Case Report on Pure Yolk sac tumor of ovary in a young female

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Abstract
Ovarian Yolk sac tumor is the second most common Malignant ovarian Germ Cell tumor (MOGT). It is seen in young women of reproductive age group, presenting commonly as abdominopelvic mass. Here we present a histopathologically diagnosed case of large ovarian Yolk sac tumor in a 21 year's old female who presented with abdominopelvic pain and swelling, raised AFP levels. She was treated with conservative surgery and currently undergoing chemotherapy.

Introduction
Yolk sac tumor is the second most common germ cell neoplasm after dysgerminoma. It accounts for 1% of all ovarian malignancies and 20% of malignant ovarian germ cell tumors2. It is common in childhood, adolescence and early adult life.3 It presents as a rapidly growing mass and is associated with high serum levels of AFP. Raised AFP levels points towards YST but confirmation by histopathological examination is necessary. It is highly malignant with early metastasis but is sensitive to chemotherapy producing remission even in advanced disease.3

Case Report
A 21year's old female presented with pain abdomen, abdominopelvic swelling and dysmenorrhea. On Ultrasonography, a large left complex adnexal mass was identified. On MRI of abdomen and pelvis, a large ill-defined lobulated solid-cystic mass lesion in pelvis on left side was identified measuring 13.2 x 8.4 cm. Radiological differentials of left Ovarian neoplasm or a degenerated fibroid were suggested. On Biochemical analysis CA-125 and hCG levels were normal however, AFP levels were raised. Patient underwent exploratory laparotomy with left salpingo-ophorectomy, right ovarian biopsy with intracolicomenteectomy and peritoneal toileting.

Grossly, Specimen of ovarian mass was received in multiple pieces (14x9x5) cm. Outer surface of one of the pieces showed attached fallopian tube with fimbrial end measuring 4.5 cm with lumen dilated. Cut surface of ovarian mass was solid, friable, gray white to gray brown with necrotic and mucoid areas. Also received a part of omentum (13 x 8 x 1.5) cm, no lymph node identified.

Right ovarian biopsy was also submitted. On Microscopic Examination of the left ovarian mass, tumor cells were arranged in glands, strands, microcyst, macrocyst and alveolar pattern. These cells revealed moderate to marked
pleomorphism with hyperchromatic nuclei, conspicuous nucleoli and moderate amount of clear to eosinophilic cytoplasm. Focal areas of sheets and nests of tumor cells were seen.

Additionally, atypical mitoses (6-7MF/10HPF), abundant intracellular and extracellular hyaline globules, focal myxoidstroma, inflammatory infiltrate, occasional Schiller-Duval bodies and areas of necrosis and hemorrhage seen. No other component of Germ cell tuor identified.

Left fallopian tube, omentum and right ovary were free from tumor.

A histomorphological diagnosis of Yolk Sac tumor, left ovarian mass was suggested and IHC was advised for confirmation.

On IHC (Pvt. Lab) the tumor cells were immunoreactive for GATA3, BER-EP4, AFP, SALL4 and negative for CK7/20, Heppar-1, ER, PAX8, WT1, SATB2, RCC, CD56, TTF1, EMA conforming the diagnosis of Yolk Sac tumor.

Discussion

Malignant Ovarian germ cell tumors (MOGT) account for 3-5% of all ovarian malignancies. Yolk sac tumor is the second most common MOGT. Also known as Endodermal sinus tumor, it is known to occur especially in young females of reproductive age group as a rapidly growing pelvic mass.
The etiology behind YST is unknown. However, studies have suggested that hypermethylation of the RUNX3 gene promoter along with overexpression of GATA-4, a transcription factor regulating differentiation and function of yolk sac endoderm, may play important role in its pathogenesis.\(^5,6\)

Clinically identified with raised serum AFP levels which have diagnostic as well as prognostic significance during treatment.\(^7\) Patient usually presents with abdominal swelling or mass or abdomino-pelvic pain.\(^8\)

Grossly, YST are usually unilateral, large, encapsulated, lobulated gray brown to gray yellow with areas of hemorrhage and cystic changes.\(^3\)

Histologically, YST can be Pure or mixed. YST exhibit multiple pattern on microscopy including microcystic or reticular, endodermal sinus, alveolar, macrocystic, glandular, solid, papillary or hepatoid. The tumor may show Schiller-Duval bodies i.e central vessel surround by tumor cells, present in approximately 50% and if found, they are pathognomonic.\(^9\)

It is highly malignant with early lymphatic spread. Fertility-sparing surgery is as effective as radical surgery. Combined chemotherapy improves the survival rate of the patient. Both surgery and chemotherapy are important prognostic factors.\(^8\)

Reference