Case Report

Mucinous Cystadenocarcinoma of Ovary in Preadolescence: An Ordinary Tumor but at an Unexpected Age

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ABSTRACT

Malignant epithelial ovarian tumors are common in adult women, but are rare in childhood. Especially, mucinous ovarian cystadenocarcinoma are extremely rare in pediatric age group, only a limited number of cases are reported in this age group till date with no case reported in preadolescence age in the literature. We describe a case of mucinous cystadenocarcinoma in eleven year old premenarchal girl presenting with symptoms of abdominal distension and respiratory distress from last three months.

Key words: Cystadenocarcinoma, mucinous, ovary, preadolescent

INTRODUCTION

Malignant ovarian mass is a rare entity in children. Norris and Jensen found that < 1% of epithelial carcinoma occur below 20 years of age.[1] The mucinous tumors principally occur in middle adult life and are uncommon before puberty. We are presenting a case of mucinous cystadenocarcinoma in 11-year-old premenarchal girls with complaints of abdominal distension and distress from the last 3 months.

CASE REPORT

An 11-year-old premenopausal girl presented with 3 months history of abdominal distension and respiratory distress. On physical examination, a large abdominal mass was palpable predominantly on left side extending above the umbilical region. Ultrasound (USG) examination shows ovarian mass (12.5 cm × 11 cm × 7 cm) with both solid and cystic components. Multiple loculations with thin-walled septa were seen in the tumor [Figure 1]. There was mild ascites along with mild bilateral pleural effusion. The levels of serum cancer antigen 125 (CA-125), serum lactate dehydrogenase (LDH), and alpha fetoprotein were 179.4 U/ml, 715.9 IU/L, and 0.96 ng/ml, respectively. The patient underwent left salpingo -oophorectomy along with multiple site biopsies of peritoneum, omentum, and pelvic and para-aortic lymph node sampling was done. Grossly, the outer surface of mass was smooth with intact capsule and cut surface having multilocular cyst with solid areas [Figure 1]. The cyst contained mucinous material. Histological examination revealed mucinous carcinoma with moderately differentiated (G2) and lympho-vascular invasion. The fluid cytology had no malignant cells. The omentum, multiple peritoneal biopsy and lymph node specimens had no evidence of malignancy (stage Ia). The immunohistochemistry study showed cytokeratin 7 negative and cytokeratin 20 focally positive (in 5% of tumor cells) [Figure 1]. The postoperative CA-125 and LDH level was normal. In view of lack of consensus, no adjuvant therapy was planned. At 1 year follow-up visit, the tumor marker level, USG abdomen and pelvis study were normal.

DISCUSSION

The mucinous cystadenocarcinoma of the ovary accounts for 5–10% of all ovarian mucinous tumors even in adults.

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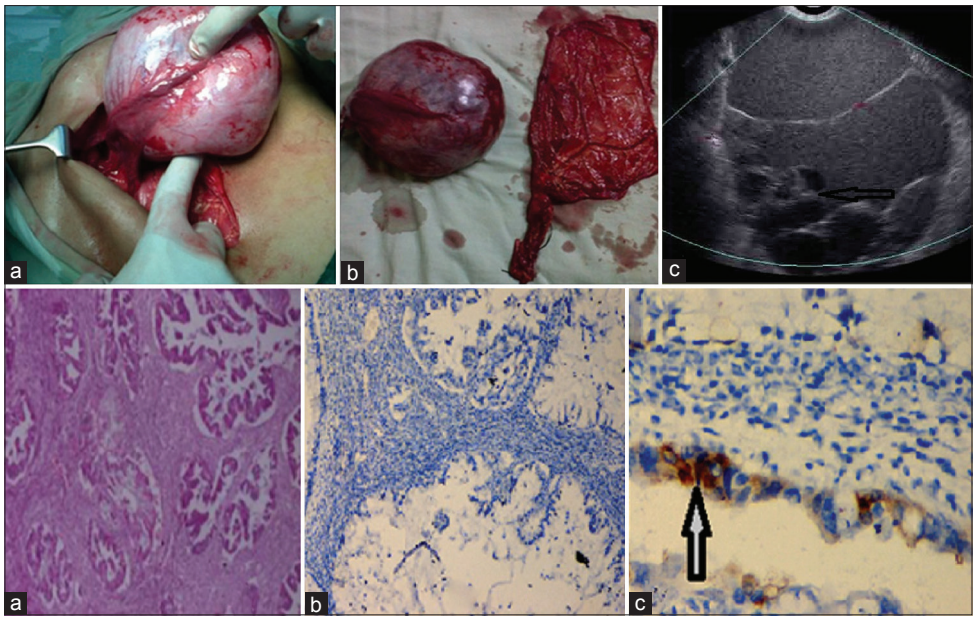

Figure 1: Above (a) intraoperative picture of ovarian mass. (b) Postoperative ovarian mass and excised omentum. (c) Ultrasound shows ovarian mass with both solid, cystic components and loculations with thin-walled septa. Below (a) Histopathological examination showing moderately differentiated mucinous carcinoma with lymph vascular invasion. (b) Immunohistochemistry study showing cytokeratin 7 negative. (c) Focally positive cytokeratin 20 (in 5% of tumor cells)

The incidence of malignant ovarian tumors in < 20 years age group is 22.6% in Indian scenario.[2] The mucinous tumors are filled with a mucus-like material, their name was given on the basis of this property; this mucus is produced by mucus-secreting goblet cells very similar to the cells lining normal intestine. These tumors may become very large. The cystadenocarcinomas contain a more solid growth pattern with the hallmarks of malignancy: Cellular atypia and stratification, loss of the normal architecture of the tissue, and necrosis. The appearance can look similar to colonic cancer. The mucinous ovarian cancer sometimes associated with pseudomyxoma peritonei, where the tumor associated with extensive mucinous ascites and adhesions. Distant metastases are rare and survival is 95% for stage I and 32% for stages II or greater. In early-stage ovarian cancer, lymph node dissection is necessary to make a meticulous staging according to the International Federation of Gynecology and Obstetrics classification and to select an adequate adjuvant therapy.[3] The effect of lymph node dissection on progression-free survival and overall survival in patients with advanced ovarian cancer is still unknown. The prognostic factors for stage I tumors are infiltrative invasion, high nuclear grade, tumor rupture.[4] In early stage (1a and b) mucinous tumors, there is a questionable role of adjuvant chemotherapy or radiotherapy. Hess *et al*. reported that advance mucinous ovarian cancer had a worse outcome as compared to nonmucinous type, with advanced nonmucinous living 3 times longer than those with mucinous pathology.[5] If the adjuvant treatment required, 5 fluorouracil (5-FU) with oxaliplatin- or irinotecan-based chemotherapy can be planned rather than platinum based agents as mucinous ovarian cancer generally do not respond to platinum agents.[6] Sato *et al*. found that the combination treatment of oxaliplatin and 5-FU had marked cytotoxic effects on mucinous adenocarcinoma of the ovary cells, even on cell types known to be resistant to conventional platinum and taxane-based chemotherapy.[7] Shimizu *et al*. conducted a phase II trial in platinum refractory mucinous carcinoma and showed a response rate of 52% and median overall survival of 15.3 months with irinotecan and mitomycin.[8] In our patient, adjuvant chemotherapy was not planned in view of favourable histologic factors and limited data on benefit of chemotherapy in such settings.

CONCLUSION

Mucinous cystadenocarcinoma is rarely encountered in pediatric age group. These mucinous ovarian cancers tend to be detected earlier due to their larger sizes than the more serous type of ovarian epithelial cancer, leading to a generally favorable prognosis. In early stage ovarian cancer, systematic lymph node dissection is required for meticulous staging, which is helpful for adjuvant treatment planning and to explain prognosis. The role of adjuvant chemotherapy in this clinical setting remains doubtful and needs to be investigated by well-designed prospective trials.

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