Axial T2-weighted MRI shows a heterogeneous mass, well-circumscribed, with regular contours, revealing a low-signal intensity outer rim and central intermediate-signal intensity nodules alternate with high-signal intensity cystic areas. This pseudolobular pattern characterises this tumour.

Origin: Cunha TM, Department of Radiology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal
Description: Coronal T2-weighted MR image shows the pseudolobular pattern that characterises this tumour: intermediate-signal intensity nodules that alternate with the high-signal intensity cystic areas.

Origin: Cunha TM, Department of Radiology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal
Description: Sagittal T2-weighted MR image shows the pseudolobulation of the tumour.

Origin: Cunha TM, Department of Radiology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal
Figure 2

**a**

**Description:** Non-enhanced T1-weighted MR image reveals a mass with homogeneous low-signal intensity. **Origin:** Cunha TM, Department of Radiology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal

**b**

**Description:** Fat-suppressed image, after gadolinium enhancement, shows avid enhancement of the periphery of the mass. **Origin:** Cunha TM, Department of Radiology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal
**Figure 3**

**Description:** Yellowish tone solid mass, soft in consistency and slightly lobulated, with a central oedematous area. **Origin:** Félix A, Department of Pathology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal
Description: Pseudolobulation is created by cellular and fibrous areas (H&E 1x). Origin: Félix A, Department of Pathology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal
**Description:** Interface between cellular and fibrous areas (H&E 10x). **Origin:** Félix A, Department of Pathology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal

**Description:** Cellular area with large polyhedral and spindle tumour cells (H&E 40x). **Origin:** Félix A, Department of Pathology, Istituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal