Case Report

Mucinous Borderline Tumour of The Ovary in A Premenarchal Girl

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Keywords: *Borderline Tumor; Mucinous tumor; Ovary; Premenarchal*

ABSTRACT

Epithelial ovarian tumors are common in adult women, but rare in children. Mucinous cystadenoma (MCA) are rare, with only 16 cases in premenarchal girls reported to date. Low grade (borderline) tumors are even exceptionally rarer. To the best of our knowledge, only few cases of an ovarian borderline intestinal-type mucinous tumor in premenstrual girl have been reported in literature. We present a rare case of an ovarian mucinous borderline tumor in a thirteen year old girl. The histological features of low grade borderline tumors, rapid enlargement of the ovarian mass, the young age and the difficulty in diagnosis, make this case interesting for publication.

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Introduction

Ovarian neoplasm, though unusual in the pediatric age group, if present the majority are of germ cell origin. [1] Mucinous cyst adenoma of the ovary constitutes about 15% of ovarian tumors. [2, 3] It is more common in women between 20 and 40, but is rare in teenagers and exceptional in pre-menstrual girls. [4] Though mucinous borderline tumors can be seen in a wide range from 13-88 years, but still these are extremely rare in young children. [5] As far as we know, though approximately 16 cases of benign mucinous cyst adenoma have been reported in young, only a few case reports of borderline mucinous tumor exist. [6, 7] Here the authors present a rare case of an ovarian mucinous borderline tumor in a thirteen year old girl.

Case Report

We report the case of a thirteen year old girl who presented to our hospital with an increase of abdominal girth during last 20 months. It was associated with mild on & off pain in the epigastric region. Her menarche was not attained and she denied the use of any illicit drugs. There was no significant family history. Physical examination revealed pale skin, a temperature of 36.1°C, respiratory rate 22 breaths/minute, pulse rate 78/minute, blood pressure at 180/140 mmHg, normal cardiac and respiratory sounds. Her abdomen was distended, tense due to ascites and abdominal girth measured 130cm, with dullness on percussion and superficial dilated veins. She was admitted to surgical ward for diagnosis and treatment. Laboratory test revealed normal complete blood count, kidney and liver function test. CEA, Alpha fetoprotein and CA-125 were with in normal level. Ascitic fluid cytology revealed total leukocyte count of approximately 250/cmm with predominantly lymphocytes & occasional neutrophils & mesothelial cell. The chest radiograph showed an upward compression of her diaphragm. Abdominal ultrasonography study showed massive ascites along with a huge multiloculated abdominal mass. The origin of this mass could not be delineated due to massive fluid (Figure1A). Computed tomography (CT) showed heterogenous enhancing large cystic lesion of size approx 30x18 cm arising from left adnexa. There was heterogenous enhancing solid component of 6x6 cm size with multiple enhancing fine and thin septations (2 to 3 mm thickness). The mass was displacing bowel laterally. Ultrasonography showed moderate ascites and no focal lesion was seen in liver. There was no fat component or calcification. No abnormal lymphadenopathy was noted. (Figure 1B) So clinico-radiologically, the provisional diagnosis of malignant germ cell tumor was kept. Preoperatively temlisartan and amlodipine was given to control the B.P of the patient. Left salpingo-oophorectomy was performed.

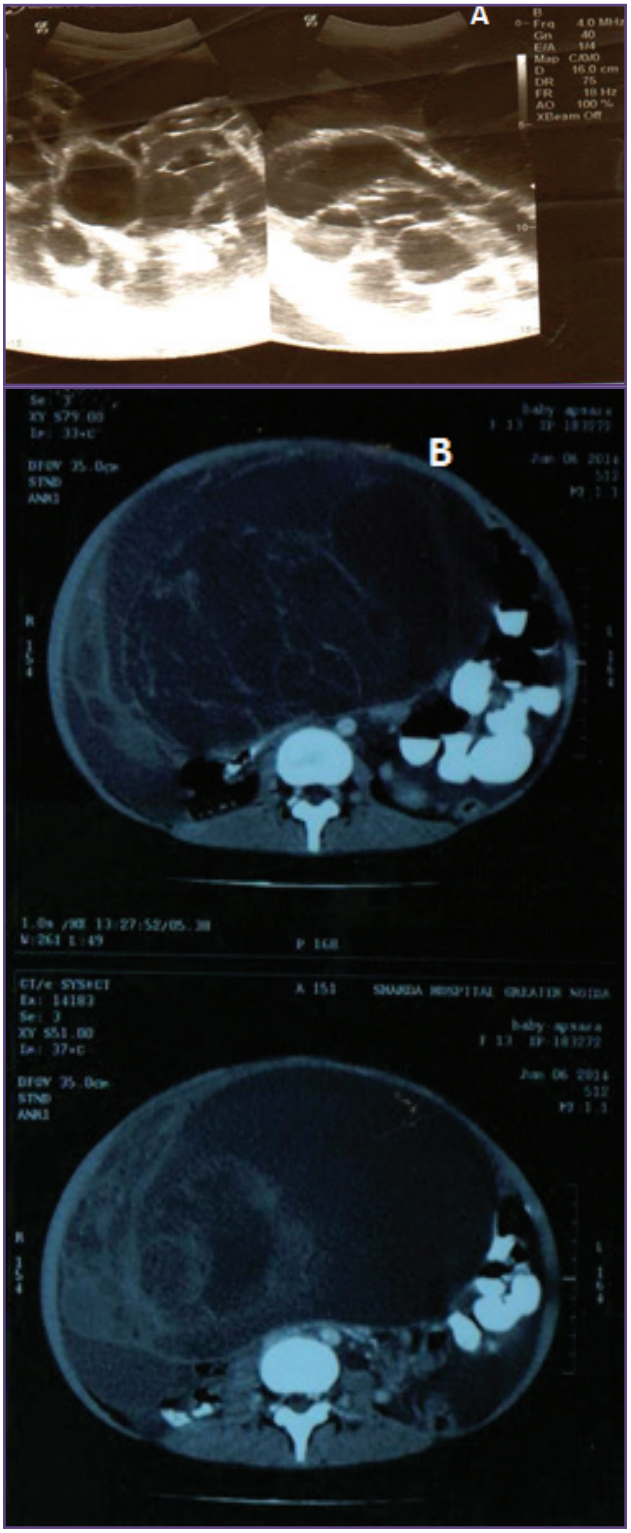

Fig. 1: (A) USG: A complex, multiseptate predominantly cystic mass extending from the pelvis into the upper abdomen. (B) CT SCAN: Complex multiseptate mass arising from the left ovary and a moderate amount of ascites.

(Figure 2A). There was no adverse outcome on surgery. Specimen received in the pathology department, showed an ovarian mass and measured 30x25x10cm and weighed 7 kgs. External surface was smooth and glistening. Cut surface showed a large multiloculated cyst filled with mucin. (Figure 2B). Thirty five sections were taken based on CAP protocol and were subjected to careful microscopic examination. Microscopically the tumor sections showed cysts, glandular and papillary structures lined by epithelium of intestinal type. The epithelium was focally stratified up to three layers and showed mild to moderate atypia at places. No definite stromal invasion was seen. Mitotic figures were rare. Focal areas of necrosis and mild chronic inflammation were seen. (Figure 3, 4, 5) The final diagnosis of borderline mucinous tumor, intestinal type (left ovary) was rendered. Postoperative course was uneventful. No evidence of recurrence has been seen during one year postoperatively.

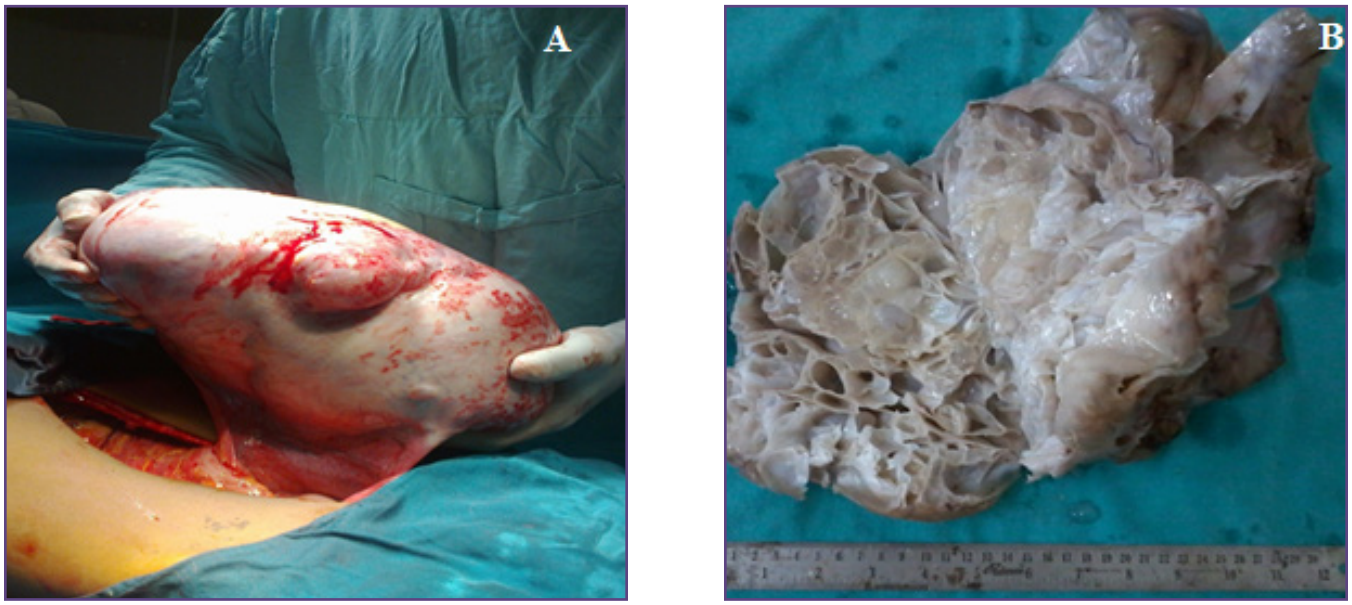

Fig. 2: (A)Tumor delivered from abdominal incision while performing left salpingo-oophorectomy.(B) Cut section showing multilocular mucinous with multiple small cysts filled with mucus material in their lumens.

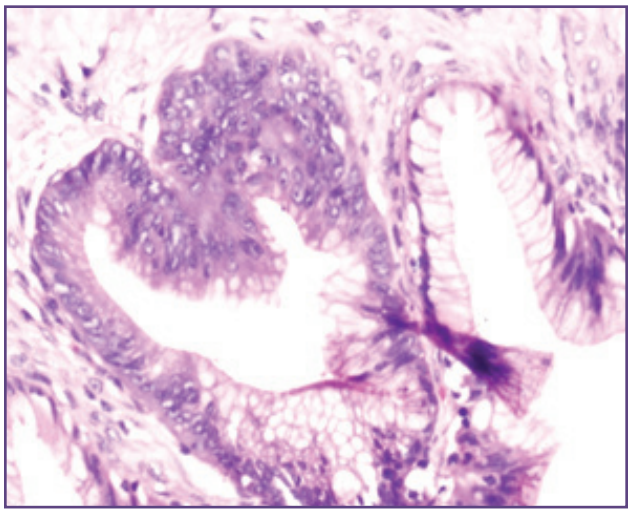

Fig. 3: The papillae were lined by mucinous intestinal type of epithelium displaying stratification at focal areas, hyperchromasia and mild to moderate nuclear atypia (H&E, 40X).

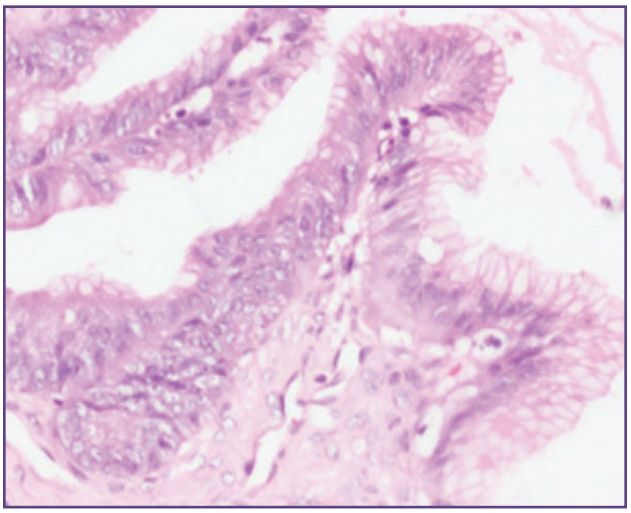

Fig. 4: The central delicate stromal cores supporting intraluminal projections along with focal epithelial proliferation at places (H&E, 40X)

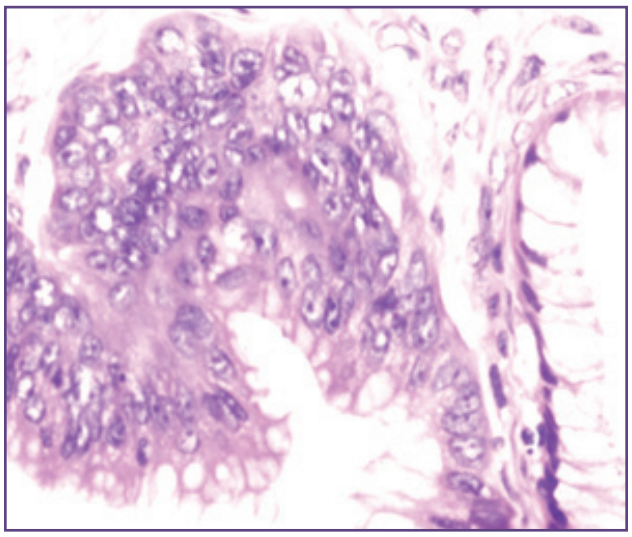

Fig 5: Section showing focus with borderline morphology displaying stratification, moderate nuclear atypia and occasional mitotic figures (H&E, 100X).

Discussion

Ovarian masses in children are an uncommon occurrence. They represent less than 2% of all tumors in girl less than 16 years of age.[1]Mucinous tumors of the ovary including borderline tumors occur in middle adult life and are extremely rare prior to menarche.[2-7]Mucinous borderline tumors of low malignant potential are usually large, multilocular cyst. These exhibit an epithelial proliferation of mucinous type cells greater than that seen in benign counterparts but without stromal invasion. The epithelial components resembling intestinal epithelium almost always contain goblet cells, usually contain neuroendocrine cells and rarely contain Paneth cells. In borderline areas the cells lining the cysts are stratified (usually not more than 3 layers) and may form filliform intracystic papillae with at least minimal stromal support. There is mild nuclear atypia and few mitotic figures are also seen. [9, 10, 11] In our case microscopic examination of huge ovarian mass showed multilocular cyst lined by papillary structures displaying columnar epithelium with stratification up to 3 layers. Fair number of goblet cells and mild to moderate nuclear atypia was evident. Bearing these features in mind, the final diagnosis of ovarian borderline mucinous tumor, intestinal type was given. The patient was discharged on 12th day after removal of sutures. She is doing fine for last two months and has put on 1Kg weight without any treatment.

Surgical management to excise all visible tumor tissue remains the keystone of therapy. Even in advanced disease there is no proven benefit of adjuvant chemotherapy or radiotherapy.[12]

Conclusion

This is a case report of primary retroperitoneal mucinous borderline tumor in teenager who was initially thought to be a surgical case of retroperitoneal mass. On ultrasonography the origin of this mass could be delineated from ovary. Later further on CT scan it was suspected to arise from ovary, considering the age of patient the diagnosis of malignant germ cell tumor was kept. During the surgery the origin was confirmed to be from the ovary. On histopathological examination final diagnosis of mucinous borderline tumor was rendered. Knowledge of the clinical presentation, imaging, and histologic characterization of this entity can lead to correct diagnosis and appropriate treatment as long term follow up is required in such cases.

Acknowledgement

We authors thank Mr. Ankit Singh, histopathological technician, for providing technical help.

Funding

None

Competing Interests

None declared

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