International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Annal A et al. Int J Reprod Contracept Obstet Gynecol. 2021 Feb;10(2):758-760
www.ijrcog.org

pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20210072

Case Report

The treatment approach of a 12 kg giant ovarian mucinous cystadenoma in a 13-year-old: a case report

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Received: 20 December 2020
Accepted: 11 January 2021

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ABSTRACT

Epithelial ovarian tumours are rare in the adolescent age group, accounting for less than 15% of overall ovarian malignancy. Of this, one-fourth of them are mucinous in nature. Mucinous cystadenomas, being precursors of borderline and invasive ovarian tumours, can reach to size as large as 15-30 cm in diameter without having malignant potential. Also, intraoperative management of such large abdominal mass can be challenging. We present a case of a giant unilateral 12 kg mucinous cystadenoma in a 13-year-old young girl, that grew over a period of 1 year. She underwent left salpingo-oophorectomy after decompression of the mass without any complications.

Keywords: Giant mucinous cystadenoma, Ovarian neoplasm, Paediatric ovarian malignancy

INTRODUCTION

Epithelial ovarian tumours account for 65-75% of overall ovarian malignancies of which mucinous cystadenomas make up to one-third of the cases.[1] It most commonly occurs in middle-aged women and is rarely diagnosed among adolescents. Most of the ovarian masses diagnosed in the younger age group are germ cell tumours. Mucinous tumours are encountered in less than 10% in women younger than 20 years.[2] It grossly presents as multiple cysts of variable sizes that are notorious for reaching large dimensions without necessarily indicating malignancy. Decompression of such large abdominal mass can be challenging. We present such a case of a 13-year-old adolescent girl with giant mucinous cystadenoma of 40 cm, radiologically suspected to be malignant, but histopathology turned out to be benign, managed without any complications.

CASE REPORT

A 13-year-old girl presented with a six-month history of abdominal pain and distension to the gynaecology department. She attained her menarche at the age of 12, and her menstrual cycles were regular. There was no history of malignancy in the family. Her previous medical and surgical history were unremarkable. On examination, her general condition was good. Her per abdominal examination revealed large tense cystic mass extending from pelvis to xiphisternum.

Her tumour markers were CA 125: 33 U/ml, CA 19-9: 1.87 U/ml, carcinoembryonic antigen: 2.44 ng/ml, Alpha-fetoprotein: 1.1 ng/ml, serum lactate dehydrogenase: 213 U/L and beta HCG: 0.1 mIU/ml. Patient's routine blood analysis and renal function test were within the normal range. Her abdominal ultrasound demonstrated a large multi-septate cystic mass reaching 36 weeks size of pregnant uterus. Computed tomography of abdomen and pelvis showed a large thick-walled cystic lesion of 36x29x20 cm filling almost entire abdomen and pelvic cavity with multiple internal septations. The lesion appeared to arise from left ovary. There were no solid areas. The post-contrast study showed enhancement of wall and septation. Mild ascites and anterior wall oedema with moderate bilateral hydroureteronephrosis were

noted. Based on the imaging diagnosis, the tumour was suspected to be malignant cystic ovarian neoplasm.

A multidisciplinary tumour board was convened, and the decision for open left salpingo-oophorectomy was taken. Laparotomy was performed with midline vertical incision. Intraoperatively minimal ascites with left large ovarian mass filling the entire abdominal cavity, with an intact capsule was noted. No obvious implants or metastasis was found on diaphragmatic surface, liver, stomach, colon, abdominal wall and pelvis. The cyst wall was punctured with precaution, and around 12 litres of mucinous content was drained without any spillage into the abdominal cavity. The patient's hemodynamic was monitored, and adequate fluid replacement with crystalloids (0.9% normal saline and Hartmann's solution) was commenced. To prevent hypovolemic shock resulting from excessive fluid loss, fluid resuscitation with colloids (500 ml of hydroxy ethyl starch) was started. Left salpingo-oophorectomy was done, and the specimen was sent for frozen section. An intraoperatively frozen biopsy was reported as mucinous cystadenoma of left ovary without solid components. On histopathology, macroscopic examination of the tumour revealed white, smooth surface without any solid areas or papillary projections. The diagnosis was confirmed to be benign surface epithelial ovarian neoplasm, morphologically consistent with mucinous cystadenoma of the left ovary.

The postoperative course was uneventful, and the patient was discharged on 4th postoperative day. A clinal follow up after six months was found to be normal.

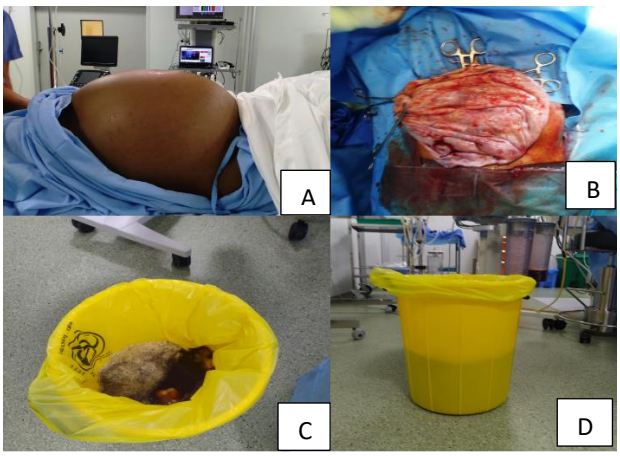

Figure 1: (A) Grossly distended abdomen before surgery; (B) intra-operative finding of capsule of left ovarian mass after drainage of mucinous fluid; (C and D) 12,000 cc of dark haemorrhagic mucinous content of left ovarian mass.

DISCUSSION

Ovarian tumours constitute 1% of all childhood malignancies and 8% of abdominal masses in children, of which 10-30% can be malignant.[2] Germ cell tumours are the most common ovarian malignancies in the adolescent age group. Epithelial ovarian tumour accounts for less than 10% below 20 years and even more rare before menarche.[3] Among the ovarian epithelial cystadenomas, three-fourth are serous, and one fourth are mucinous in nature. These epithelial tumours can be benign, borderline, or malignant. The ovarian mucinous borderline tumour was included in the FIGO classification of cancer in 1971 as those with proliferating epithelial cells and nuclear stratification but without stromal invasion.[4]

These tumours are generally asymptomatic in early stages but can cause pressure symptoms over time because of the mass effect on the surrounding organs, manifesting outwardly as abdominal distension. Thus, they are diagnosed late, especially in people with low socioeconomic status and lacking awareness. Abdominal pain, nausea, menstrual irregularities, constipation, and increased frequency of micturition are the frequent symptoms and torsion, rupture or haemorrhage are the most common complications of these cysts. In our case, the patient was asymptomatic with long-standing abdominal distension.

Transabdominal sonography is the initial modality of choice in adolescents. CT/MRI pelvis is used for additional information like the extent or nature of the lesion. Serology tumour markers (AFP, LDH, CA-125, inhibin and beta HCG) help narrow down the type of ovarian tumour. Increase in CA-19-9 can be non-specific as it can rise with rupture or inflammation of large masses. Differential diagnosis of large abdominopelvic masses in this age group includes cystic lesions of the liver and biliary system like mesenchymal hamartoma, choledochal cyst; congenital cysts of spleen or pancreas, hydronephrosis or multi cystic dysplastic kidneys, gastrointestinal lesions such as mesenteric or omental cyst, genitourinary urachal cyst and ovarian tumours like teratoma, cystadenoma, functional cysts. The most remarkable previous description of the giant ovarian cyst was Spohn et al who described a mass of mucinous cystadenoma of 148 kg in 1922.[5]

A multidisciplinary team of gynaecologist, anaesthetist, dietician and psychologist should be involved preoperatively. Intraoperative diagnosis of borderline mucinous cystadenoma is misleading and can have sensitivity as low as 50%.[6] The challenging part of these large-sized tumours is management during the course of the surgery as it can be potentially dangerous if not managed correctly. The massive size of the mass could compromise the patient's haemodynamic and pulmonary function, which emphasizes monitoring of central venous pressure as there is marked fluctuation of venous return during the intraoperative period. Studies have shown splanchnic dilatation and pooling of venous blood following sudden decompression of large abdominal masses.[7] Appropriate fluid management with crystalloid

and colloid solutions helps stabilize the central venous pressure. Considering potential malignant risks, spillage during drainage can cause upstaging of the tumour and adhesions in the peritoneal cavity. Therefore en-bloc resection if possible, is a preferable treatment option. In our case, even though en bloc was not possible owing to the massive size of the tumour, drainage of content was done without spillage and complication with proper monitoring.

Borderline mucinous cystadenomas have a 5-year survival rate of 98% with a low recurrence rate of 6%.[8] Due to the good prognosis of these cases, oophorectomy as treatment is necessary and sufficient. If proven, biopsies of the contralateral ovary are not required as most of them are stage 1 tumours. Long term follow-up is advised as cases of recurrence until 20 years have been reported.[9]

CONCLUSION

Mucinous cystadenomas can grow to massive dimensions without having malignant potential and pose a challenge during surgical management. The diagnosis may be missed or delayed in children or adolescents due to rarity and diagnostic dilemma. Borderline tumours diagnosed intraoperatively by frozen biopsy can be treated by fertility-preserving surgery after careful intraperitoneal exploration. A multidisciplinary approach with collaboration with anaesthetist, dietician and psychologist aids in minimizing the risk of complications.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Annal A, Khalid ZA, Francis S. The treatment approach of a 12 kg giant ovarian mucinous cystadenoma in a 13-year-old: a case report. Int J Reprod Contracept Obstet Gynecol 2021;10:758-60.