GIANT MUCINOUS OVARIAN CYSTADENOMA IN 13-YEAR-OLD PREMENARCHAL GIRL

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Abstract

Ovarian tumors represent less than 2% of all tumors in girls under 16 years of age. Mucinous cystadenomas (MCs) generally occur in adult life and are an extremely rare finding before menarche. To the best of our knowledge, only 19 cases have previously been reported in the literature. We present the rare case of a 13-year-old premenarchal girl who was admitted to the hospital with lower abdominal pain, fever and a palpable abdominal mass. Abdominal ultrasonography disclosed a multilocular cystic mass measuring 10x30x25 cm that virtually occupied the entire abdomen. Neither ultrasonography (U/S) nor magnetic resonance imaging (MRI) could identify the origin of the mass, due to its enormity. At laparotomy, the giant tumor was found to originate from the left ovary, which contained a small amount of gelatinous peritoneal fluid. Unilateral oophorectomy with tumor removal was performed. Histology revealed a benign mucinous cystadenoma of the left ovary. Regular follow-up with ultrasonography has shown no sign of recurrence. Albeit a rare entity, this diagnosis should be considered in young girls aged 11-15 years who present with a very large abdominal mass.

Key words: ovary, mucinous cystadenoma, premenarchal girl

Introduction

Ovarian tumors account for less than 2% of tumors in young girls under 16 years of age[1]. Mucinous cystadenomas (MCs) are benign and represent 15% of ovarian tumors. They contain mucinous fluid and differ in size[2]. Usually unilateral, they are more prevalent in women aged between 20 and 40 years but have also been reported in postmenopausal women. Their incidence is exceptionally rare in young, premenarchal girls[3]. Like serous cystadenomas, MCs are considered as non-functional childhood tumors. They vary greatly in diameter, often occupying a considerable part of the peritoneal cavity and are described as encapsulated, multilocular cystic lesions with a smooth surface, without adhesions to neighboring organs[4]. A mucinous cystadenoma can cause discomfort, particularly if it compresses adjacent organs, such as the bladder, rectum and ureters. Other complications include torsion, suppuration and perforation. Malignant transformation is not common; it has been reported in just 5-10% of cases. Herein, we describe the rare case of a young, premenarchal girl aged 13 years who presented with a mucinous ovarian cystadenoma.

Patient-method

A 13-year-old girl presented to the outpatients department at Aglaia Kyriakou Children's Hospital with a 4-day history of intermittent lower abdominal pain, bloating and a fever reaching 38.5°C. The patient reported that for the past 4 months, she had experienced recurrent lower abdominal pain, constipation and a gradual increase in swelling of the abdomen.

Clinical examination revealed considerable abdominal distension and a large, palpable mass that occupied the entire abdominal cavity (pic 4), thought most likely to be a cyst.

Laboratory tests showed WBC: 7.4x10/μL (64%NE, 27%LE), CRP: 92mg/L, LDH: 212U/L, β-HCG: 0.0U/L and AFP: 1.7ng/mL.

Ultrasonography and MRI (pics 1,2,3) identified a sizeable multilocular cystic mass that occupied the entire abdomen and lesser pelvis, extending upward between the abdominal paracolic sulcus toward the anterior liver surface.

A mild dilatation of the pyelocaliceal system of both kidneys was also detected, while a chest x-ray showed elevation of the diaphragm. It was not possible to identify the origin of the mass, due to its enormity.

At laparotomy, a huge multilocular cystic mass was found to arise from the left ovary (pic 5). The cyst had caused a slight tear to the lower part of the ovary, from which exuded gelatinous fluid. Sample fluid was obtained for cytologic examination, which proved negative for cancer cells. The patient underwent en bloc resection of the mass with ipsilateral oophorectomy; the fallopian tube was preserved (pic 6). This procedure was followed by an appendectomy.

The patient's postoperative course was uneventful. She was discharged from hospital on the 9th postoperative day. Histologic examination of the mass confirmed a benign ovarian cystadenoma, measuring 40cm at its greatest diameter and weighing 11.5 kg.

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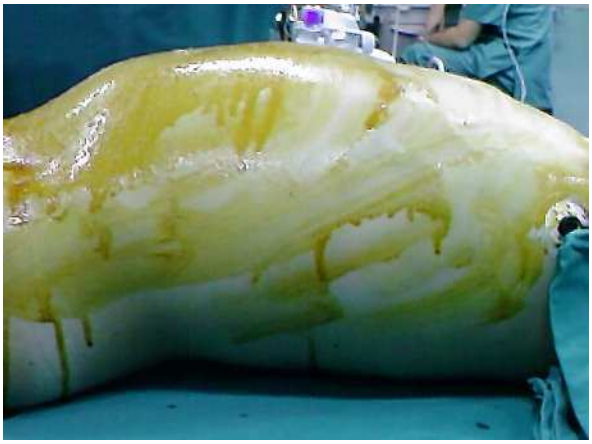
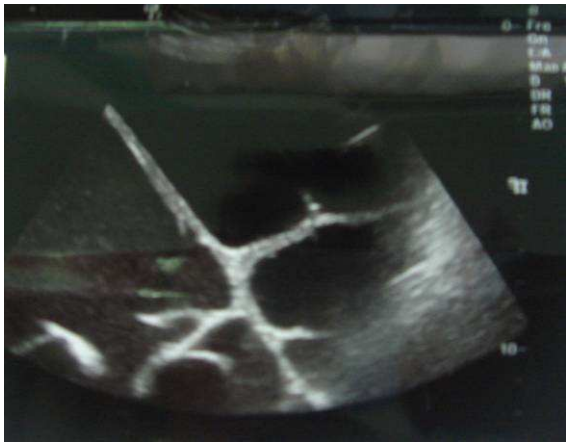
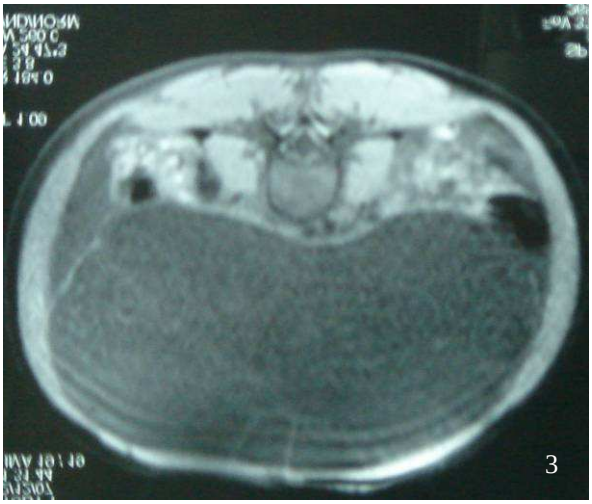


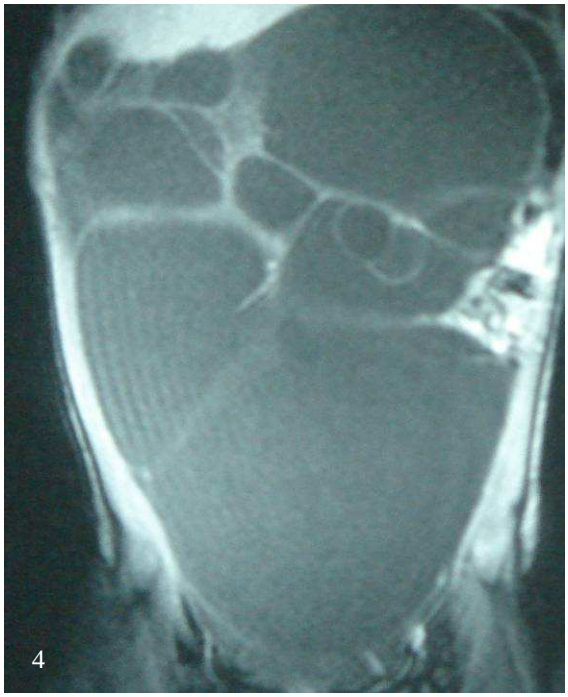
Picture 1: Patient showing the abdominal distention.

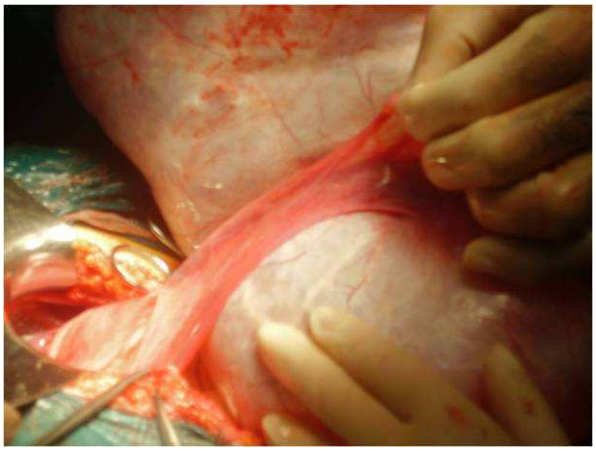


Picture 2: Ultrasound scan showing a large cystic mass containing multiple septa.

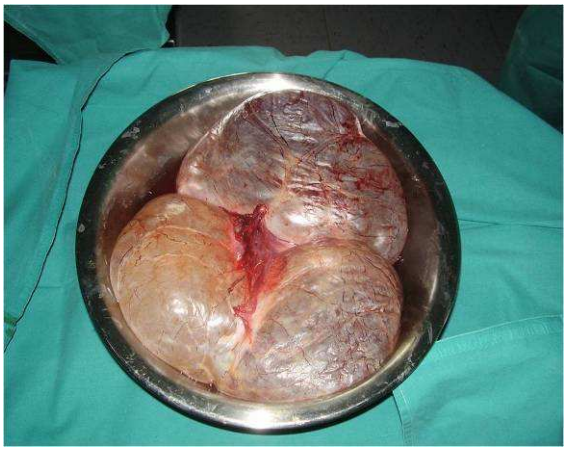


Picture 3,4: MRI scan of the abdomen.





Picture 5: Large multicystic tumor originating from the left ovary.



Picture 6: Mucinous cystadenoma of the left ovary.

Discussion

Ovarian tumors in children constitute an unusual entity[1,5,6]. Sixty-four percent of these tumors are malignant and derive from mesenchymal cells[1,6], while just 10-17% stem from epithelial cells. The most common of epithelial tumors are benign cystadenomas, of which 75% are serous and 25% mucinous. The latter constitute an extremely rare finding in premenarchal girls[1]. Mucinous cystadenomas are sizeable tumors, often multilocular, containing sticky gelatinous fluid. Microscopically, they comprise a tall columnar epithelium that produces mucus. Histopathologically, MCs fall into three categories[9,10]:

1) Benign cystadenomas without cellular stratification and without stromal infiltration
2) Borderline malignancy with cellular stratification up to 2-3 layers, without stromal infiltration
3) Mucinous carcinoma with extensive cellular stratification exceeding 3 layers, and stromal infiltration

Prognosis is favorable, with malignant transformation occurring in only 5-10% of cases. In 2003, Morowitz et al presented a 14-year retrospective study of epithelial ovarian tumors in patients with a mean age of 13.9 years. Borderline malignancy was reported in just 4 cases of serous cystadenomas; no malignant transformation was noted for MCs[6]. In 1993, Skinner et al reported a case of MC in a 13-year-old girl[11]. Deprest et al, in a literature review among 2,225 ovarian tumors, just one 12 year-old patient presented with MC of borderline malignancy[12]. At the same review, Deprest described only 4 cases of epithelial carcinoma in premenarchal girls. Flosho et al reported a similar case[13]. In 1984, Morris et al described 4 cases of young girls under 15 years of age and 6 cases of malignant cystadenomas out of 172 cases of ovarian tumors in patients under 15 years of age [10].

The main clinical symptoms of ovarian tumors include chronic lower abdominal pain and, in 55% of cases, a palpable pelvic mass. Among the more serious complications is ovary torsion, which demands urgent surgical intervention[14].

Ultrasonography is the examination modality of choice. In cases where it is difficult to identify the origin of the mass, computed tomography (CT) and MRI can help establish diagnosis. In the case of the patient presented herein, it was not possible to identify the origin preoperatively due to the enormity of the mass. Transvaginal U/S would appear to be more accurate[15].

The preferred treatment for MCs is cystectomy, either with preservation of ovarian tissue or an oophorectomy. In cases of suspected borderline MCs, in line with the Clinical Practice Guidelines of the Society of Obstetricians and Gynecologists of Canada (SOGC), treatment includes resection of all visible disease, an omental biopsy and appendectomy [16]. According to the International Federation of Gynecology and Obstetrics and the American College of Obstetricians and Gynecologists (ACOG), treatment can either involve a salpingo-oophorectomy or just a cystectomy if the patient wishes to remain fertile[17,18]. There are several reports in the literature of patients managed by simple cystectomy showing excellent fertility rates ranging from 40-70%[19]. The rate of recurrence in cases of fertility preservation varies from 12-37.5%[20]. Infiltration at the surgical margin of the resected tumor is conducive to an increase in the risk of recurrence[21]. As reported by the ACOG, laparoscopic surgery is contraindicated if malignancy is suspected. Postoperatively, the SOGC proposes annual follow-up with ultrasonography and testing for CA-125 only in the presence of poor prognostic factors[16]. In contrast, the International Federation of Gynecology and Obstetrics recommends annual U/S for all patients in whom the ovary was preserved[17,18]. Unfortunately, it has not yet been possible to determine the required duration of follow-up with any degree of certainty, given that recurrences have been reported as late as 10 years after surgery, particularly in cases of borderline MCs[22]. A recurrence rate of up to 60% has been reported for borderline MCs, mainly within the first year[23].

Conclusions

The mucinous ovarian cystadenoma is a rare disease among premenarchal girls under 15 years of age. Oophorectomy is considered the treatment of choice. While ovarian preservation for fertility reasons is considered a safe method, it demands a far more rigorous follow-up after surgery. In cases that do not present a risk for malignancy, follow-up with U/S is adequate.

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