

An adolescent with an asymptomatic adnexal cyst: To worry or not to worry? Medical versus surgical management options

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Summary. Paraovarian cysts or paratubal cysts (PTCs) arise from either the mesothelium or from paramesonephric remnants. These present as either adnexal mass or as an incidental finding. Diagnosis is usually established on ultrasound and it is important to differentiate these from ovarian cysts. Typically PTCs appear as simple cysts by ultrasound and are indistinguishable from ovarian cysts if one does not recognize the extraovarian location. Occasionally, PTCs have internal echoes due to hemorrhage. PTCs are usually asymptomatic and benign. The differential diagnosis includes a simple ovarian cyst, peritoneal inclusion cyst and hydrosalpinx. Malignant changes have been reported in about 2% to 3%, and it should be suspected if papillary projections are present. PTCs management depend upon the presence and severity of the symptoms, the cyst size and US characteristics, CA 125 results, age of the patient and the risk of malignancy. Simple PTCs can be expected to regress and may be managed expectantly. When surgery is indicated, a joint multidisciplinary management by the paediatric surgeons and trained paediatric gynaecologists should be the gold standard. (www.actabiomedica.it)

Key words: paraovarian cysts, paratubal cysts, diagnosis, treatment, complications

Introduction

In gynecology, the adnexa refer to the region adjoining the uterus that contains the ovary and fallopian tube, as well as associated vessels, ligaments, and connective tissue. Pathology in this area may arise from the uterus, bowel, retroperitoneum, or metastatic disease from another site, such as the breast or stomach (1, 2).

The adnexal cysts may be classified as paratubal or paraovarian depending on their proximity to either the tube or the ovary (1-4). Both are usually used synony-

mously and have been reported in all age groups, beginning from premenarchial period up to menopause (1).

The incidence of PTCs is not clearly known. In an Italian population an incidence of 29 per 1,000 (~3%) has been reported with a peak age of occurrence in the third and fourth decades of life (3). In pediatric and adolescent population, a much higher incidence of PTCs was reported (7.3%) (5).

PTCs usually range in size from 2 to 8 cm. The smaller cysts are most commonly found in middle-aged women (in the 30 to 40 years age group), and are often indistinguishable from simple ovarian cysts.

Larger PTCs (up to 20 cm) tend to develop in younger women, quite often during pregnancy, at which time they have a tendency to grow rapidly (1-4).

The aim of this report is to present an adolescent referred to our adolescent outpatient clinic for a persistent “large unilocular ovarian cyst”. The most common types of extraovarian masses are reviewed with emphasis on diagnosis and management.

Case presentation

A 15-year-old virgin adolescent was referred, in March 2013, for persistent “large unilocular ovarian cyst” (4.3 x 4 cm) contiguous with, but not clearly separate from, the ovary. The cyst was found incidentally on pelvic sonogram 6 months before, because of menstrual irregularity and was treated for 3 months by her general practitioner with a contraceptive pill without success.

Her family and medical histories were unremarkable. Menstrual cycles were irregular with no dysmenorrhea. Her breast and pubic hair were at Tanner stage 5. The patient standing height was 158 cm (26th centile), weight 56 kg (63th centile) and body mass index (BMI) 22.4 Kg/m². Her pulse rate was 88 beats/minute and blood pressure 105/60 mmHg. Abdominal examination was negative. No signs of peritoneal irritation were noted. The patient did not have hirsutism, acne or galactorrhea.

Her lab investigations at the time of presentation were as follows: estradiol: 84 pmol/L; follicle-stimulating hormone (FSH): 4.0 IU/L; luteinizing hormone (LH): 5.4 IU/L; prolactin: 15.1 ng/ml; testosterone: 1.1 nmol/L and thyroid-stimulating hormone (TSH): 3.5 mIU/L. The routine urine, haematological and biochemical profiles, and CA-125 level were within the normal range.

Ultrasonographic (US) scan of the pelvic region showed a normal sized uterus, with a thin and regular endometrial lining. The left ovary was normal. In the right adnexal region a unechogenic thin-walled cystic mass (4 x 3.8 cm) with regular contours was visible (Figure 1). The right ovary was not visible. No free abdominal fluid was noticed. She had her last menstruation 24 days back.

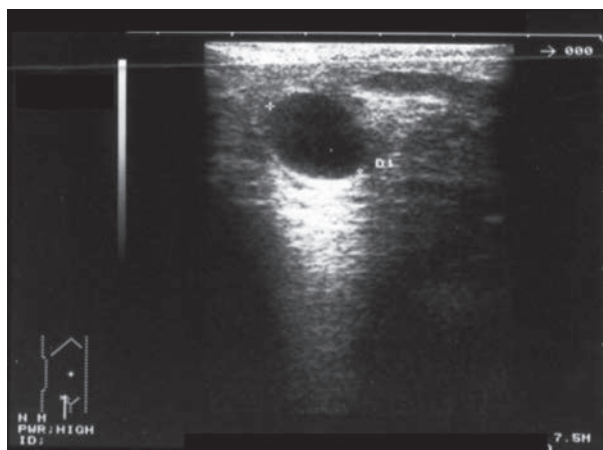


Figure 1. Ultrasound image of the right adnexa. An unechogenic thin-walled cystic mass (4 x 3.8 cm) with regular contours is visible. No free abdominal fluid was noticed

Abdominal magnetic resonance imaging (MRI) showed a cystic lesion (4 x 3.5 cm in size) in the abdomen that was separate from the right ovary and thought to be a paraovarian cyst.

The patient was discharged with a diagnosis of paraovarian cystic mass and managed conservatively. She was followed up clinically and by US every 6 months. The patient responded well to conservative management, and a significant regression in the size of the cystic lesion was observed at the end of the 12-month follow-up. A complete resolution was observed after 24 months without any need for surgical intervention. At last clinic visit her menstrual cycle intervals were between 24 to 35 days.

Discussion

PTCs are not uncommon. They rarely cause symptoms and therefore are usually incidentally found. The symptoms occur when they grow excessively, or in case of hemorrhage, rupture, or torsion. They arise from either the mesothelium or from paramesonephric remnants or from the invagination of the tube's serosa (creating a mesothelial cyst). The mesonephric ducts begin to develop at 20-30 days of gestation and contribute to the development of the male reproductive excretory system (vas deferens, epididymis and seminal vesicles) (Figure 2). In females, these ducts remain as

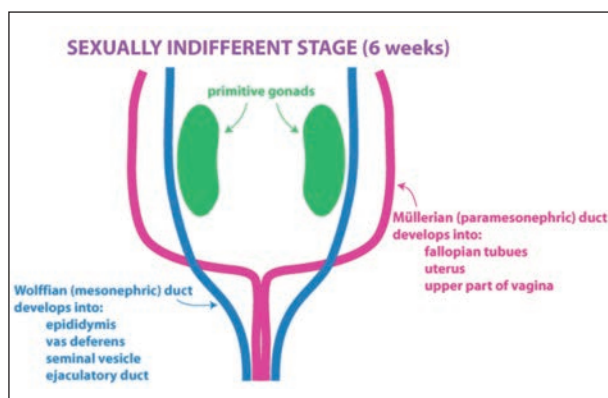


Figure 2. Schematic representation of paramesonephric and mesonephric ducts at 6 weeks of gestation

vestigial structures and are often located in the broad ligaments. Parts of the epithelial lining may unusually remain active and continue to proliferate resulting in cystic masses (6).

In one large study, laparoscopic evaluation showed that these cysts were paratubal in 40% of patients and paraovarian in 60%. They were unilateral in 67.7% and bilateral in 15.3%. More than one small cyst occurred on one side in the remaining 17% of patients (7).

Small PTCs are usually asymptomatic and may disappear spontaneously. However, patients with larger PTCs frequently complain of pelvic pain, usually on

one side, irregular periods, abnormal uterine bleeding, and dyspareunia. These symptomatic PTCs may need a laparoscopic procedure for histological diagnosis and treatment (8, 9).

Diagnosis is usually established by US imaging. It is important to differentiate these PTCs from true ovarian cysts. US can accurately diagnose PTCs in 87.5% of cases. When the mass is large or cannot be visually separate from the ovary an additional MRI may be necessary (10-12).

The majority of these cysts are benign. However, in rare cases borderline tumours and carcinomas have been reported arising from paratubal and paraovarian masses (1-2% of cases) (1, 2, 5, 13). In a retrospective study of symptomatic cysts by Savelli et al. (14) an incidence of 3-5% of malignancy was reported.

Certain sonographic findings (mural nodules, internal blood flow, papillary excrescences and ascites) are more pertaining to malignancy than others (15-21).

MRI may be useful when the ipsilateral ovary is not visualized separately, in the presence of complex paraovarian masses or suggestive US features of neoplasia. MRI offers better delineation of the mass and accurate evaluation of its vascularity (22).

The differential diagnosis of paraovarian and paratubal cysts includes peritoneal inclusion cysts and hydrosalpinx (23, 24) (Table 1).

Table 1. Ultrasound (US) differential diagnosis of simple paraovarian cysts, paraovarian cystadenomas, hydrosalpinx and peritoneal inclusion cysts

| Paraovarian cyst mass | US characteristics |
|----------------------------------|--|
| Symple paraovarian cyst | Paraovarian cysts can show a wide range of sonographic features. Sonographically they are usually thin-walled, smoothly marginated, unilocular cysts. Occasionally, paraovarian cysts have internal echoes due to hemorrhage. |
| Paraovarian cystadenoma | Paraovarian cystadenomas or cystadenofibromas are uncommon but should be considered when an extraovarian cyst contains a mural nodule or septation. |
| Hydrosalpinx | A hydrosalpinx should be considered when one encounters an elongated cystic mass with a tubular shape, with a partial septation and multiple small nodular areas along the wall because of thickened endosalpingeal folds. |
| Peritoneal inclusion cyst | Peritoneal inclusion cysts are multilocular cystic masses with an irregular, star like morphology and no proper wall; septations are multiple and free to oscillate when moving the probe (flapping sail sign). On sonography, the presence of the ovary inside a large, ovoid or irregular, anechoic cyst is characteristic of a peritoneal inclusion cyst. |

Conclusions

Paraovarian or paratubal cysts (PTCs) constitute about 10% of adnexal masses. Although they are not uncommon, they rarely cause symptoms and are usually incidentally found. The symptoms occur when they grow excessively, or in case of hemorrhage, rupture or torsion. Although malignancy is rare, borderline paratubal tumors have been reported in the literature.

PTCs management depends upon the presence and severity of the symptoms, the cyst size and US characteristics, CA 125 results, age of the patient and the risk of malignancy. US imaging is currently considered as the first-line imaging technique for discriminating between benign and malignant adnexal masses. However, this technique is highly dependent on the expertise of the examiner (25-27).

Simple PTC can be expected to regress and may be managed expectantly. US morphological and functional properties must be periodically monitored as an alternative to surgery since malignant transformation is rare. However, "a cautious decision is necessary because larger and prospective patient series are needed to more definitely answer the question of which patients can be managed expectantly, and which patients need surgical management" (28).

When surgery is indicated, enucleation of the cyst from the mesosalpinx, with careful avoidance of cyst rupture or damage to ovary and fallopian tubes is recommended (29, 30). Joint multidisciplinary management of girls by the paediatric surgeons and trained paediatric gynaecologists should be the gold standard.

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