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Case Report

Mucinous cystadenocarcinoma of ovary in 16 years old postmenarchal patient-A rare case report

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ABSTRACT

Malignant epithelial ovarian tumor is a rare entity in children. Especially, mucinous ovarian cystadenocarcinoma is exceedingly rare in pediatric age group, only a limited number of cases are reported in this age group till date with less than 5 cases reported in adolescence age in the literature. We describe a case of mucinous cystadenocarcinoma in a sixteen-year old postmenarchal girl presenting with complaints of abdominal distension and irregular periods from last three months. Management of such cases is tricky as conservative approach involving sparing fertility of the patient is adopted. This case is presented for its rarity and unique presentation.

1. Introduction

In children, epithelial ovarian tumors are mostly benign in nature, comprising 15-20% of cases[1] and approximately 1-2% of all childhood malignancies.[2,3] An estimated incidence of ovarian neoplasms is 2.6 cases per 100,000 per year.[4] After reviewing current literature, it has been seen that malignant epithelial tumors of the ovary are extremely uncommon with <1% of incidence below 20 years of age as reported by Norris and Jensen.[5,6] Worldwide, Only few cases are documented with ovarian mucinous cystadenocarcinoma in the paediatric population. We present a case of sixteen year old postmenarchal girl with ovarian mucinous cystadenocarcinoma.

2. Case Report

A sixteen year old girl presented to gynaecology out patient department at a tertiary care hospital with complaints of abdominal distension and irregular periods for past three months. Abdominal distension was insidious in onset and gradually increasing in size with the presence of abdominal pain for past one week. Patient had irregular menses with polymenorrhoea (3-4 d/ 15-20 d) for past three months. General physical examination was normal. Past history was unremarkable. Abdominal examination showed generalised abdominal distension, 30 weeks size, cystic to firm in consistency with restricted mobility, without any fluid thrill. No hepatospleenomegaly was noted. Vaginal examination revealed uterus in anteverted position with free fornices.

Tumor markers were evaluated and showed normal CA125 level of 18 U/L (normal, 0 to 35 U/L). Complete hemogram, liver function test and coagulation profile were normal. USG whole abdomen and contrast enhanced computed tomography scan of abdomen revealed large, well circumscribed, multilocular, midline abdomino-pelvic cystic lesion predominantly on right side, measuring 23.4cm (SI) * 16.5cm (TR) * 9.0cm (AP), extending from the pelvis upto the epigastrium superiorly with enhancing septas and solid components within suggesting of ovarian cystadenoma. The mass was causing compression of

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lower ends of bilateral ureters thereby causing upstream hydroureteronephrosis (right >left) (Figure 1).

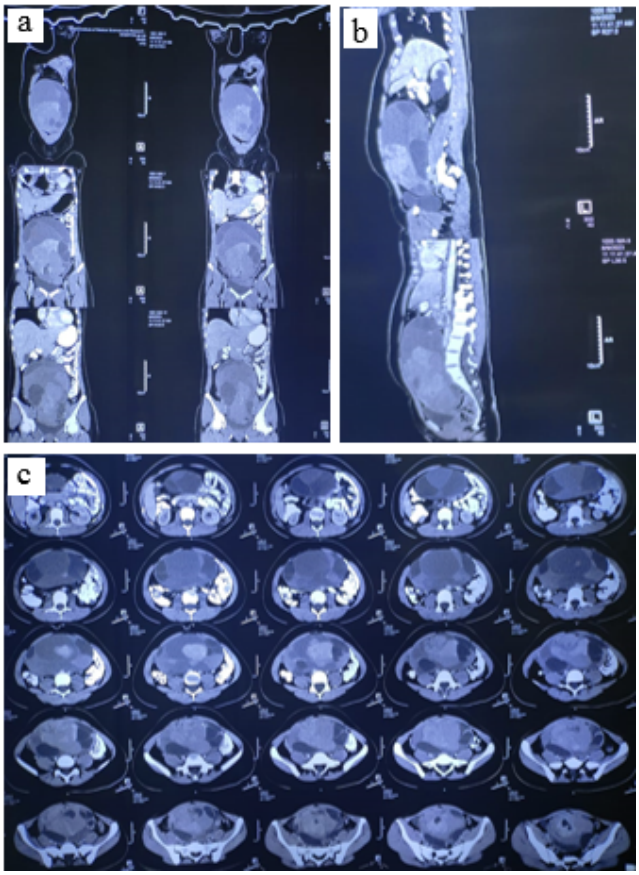

Figure 1: (**a**), (**b**) and (**c**) shows large, well circumscribedabdomino- pelvic cystic lesion predominantly on right side reaching upto the epigastrium superiorly.

Patient underwent laproscopic ovarian cystectomy. Intraoperatively, a large right sided ovarian tumor (~20 weeks size) with intact capsule was noted however surrounding peritoneum and contralateral ovary appeared unremarkable. Two thousand five hundered milliliters of ascitic fluid (mixed consistency with occasional caseous material mixed with blood) was aspirated and sent for cytological examination for malignant cells. Right sided salpingo-oophorectomy was done and sent for histopathological examination and ovarian cystic fluid sent for cytology.

Ovarian cystic fluid cytology was suggestive of epithelial neoplasm. Histopathological examination findings revealed an enlarged ovary measuring 20*14*5 cm with intact capsule and smooth external surface with no papillary excrescences seen. On cut section, mucinous fluid was recovered with multiloculated cystic and solid necrotic areas within. Microscopic sections revealed tumor arranged in the form of multiloculated cysts lined by tall columnar epithelial tumor cells which showed nuclear pleomorphism, immature nuclear chromatin, prominent nucleoli and moderate cytoplasm exhibiting vacuolation. Large pools of extracellular mucin was present. No perineural invasion and lymphovascular invasion was identified. Right fallopian tube and omentum was free of tumor and ascitic fluid was negative for malignant cells. Thus, the final diagnosis of Malignant Mucinous Cystadenocarcinoma was rendered.

Postoperative course of the patient was unremarkable with CA125 level of U/L (normal, 0 to 35 U/L) , beta human chorionic gonadotropin level of 1.20 mIU/ml (normal, <5 mIU/ml), alpha fetoprotein level of 1.65 ng/ml (normal, <10 ng/ml), carcinoembryonic antigen level of 0.89 ng/ml (normal, <3ng/ml) and lactate dehydrogenase level of 373 U/L (normal, <247 U/L). Immunohistochemistry studies showed tumor cells positive for CK7, CK20, CDX2 and negative for SATB2, PAX8, SALL4, AFP, Glypican 3. p53 was positive in tumor cells and Ki 67 was ~20% (Figure 2). The final diagnosis was confirmed as Mucinous Cystadenocarcinoma of the right ovary.

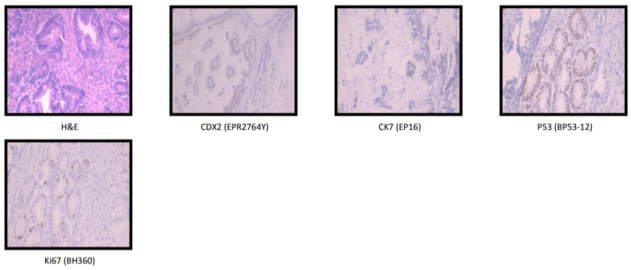

Figure 2: Immuno histochemistry study showing tumor cells with tufting and architectural complexity with papillae and glandular formation, also nuclear pleomorphism, fine nuclear chromatin, prominent nucleoli and moderate cytoplasm exhibiting vacuolation could be seen at places. The tumor cells are positive for CK7, CK20, CDX2 and P53. Ki67 was approximately 20%.

Post-operative Whole Body Positron Emission Tomography and Computed Tomography (PET-CT) scan was suggestive of small cystic lesion noted in right adnexal region with minimal peripheral FDG uptake (measuring ~1.3*1cm) ? residual part of primary lesion (Figure 3). Ultrasound whole abdomen was done which also suggested a cyst like area with thick walls in right adnexa measuring ~ 1.3*1.25 cm that could be considered post operative changes or a residual lesion. Based upon the intra operative findings and histopathological examination suggesting intact capsule of the tumor, patient is kept on close follow up on with serial USG and tumor markers.

3. Discussion

Mucinous cystadenocarcinoma of the ovary is seen in only 5-10% of the cases of all adults among all the ovarian malignant tumors. The incidence of all types of malignant ovarian tumors in < 20 years age group is 22.6% in

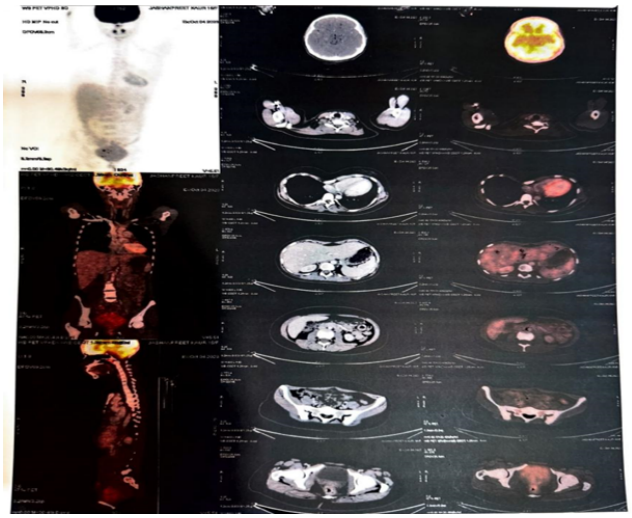

Figure 3: PET CT scan showing minimally metabolic small cystic lesion in right adnexal region measuring ~1.3*1.0 cm. Post surgery inflammatory changes seen in lower anterior abdominal wall with focal nodular thickening along peritoneum.

Indian scenario.[7] Mostly, the patients initially ignore the symptoms that leads to advancement of the disease at the time of diagnosis. Also, the childhood tumors are much more aggressive than their adult counterparts and disease progression is also higher despite treatment.[8]

According to literature, most of the cases of ovarian adenocarcinoma are reported in pre-menarchal girls or girls <15 years of age. Our patient was however post menarchal with age of 16 years. This difference can be explained on the basis of international variability of attaining menarche. Our patient was an Asian girl girl whereas most of the other studies have been reported in American or European girls.

Tumor markers and radiology in association serve as an important tool in the diagnosis of ovarian cancers.[9] CA-125 is the tumor marker for epithelial ovarian tumors however its utility is controversial. Only 50% of patients with Stage I disease have elevated serum CA-125 levels (>35 U/ml), despite of 80% of all ovarian cancer patients having increased CA-125 levels. Non malignant conditions eg liver cirrhosis, endometriosis, first trimester pregnancy, pelvic inflammatory disease and pancreatitis can also present with elevated CA-125 levels. It could also be increased in 40% of the patients with advanced intra abdominal non-ovarian malignancy.[10] Therefore, the raised CA-125 levels should be interpreted carefully and in collacoration with radiology. It is believed that if levels are raised at the time of diagnosis, they can be used as a diagnopstic as well as prognostic marker at follow up for detecting residual or recurrent disease.[11]

In this case also, normal CA 125 levels and large abdominal mass on Contrast Enhanced Computed Tomography scan were suggestive of most likely diagnosis of ovarian cystadenoma but later on, on histopathological studies and immunohistochemistry, it came out to be a rare case of Mucinous Cystadenocarcinoma of ovary. Patient's relatives were advised explorative laprotomy, but they insisted for laproscopic surgery and hence taken up for same.

The intra-operative examination of the other ovary, uterus, fallopian tube, pouch of douglas, omentum and peritoneal surfaces is mandatory as it can often lead to upgradation of the stage of the tumor if macroscopic deposits are seen during surgery.[12] In our patient, all these structures appeared grossly normal and histopathological examination also confirmed the same. Hence, the patient was diagnosed as Stage Ia Ovarian cancer by FIGO (International Federation of Gynaecology and Obstetrics 2018) guidelines. Some authors also recommend a prophylactic wedge biopsy of the uninvolved ovary or biopsy from peritoneal surfaces or lymphnode dissections; however this was not performed in this present case.

In Shankar et al study, the patient underwent left sided oophorectomy for cystadenocarcinoma and received chemotherapy for the same. She presented with abdominal pain after 15 months of her surgery; imaging revealed a large mass arising from the pelvis. On second laprotomy, a solid cystic mass arising from right ovary was excised aling with right fallopian tube. Pathology confirmed a mucinous cystadenocarcinoma of the right ovary without any evidence of local or distant metastasis. But, months later, she presented again with abdominal pain, and after investigation, the patient was diagnosed with metastatic cysadenocarcinoma.[8] In other case report by Hernandez et al, after removal of left ovarian cystadenocarcinoma, the patient had no evidence of disease at 22 months.[13]

The prognostic factors for Stage I tumors are infiltrative invasion, high nuclear grade and capsule rupture.[14] In early stage Mucinous tumors, role of adjuvant chemotherapy or radiotherapy is controversial. Hess et al. concluded that advance mucinous ovarian cancer had a worse outcome as compared to advance non mucinous type, with overall survival 3 times more in advanced non- mucinous type tumors.[15] 5 Fluorouracil (5-FU) with oxaliplatin or irinotecan based adjuvant chemotherapy can be planned rather than platinum based agents as mucinous ovarian cancers are generally refractory to platinum agents.[16] Sato et al reported that 5-FU and Oxaliplatin in combination had marked cytotoxic effects on mucinous adenocarcinoma of ovary, even on cells which are resistant to conventional platinum and taxane based chemotherapy.[17] Shimizus et al. conducted a phase II trial in platinum refractory mucinous carcinoma and found a response rate of 52% and median overall survival of 15.3 months with Irinotecan and Mitomycin.[18] Blom and Tokildsen administered intraperitoneal phosphorus in the paediatric patient with mucinous cystadenocarcinoma;[19] whereas Gribbon et al.

gave intraperitoneal radiotherapy in their two cases of cystadenocarcinoma.[20]

Gupta et al also reported a case of Stage III mucinous cystadenocarcinoma of ovary with omental metastasis in a post menarchal 14 year old girl in which they gave systemic chemotherapy with cisplatin and paclitaxel.[21] A study by Aggarwal et al[22] found that low malignant recurrences have been reported more than ten years after initial surgery even in adult patients. Hence, close follow up is important for young patients also to monitor recurrence. Our patient is also kept on close follow up as according to the intraoperative findings and histopathological report, the capsule was intact and there are more chances that the cystic fluid seen on PET CT scan and ultrasound whole abdomen to be due to post operative changes. This type of malignancy in paediatric age group is unpredictable and has limited information.

4. Conclusion

Mucinous cystadenocarcinoma of ovary is rarely encountered in children. We have presented a case of 16 years old girl with ovarian mucinous cystadenocarcinoma that was treated surgically with unilateral salpingo-oophorectomy of right side. As the patient's histopathological report is suggestive of stage Ia ovarian carcinoma, Patient is now being planned for regular close follow up with serial investigations. Because of the rarity of this malignancy, future studies are needed to establish optimal understanding of natural history and management of such histopatholoigical variety of ovarian tumors in this age group.

5. Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

6. Source of Funding

None.

7. Conflict of Interest

None.

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