Yolk Sac Tumor of the Ovary in a Four-Year-Old Girl: A Rare Case

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Citation

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Abstract

Germinal cell tumors form a minority of all malignancies of the ovary. Ovarian germ cell tumors are still rarer, especially in childhood. The common age group affected by yolk sac tumors is 11 to 24 years. Here, we present a case of yolk sac tumor in a four-year-old girl with the aim that this will effect an early diagnosis by a vigilant clinician. Prognosis of yolk sac tumors is highly stage-dependent and an early diagnosis can result in a drastic difference in the final outcome of the treatment of this highly aggressive disease.

INTRODUCTION

Germinal cell tumors represent only 2-5% of all cancers of the ovary.[1] Ovarian germ cell tumors are rare in childhood. Yolk sac tumors (YST), also called endodermal sinus tumors (EST) are the second most common germ cell tumors of the ovary.[2] The most common age group affected by YSTs is between 11 and 24 years. We conducted an extensive search in the Pubmed database and the youngest patient of the disease was 9 years old.[2] Here, we present a case of a four-year-old girl with yolk sac tumor. Besides the rarity of the tumor, the importance of the fact, that a vigilant and informed clinician can make an early and timely diagnosis of this condition even in girls of such a young age and make a drastic difference in the final outcome of the treatment, compels us to present this case.

CASE HISTORY

A four-year-old girl presented with complaints of progressive distention of the abdomen for three months. On examination, she was otherwise normal. Abdominal examination revealed a freely mobile abdominal mass. Her routine hematological profile and liver function tests were within normal limits. X-rays of chest and abdomen were unremarkable. Computed tomography of the abdomen revealed a large cystic mass arising from the ovary occupying most of the peritoneal cavity displacing the bowels to the left, with thin walls and enhancing thick septae (Fig. 1, 2).

Figure 1

Figure 1: Coronal section of abdominal CT scan showing a large ovarian mass.

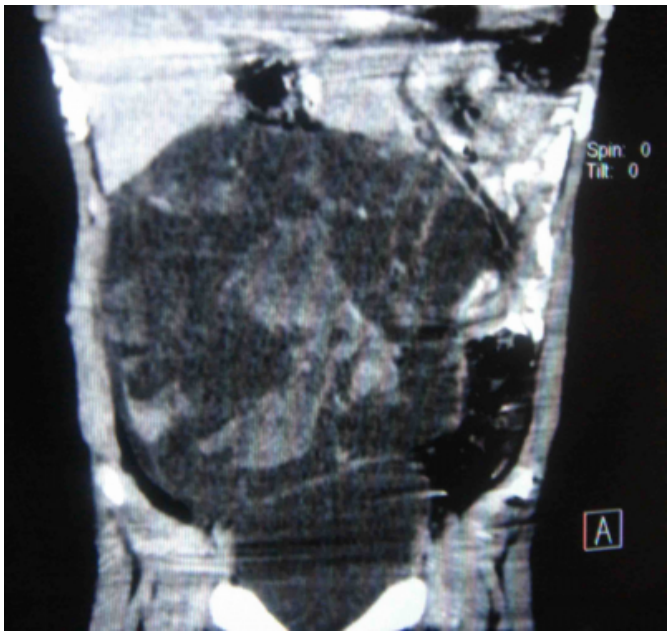
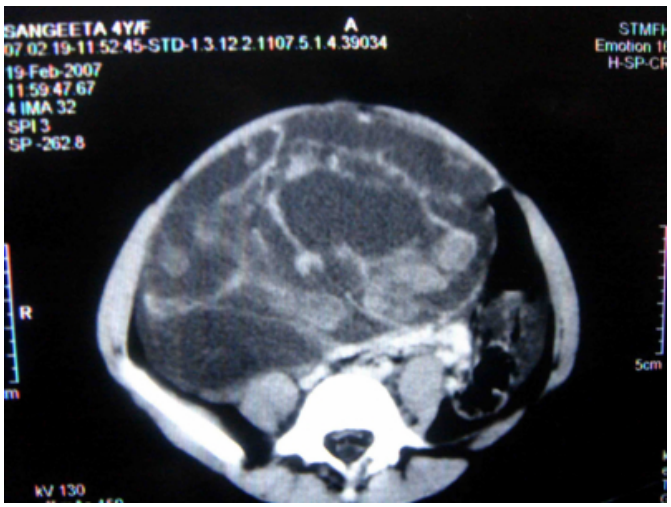


Figure 2

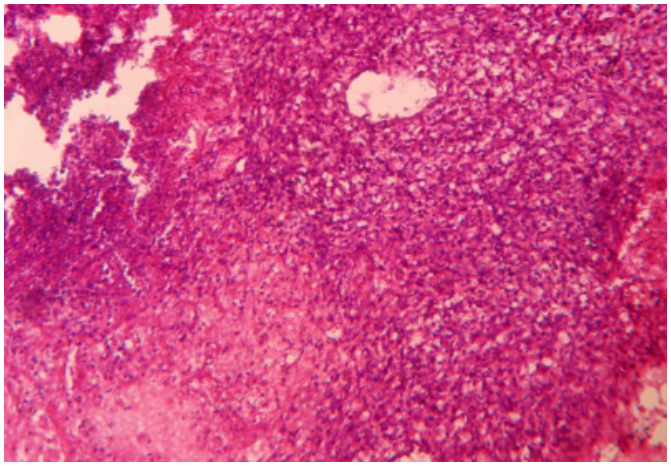
Figure 2: Transverse section of abdominal CT scan showing a large ovarian mass.



The serum alpha-fetoprotein level was found to be raised. Exploratory laparotomy revealed a large right ovarian cystic mass and right salpingo-oophorectomy was performed. Left ovary, uterus and the other intra-abdominal organs were normal. Cut section of the mass showed multiple small cystic areas along with solid areas. Peritoneal fluid cytology was free of malignant cells. Histopathology showed areas of reticular micro-cystic spaces lined by a single layer of flattened cuboidal cells. At places, Schiller-Duval bodies were also seen along with hyaline globules (Fig. 3).

Figure 3

Figure 3: Photomicrograph showing areas of reticular micro-cystic spaces lined by a single layer of flattened cuboidal cells. (10x; H & E stain)



The post-operative period was uneventful. The patient was discharged on the eighth post-operative day with the advice to come for chemotherapy. The patient did not report for follow-up for three months. After three months she reported with gross abdominal distention and respiratory distress. Investigations revealed ascites and bilateral pleural effusion. The patient succumbed to her disease within a day of her admission.

DISCUSSION

Yolk sac tumors of the ovary are rare and highly malignant tumors of utmost importance occurring in children and young adults.[3] They are classified into four patterns: endodermal sinus pattern, polyvesicular vitelline pattern , hepatoid pattern and glandular pattern. It is not too much to say that the Schiller-Duval body is a symbol of yolk sac tumors.[4] Serum alpha-fetoprotein is a useful marker for the diagnosis and management of YST.

YSTs are highly aggressive tumors with poor prognosis and were almost always fatal till the refinement of chemotherapeutic regimens in the past few decades. Survival rates have improved substantially, especially for stage I and II disease. Keeping the fact in mind that YST occurs in young age, fertility of the patient usually needs to be preserved and the optimal surgery done is laparotomy with unilateral salpingo-oophorectomy, peritoneal cytologic studies, omentectomy, multiple peritoneal and abdominal biopsies and resection of all visible disease. Three to four courses of post-operative chemotherapy consisting of bleomycin, etoposide and cisplatin are given keeping in consideration the bulk of residual disease.

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