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Ovarian yolk sac tumor: A case report

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Abstract
Yolk sac tumors (YST), also known as endodermal sinus tumors, are rare and rapidly developing neoplasm presenting in young females. They are second most common germ cell tumor after dysgerminoma. As they present in early age and have rapid malignant potential, fertility preservation is an important concern in such patients. Hereby we report a case of Yolk sac tumor in a 22 yrs. old female, who underwent fertility preserving staging laparotomy followed by 4 cycles of BEP, and is doing well.

Keywords: Yolk sac tumor, germ cell tumor, fertility preservation, Alpha fetoprotein

Introduction
Yolk sac tumors (YSTs) are defined as tumors that resemble the yolk sac, allantois, and extra embryonic mesenchyme. Yolk sac tumors are rare and a subtype of germ cell tumor, and present as rapidly developing abdominal mass in young women, typically between 18 yrs. to 30 yrs. of age. YST are second most common malignant germ cell tumors after dysgerminomas and account for only 1% of all ovarian malignancy [1]. The tumor metastasizes fast and intrudes all intra-abdominal structures and retroperitoneal lymph nodes. Early diagnosis, treatment with surgery and multi-agent chemotherapy are associated with high cure rates of 81.2-90.0% even in advance disease [2].

Case Report
A 22 yrs. old unmarried female, known case of Autism Spectrum Disorder (ASD), presented to OPD with complaints of diffuse abdominal pain and large abdominal mass. She had associated loss of weight and appetite.
On abdominal examination, a mass corresponding to 30 wks gravid uterine size was palpable with hard consistency, well circumscribed with irregular margins, non-mobile and not distinguished from adnexa separately.

Investigations
Ultrasonography revealed a large, complex cystic lesion measuring 19 × 19 × 12 cm with internal echoes and intra-septal vascularity, in left adnexa extending to midline, left ovary not visualised, with ORADS class 4.
CECT (Abdomen + Pelvis + Thorax) revealed (Fig. 1):

- Large, well defined, thick walled heterogeneously enhancing solid cystic lesion noted arising from left adnexa measuring 20 × 19 × 12 cm, probably neoplastic ovary.
- Multiple heterogeneously enhancing lesions (<1 cm) involving pre/para-aortic lymph nodes, omentum and mesentery adjacent to lesion.
- Moderate ascites and mild right sided pleural effusion.
- Probable FIGO Stage IIIB - IVA.

Serum Alpha Fetoprotein was raised (3, 10, 000 ng/mL). Rest tumor markers were within normal limits.
Complete Hemogram showed hemoglobin 9.2 gm %. Other parameters were normal; random blood sugar, liver function test, and renal function test were normal.

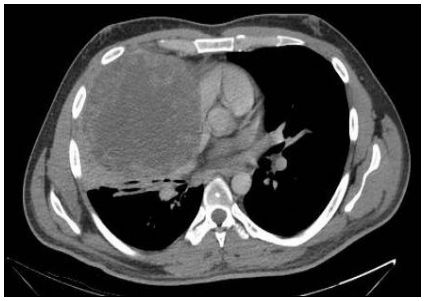

Fig 1: CECT plate showing large adnexal mass.

Treatment

Exploratory staging laparotomy was performed, after taking due informed consent and fitness from anesthesia point of view.

Intra-operative finding (Fig. 2) was a large solid cystic mass with intact capsule, adherent to omentum, uterus and anterior abdominal wall, arising from left ovary. Left sided salpingo-opherectomy was undertaken with infra-colic omentecomy and para-aortic lymphadenectomy.

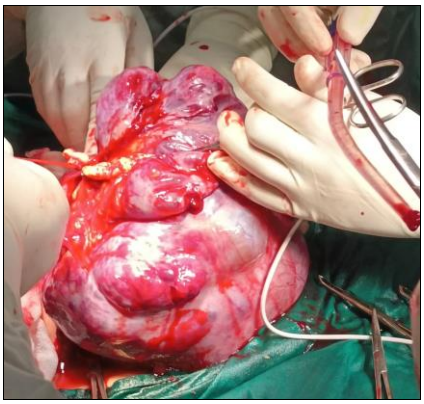

Fig 2: Intra-operative dissection of adnexal mass (YST).

22 × 22 × 12 cm mass was excised and sent for Histopathology (Fig. 3). Microscopy revealed tumor cells arranged in solid, reticular patten with prominent nucleoli and atypical mitiosis, large areas of hemorrhagic necrosis. Schiller Duval bodies noted, suggestive of Yolk Sac Tumor (Fig. 4).

Immunohistochemistry revealed OCT-4 to be negative. Retro-peritoneal lymph nodes showed microscopic metastases, while peritoneum was negative for metastatic deposits. The surgical staging of the tumor was Stage III. Post-operative recovery of the patient was good.

Patient was administered adjuvant chemotherapy - BEP regime, 4 cycles (Bleomycin, Etoposide, Cisplatin) and patient is following up regularly in Gynecology OPD.

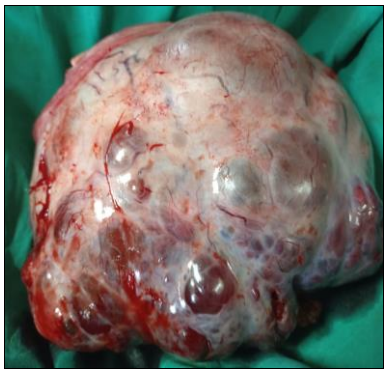

Fig 3: Ovarian mass specimen (sent for HPE) [22 × 22 × 12 cm]

Discussion

Ovarian germ cell tumors constitute 15 to 20% of all the ovarian tumors [3], originating from the primitive germ cell and gradually differentiating to mimic tissues of either the embryonic origin like ectoderm, endoderm and mesoderm or of the extraembryonic tissues like the yolk sac and trophoblast [4, 5].

Yolk sac tumour (YST) is usually seen in adolescents and young adults, between 18 to 30 years of age [4]. Most common symptoms are pelvic pain, menstrual abnormalities and abdominal mass. Sometime it may present with emergencies like torsion, hemorrhage and capsule rupture [6]. The tumour is almost always unilateral, but with rapid growth and extensive intra-abdominal spread leading to poor prognosis, if detected late. Involvement of omentum, abdominal peritoneum and serosal surfaces of the bowel has been reported in 30% of the cases [7]. In advanced stages, retroperitoneal lymph nodes and liver parenchyma are also involved.

The etiology of YST is unknown. Hypermethylation of the RUNX3 gene promoter and overexpression of GATA-4, a transcription factor that regulates differentiation and function of yolk sac endoderm, may play important roles in the pathogenesis of YSTs. But these hypotheses have not been proved [8, 9].

Pre-operative diagnosis is difficult, as yolk sac tumours do not have a specific radiological image[7]. Elevated AFP levels is the hallmark of this tumour and rapid decline in serum levels of AFP indicate absence of residual tumour after surgery. Other markers of YSTs have been reported recently. The prominent one is Glypican 3. It is a membrane-bound heparan sulfate proteoglycan. Glypican 3 is more sensitive but less specific than AFP, as Glypican 3 can be detected in some immature teratomas, in addition to liver and choriocarcinoma [10].

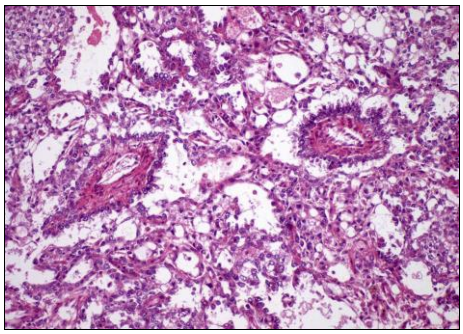

Fig 4: Microscopic slide showing Schiller Duval bodies along with tumor cell nests.

The diagnosis is histopathological. Histologically, the malignant tissue resembles the structure found in early embryonic development – the Schiller Duval bodies [11].

Complete surgical excision is the standard management for these tumours [7]. Fertility-sparing surgery is often possible, as the tumours are unilateral. According to Nishio *et al*, fertility sparing surgery with adjuvant chemotheraoy is possible at all stages of YSTs [12]. The BEP chemotherapeutic regimen has proved to be efficacious in treating Germ cell tumors since its introduction in the 1980s. The number of cycles and choice of chemotherapy regimen is based upon the histology and stage of disease. Toxicity encountered with BEP regimen - hair loss, myelosuppression, peripheral neurpathy - are uncommon and short lived, as the standard duration of BEP is only 3-4 cycles [3].

Factors related to good prognosis are [7]:

- No ascites at presentation
- Stage I disease
- Less than 42 days to AFP normalization

- Fertility-sparing surgery.
- Serum AFP half-life less of 10 days.

In a case series of 52 patients with yolk sac tumor by Rouge *et al* in evaluating long term fertility results, 97% patients had regular menstruation after achieving complete remission and 75% became pregnant who attempted conception. Only one patient suffered ovarian dysfunction [13]. In a restrospective study by Kojimara *et al*. 83% resumed menstrual cycles and were potentially fertile and 11% had live birth rate [2].
Follow-up plan for these patients include repeating serum AFP at regular intervals and an annual pelvic ultrasound to detect contra-lateral occurrence in patients who underwent conservative surgery [1].

Conclusion

Our case highlights the importance of fertility preserving treatment for Yolk Sac Tumor in young women, and we have provided our experiences and approach for treatment in such cases, underlining the standard care plan. Fertility preservation is an important concern in these patients, and can be achieved by early diagnosis and appropriate management.

Conflict of Interest: None.

Source of support: Nil.

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