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Large Bilateral ovarian Mucinous cystadenoma in an adolescent: Case Report and literature review

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Abstract: Ovarian tumors are rare, account for less than 2% of all tumors in pediatric and adolescent age group, the epithelial type accounts 10-28% out of them. Mucinous cystadenoma considered to be the second most common subtype of epithelial ovarian tumors after **serous** cystadenoma. It tends to be presenting with nonspecific signs and symptoms of lower abdominal pain, fulness and progressive distension, or completely asymptomatic. It can reach huge size and can be benign, borderline or malignant. Primary mucinous ovarian tumors are the second common origin with the appendix as first common site of this histological type. Also, it tends to be unilateral ovarian involvement with risk of 10 % to be bilateral. We are presenting a rare case of bilateral large primary mucinous cystadenoma in a 14 years old adolescent girl with nonspecific symptoms of abdominal pain and distension who was found to have bilateral ovarian mass which was found to be primary ovarian mucinous cystadenoma in both ovaries of endocervical type by histological examination after successful laparoscopic bilateral ovarian cystectomy.
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Key words: Mucinous cystadenoma, Adolescent ovarian tumors, Bilateral ovarian mucinous cystadenoma.

1. Introduction:

Ovarian tumors are quite rare in the pediatric and adolescents age group, representing less than 2% of all tumors while it is about 6% in other age groups. The epithelial type account for about 8-10% of all ovarian tumors in adults, and 10 - 28% in pediatric and adolescents [1,2]. There are more than twelve histological subtypes of epithelial tumors the commonest are serous and mucinous cystadenoma, the mucinous subtype is the second most common tumor of the epithelial tumors, it represents 10-15% of ovarian neoplasm, and they are divided into three categories: benign, borderline and malignant. The Benign mucinous neoplasms include mucinous cystadenoma and mucinous adenofibroma, represent 80% of cases [3]. Primary mucinous cystadenomas are not typically seen in the ovaries, they are most commonly appearing in the appendix, followed by ovaries then the pancreas [4]. in addition, they are substantially unilateral, only 21.3% showed bilateral involvement [5].

In this case report, we present a 14-year-old young girl with bilateral primary mucinous cystadenoma. detailed discussion of diagnosis and treatment strategy as well as therapeutic regimen for this scarce congenital ailment are included.

2. Case:

14-year-old female presented to OPD with history of recurrent abdominal pain for a period of 5 weeks associated with vomiting sometimes, no relieving or aggravating factors not been investigated before, she is a known case of epilepsy on treatment, Menarche at the age of 12.

On examination patient was vitally stable not in pain when seen in clinic palpable pelviabdominal mass that cannot be assessed properly due to mild tenderness with palpation. Ultrasound and MRI shows bilateral ovarian cysts, right side measuring 5.8 x 2.7 cm with septation and large midline/left cyst measuring 9.6 x 8.8 with internal incomplete septation, all tumor markers including B-hCG, LDH, aFP, CA-125, CEA and CA-19.9 were negative. After completing necessary examination, she was taken for laparoscopy. Intra-operative finding was 2 large smooth wall ovarian cyst on both ovaries (Fig.1),

normal abdominal organs, and no peritoneal mucinous material a successful laparoscopic bilateral ovarian cystectomy (Fig.2). Both ovaries were successfully spared (Fig.3). Postoperative histopathology report showed mucinous cystadenoma, Endocervical subtype with no evidence of atypia or malignancy. 6 weeks post-operative patient was having no complain and by ultrasound both ovaries of normal size. Follow up for one year patient was having regular periods no complications and no recurrence of the cyst.

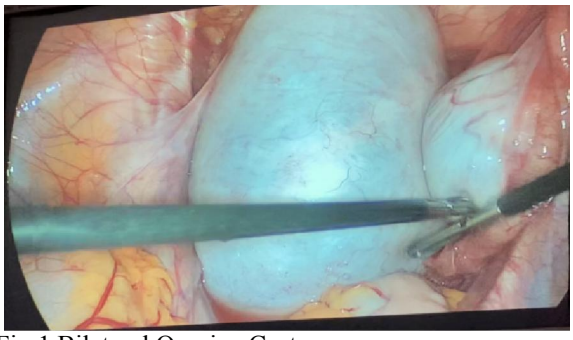

Fig.1 Bilateral Ovarian Cyst

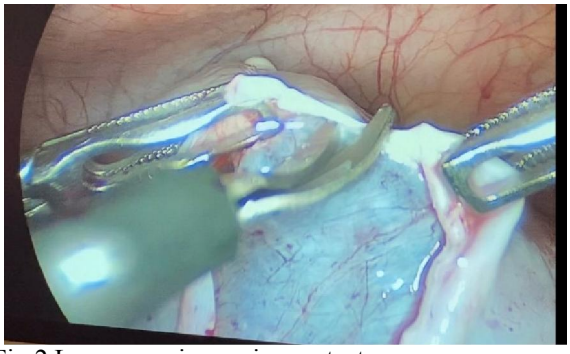

Fig.2 Laparoscopic ovarian cystectomy

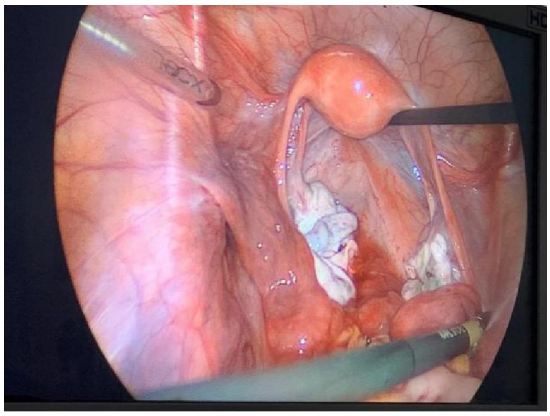

Fig. 3 Immediate post-operative Bilateral Ovarian Sparing

3. Discussion:

Mucinous subtype of epithelial ovarian tumors is considered to be rare and less common than serous subtype in all age groups and rarer in pediatrics and adolescents, it represents around 10-15% of epithelial tumor. The mucinous neoplasm is benign in 80% (mainly mucinous cystadenoma and mucinous fibroadenoma), borderline in16%, and malignant in 4% [3]. Primary mucinous cystadenomas are commonly involving appendix, ovary, and pancreas [4]. Our patient was an adolescent with primary ovarian mucinous cystadenoma of endocervical type which considered to be a rare presentation and also bilateral involving both ovaries with the same pathological type and it is known that it is usually as unilateral with a risk of less than 10% to be bilateral [6].

Histologically these neoplasms are lined by mucin-secreting columnar cells with fibrocollagenous changes in the stroma that's resemble cells of the intestine, which makes it difficult to identify the tumor origin, but it was not the condition in our patient as it was of the rare endocervical type. Mucinous tumor can become particularly large with increased risk of rupture, dissemination, implantation and growth of mucin-secreting epithelium in the peritoneal cavity and produce mucinous ascites (pseudomyxoma peritonei) [1,4]. The ovarian cystadenoma has good prognosis with a 5-10% risk of malignant transformation

Signs and symptoms of MCA are nonspecific and commonly they present with progressive abdominal distension or pelvic-abdominal mass which may involve the entire abdominopelvic cavity [5]. It can also present with acute or chronic diffused abdominal pain, fullness, and menstrual cycle disorders [7]. Due to huge size that mucinous cystadenoma can reach it may contain foci of low or undifferentiated epithelium which make surgical frozen pathological diagnosis difficult with a 17% risk of misdiagnosis. That's why the definitive diagnosis is made by histopathology postoperatively [1].

Proper preoperative evaluation of pelviabdominal masses include detailed history, full examination and a thorough review for radiological and laboratory workup [8] is very important for good outcome. Biomarkers such as Cancer antigen 125 (CA-125), CA 19.9, carcinoembryonic antigen (CEA) although not specific but are of great interest because they are easy to obtain and tracked. Normal CEA and CA 19.9 can exclude intestinal or pancreatic neoplasia [4,6,8]. An ultrasound is the first line of radiological investigation, when there is a doubt, CT or MRI scan can be performed [5].

Surgical approach can be individualized and determined by patient characteristics, clinical findings, radiological and biomarkers studies {2,6}. The gold standard surgery for benign and borderline ovarian masses in children and adolescent is cystectomy with ovarian sparing either laparoscopy or mini-laparotomy [4,6]. Laparoscopy is the preferred approach for ovarian mass [6]. But in adolescent patient with large ovarian mass laparoscopy can be controversial due to risk of cyst rupture intraoperatively, limited visualization, and difficulty in extracting the tumor without any spillage [2]. In this situation, a mini-laparotomy can be performed as the preferred option [6], this option has been offered to our patient and discussed with the parents the risk and benefit of each modality and they preferred to go with laparoscopic approach.

The size of tumor is not indicative for malignant potential even if it reaches 30 cm sometimes but it has a concern for rupture recurrence and risk of oophorectomy [8]. Despite these tumors are rare, recurrence occur more frequently (11%) [4]. Therefore, Long term follow up with imaging and tumor marker is recommended due to the possibility of late relapse [2,4], our patient is following now for one-year post-operative with good regular menstrual cycle, no pain and no evidence of recurrence by ultrasound and tumor markers.

4. Conclusion

In conclusion we are reporting a rare presentation of primary ovarian mucinous cystadenoma of endocervical type. with bilateral ovarian involvement how had been successfully treated by laparoscopic ovarian cystectomy and ovarian sparing with one-year follow-up till reporting this case with no recurrences and asymptomatic with regular period. Further follow-up for this adolescent will be continued.

Conflict of interest

The authors have no conflict of interest

Disclosure

The authors did not receive any type of commercial support either in forms of compensation or financial for this study and have no financial interest in any of the products or devices mentioned in this article.

Ethical Approval

Obtained

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