OVARY

Functional ovarian lesions in children and adolescents: when to remove them

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Abstract

Study objective. Functional ovarian lesions represent 45% of all pediatric adnexal abnormalities. Their surgical management, even if frequent, is not clear, especially in pediatric age.

Materials and methods. We retrospectively reviewed 22 pediatric patients surgically treated for functional ovarian lesions from 2000 to 2006. The following characteristics were analysed: age, size of the lesion, ultrasound (US) aspect and clinical presentation.

Results. The average age was 16.1 years of age (range: 6 months–18 years). Of the 22 functional lesions, 12 (55%) were follicular cysts and 10 (45%) corpus luteum ones. The average size was 6.7 cm (range: 5.1–33 cm). US scan showed simple lesions in 10 cases (45%) and complex ones in 12 cases (55%). In 16 girls (72.8%) the presenting symptom was abdominal pain while 2 patients (9%) presented abdominal distention. In the other 4 patients (16.2%) the lesion was found accidentally during US examination.

Conclusion. In pediatric subjects, functional ovarian cysts rarely required surgical intervention, though no complications or disorder recurrence were reported. To preserve ovarian function, conservative surgery has to be performed whenever feasible.

Keywords: Ovary, pediatric age, surgery

Introduction

Pediatric ovarian lesions are estimated to occur at a rate of appreciatively 2–5 cases per 100,000 girls per year [1–3]. These lesions, although rare, span a spectrum of pathologies from functional non-neoplastic ovarian cysts to ovarian torsion and from benign tumors to highly aggressive neoplasms. Functional ovarian lesions (FOL), such as follicle cysts and corpus luteum ones, are the most frequent abnormalities both in adults and in children where these represent about 45% of all adnexal pathologies [4,5]. Occurrence aged displays two peaks: fetal/neonatal, due to maternal hormonal stimulation; and perimenarchal, a consequence of the dysfunctional ovulation characterising the early period following menarche [6]. Ovarian cysts are frequently detected in the uterus by pre-natal ultrasound (US); about 30% of fetuses have follicular ovarian cysts greater than 1 mm in diameter as a consequence of maternal and placental hormonal stimulation [7]. Beyond the newborn period, ovarian cysts in pre-pubertal children are unusual and the presence of an ovarian lesion has to be carefully evaluated and treated as potentially malignant. In post-menarcheal girls both follicular and corpus luteum cysts are common due to anovulatory cycles. Functional ovarian cysts, as the other ovarian lesions, come to surgical attention in a variety of ways; some are detected incidentally during prenatal or postnatal US examination, whereas others present themselves insidiously as painless abdominal swellings. Most patients suffer from acute or chronic abdominal pain. Acute symptoms may be caused by torsion, bleeding or rupture of a large cyst. Constipation, nausea, vomiting as a well as fatigue are also reported [7]. More rarely, there may be symptoms of precocious puberty or of virilisation [7,8]. Even if frequent, their management (surveillance, medical therapy or surgical enucleation) is not clear, especially in pediatric age and this is the reason of this study which aim is to outline the characteristics of FOL that should be surgically treated.
Materials and methods

This study included all pediatric subjects undergone surgical intervention for FOL in the Surgical Institutions of the University of Pisa from 1st January 2000 to 31st December 2006. After reviewing the pathology reports, the medical records of the patients were retrospectively reviewed. Specific data was collected on each patient included for clinical presentation, age at operation, imaging studies (ultrasonography (US), computerised tomography (CT) and magnetic resonance imaging (RMN) scans), serum data (CA-125, α-fetoprotein and β-human chorionic gonadotropin), surgical route and outcome. The data have been reported as mean (± standard deviation). After the local Institutional Review Board approval, the informed consent was obtained from all parents prior to the study. Abdominal US was performed as the first diagnostic test for all patients to characterise ovarian diseases in simple, complex or solid ones [5,9].

Results

In the 7-year period, 67 consecutive pediatric patients were surgically treated for ovarian lesions (e.g. mature cystic teratoma, cystoadenoma and yolk-sac tumour). Of them 22 girls (32.8%) were affected by functional cysts.

At a first surgical evaluation, the 22 girls were 16.1 ± 1.6 years of age (range: 6 months–18 years); 19 girls (86.3%) were post-menarcheal with a menarcheal age of 12.3 ± 1.7 years of age. Four patients (18.1%) presented a personal history of disruption of the menstrual cycle or dysmenorrhea. One patient presented with a familiar history of hemorrhagic corpus luteum cysts, her mother had been surgically treated for the same disease. No patients presented clinical characters of precocious puberty, skin pigmentation or other known syndrome markers at physical examination. Height, weight and body mass index were 0.7 ± 0.41, 0.6 ± 0.28 and 0.4 ± 0.26 standard deviation score (SDS), respectively.

Abdominal US was performed as first diagnostic test for all patients to characterise ovarian diseases. US examination showed unilateral lesion in all our patients: complex ovarian masses in 10 girls (45.0%) and simple ovarian masses in 12 cases (55.0%), respectively. The average size was 6.7 cm (range: 5.1–33 cm). An additional abdominal CT scan was also performed in 7 patients (31.8%) to better define the complex lesions as well as to exclude associated pathologies.

Serum levels of tumour markers were included in our normal range for all cases (normal values: CA 125 < 35 U/mL, α-fetoprotein < 15 ng/mL, HCG 0 ng/mL).

Primary presentation that led to operation were persistent abdominal pain in 13 patients (59%), acute abdominal pain in 3 patients (13.7%) – due to ovarian torsion and suspected cyst rupture – and abdominal distention in 2 patients (9%), respectively. In the other 4 girls (18.3%) the lesion was found accidentally during pre-natal (1 case − 4.6%) and post-natal (3 cases − 13.7%) US scan.

Nineteen patients (86.4%) were surgically treated after an ongoing observation of 5.3 ± 1.2 weeks (range: 4–8 weeks), with several US scans. Three patients (13.7%) were treated in urgency, two with a cyst complicated by ovarian torsion and the other with a suspected rupture of the lesion where we found intrapelvic blood at the surgery view.

Six patients (27.2%) underwent operative laparoscopy: excision of the cyst was performed in all cases.

Sixteen patients (72.8%) underwent laparotomy: simple enucleation of the cyst with the preservation of functional ovarian tissue was performed in 12 cases (54.6%), oophorectomy in 3 cases (13.7%) and salpingo-oophorectomy in one case (4.5%). Incidental appendectomy was performed in 5 patients (22.7%) with no associated complications (Table I).

The exclusion of the US features suggestive of ovarian cancer (papillary projections, presence of an entirely solid tumour, maximal diameter of solid component, irregular internal cyst walls, acoustic shadows, a colour score of intratumoural blood flow), according to the maximum diameter of the

| Table I. Presentation and surgical procedures of patient with functional ovarian lesions. |
|-----------------------------------------------|-----------------------------------------------|-----------------------------------------------|-----------------------------------------------|
| **Age (range)** | **Clinical presentation*** | **US aspect** | **Surgical approach** | **Surgical technique**** |
| Follicular cysts | 12 (55%) | Abdominal pain 7 (31.8%) | Simple – 10 (45.5%) | Laparotomic – 10 (45.5%) | Cistectomy – 11 (50%) |
| | | Incidental finding 3 (13.7%) | Complex – 2 (9%) | Laparoscopic – 2 (9%) | Oophorectomy 1 (4.5%) |
| Corpus luteum cysts | 10 (45%) | Abdominal pain 9 (41%) | Simple – 6 (27.2%) | Laparotomic – 1 (4.5%) | Cistectomy – 7 (32%) |
| | | Incidental finding 1 (4.5%) | Complex – 4 (18.3%) | Laparoscopic – 2 (9%) | Oophorectomy – 1 (4.5%) |

*In 2 girl all of them of the follicular cyst group presented with abdominal distention.

**In 1 girl with corpus luteum cyst a salpingo-oophorectomy was performed.
lesion (<10 cm), serum CA125 <35 IU/mL and the absence of pelvic endometriosis or pelvic adhesions, allowed the decision of laparoscopic surgery.

The histological examination revealed the following: in 12 cases (55%) a follicular cyst was found, 9 (75%) in post-menarcheal girls and 3 (25%) in pre-menarcheal girls, a corpus luteum cyst was detected in 10 post-menarcheal girls (45%), in the right ovary in 6 patients (60%) and in the left in 4 patients (40%); one girl had an ipsilateral serous cystoadenoma also whereas another patient presented a contralateral paraovarian cyst also.

After conservative surgery, ovarian size and viability, as assessed by 6-month US examination, were normal. At a follow-up of 24.5 ± 12.8 months (range: 9–53 months) no recurrences occurred and regular menses cycles of 28–31 days were reported in post-menarcheal girls.

Discussion

Pediatric ovarian lesions, although rare, span a spectrum of pathologies from functional and so non-neoplastic ovarian cysts to ovarian torsion and from benign tumours to highly aggressive neoplasms [2–4,8]. Even if their frequency is probably underestimated, functional ovarian cysts represent the most common ovarian lesion in children and adolescents constituting 17.1–43.6% (mean: 29.5 ± 9.6) of all surgically treated ovarian abnormalities in pediatric age [9–11]. Follicular cysts constitutes 10.0–17.2% (mean: 12.1 ± 3.0) whereas corpus luteum constitutes 14.2–26.4% (mean: 17.6 ± 5.1) (Table II) [1,4,8,10,12–14].

Small ovarian cysts are detected incidentally in 2–5% of pre-pubertal children, the major part in neonatal than in childhood due to maternal and placental hormonal stimulation [7–9]. The foetal incidence (30%) is, in fact, disproportionately high considering that the neonatal one is classically 1 per 100,000 births. This is considered the consequence of cysts spontaneous involution or their silent torsion either late in pregnancy (more frequently) or shortly after birth [7].

The incidence of ovarian cysts tends to decrease in early childhood as a consequence of the low levels of gonadotropin and estril but their incidence is increased in the adolescences due to the beginning activity of the ovary. The early period following menarche is commonly associated with dysfunctional ovulation so ovolatary cysts are often seen as the result of aborted ovulations or corpus luteum persistence and this is the reason why both follicular and corpus luteum cysts are extremely common in this period of life.

FOL in childhood and adolescence, as supported by our study and recent literature [10,11,13], may be asymptomatic or associated with menstrual irregularities, just in post-menarcheal girls, abdominal pain, urinary frequency, constipation or pelvic discomfort. In adolescents, it is essential to elicit a full history including details of the menstrual and sexual history [15–18]. The differential diagnosis widens to include a variety of acquired reproductive disorders such as pregnancy, sequels of sexually transmitted diseases or endometriosis [10,18].

The pre-operative diagnostic work-up of ovarian pathologies in children and adolescents includes US scan and blood samples for tumour markers [15,19]. The diagnosis is greatly aided by the use of imaging; the widespread availability and US use has resulted in higher detection rate of functional cysts [10,18].

Trans-abdominal US is a non-invasive tool and it may be helpful in differentiating ovarian pathologies from appendicitis and other acute surgical conditions. US examination in skilled hands is an excellent tool for distinguishing benign tumours and FOL from malignant adnexal masses. Experienced US examiners usually use the ‘pattern recognition’ method, a subjective evaluation of the grey-scale US image, often supplemented by colour-, power- or spectral-Doppler US, to make a diagnosis of an adnexal mass [20,21].

Different morphological scoring systems are also used to identify malignant ovarian masses; the DePriest and Ueland one seems to be a very useful procedure for differentiating benign from malignant ovarian tumours in children [17,22,23].

US has proven to be very valuable not only in diagnosis, but also in allowing observation of these lesions over time. When feasible, it may be useful to perform a trans-vaginal scan also [10,23,24].

Hemorrhage in an ovarian lesion can lead to a diagnostic dilemma; in fact it may appear ultrasonographically complex or solid [10,12,17]. Balan [10] reported a patient where an ovarian lesion was considered as highly suspicious of malignancy at

| Table II. Incidence of pediatric functional ovarian cysts: a review of the literature. |
|---------------------------------|-----------------|-----------------|---------------|-----------------|-----------------|---------------|
| Surgically treated ovaries     | 34              | 140             | 106           | 47              | 40              | 129           | 67             |
| Functional cysts              | 14 (41.2%)      | 61 (43.6%)      | 26 (24.6%)    | 12 (25.5%)      | 10 (25.0%)      | 14 (17.1%)     | 22 (32.8%)     |
| Follicular                     | –               | 24 (17.2%)      | 11 (10.4%)    | 5 (10.6%)       | 4 (10.0%)       | –             | 12 (17.9%)     |
| Corpus luteum                  | –               | 37 (26.4%)      | 15 (14.2%)    | 7 (14.9%)       | 6 (15.0%)       | –             | 10 (%)         |
US that resulted in hemorrhagic corpus luteum cyst after surgery.

Difficulties in identifying lesion histology before surgery is due to the fact that an histological lesion corresponds to several US pictures and vice versa. Moreover sometimes, even macroscopically, may be difficult to define the exact nature of a lesion, as reported by Doret et al. [25,26].

The accuracy of US, CT and MRI scans in diagnosis of pelvic pathologies are 77%, 87% and 97%, respectively, in accordance with Balan et al. [10]. Ten gonads (10.0%) presented as US complex lesions in our series. In presence of complex or solid lesions it is compulsory to perform other investigations such as CT or MRI scans, to distinguish ovarian disorders from other diseases such as acute appendicitis, appendix abscess, tubo-ovarian abscess or hydrocolpos/hematocolpos [10,23,27,28].

Though US, CT and MRI can help in the diagnosis, only surgery and histopathological analysis allow precise identification of the exact nature of a complex lesion [1,3,27,29]. Ovarian masses in fact cover a wide range of pathologies, from more or less secretory tumours and benign but complex anomalies to highly aggressive and invasive tumours. Generally, recognition of the precise histological type of the lesion goes beyond mere histopathological classification, prognosis and treatment depend on it [29].

The size of the tumour is not an indication of its potential malignancy. In some voluminous tumours, the upper pole of the mass extends beyond the pubic symphysis and develops within the abdomen. A mass may be also totally asymptomatic and revealed by US [5,30].

According to literature, guidelines for the treatment of adolescents FOL do not exist, but a few parameters must be considered: the age of the patient, the clinical presentation, the size and the US aspect of the cyst. Therapeutic options include simple surveillance, hormonal therapy, percutaneous drainage and surgical enucleation [23,26]. Follicular cysts, as they are hormonally driven, tend to resolve spontaneously in a few weeks (4–6 on average) and just a small number of them requires surgical treatment [15,28,30]. In the presence of asymptomatic cysts under 5 cm in diameter and normal US-based and biochemical parameters, ongoing surveillance is a reasonable option [15,19]. Surgical indications, although not absolute, include cysts bigger than 5 cm, a failure of the cyst to resolve or decrease in size spontaneously, complex or solid lesions indicative of suspected malignancy, severe persistent abdominal pain and complications such as ovarian torsion, hemorrhage or infarction [10,13,19].

Final treatment is defined by the diagnosis, but clearly significance is placed on future reproduction: in order to preserve ovarian function, we performed conservative surgery whenever feasible [15,30]. When all clinical, radiological, biological and endoscopic criteria of benignity are met and the organic mass is small in size, lesion excision can be envisaged, either within the abdomen by laparoscopy or outside the abdominal cavity after externalisation of the adnexa by a short incision at the level of an iliac fossa [30].

Conservative treatment consists in the enucleation of the cyst with ovarian reconstruction so as to preserve reproductive function as far as possible [11,28]. In our series a conservative surgery has been performed in 18 cases (81.8%).

With a large lesion it may be impossible to preserve ovarian tissue and oophorectomy is then necessary, conserving, if possible, the ipsilateral Fallopian tube [30]. Oophorectomy is also the choice procedure in ovarian torsions [28,31]. Some authors reported that detorsion may result in conserving follicular function independently from the pre-detorsion size and from the colour of the ovary, even if this is seen to be black [1]. There is an important debate about the importance of prophylactically fixing the contralateral ovary [1,15,18].

It is not clear if oophorectomy affects the fertility of these young patients [1,31]. It is known that women with a single gonad are more prone to infertility that controls. However, it is reported that patients who have had an oophorectomy have the same pregnancy rate as the general population [32]. Furthermore, in animals it has been shown that there is a compensation by the contralateral ovary consisting in a functional and an anatomic hypertrophy but it has not been demonstrated in humans [19,32].

Until now, literature shows that in pediatric practice conventional surgery is the method of choice for the treatment of gonadal pathology in infancy and childhood and it still holds a considerable place, although the present trend is toward increasing the use of laparoscopy [33]. In recent years, following the experience with adult patients, laparoscopy has been used in the management of presumed benign ovarian cysts in children [33–35]. Literature also shows that laparoscopy is associated with fewer adhesions than conventional open surgery but most pediatric surgeons continue in choosing laparotomy for large cysts due to technical difficulties and the possibility of malignancy [18,35–38].

In conclusions, US and CT may be helpful in differentiating FOL from other surgical conditions characterised by abdominal pain but, as confirmed in our experience, a certain diagnosis may only be made at the time of surgery. Indications for intervention, as showed in our experience, included the detection of complex or solid cysts indicative of suspected malignancy, severe or persistent abdominal pain and complications and of persistent simple cysts greater than 5 cm in diameter. The cut off of 5 cm for surgery indication is controversial; some authors, in fact, suggest that in a stable patient with an appearing benign, cystic, unilocular lesion under
10 cm in diameter the simple surveillance can be considered an acceptable option [38].

Finally, the goal of surgical management of pediatric is to remove the lesion preserving the underlying ovary in order to optimise the conservation of steroidogenesis and fertility.

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References


