Case 10244
Small cell carcinoma of the ovary

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Section: Genital (Female) Imaging
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Patient: 15 year(s), female

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Clinical History
A 15-year-old girl, with no relevant personal or medical history was admitted at the emergency department with a large, painless mass in the hypogastric region, extending to the umbilical region. Laboratorial evaluation revealed an elevated calcium level (15.5 mg/dl) and a decreased phosphor level (1.9 mg/dl). Tumour markers were negative.

Imaging Findings
Pelvic and abdominal ultrasound demonstrated a 12x10 cm heterogeneous pelvic mass, with smooth borders and mixed structure, with solid and cystic areas. The origin of the mass could not be determined by ultrasound, however, a relation with the left ovary was suspected. Abdominal organs were normal, except for a slight hydronephrosis in the left kidney.
Pelvic MRI showed a median mass with an approximated volume of 600cc, which suggested a primitive lesion of the left ovary. It had well defined, regular borders, with a peripheral, heterogeneous solid component, and a central cystic area, with multiple thin septa and a mildly increased T1 signal without fat suppression, compatible with a protein-mucinous component.
Surgery was performed. Pathological examination demonstrated a small cell carcinoma of the ovary and staging CT revealed no visible metastases. Final tumour staging were stage IA (T1aN0M0).
Discussion

Small cell carcinoma of the ovary is a rare malignant tumour arising primarily in young women, with an average age at presentation of 24 years [1, 2]. The histogenesis of this tumour is unknown, as the tumour cells cannot be subtyped as surface epithelial, germ cell, sex-cord or neuroendocrine cells. It is the most common undifferentiated ovarian carcinoma in young women, and it should be included in the differential of ovarian tumours in this age group, along with well differentiated tumours [2, 3].

Clinical presentation includes pelvic mass, abdominal pain, nausea, vomiting, menstrual irregularities and infertility. Approximately two thirds of patients with ovarian small cell carcinoma have hypercalcemia. Among those patients, only a minority have symptoms related to the hypercalcaemia, such as hypertension, fatigue, polydipsia, obstipation, abdominal and lumbar pain, as well as renal insufficiency [3].

MRI is the method of choice for imaging pelvic masses. Images must be obtained in two or more planes. Acquisition of both T1 and T2 weighted images and fat-saturated T1 images are essential. In this case, the increased T1 signal intensity of the lesion could represent fat, haemorrhage or mucin. As the lesion didn't lose signal intensity after fat saturation, fat could be excluded, and endometriomas and hemorrhagic cysts are often dark on T2. High viscosity mucin can be bright on T1 weighted images, whether low viscosity mucin is dark. Combining image findings on T1, T2 and fat saturation sequences, a mucinous content of the lesion could be suspected. Gadolinium-enhanced T1-weighted images could help characterise the internal architecture of the cystic components [4, 5].

There are no specific MRI signal intensity characteristics for most malignant tumours. Thus, surgery is mandatory when a suspicious adnexal mass is found. Concerning small cell carcinoma of the ovary, therapy is primarily surgical, with adjuvant treatment reserved for advanced-stage disease [2].

Small cell carcinoma of the ovary has a poor prognosis. The most important prognostic factor is the stage of the tumour at the time of diagnosis. More than 50% of stage IA patients die within 2 years and 33% have an average 6-year disease free survival. Among the stage IA group, patients with tumour size less than 10 cm have a better prognosis, compared with those with larger tumours [2, 3].

Ovarian small cell carcinoma is a rare malignant tumour with poor outcome. Young age at presentation, hypercalcemia and imaging findings are essential diagnostic features.

Final Diagnosis

Small cell carcinoma of the ovary

Differential Diagnosis List

Mucinous cystadenoma / cystadenocarcinoma, Serous cystadenoma / cystadenocarcinoma, Endometriode carcinoma, Clear cell carcinoma, Teratoma, Dysgerminoma, Endometrial sinus tumour, Embryonal carcinoma, Granulosa cell tumour (juvenile type), Sertoli-Leydig cell tumour

Figures
Figure 1 Abdominal and pelvic ultrasound

Longitudinal plane in the pelvis demonstrating a heterogeneous mass with smooth borders and mixed structure, containing solid and cystic areas.

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Area of Interest: Genital / Reproductive system female;
Imaging Technique: Ultrasound;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

Transversal plane in the pelvis demonstrating a heterogeneous pelvic mass with smooth borders and mixed structure, containing solid and cystic areas.

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Area of Interest: Genital / Reproductive system female;
Imaging Technique: Ultrasound;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;
During the abdominal scan, a moderated hydronefrosis was noted in the left kidney

Area of Interest: Kidney;
Imaging Technique: Ultrasound;
Procedure: Diagnostic procedure;
Special Focus: Obstruction / Occlusion;

Figure 2 Abdominal and pelvic MRI

Axial T1: primitive lesion of the left ovary with well defined, regular borders.

Area of Interest: Genital / Reproductive system female;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

Axial T2: primitive lesion of the left ovary containing a peripheral heterogeneous solid component and a central cystic area, with multiple thin septa.

Area of Interest: Genital / Reproductive system female;
Imaging Technique: Ultrasound;
Sagittal T2: primitive lesion of the left ovary containing a peripheral heterogeneous solid component and a central cystic area, with multiple thin septa.

Coronal T2: primitive lesion of the left ovary containing a peripheral heterogeneous solid component and a central cystic area, with multiple thin septa.
Figure 3 Abdominal and pelvic MRI

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<th>Area of Interest:</th>
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Axial T2 fat saturation image demonstrating that the lesion contains no fat, but probably a proteic-mucinous material.

Figure 4 Abdominal and pelvic MRI

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Sagittal T2 fat saturation image demonstrating that the lesion contains no fat, but probably a proteic-mucinous material.
Axial T2: normal right ovary (yellow arrow).

Area of Interest: Genital / Reproductive system female;
Imaging Technique: Ultrasound;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

Sagittal T2: normal right ovary (yellow arrow).

Area of Interest: Genital / Reproductive system female;
Imaging Technique: Ultrasound;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

Figure 5 Abdominal and pelvic MRI
Axial T2: the mass is causing homolateral ureterohydronephrosis (yellow arrow).

Area of Interest: Genital / Reproductive system female;
Imaging Technique: Ultrasound;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

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.

Ovarian Neoplasms [C04.588.945.418.685]
Tumors or cancer of the OVARY. These neoplasms can be benign or malignant. They are classified according to the tissue of origin, such as the surface epithelium, the stromal endocrine cells, and the totipotent germ cells.

References


Citation

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