A Rare Case of Mucinous Borderline Tumor in Adolescent Age Accompanied by Complete Vaginal Obstruction

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Abstract- Ovarian tumors are rare in childhood and adolescent age. A 14-year-old girl presented with abdominal distention and mild cyclical abdominal pain since 3 months ago. There was an abdominal distention, and huge firm mass was palpated from pelvis to epigastric region. Abdominal ultrasonography revealed normal uterus and large multiloculated adnexal mass with multiple fine septations. Laparotomy was performed, and the ovarian mucinous borderline tumor was reported in frozen section biopsy. Exploration of other abdominopelvic organs revealed no other pathological signs. The final pathological report showed the right ovarian mucinous borderline tumor. Although the mucinous ovarian borderline tumor is a rare condition in adolescent age, pelvic mass, especially with solid or nodular component, must arise this diagnosis, and exploratory laparotomy with comprehensive surgical staging with regard to fertility preservation is warranted.

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Introduction

Ovarian tumors are rare in childhood and adolescent age. The incidence of ovarian malignancies in these ages is about 1%, and the most common pathologies in these ages are non-epithelial tumors (1). The mean age of diagnosis for epithelial ovarian cancer is about 60 years (2). Borderline ovarian tumors are diagnosed about 10 years earlier than ovarian cancer (3). These tumors are non-invasive and show much epithelial proliferation and cytological atypia than benign ovarian tumors, but less than malignant ones (4). The imperforated hymen is the most common obstructive congenital lesion of the female genitalia; although complete obstruction of the vaginal orifice by the hymen is a rare condition which occurs in 0.05 to 1% of female newborns (5). Although most commonly, this is not diagnosed until menarche at age 13-15 years; delay in diagnostic and therapeutic interventions may cause chronic pelvic pain, hematometra and even hemoperitoneum (6). We present a rare case of mucinous borderline tumor in adolescent age accompanied by complete vaginal obstruction.

Case Report

A 14-year-old girl who presented with abdominal distention and mild cyclical abdominal pain since 3 months ago was referred to an academic hospital for diagnosis and treatment in September 2016. She complained of epigastric fullness and difficult micturation. She had no past medical history except for abdominal distention and lack of menstrual cycle (primary amenorrhea). She didn't use any medication before. In her family, there was no history of primary amenorrhea. At the physical examination, there was an abdominal distention, and huge firm mass was palpated from pelvis to epigastric region. The mass was not tender. We examined her secondary sexual trait for discovering the cause of primary amenorrhea; development of breast, pubic and axillary hair were normal. Pelvic examination showed complete vaginal obstruction with bulging of the bluish hymenal membrane at Valsalva maneuver.

The informed consent was obtained from the patient for publication of this case report and accompanying images.

Rrectal examination showed a huge mass was palpated across and above the rectovaginal septum. Abdominal ultrasonography revealed normal uterus and

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large multiloculated adnexal mass (22×15 cm) with multiple fine septations. So, we decided to obtain abdominopelvic magnetic resonance imaging (MRI) by Gadolinium Contrast agent to confirm the diagnosis, so it performed to evaluate genital and urinary tract anomalies more accurately.

It showed the vagina was filled up with an echogenic thick fluid and large heterogeneous cystic mass of about 19×19×11 cm in size arising from right adenex. The cyst was multiloculated and had septation and solid components that enhancing the contrast material (Figure 1,2,3).

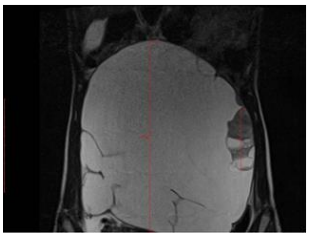

Figure 1. Coronal T2 weighted image shows a cystic lesion with twin septations and a peripheral low signal intensity mass lesion

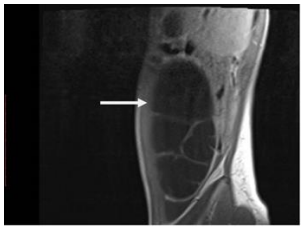

Figure 2. Sagittal T1 weighted images show a cystic lesion with multiple thin septations

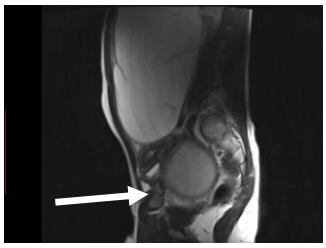

Figure 3. Sagittal T2 weighted images show the fluid collection in vagina compatible with the hematocolpos

A large hematocolpos was seen, but the uterus was completely normal. There was no pathological finding in the gastrointestinal and urinary tract. Lymphadenopathy and ascitis also were not seen.

Tumor marker showed normal carcinoembryonic antigen (CEA), α-fetoprotein (αFP), human chorionic gonadotropin (hCG) and CA 19-9, but CA125 was highly elevated (113 u/ml). Blood analysis, renal and liver function tests were also normal. With regard to the presence of huge abdominal mass and diagnosis of the imperforated hymen, the surgery was planned. At surgery room, in the dorsal lithotomy position, a vertical incision was made on the hymen membrane, and plenty of sticky dark blood was drained from the vagina. The mucosal leaves of the hymen were sutured to the introital edge with interrupted, delayed absorbable sutures. Then, the abdomen was opened with a supraumbilical midline incision. A huge multicystic mass in the size of 20 cm occupied the abdominal cavity from pelvic up to diaphragm. The cyst was multiloculated, its external surface was smooth, and its origin was from right ovary. The left adenex was normal. Decompression of the cyst was done by controlled drainage. Then, ovarian mass excision was done and sent to pathology for frozen section biopsy. The ovarian mucinous borderline tumor was reported by the attending pathologist. Exploration of other abdominopelvic organs revealed no other pathological signs. No pelvic or paraaortic lymphadenopathy was seen, then right salpingo-oophorectomy was done. Left ovary and uterus were normal.

After right salpingo-oophorectomy, the surgical staging was completed by infracolic omentectomy and multiple biopsies from para colic peritoneum, anterior and posterior col de-sac, intestinal mesentery and vesical peritoneum. According to the histopathology report of mucinous type tumor, appendectomy was also performed.

The patient withstood the surgery well with no intraoperative or major post-operation complication, just nausea and vomiting observed until the second day of surgery. Topical conjugated estrogen cream was prescribed for better hymenal epithelialization up to 14 days after surgery. The patient was discharged on the 3rd day after surgery with good general condition.

The final pathological report showed right ovarian mucinous borderline tumor (Figure 4).

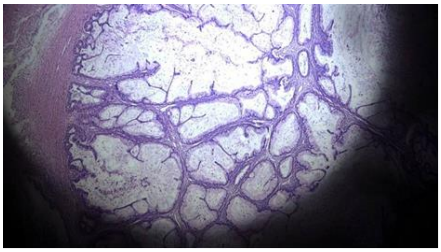

Figure 4. Image demonstrating the ovarian tissue with cysts containing complex gland with mucosecretion and psudostratification nuclei with mild atypia without evidence of stromal invasion (magnification ×10)

All other biopsies were free of tumor. It was reviewed by another expert pathologist for precise diagnosis. The

patient instructed to every three months follow-up for physical examination, ultrasonography, and tumor markers analysis.

Discussion

Ovarian tumors are rare during adolescent age and presented less than 2% of tumors in young adolescents age (less than 16 years) (7). Borderline ovarian mucinous tumors occur in adult age and are extremely rare before menarche (8,9). Mucinous borderline tumors of low malignant potential are usually large, and multiloculated (10), as in the current patient ultrasonography revealed a large multiloculated mass (20 cm in size) with solid and nodular components which emitted malignant tumor.

Large abdominal mass usually causes abdominal distention with or without pain or other organ compression symptoms. In this case, abdominal distention with mild cyclical hypogastric pain was the main symptom. Since she had an imperforated hymen, the cyclical pain was contributed to this anomaly.

The current patient presented as a case of primary amenorrhea that in some other case reports, amenorrhea was reported as a symptom which accompanied to ovarian mucinous borderline tumors. In 2 case reports, both cases had the secondary amenorrhea; one case was a 13-year-old adolescent girl and the second one was a 37-year-old woman with secondary amenorrhea accompanied by ovarian mass (11,12). Maybe the ovarian tumor had a dysfunctional effect on the ovarian hormone production. Precise imaging modalities reported by an expert radiologist could make an accurate diagnosis of ovarian neoplasm. In the current case, reporting of a solid component and nodule in the multiloculated huge cystic mass (in MRI) propound the possibility of neoplastic nature in the mass. In some other studies also, the presence of solid components in borderline mucinous ovarian tumors were reported in the imaging modalities (11). Ahmed *et al*., reported a case of borderline ovarian mucinous tumor in a 37-year-old woman. The tumor had multiple papillary structures along with a small nodule, pathological evaluation showed sarcoma-like mural nodules, but in the current case, the nodule and solid component didn't show any sarcomatoid changes (12).

In patients who want to preserve their fertility, conservative surgery consisting of unilateral salpingo-oophorectomy and appropriate staging is recommended by exploration of the entire abdominal cavity with peritoneal washing, infracolic omentectomy and multiple peritoneal biopsies (13).

It is challenging to determine whether an ovarian mucinous tumor is a primary lesion or a metastatic implant from the gastrointestinal tract, particularly the appendix which can produce mucinous tumors (14). So, careful evaluation of gastrointestinal tract is required to rule out the possibility of metastatic cancer to the ovaries. Also appendectomy is recommended as a part of this comprehensive evaluation; as we performed in the current case.

However, precise radiological evaluations didn't show any other pathologic origin from the gastrointestinal tract before the surgery. We explored all the abdominopelvic organs during laparotomy, and comprehensive surgical staging was performed to identify the worst degree of epithelial proliferation. One study showed that tumor stage≥IC and age<45 years were associated with tumor recurrence (15). Our patient was recommended for regular follow-up every 3 months to detect early tumor recurrence by physical examination, ultrasonography and tumor markers.

Although the mucinous ovarian borderline tumor is a rare condition in adolescent age, abdominal distention with pelvic mass especially with solid or nodular component must arise this diagnosis, and exploratory laparotomy with comprehensive surgical staging with regard to fertility preservation is warranted.

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