Open Journal of
Clinical & Medical
Case Reports

Volume 5 (2019)
Issue 6

ISSN: 2379-1039

Ovarian yolk sac tumor in a 43 year old woman: A case report

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Abstract

Yolk sac tumor is a type of malignant ovarian germ cell tumor (MOGCT) and is considered rare. It is also known in the literature as endodermal sinus tumor, and is widely regarded as the second most common type of ovarian germ cell tumor, after dysgerminoma. Yolk sac tumor is the second most common malignant ovarian germ cell tumor and is a highly malignant tumor that metastasizes early.

This is a case report of a unique case of ovarian yolk sac tumor diagnosed in a 43-year-old woman. With this case, the old age should not preclude a clinical suspicion of ovarian yolk sac tumor.

Keywords

ovarian yolk sac tumor; germ cell tumor

Introduction

Yolk sac tumor is a type of malignant ovarian germ cell tumor (MOGCT) and is considered rare [1,2]. It is also known in the literature as endodermal sinus tumor, and is widely regarded as the second most common type of ovarian germ cell tumor, after dysgerminoma. Yolk sac tumor represents approximately 15 to 20 percent of all ovarian tumors. However, it accounts for less than 5 percent of all malignant ovarian tumors [3]. It is a highly malignant tumor that metastasizes early and invades the intraabdominal structures [2]. This type of tumor are usually observed in children and young adults. In light of this, upto our knowledge and literature review, there is no similar cases reported from Saudi Arabia [4]. Serologic and immunohistochemically α-fetoprotein (AFP) expression characterize this tumor [1,5]. AFP is also useful for monitoring the recurrence of ovarian yolk sac tumor [5]; elevated serum AFP levels during chemotherapy indicate a poor prognosis [2,6]. This is a case report of a unique case of ovarian yolk sac tumor diagnosed in a 43-year-old patient.

Case Report

A 43-year-old female presented to Emergency Department complaining of a vague lower abdominal pain for two weeks. She denied any history of loss of appetite, abdominal distention, change of bowel or urinary habits and no history of significant weight loss. She is medically known case of renal stone and hypothyroidism on levothyroxine. She has no history of any prior surgeries. She has an obstetric history of seven full term normal vaginal deliveries, last one was 18 months prior to her presentation. She has a regular menstrual cycle with average flow, and she never used contraception pills.

On Examination patient was vitally stable. Abdomen was soft with no tenderness. There was a palpable mass on the right side, below the umbilical level, measuring almost 10 cm.

The patient underwent a Computerized Tomography KUB (Kidneys, Ureters and Bladder) in Emergency Department due to her history of renal stone. The CT KUB revealed an incidental finding of a large abdominopelvic well defined mass, likely originates from right adnexa/ovary and was not seen in a prior CT scan done in 2017. There was no ureteric stone. Baseline investigations were sent including tumor markers that showed an elevated levels of Alphafetoprotien (AFP) at 39607 ng/ml and elevated CA 125 at 136.1 U/ml. Other tumor markers were all within normal limits.

CT Chest, abdomen, and pelvis was done and reveled, an abdominopelvic mass occupying mainly the midline with apparent solid cystic component with septations inside the mass measuring 13.3 x 9.12 x 15.1 cm and faint tiny solitary suspicious nodule at the left upper lung lobe with the possibility of early metastasis.

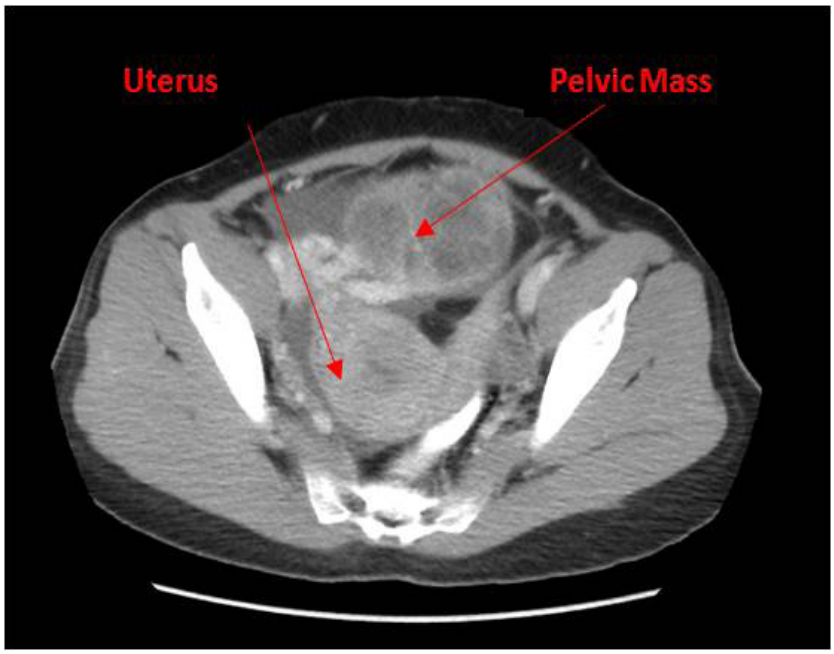

Figure 1: CT scan image. Post contrast axial image show a mass located in the pelvis, anterior to the uterus

The patient was seen by gynecology oncology and as the patient's age does not typically fit the malignant germ cell of the ovary, the AFP was repeated. The repeated AFP came back again elevated at 32681 ng/ml. The patient was counseled for surgery to include midline laparotomy with right salpingooophorectomy.

She had unremarkable postoperative period. The histopathological results revealed a yolk sac tumor (endodermal sinus tumor) of the right ovary with no involvement of the ovarian surface of fallopian tube surface. Peritoneal ascitic fluid was negative for malignancy.

Immunostains confirmed the diagnosis where the tumor cells were positive for AFP and negative for CD30, CD117, OCT 4, PLAP. The patient was referred to medical oncology for adjuvant chemotherapy.

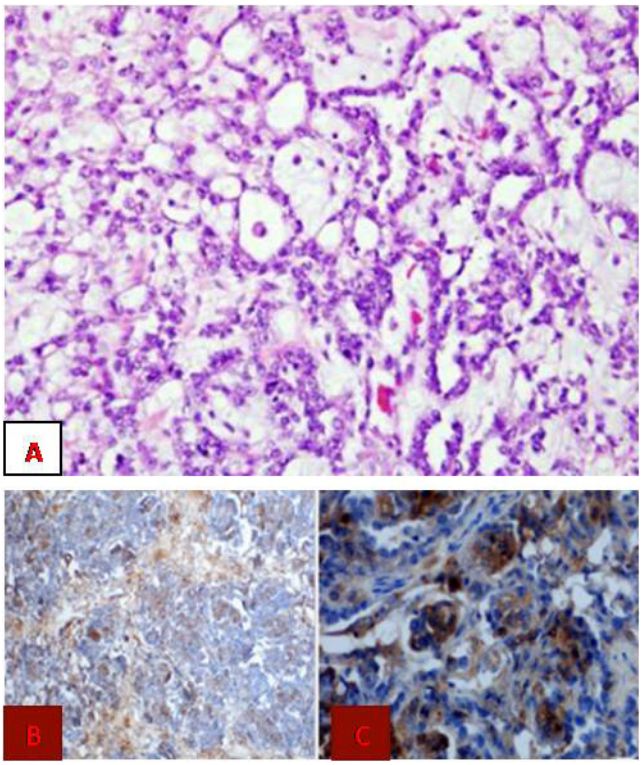

Figure 2: Photomicrographs of Yolk Sac Tumour. (A) Schiller-Duval body: central blood vessel enveloped by germ cells within a space. similarly lined by germ cells, resembles glomerulus. (B and C) Immunoperoxidase stain to AFP shows cytoplasmic positivity

Discussion & Conclusion

Ovarian yolk sack tumor is a type of malignant ovarian germ cell tumors and is generally rare [1-3]. The peak incidence is found in young women or adolescent girls, the median age at diagnosis being reported 19 years [2]. We report an exceptional case of ovarian yolk sac tumor diagnosed in an elderly patient. Rare similar cases were previously reported in post-menopausal patients [7-9]. Thus, the old age should not preclude a clinical diagnosis of ovarian yolk sac tumor.

Yolk sac tumor has a unique genetic background and thought to be caused by the hyper-methylation of the RUNX3 gene promoter [10], and the overexpression of the transcription factor GATA-4 gene which

normally regulates the differentiation of yolk sac endoderm [11]. The majority of cases present with abdominal pain and swelling as symptoms at the time of diagnosis [12], as it was in this case.

The tumor is grossly glistening on the surface while cystic and hemorrhagic on cut sections. Histologically, the pathognomonic morphological findings are the Schiller-Duval body, consisting central blood vessel surrounded by germ cells within a space lined by germ cells mimicking glomeruli. The histological patterns of yolk sac tumour are reticular or macro cystic, polyvesicular vitelline, microcytic, endodermal sinus or festoon or micro papillary, alveolar-glandular (with endometrioid variant), solid, hepatoid and myxoid patterns [8,12].

The tumor may show pure yolk sac tumor features or in combination with features of other germ cell tumors [12].

Tumor markers such as AFP and CA-125 are elevated; while the immunohistochemistry antibodies to confirm yolk sac tumor include AFP, CD10 and alpha-1-antitrypsin [13]. AFP immunosperoxidase stain was strongly positive in tumor cells in this case.

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The poor prognostic factors include tumor size > 2 cm, stage > I and presence of malignant ascites [2,12,14]. Yolk sac tumor diagnosed in early stages could be successfully treated with surgery combined with bleomycin, etoposide and cisplatin chemotherapy which are known to exhibit good outcomes in ovarian yolk sac tumors [6].

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Manuscript Information: Received: February 28, 2019; Accepted: April 23, 2019; Published: April 30, 2019

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Citation: Chamsi AT, Sadoun D, Shahed S, Wadi K. Ovarian yolk sac tumor in a 43 year old woman: A case report. Open J Clin Med Case Rep. 2019; 1537.