Original Article

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Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.104726
Quick Response Code:

Ovarian tumours in children: A review of 18 cases

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ABSTRACT

Background: To review the experience of Children's Hospital of Rabat in managing ovarian tumours in children. **Materials and Methods:** There were 18 patients between 2 and 15 years of age who presented with an ovarian tumour at Children's Hospital of Rabat between January 2000 and December 2008. Data collected from the hospital medical records included age at diagnosis, patient's history, presenting complaints, radiological examination, tumour markers, management, operative procedure, histopathological examination and outcome of the patients. **Results:** The most common presenting complaint was abdominal pain in 10 (55%) patient. 77% of ovarian tumours were germ cell tumours; 71% of these were teratomas which were benign in 66% of cases. Unilateral salpingo-oophorectomy was the most common surgical procedure performed in 15 patients (83%) through laparotomy. Laparoscopic ovarian cystectomy was carried out in 2 (11%) patients with benign cystic teratoma. Of the 7 (39%) patients with malignant tumours, three received postoperative chemotherapy. Outcome was good in most cases. There were no cases of resistance to treatment, or death. **Conclusion:** Early diagnosis of ovarian tumours in children and adolescents is important. Since most of these tumours are benign, surgical treatment should be conservative to minimise the risk of subsequent infertility, while the treatment of malignant tumours should include complete staging, resection of the tumour, postoperative chemotherapy when indicated, to give the patient a chance for future childbearing.

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Key words: Children, fertility, germ cell tumours, ovarian preservation, ovarian surgery, ovarian tumours, teratoma

INTRODUCTION

Ovarian tumours are uncommon lesions in children. Their actual incidence in young girls is unknown, because of recruiting bias, lack of a national registry of tumours and the wide spectrum of gonadal pathologies ranging from highly aggressive malignant tumours to benign cysts. It has been estimated that the approximate incidence of ovarian tumours is about 2.6 cases annually/100 000 girls, and malignant ovarian tumours make up about 1% of all childhood cancers.[1] These lesions may have multiple presentations. As ovarian masses are relatively rare and management occasionally unclear, we retrospectively reviewed the experience of Children Hospital of Rabat in order to evaluate their epidemiology, pathology, treatment and outcome.

MATERIALS AND METHODS

We retrospectively reviewed 18 cases of ovarian masses presenting to the Children's Hospital of Rabat between January 2000 and December 2008. Data collected from the hospital medical records included age at diagnosis, patient's history, presenting complaints, radiological examination, tumour markers, management, operative procedure, histopathological examination and outcome of the patients.

Morphologic characteristics of tumours were assessed with imaging, ultrasonography, computed tomography, and, in one case, magnetic resonance imaging (MRI).

Unilateral salpingo-oophorectomy (USO) through laparotomy was the most common surgical procedure.

Laparoscopic ovarian cystectomy was carried out in 2 patients with benign cystic teratoma.

Benign tumours were treated with only surgery. Ovarian-sparing tumourectomy has been performed when macroscopic appearance allowed separating ovarian parenchyma from tumour.

Patients with malignant tumours were treated according to the protocol of the French Society for Pediatric Oncology: TGM (Tumeurs Germinales Malignes) 95 protocol. USO was performed by laparotomy with adequate staging including abdominal and pelvic exploration, peritoneal washings, biopsies of suspicious areas and lymph node sampling. After surgery, tumours were staged according to the postoperative classification of French Society for Pediatric Oncology.

Chemotherapy, following the indicated protocol, was administered when resection was microscopically incomplete. In cases when a huge tumour was preoperatively assessed to be unresectable, patients were submitted to chemotherapy before surgery.

RESULTS

Median age of the patients at surgery was 11.2 years (range, 2–15 years). No tumour was detected antenatally. Four tumours presented prior to 8 years of age and 1 (25%) was malignant. Fourteen tumours presented after 8 years of age and 6 (43%) were malignant.

Five patients were mature. Consanguinity was found in 4 cases. A family history of cancer was noted in 2 patients.

The most common presenting complaint was abdominal pain in 10 (55%) patients, abdominal swelling in 3 (16%) cases and menstrual disorders in 1 (5.5%) patient. In 2 cases (11%) of 18, ovarian tumours were revealed by abrupt onset of abdominal pain mimicking ovarian torsion. In 4 cases (22%), tumour was discovered incidentally during routine clinical examination or investigation for extra abdominal disease or pubertal disorder. An abdominal mass was palpable on examination in 82% of cases (n = 15).

All the patients had ultrasound examination. The ovarian origin was evoked in 14 patients (77%). Seven had complex mass, of those 3 (43%) were benign and 4 (57%) were malignant. Eight patients showed a cyst, of those 7 (88%) were benign and 1 (12%) was malignant. Two had a solid mass, both of them were malignant. The mean size of the ovarian tumour was 136 mm/84 mm (range, 30 mm/24 mm-261 mm/144 mm).

Abdominal computerised tomography (CT) scan was performed in 83% of cases (n = 15) and MRI in 1 patient, where further evaluation of the nature of the ovarian tumour was necessary, and to evaluate the extent of the disease in suspected malignant cases. Chest radiography was performed in all cases. No cases of lung metastasis were noted.

Levels of tumour markers were elevated in 4 cases of malignant tumours. Alpha fetoprotein (AFP) was elevated in 3 cases (immature teratoma, vitelline tumour, and one case of unspecified malignant germ cell tumour). Human chorionic gonado-trophic hormone (B HCG) was elevated in one patient with dysgerminoma.

In the group of benign tumours, mature teratoma was the most common—7 cases/9 (77%). The two patients with mucinous cystadenoma were mature.

Massive oedema of the ovary and Haemorrhagic cyst are histologically pseudotumoural lesions, but they were initially considered and treated as benign ovarian tumours based on data from investigations.

All malignant lesions were germ cell tumours. Immature teratoma was the most common malignancy—3 cases/7 (42%). In one case, diagnosis of malignant germ cell tumour was done on data from imaging and markers. Histopathologic characterisation was not possible because the tumour was treated with chemotherapy before surgery.

USO was performed in 15 patients (83%). Most of patients (10, 55%) underwent laparotomy through a Pfannenstiel incision, and 6 (33%) patients had a midline laparotomy.

Two patients with large pelvi-abdominal benign tumours had removal of the tumour in-toto by USO as the contralateral ovary was normal. Both of them were found to be mucinous cystadenoma (cases 6 and 11) [Table 1]. Of the 7 patients with mature teratoma, two had unilateral ovarian cystectomy (cases 1 and 8) [Table 1]. USO was carried out in the five remaining patients (cases 2, 3, 4, 7 and 10) [Table 1].

The two patients with pseudotumoural lesions (Massive oedema of the ovary and Haemorrhagic cyst) underwent USO because of suspicion of malignancy on radiological examination (cases 5 and 9) [Table 1].

The outcome was satisfactory in most cases of benign lesions (8 cases/11). Recurrent contralateral mass occurred in one patient with mature teratoma 3 months after surgery. Two patients were lost to follow up.

Among the 7 patients with malignant tumours, 4 underwent primary surgery. USO was performed in 6 cases by laparotomy. Six tumours were stage pS I, and 1, stage pS IIIb. Three patients were submitted to chemotherapy before surgery, and 3 have received postoperative chemotherapy.

One of the 2 patients with dysgerminoma had a left salpingo-oophorectomy (case 4) [Table 2]. The patient had no postoperative chemotherapy since the tumour was localised, stage pS I. Ten months after surgery, the patient had a left lateroaortic lymph node recurrence for which lymphadenectomy was performed and postoperative multiple drug chemotherapy consisting of VP16, ifosfamide and cisplatin (VIP) regimen was started. She received 5 cycles with satisfactory response. She is alive and well for a follow-up of 6 years.

The second patient with dysgerminoma (case 7) [Table 2] had an advanced tumour that was unresectable and adherent to the uterus. Surgical biopsy was performed for histological diagnosis. Then, the patient received 4 courses of vinblastine, bleomycin and cisplatin (VBP) according to TGM 95 protocol high risk. Surgical resection was incomplete because of the inability of cleavage of the tumour from uterus.

Table 1: Details of the 11 patients with benign tumours

Year of treatment	Age (yrs)	Diagnosis	Presenting complaint	Operation performed	Outcome
2000	14	Dermoid cyst	Discovery during staging of a cavum cancer	Cystectomy	Lost to FU
2000	7	Dermoid cyst	Abd pain and mass	LSO	Well-2 years FU
2001	13	Dermoid cyst	Abd pain	RSO	Well-1 year FU
2001	13	Mature teratoma	Abd pain and mass	LSO	Well-2 years FU
2003	13	Massive edema of the ovary	Abd mass	RSO	Lost to FU
2003	15	Mucinous cystadenoma	Abd pain and mass	RSO	Well-4 years FU
2005	15	Mature teratoma	Abd mass	RSO	Recurrent contralateral mass of the ovary 3 months after surgery (Conservative treatment in a service of gynaecology)
2006	7	Dermoid cyst	Abd pain	Cystectomy	Well-6 months FU
2007	12	Hemorrhagic cyst	Abd pain	Left oophorectomy	Well-6 months FU
2008	10	Dermoid cyst	Abd pain	LSO	Well-1 year FU
2008	14	Mucinous cystadenoma	Abd pain	LSO	Well-2 years FU

Table 2: Details of the 7 patients with malignant tumours

Year of treatment	Age (yrs)	Diagnosis	Presenting complaint	TM	Preop chemotherapy	Operation performed	Postop chemotherapy	Outcome
2000	2	Immature teratoma	Abd pain and mass	AFP	-	RSO		A/W-1 year FU
2001	9	Immature teratoma	Abd pain	-	-	RSO		A/W-1 year FU
2004	11	Unspecified MGCT	Abd pain and mass	AFP	4 VIP	LSO		A/W-4 years FU
2004	10	Dysgerminoma	Abd pain	BHCG	-	LSO	-	Left latero-aortic lymph node recurrence 10 months after surgery (lymphadenectomy + 5 cycles of VIP) A/W-6 years FU
2005	14	Vitelline tumour	Abd pain and mass	AFP	3 VBP	LSO	1 VBP	A/W-5.5 years FU
2007	8	Immature Teratoma	Abd mass	-	-	RSO	3 VBP	A/W-3.5 years FU
2008	14	Dysgerminoma	Abd pain	-	4 VBP	Incomplete resection	4 VIP	A/W-3years FU

Yrs: Years; TM: Tumour markers; abd: Abdominal; A/W: Alive and well; RSO: Right salpingo-oophorectomy; LSO: Left salpingo-oophorectomy; MGCT: Malignant germ cell tumour; VBP: Vinblastine, bleomycin and cisplatin; VIP, VP16: Ifosfamide and Cisplatin; FU: Follow up

Postoperative chemotherapy with 4 cycles of VIP showed good response with regression of residual tumour. The patient is alive and well for a follow-up of 3 years.

The 3 patients with immature teratoma (cases 1, 2 and 6) [Table 2] were treated with USO followed by chemotherapy in one of them, according to TGM 95 protocol standard risk (3 cycles of VBP). They were free of disease at a mean follow-up of 2 years.

The patient with vitelline tumour (case 5) [Table 2] was treated by primary chemotherapy according to TGM 95 protocol standard risk, with a good response, followed by left USO. A cure of VBP was added after surgery because of the presence of alive tumour tissue in the specimen. She is alive and well for a follow-up of 5.5 years.

The patient with unspecified MGCT (case 3) [Table 2] underwent left USO after a preoperative chemotherapy according to TGM 95 protocol high risk (4 cycles of VIP). She is free of disease at a follow-up of 4 years.

DISCUSSION

Ovarian tumours generally have been considered rare in children and adolescents. The incidence of 2.6 cases annually/100 000 girls[1] concerns only tumours and does not include functional lesions (cysts, adnexal torsion…), which are as frequent as tumours.[2,3] The results of our study confirm the rarity of ovarian tumours reported by different authors.[4-6]

The presenting complaints in patients with ovarian masses are nonspecific, making diagnosis difficult. Acute or chronic abdominal pain and abdominopelvic mass are the most common initial symptoms.[1] Endocrine signs can also reveal some secreting ovarian tumours (precocious puberty, virilisation signs…).

Ultrasound is a helpful diagnostic tool for ovarian lesions. It was performed in all our patients and was accurate in diagnosis of ovarian pathology in 14 patients (77%). However, it could not distinguish between benign and malignant tumours.[1] In our series, malignancy was suspected in 3 patients, among them only one had a malignant tumour on pathological examination. Solid ovarian masses on ultrasound were all malignant, whereas cystic tumours were, in most cases, benign (88%).

CT scan and MRI are useful in case of undetermined mass, when we suspect malignant ovarian tumour, and before any surgery. They give additional information as to the nature of the tumour and pelvic and para-aortic lymph nodes, which add to the accuracy of the diagnosis of ovarian malignancy.

Tumour markers are essential for diagnosis and follow-up of ovarian tumours. Their rate may increase several months before the clinical or radiological detection of recurrence. In our study, four malignant germ cell tumours of a total of 7 were secreting.

Most ovarian masses in children are benign.[7] The incidence of benign tumours in our study was 61% and mature teratoma was the most common among them (77%), which is consistent with other reports.[1,3,8] Among the mature teratomas, we found, in accordance with literature data,[9] a clear predominance of dermoid cysts (71%).

Epithelial tumours are rare in children and adolescents, and are usually benign. Serous cystadenomas are the most frequent, followed by mucinous cystadenomas.[2] The two cases of epithelial tumours in our series were, in discordance with literature, mucinous cystadenomas.

Pseudotumoural lesions are as frequent as tumours. They are dominated by functional cysts which may be simple or complicated.[2] We had 2 cases of pseudotumoural lesions in the series: haemorrhagic cyst and massive oedema of the ovary which is a rare pseudotumoural lesion, occurring primarily in adolescents and young women and often confused with malignant tumours.[10]

Malignant tumours of the ovary are infrequent in children and comprise about 3% of all childhood cancers. The risk of malignancy increases with age.[11] They account for 20 to 25% of ovarian tumours.[11] Germ cell tumours are the most common malignancy; epithelial cell tumours are less likely.[4,5,12] This is the opposite of what is seen in adult patients. Immature teratoma, dysgerminoma and mixed germ cell tumour are the most common subtypes.[5,12] All the malignant ovarian tumours found in our study were germ cell tumours, among them immature teratoma was the most common (42%), followed by dysgerminoma (28%).

In differentiating patients with benign tumours from malignant ones, we found that age, presenting symptoms, and size of the tumour were not predictive, but imaging characteristics of the tumour and tumour markers were useful. On imaging, the presence of more

solid components in the tumour was a definite feature that predicted malignancy.

Surgical approach, laparotomy *vs* laparoscopy, depends on the level of preoperative suspicion of malignancy. The laparoscopic removal of selected ovarian tumours is a well-established procedure, but controversy still exists about selection of tumours that can be removed by this method.[1] Laparotomy allows reducing the risk of spillage and is mandatory in case of malignant tumour.

Surgical management of benign ovarian masses should be conservative after exclusion of a malignancy. Cystectomy and partial oophorectomy should be carried out when possible to spare as much normal ovarian tissue as possible to maintain stable reproductive and endocrine function.[13] In cases of dermoid cyst, cystectomy should be careful to avoid spillage during surgery which can cause a chemical peritonitis and adhesions with the uterus. Oophorectomy is required when voluminous size or macroscopic appearance does not allow separating ovarian parenchyma from tumour.

In cases of malignancy, the treatment is mainly based on surgery, with or without chemotherapy. The surgical approach must consider both recovery and fertility preservation. Primary surgery is indicated in cases of emergency, in cases of non-secreting tumours to make diagnosis and proper treatment, and in cases of localised tumours which allow complete resection without mutilation. Salpingo-oophorectomy is performed with adequate staging including abdominal and pelvic exploration, peritoneal washings, biopsies of suspicion areas and lymph node sampling. In our study, malignant ovarian tumours were staged according to the postoperative classification of French Society for Pediatric Oncology, which is specific to paediatric malignant germ cell tumours.

Primary chemotherapy is indicated when either biopsy revealed malignant tumour (cases of non-secreting tumours) or tumour marker levels are elevated, and huge tumour is assessed to be unresectable. Surgery is then required to remove the affected organ, even if there is no residual tumour and even if the markers are negative.

The indication of postoperative chemotherapy depends on the initial extension, the possibility for removal of the tumour, the level of tumour markers and histological type. Current recommendation suggests that stage pS I tumours do not require chemotherapy beyond surgical resection, which has excellent survival rates.[1] If there is a residual tumour after postoperative chemotherapy and if markers have normalised, surgical removal of the residue is required.[14]

The prognosis of ovarian tumours in children is excellent if they are detected in an early stage. The risk of recurrence of benign tumours in the contralateral ovary is about 10%, which requires a regular ultrasound monitoring, throughout the childbearing.[15] Malignant tumours have a good prognosis for survival if they are properly treated. The most common histological types have a low metastatic potential with high-recovery rates.[16] Early diagnosis and the use of cisplatin have a major role in determining the prognosis of ovarian malignant tumours in children.[16]

CONCLUSION

Ovarian tumours are rare in children. Most are benign. The risk of malignancy increases with age. Germ cell tumours are the most frequent malignant tumours in children. Surgical treatment is conservative for both benign and malignant lesions. It is important for an early diagnosis to be established in order to reduce the risk of ovarian torsion with possible loss of the ovary and to improve the prognosis in patients with malignant tumours. In Morocco, to fully appreciate the various aspects of ovarian tumours in children, we must register all tumours, including benign ones, and encourage multicentre studies.

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Cite this article as: Ammor A, Kisra M, Oulahyane R, Kababri M, Maalmi N, Cherkaoui A, *et al*. Ovarian tumours in children : A review of 18 cases. Afr J Paediatr Surg 2012;9:231-6.

Source of Support: Nil. **Conflict of Interest:** None declared.

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