International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Rare Case of Mucinous Cystadenocarcinoma in a Young Patient

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Abstract: ***Epithelial ovarian tumors can occur in girls as young as 15 years, but the median age at diagnosis is 63 years, and most cases are diagnosed in women 55 - 64 years of age. Also, benign tumors are more common in young age compared to malignant lesions. Here we present a rare case of a large mucinous adenocarcinoma in a 25yr old woman.***

Keywords: Epithelial, ovarian, mucinous, adenocarcinoma

1. Introduction

Epithelial ovarian tumors are rare in children comprising 15% to 20% of cases [1]. Not surprisingly benign cystadenomas are common in younger age group and malignant lesions are rare. This case is presented for its rarity and unique presentation of mucinous cystadenocarcinoma in a young female.

2. Case Discussion

25 years, unmarried female reported to gynae OPD with H/O pain abdomen for one year. It was dull generalized ache, with no associated menstrual, urinary or bowel complaints. On P/A - mass was felt arising from pelvis and occupying whole abdomen, mobile from side to side, non tender. There was no ascites

Investigations:

- CA125 – 44.60U/ml
- Rest all WNL

USG abdomen and pelvic organ
Imp - Multiseptated cystic lesion showing papillary projections likely left ovarian origin? Mucinous cystadenocarcinoma

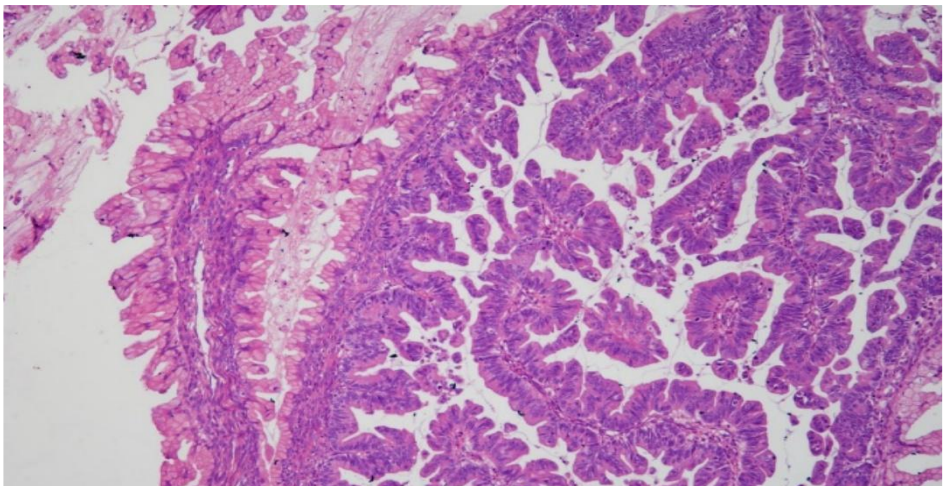
MRI Pelvic Organ at IGMC
Imp - large multiseptated cystic abdominopelvic lesion arising from left ovary with presence of papillary projections and enhancing solid nodule? Mucinous cystadenocarcinoma

Per operative finding
Grossly - Cystic mass of size approx.28*20cm, tense, smooth surface, transparent
On cut sec – there was e/o drainage of 900cc of mucoid, brownish fluid from the cyst. e/o 2 solid areas over the cyst wall mx 2*1cm each with no e/o multiple loculations or papillary excrescences

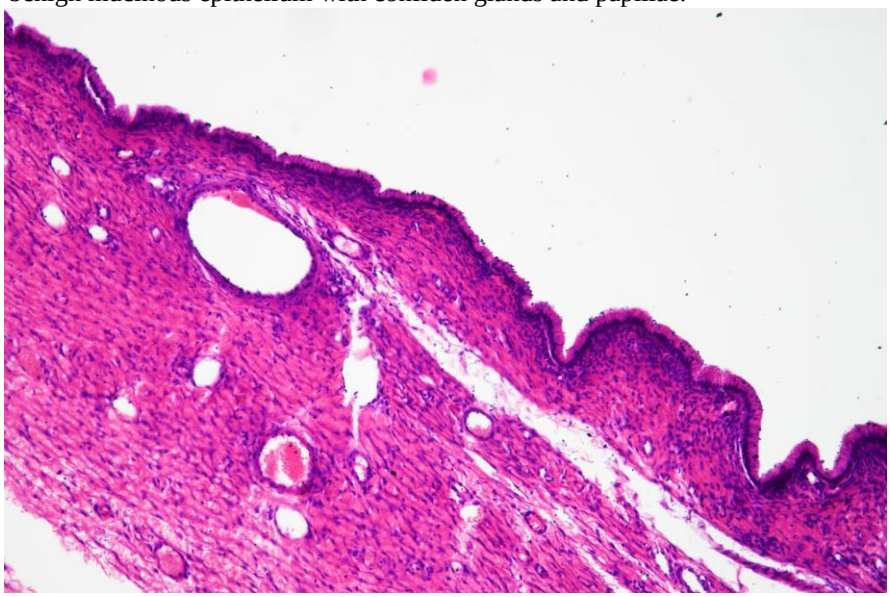
HPE - Intracystic mucinous carcinoma of confluent glandular/expansile type, capsule is intact no invasion seen

Figure 1 (a, b): H&E (100x) image of the same mass

a) Section from solid focus revealing benign mucinous epithelium merging with confluent glands lined by stratified epithelium with nuclear atypia.



b) Cyst wall lined by benign mucinous epithelium with confluen glands and papillae.



PET report -

1) Faintly FDG avid/ non FDG avid ill defined soft tissue density areas with adjacent fat stranding are noted in left adnexal region? Post surgical changes
2) FDG avid ill defined irregular soft tissue density with adjacent fat stranding in the left pelvis anteriorly? Post surgery changes? Residual disease
3) Mildly FDG avid well defined well capsulated cystic structure in the right adnexal region? Functional ovarian cyst/ Graafian follicle
4) Few faintly subcentimeter (SAD) left external iliac nodes
5) Mild ascites

Post operative period was uneventful and currently patient is on regular follow up with CA - 125 and CEA levels and recent PET - CT showed no growth.

3. Discussion

Mucinous ovarian tumors are rare in children <15 years of age with <50 cases reported in world literature [1 - 10]. Majority of these are benign/borderline tumors with frequency of carcinoma still rarer with only twelve cases reported at a young age of <15 years [2 - 5, 7, 9, 10]. Morwitz et al evaluated the histopathology of 240 patients with ovarian masses during 14 years [10], and only three cases were found to have cystadenocarcinoma

Mostly, patients present with vague symptoms that are initially ignored by them. This often leads to advancement of disease before the lesion gets diagnosed. Furthermore, it is believed that childhood tumors are far more aggressive than their adult counterparts and progress to advanced disease despite treatment [7].

Most of the cases of adenocarcinoma reported in the literature are pre - menarchal though some of them hover around the age group of 14 years. Tumor markers and radiology in collaboration serve as an essential tool in the diagnosis of ovarian cancers [6]. CA 125 has been widely used as a marker for epithelial ovarian tumors however its utility is debatable. Although elevated serum CA 125 levels (>35 U/mL) have been found in more than 80% of ovarian cancer patients, only 50% of patients with stage I disease have elevated levels. Furthermore, CA 125 is also raised in approximately 1% of healthy control subjects, liver cirrhosis, endometriosis, first - trimester pregnancy, pelvic inflammatory disease, pancreatitis, and in 40% of patients with advanced intra - abdominal non - ovarian malignancy [11]. Therefore, its raised levels must always be interpreted with caution and in conjunction with radiology. It is believed that if levels are initially raised at the time of detection they can be used as a marker for identifying residual or recurrent disease later at follow up [6, 8]. Sometimes mass can be very big creating confusion as to the exact source of origin even on radiology. Raised CA 125 levels in collaboration with the computed tomography scan report lead clinicians to believe that they were dealing with an ovarian mass probably epithelial in origin and explorative laparotomy has to be performed. The intra - operative examination of the other ovary and surrounding structures is important as it can often lead to upgradation of the stage of the tumor if macroscopic deposits are visualized during surgery. Though the adult staging protocols dictate mandatory lymph node dissections and biopsies of peritoneal surfaces; these procedures are often omitted in pediatric cases unless gross metastatic disease is present [10]. Given the significant incidence of bilateral disease, some authors have also recommended a prophylactic wedge biopsy of the uninvolved ovary at the time of initial debulking surgery or biopsy from grossly suspicious foci. Intraoperative frozen section can often aid in the diagnosis and further management of the patient. However, it can often be challenging to report a carcinoma in a minor age group without definitive infiltration into the stroma, rendering an equivocal report.

Guidelines of surgical treatment of malignant ovarian tumor with metastasis is an aggressive surgery in adults comprising of total abdominal hysterectomy with bilateral salphingo - oopherectomy along with tumor debulking/cytoreduction. Furthermore, some studies have compared fertility sparing surgery with radical surgery in borderline ovarian tumors [12]. In these studies, though the recurrence rate was some - what higher in the fertility sparing group as compared to

radical group, these recurrences were amenable to salvage by subsequent surgeries. A study done by Aggarwal et al. [13] pointed out that low malignant recurrences have been reported more than ten years after initial surgery even in an adult patient population. Therefore, like in adults; young patients should also be kept under close follow up to monitor recurrence which should be treated with another salvage surgery. Various adjuvant regimens ranging from single agent (carboplatin) [14] to multi agent regime have been tried for the treatment of malignant ovarian neoplasms. More recently, combination of hexamethyl melamine, doxorubicin, and cisdiamminedicholoroplatinum with/without methotrexate has been used successfully in adults for the treatment [2, 15]. Whereas, Blom and Torkildsen [3] administered intraperitoneal phosphorus in the pediatric mucinous cystadenocarcinoma that they encountered; Gribbon et al. [4] gave intraperitoneal radiotherapy in their two cases of cystadenocarcinoma. Prognosis of ovarian cancers presenting at younger age remains variable and depends on the stage of presentation. Most of the cases reported in the literature have had bad prognosis with almost all the patients dying within five years of detection of the lesion.

4. Conclusion

Epithelial ovarian tumors rarely occur in children <15 years of age, and are always almost benign. Malignant neoplasms are exceedingly rare however should always be kept in mind especially in cases with raised CA 125 levels suggesting non germ - cell origin. Since signs and symptoms are vague, patient often presents late and at an advanced stage.

Because of the rarity of this malignancy, future studies are needed to establish optimal understanding and management. In early stage, prognosis remains good but advanced staged tumors have a poor prognosis as mucinous tumors are relatively chemo resistant. Hence, cytoreduction should be optimum. Fertility sparing surgery should be considered in young patients in stage I but not beyond stage Ic3. Preoperative optimization, anticipation of complications and monitoring in intensive care unit are crucial for a fair outcome

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