A 9-kg Ovarian Mucinous Cystadenoma in a 14-Year-Old Premenarchal Girl

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Conflict of interest: None declared

Patient: Female, 14
Final Diagnosis: Ovarian mucinous cystadenoma
Symptoms: Abdominal enlargement • abdominal pain • constipation
Medication: —
Clinical Procedure: —
Specialty: Obstetrics and Gynecology

Objective: Rare disease
Background: Although ovarian tumors are most commonly observed in adults, they relatively rarely occur in children. The majority of ovarian masses encountered in the premenarchal or childhood stages are non-neoplastic lesions such as benign functional cysts. Epithelial tumors account for 8–10% of all ovarian tumors and are histologically classified as mucinous or serous. The most common benign epithelial ovarian tumor is cystadenoma.

Case Report: We report the case of a 14-year-old premenarchal girl with chronic abdominal pain, constipation, and abdominal enlargement. A computed tomography detected a huge left ovarian cystic tumor. A 9-kg ovarian tumor was removed surgically. Pathology showed a benign mucinous cystadenoma (MCA).

Conclusions: Ovarian neoplasms in children present a diagnostic quandary, and very often the diagnoses are missed or delayed. When the diagnosis is made, a prompt and fertility-preserving surgical treatment must be performed and followed to prevent recurrence.

MeSH Keywords: Abdominal Pain • Cystadenoma, Mucinous • Menstruation • Ovarian Neoplasms

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Background

Although ovarian tumors are most commonly observed in adults, they relatively rarely occur in children [1]. Most ovarian masses encountered in the premenarchal or childhood stages are non-neoplastic lesions such as benign functional cysts. The clinical signs and symptoms of ovarian masses are usually non-specific. Therefore, making the correct diagnosis preoperatively is sometimes difficult, and early management may be necessary to save the patient’s life and fertility. Epithelial tumors account for 8–10% of all ovarian tumors and are histologically classified as mucinous or serous. The most common benign epithelial ovarian tumor is cystadenoma, of which 75% are serous and 25% are mucinous cystadenoma [2,3]. We present here the case of a premenarchal 14-year-old girl with a giant ovarian mucinous cyst adenoma.

Case Report

A 14-year-old premenarchal girl was admitted with a 4-month history of chronic abdominal pain, constipation, and abdominal enlargement. Although she had palpated a mass in her abdomen 1 month before presentation, it had not been noticed by her parents. Her physical examination showed abdominal distension and a hard mass extending from the pelvis to the xiphoid process. Abdominal ultrasound demonstrated a giant multiloculated cystic mass extending from the pelvis to the level of the processus xiphoid. CT showed an enormous mass occupying almost the entire abdomen (Figure 1). Neither USG nor CT could identify where the mass had originated from. The tumor markers CA 19-9: 237 U/ml (0–33 U/ml) and CA-125: 144 U/ml (0–16.3 U/ml) were elevated, but CEA, α-fetoprotein, and human chorionic gonadotropin levels were within the normal range. The patient’s routine blood analyses and renal functions were normal: white blood cells 9.43×10³/mm³ (4.3–10.8×10³/mm³); hemoglobin 12.9 mg/dL (12–18 mg/dL); hematocrit 39% (35–53%); alanine transaminase 17 units/L (5–40 units/L); aspartate aminotransferase 18 units/L (5–42 units/L); urea 18 (10–50) unit; and creatinine 0.61 mg/dL (0.2–1.2 mg/dL).

Laparotomy was performed by midline incision. The smooth-surfaced mass filled the abdomen (Figure 2A, 2B). At surgical exploration, the tumor was found to originate from the left ovary. The right ovary was of normal size and shape. The examination of the pelvis, abdominal walls, diaphragmatic surface, and peritoneum was not indicative of implants or metastases. There was no free fluid in the abdomen. Because of the absence of normal ovarian tissue, a left salpingo-oophorectomy was performed. The tumor measured 40×25×25 cm and weighed 9 kg. Macroscopic examination revealed that the material had a smooth surface, dirty white color, and increased vascularity on the tumor surface (Figure 3). On gross examination, the cystic mass was filled and expanded, with thick tenacious mucinous material, and was multiloculate with thin septa. Histopathologic examination was remarkable for the cystic structure lined with mucinous epithelium. Cysts did not have papillary formations (Figure 4). The tumor was diagnosed as a mucinous cystadenoma of the ovary.
The early postoperative course was uncomplicated and there were no problems detected at 10-month follow-up. CA-125 and CA 19-9 levels were 140 U/ml (0–16.3 U/ml) and 10.4 U/ml (0–33 U/ml), respectively, on the third postoperative day. CA-125 level reduced in the normal range within the third postoperative month.

Discussion

Primary cysts and tumors of the ovaries are uncommon in children. One-third of these masses are non-neoplastic. Only 14% of neoplastic tumors are epithelial tumors. The tumors are usually serous and mucinous. The most common types of epithelial neoplasms encountered are benign cystadenoma, of which 75% are serous cystadenomas and 25% are MCAs. Mucinous tumors of the ovary occur mainly in the middle-aged adults and are extremely rare prior to menarche [4,5].

In general, ovarian MCAs tend to present with abdominal distension. An ultrasound scan is the first line of investigation with a palpable mass. When there is a diagnostic doubt, a CT or an MRI scan is performed. Generally, it is not possible to accomplish ovary-sparing surgery in this tumor, which is too big, despite its benign nature. The treatment for benign MCA usually has consisted of salpingo-oophorectomy and no further treatment is required [3]. Differential diagnoses of ovarian masses include mesenchymal hamartoma, choledochal cyst, hydrops of the gallbladder, congenital splenic cyst, pancreatic pseudocyst, pancreatic cystadenoma, hydronephrosis, multicystic dysplastic kidney, multilocular cystic nephroma, adrenal hemorrhage, mesenteric and omental cysts, gastrointestinal duplication cyst, meconium pseudocyst, ovarian cysts and cystic neoplasms, hematocolpos, urachal cysts, appendiceal abscess, and abdominal and sacrococcygeal teratoma [4]. In this case, neither USG nor CT could identify where the mass had originated from. Diagnosis was possible after laparotomy.

Tumor markers are used as part of the preoperative investigation for differential diagnosis in most cases, but they may be important to monitor postoperatively for complete resection of the mass, as well as relapse. CA-125 is an antigen determinant of high-molecular-weight glycoprotein. It is also specific for epithelial differentiation and is related to tumor volume; therefore, it has potential for identifying ovarian epithelial tumors. CA 19-9 has been widely used for a variety of malignancies in many tissues. It is also correlated with larger-diameter tumors like the one in the present case, because the cystic walls of larger tumors are thinner, which may cause rupture or inflammation, and more active secretion of CA 19-9. Tumor markers CA-125 and CA 19.9 are released from the damaged tumor epithelium at sites of adhesions and epithelial shearing stresses. The subsequent high local concentration of tumor markers at these sites is resorbed into the systemic circulation [7]. CA-125 and CA 19-9 levels were initially high in our patient. The level of CA 19-9 declined to normal levels by the third postoperative day and the level of CA 125 declined to normal levels within 3 months.

The usual treatment of obviously benign ovarian tumors usually consists of cystectomy or ipsilateral oophorectomy or salpingo-oophorectomy. Biopsy of the opposite ovary has been regarded as unnecessary when it is grossly normal. The therapeutic strategy was suggested to take both the cure and preservation of fertility into consideration [8]. We performed the same strategy in this case and did not take a biopsy from the contralateral ovary.

Giant ovarian tumors can cause abdominal compartment syndrome. Timely and aggressive resuscitation, prompt surgical decompression, and intensive perioperative hemodynamic management are required for patients with ovarian mucinous cystadenoma complicated by abdominal compartment syndrome [9]. We performed surgery promptly. Mucinous cystadenoma recurrence is apparently not as rare as reported in

![Figure 3. Postoperative mass appearance (40×25×25 cm).](image)

![Figure 4. Histopathologic examination (H&E ×40). The cystic structure is lined with mucinous epithelium.](image)
the literature. Intraoperative cyst rupture and cystectomy instead of adnexectomy have emerged as risk factors for recurrence [10]. Therefore, the tumor should be removed carefully and completely.

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Conclusions

In children ovarian neoplasms present a diagnostic quandary, and very often the diagnosis are missed or delayed. When the diagnosis is made, prompt and fertility-preserving surgery must be performed with the aim of preventing recurrence.