1. Abstract

Borderline ovarian tumours are rare in children and adolescents. They account for around 15% of all epithelial ovarian cancers. In 80% of cases, BOTs are diagnosed at stage I. The aim of this study is to analyse the epidemiological, clinical, evolutionary and therapeutic characteristics of borderline ovarian cancer in an adolescent girl, confirmed histologically and treated at the Mohamed IV Centre of the Casablanca University Hospital.

2. Identity

The patient was 14 years old and had undergone a cystectomy with the presence of a borderline mucinous ovarian tumour on pathology, with a peritoneal cytology fluid without abnormality. She was seen again 3 months later for abdominal distension with no other accompanying signs, and the clinical examination revealed a distended abdomen with an icicle sign without any other clinical abnormality.

A CT scan revealed an abdominal and pelvic cystic mass, probably ovarian in nature, measuring 210 x 180 x 115 mm, with a moderate amount of pelvic effusion (Figure 1).

The patient underwent a surgical exploration which revealed the presence of a moderate amount of ascites which had been sampled, and the presence of a 20 cm mass at the expense of the ovary, leading to a right adnexectomy which removed the mass, followed by an omentectomy, an appendectomy and peritoneal biopsies. Post-operative follow-up was straightforward.

The anatomopathological result showed the presence of a borderline mucinous tumour. The tumour capsule was intact, with no rupture sites, and the tube, omentum, peritoneum, appendix and peritoneal fluid were unremarkable.

The patient was referred to the oncology department for further management.

Figure 1: Right ovarian abdomino-pelvic mass [(A); (B) - axial section]. Les marqueurs tumoraux étaient negatives.
3. Discussion
Borderline ovarian tumours are rare in children and adolescents, due to the particularity of the ovary in embryogenesis. Ovarian tumours have a complex structure and composition, making adolescent ovarian tumours rare with an incidence that increases with age, rising from 0.43/100,000 cases at 1 year of age to 152/100,000 cases in 35 year old patients [1].

For reasons that are not yet clear, borderline ovarian tumours of the serous type are more common in Europe, North America and the Middle East, while mucinous tumours are more common in East Asia [1,6].

The WHO has classified these tumours into three main groups, depending on whether they develop from epithelial cells, germ cells or stromal tumours of the sex cords. Germ cell tumours account for over 70-80% of ovarian tumours [1,2], and are most common in children and adolescents. Benign epithelial tumours of the ovary are much more common in older women, but are uncommon in childhood and adolescence. Of all epithelial ovarian tumours in childhood and adolescence, approximately 10-30% are considered borderline ovarian tumours (BOTs), also known as “low malignant potential” [2,4,5].

Borderline tumours of the ovary are neoplasms of epithelial origin with low malignant potential characterised by high cell proliferation and the presence of mild nuclear atypia, but without destructive stromal invasion [2;7]. There are few reports of BOT in children and adolescents. Histologically, mucinous TOF is more common than serous TOF.

Diagnosis of ovarian tumours in children and adolescents is often difficult and delayed, as 30% of cases are asymptomatic, while 50-60% are symptomatic but non-specific, notably abdominal pain and distension [10].

Most borderline tumours are detected by ultrasound, which is recognised as an accurate method of distinguishing between benign and malignant adnexal masses [2,3].

Serum tumour markers such as CA-125 and CA-19 9 are of some value in diagnosing these borderline tumours. CA-125 is a protein associated with epithelial tumours of the ovary, often expressed at lower levels in benign tumours, and variably expressed in germ cell tumours of the ovary. However, CA-125 elevation can also be seen in children and adolescents with benign gynaecological conditions, including non-cancerous tumours, confounding the picture in this patient population [13].

CA-125 increases in both serous and mucinous borderline tumours, and CA-19-9 increases more in mucinous BOT.

In general, borderline tumours of the ovary are of low grade and, apart from surrounding invasive implantation, have a good prognosis [1].

In the presence of high-risk factors with a poor prognosis, such as invasive implantation, a regimen of paclitaxel and platinum-based chemotherapy is suggested.

Adolescent BOT has an excellent prognosis, even at an advanced stage, when managed by fertility preservation procedures [2].

A description of the natural behaviour of the different histotypes is essential for selecting the most appropriate surgical strategy. Close follow-up is important because of the high recurrence rates several years after diagnosis.

In terms of surgery, laparotomy is performed during the staging of BOT, due to the risk of tumour rupture, increased recurrence rates and reduced survival in the case of incomplete staging of the tumour, increased recurrence rates and reduced survival in the case of incomplete staging [13].

Fertility preservation procedures have been shown to be safe and effective for patients with BOT.

Early diagnosis is extremely important in all circumstances, but is most significant in young women, as it may dictate fertility preservation [10-12].

BOTs are staged according to the FIGO staging system used for ovarian carcinoma. Most BOTs have a low malignant potential and are confined to the ovaries at the time of presentation [7, 9].

When malignancy is identified or of concern, the Children’s Oncology Group recommends a staging procedure which involves removal of the tumour, collection of ascites or pelvic washings for cytology, examination and palpation of the omentum, iliac and aortocaval lymph nodes, with biopsy or removal of suspicious areas. For mucinous BOT, appendectomy is recommended [13].

Peritoneal spread is present in 10% of BOTs and is divided into two categories: non-invasive implants (nearly 85% of implants) and invasive implants (2); the mortality rate for patients with non-invasive and invasive implants is 4.7% and 34% respectively [7,8].

Studies have also shown that the presence of a gynaecological surgeon is associated with a higher rate of ovarian conservation and a reduced risk of incomplete surgical staging for malignant ovarian conservation lesions and a reduced risk of incomplete surgical staging for malignant lesions [13].

As the literature on BOT in children and adolescents is limited, further research is needed.

4. Conclusion
A borderline tumour of the ovary represents an independent group of ovarian neoplasms with atypical epithelial proliferation, which are very rare. Current knowledge of BOT in the paediatric and adolescent population comes from reports of small case series, which limits data on clinical presentation, treatment and outcome.
References