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Case Report

A 16-year-old with borderline mucinous tumor of the left ovary

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ABSTRACT

Incidence of ovarian malignancy in children and adolescents is 1 to 1.5% with non-epithelial tumors being the most common type. Ovarian mucinous tumors commonly seen in third to sixth decade, are rarely seen in adolescents. Timely diagnosis and management are important to save patient's life and fertility. We are presenting an interesting case of a 16-year-old girl with atypical borderline mucinous ovarian tumor.

Keywords: Adolescent, Mucinous borderline tumor, Ovarian neoplasms, Salpingo-oophorectomy

INTRODUCTION

Ovarian tumor is a relatively rare entity in children and adolescent population with an overall incidence of 1 to 1.5%.¹ Non-epithelial tumors such as germ cell tumors or sex cord-stromal tumors comprise the most common histological variety in this group. Epithelial tumors account for 2% of tumors in this age group. Accurate early diagnosis and treatment is crucial to save patient's life and fertility. Ovarian mucinous tumors are epithelial neoplasms seen commonly in the third to sixth decade of life.² Ovarian mucinous tumors can be classified into 3 categories based on histopathology (benign, borderline malignant, malignant). Ovarian mucinous cystadenomas of borderline malignancy in adolescents are exceedingly rare.

CASE REPORT

A 16-year-old girl came to the out-patient department of our hospital with complaints of gradual distension of abdomen since last 2 months. It was associated with generalized, dull aching pain in abdomen. She had no bowel- bladder complaints. She had attained her menarche at the age of 14 years. Her cycles were regular with average flow. She had no menstrual complaints. Her past

and family history was all unremarkable. On examination, she was moderately built and nourished. Her vital parameters were normal. No lymph nodes were palpable. Abdominal examination revealed a tense cystic mass of 34 weeks size extending up to xiphisternum occupying entire abdominal cavity, giving dull note on percussion with limited side to side mobility. Per speculum or per vaginal examination was not done. Her pre-operative blood investigations were within normal limits. Ultrasound pelvis showed large cystic lesion with multiple internal echoes, soft tissue and internal vascularity within it originating from right adnexa. Right ovary was not seen separately from the lesion. The mass was displacing bowels and kidneys laterally, features were suggestive of mucinous cystadenoma or carcinoma. MRI of abdomen and pelvis revealed a 24.6×16×10 cm, well defined peripherally enhancing multiloculated cystic lesion with locules of varying signal intensities, with thin and thick septae within, occupying abdomino-pelvic cavity in midline, more towards right side extending from S3 to D12 vertebral levels (Figure 1). Probable impression of ovarian neoplastic etiology probably arising from right ovary was made after MRI study. No evidence of abdominal metastasis was found on MRI imaging. Her tumor marker levels were as follows-Ca 125-107 IU/ML (increased), CEA-4.49 IU/ML, CA19.9-207.58 U/ML (increased),

serum LDH-437.1 U/L (increased), alfa fetoprotein-1.85 IU/ML. RMI score was 107.

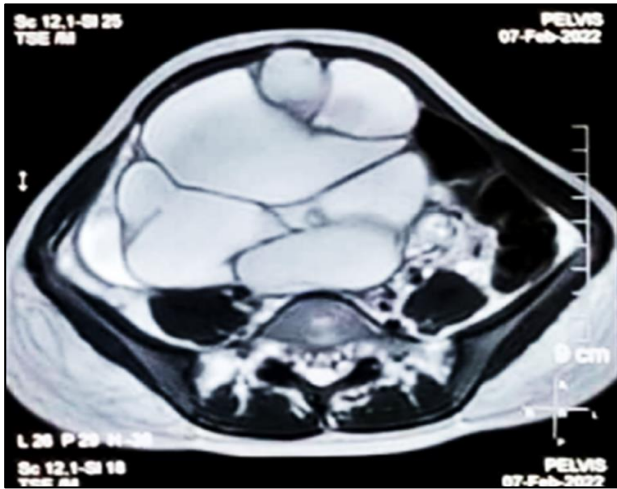


Figure 1: MRI image of the adnexal mass.

Due to high suspicion of malignancy on examination and imaging, opinion from oncologist was taken. Upper GI endoscopy was done to rule out upper GI malignancy which revealed no abnormality. Plan for Exploratory laparotomy with midline vertical incision for surgical staging followed by frozen section was made. Intraoperative examination revealed a multilobulated, fluid filled ovarian cyst of 20cm×15cm×15cm size with variable consistency, smooth surface with intact capsule with no surface excrescences, arising from left ovary. Left ovary and fallopian tube were not seen separately from it (Figure 2 and 3). Uterus and right adnexa were normal. 20cc of serous peritoneal fluid present in pelvic cavity was sent for cytological examination. Other abdominal organs including appendix and omentum were normal. Left Salpingo-oophorectomy and infra-colic omentectomy were performed. Frozen section of the cyst was suggestive of benign cystadenoma of ovary.



Figure 2: Intra-op findings: large ovarian cyst with variable consistency with intact capsule measuring 20×15×15 cm.



Figure 3: Postoperative specimen of ovarian cyst.

Post-operative period was uneventful. Histopathology of the tumor was suggestive of atypical proliferative (borderline) mucinous tumor of the left ovary. Areas of congestion and mesothelial hyperplasia were noted on the omental histopathology with no evidence of tumor implants. Patient was followed up regularly and recovery period was uneventful with normal menstrual cycle. Patient was referred to oncology center in view of borderline ovarian malignancy where she was advised regular follow up with ultrasonography and tumor markers.

DISCUSSION

Ovarian tumors are uncommon in children with an incidence of approximately 2.6 cases per 100,000 girls per year.³ Epithelial ovarian cancer is common in adults whereas in children, it comprises only 1.9% of all ovarian neoplasm.⁴ The histologic subtypes of epithelial ovarian tumors in children are serous and mucinous tumors with serous tumors being the common variety.⁵ Borderline epithelial ovarian tumors, also known as tumours of low malignant potential, form the most common histological variant in women less than 25 years of age.⁶ Patients usually present in early stage of disease. The examination, imaging study and raised tumour marker levels raise the suspicion of malignancy.⁷ Unlike germ cell tumors, epithelial ovarian tumors frequently may indicate an elevation of CA 125 level. In the case discussed here, the large mass felt per abdomen with highly elevated epithelial tumor markers (CA 19-9 and CA 125) and MRI findings suggested the possibility of malignancy. The standard treatment protocol involves Staging laparotomy with midline vertical incision followed by cystectomy and frozen section examination. If frozen section is suggestive of borderline tumor, unilateral or bilateral salpingo-oophorectomy should be done taking in consideration future child bearing potential of patient. Removal of all visible metastasis, peritoneal washing and infra-colic omentectomy is warranted. If the contralateral ovary looks

normal at exploration, biopsy or oophorectomy is not necessary, because this may lead to adhesion formation and may jeopardize future fertility. Previously appendectomy was performed for ovarian mucinous tumors, but current data suggests that it is not necessary if appendix appears normal and there is no evidence of pseudomyxoma peritonei. The aim of treatment for children and adolescents is to restore the uterus and ovarian function for conservation of fertility potential with complete extermination of disease. It is difficult to classify mucinous tumors as benign-borderline-malignant based on frozen section alone due to large tumor size and subtle histological differences. Histological features of borderline ovarian tumors are stratified epithelial cellular proliferation greater than that seen in benign tumours with varying degrees of nuclear atypia and increased mitotic activity; their lack of stromal invasion distinguishes them from invasive carcinomas. Usually, the prognosis of epithelial borderline tumors is excellent. However, in case of tumor relapse, the 5-year survival rate is approximately 81%; if the tumor becomes malignant, the 5-year survival rate reduces to 68%.⁸ Follow up for disease recurrence with the help of pelvic imaging and tumor markers (CA 125 and CA 19-9) forms an important aspect to detect early recurrence as the recurrence rate within 1 year of treatment in children and adolescents is about 75%. Follow up is necessary every 3 months for the first 2 years, every 6 months for the next 2 years and annually thereafter. As the tumor can recur for up to 20 years after initial diagnosis, long-term follow-up and evaluation of patients with borderline ovarian tumors are crucial.⁹ Careful follow up of these patients by routine surveillance pelvic imaging should be done to detect disease recurrence or progression. Monitoring of CA 125 and CA 19-9 levels for follow up in patients with borderline tumors remains a source of debate in the literature.¹⁰

CONCLUSION

Borderline ovarian tumours are a different category among ovarian tumours common in younger women and have an excellent prognosis. Histological features include degree of cellular proliferation and nuclear atypia in the absence of stromal invasion. Complete and correct staging is vital part of management. Fertility-sparing surgery can be performed successfully in early-stage disease. If in a case with presumed benign disease, a borderline ovarian tumour is diagnosed after primary surgery, histological

review and multidisciplinary approach including gynaecological oncologist are recommended.

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