ejpmr, 2025, 12(3), 237-238

SJIF Impact Factor 7.065

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Study

ISSN 2394-3211

EJPMR

AN OVARIAN YOLK SAC TUMOR WITH UNUSUALLY LOW ALPHA- FETOPROTEIN LEVELS – A CASE REPORT

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Article Received on 08/01/2025 **Article Revised on 28/01/2025** **Article Accepted on 18/02/2025**

ABSTRACT

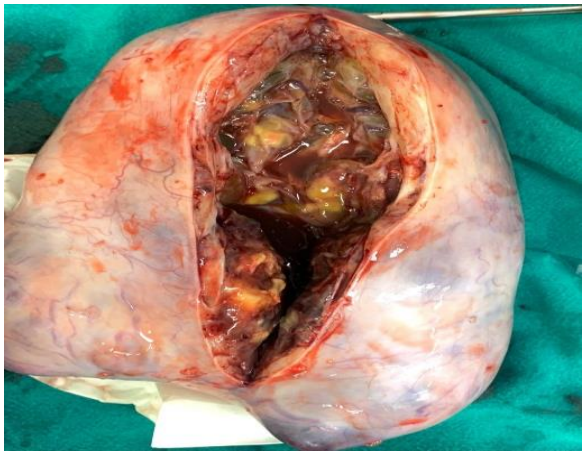
Yolk sac tumor is a rare and a highly malignant ovarian tumor of adolescent and young girls.[1] Raised alpha fetoprotein is the hallmark of the tumor and a valuable prognostic marker. Here we present a case of a 16 year old adolescent girl who presented to the Gynae OPD at Dr RPGMC Tanda with a progressively increasing abdominal lump since 2 weeks associated with pain abdomen. Routine investigations were done along with tumor markers which were normal. USG and CT scan were further done to arrive at a differential diagnosis of ovarian tumor. The patient then underwent exploratory laparotomy proceed surgical staging procced right salpingoophorectomy. She was discharged on post operative day 5 as per institutional protocol .The histopathological diagnosis was yolk sac tumor stage 1A. She was then given 6 cycles of neoadjuvant chemotherapy and had an uneventful recovery. Unusually normal levels of alpha-fetoprotein in our case make the diagnosis difficult and a high degree of suspicion is needed.

INTRODUCTION

Yolk sac tumor also known as endodermal sinus tumor is a rare malignant ovarian germ cell tumor (MOGCT), which accounts for less than 1% of all ovarian tumors characteristically expressing alpha-fetoprotein (AFP).[1] It occurs in adolescent and young adult women which makes it imperative to preserve fertility.[1] It presents with a rapidly growing pelvic mass and pain. Modalities like USG and CT scan can aid diagnosis, but confirmation can only be achieved on histopathology post exploration.

CASE REPORT

A 16 year old adolescent girl presented to Gynae OPD at Dr RPGMC Tanda with a hard bulge on her abdomen which progressively and uniformly increased in size in the last 2 weeks. It was associated with a continuous dull aching pain which had moderate to severe intensity, no aggravating or relieving factor and no radiation of pain. On examination pulse = 110bpm, BP=110/70 mmHg, temp = 101 degree Fahrenheit. On per abdomen examination there was a 16-18 weeks size abdominopelvic mass which was firm to hard in consistency, mobile from side to side, with regular margins, non-tender with lower limit not reached. There was no organomegaly. On per rectal examination same mass was felt with rectal mucosa free. On investigation her HB- was 9.3g/dL, platelet count, LFT, RFT and coagulation profile was normal. Her alpha-fetoprotein was 61.52 ng/ml, CA-125- 42.2U/ml, S.LDH- 574.4IU/L and serum beta HCG<1.2. She was advised a preliminary USG which showed a multiseptated well defined solid, cystic abdominopelvic mass -25*9cm in size, which was causing mass effect leading to bilateral hydroureteronephrosis. Uterus and left ovary was grossly normal. On CT scan -a large heterogenously enhancing solid-cystic mass lesion in the pelvis extending in the abdominal cavity measuring 13*12*20 cms causing mass effect on the surrounding structures and bilateral ureters leading to hydroureteronephrosis, lateral displacement of gut loops, psoas muscle, uterus and bilateral iliac vessels. right gonadal vein was engorged. Few lymph nodes were present in the preaortic region largest measuring upto 8.9mm. She was then taken up for exploratory laparotomy proceed surgical staging proceed right salpingo-ophorectomy. Grossly the tumor had a uniform surface with multiloculated solid – cystic areas, serous to purulent fluid with necrotic and hemorrhagic areas seen in it. It was sent for histopathological examination. The patient was discharged on post operative day 5 as per institutional protocol. She later visited the Gynae OPD with her histopathological report showing characteristic Schiller – Duval bodies and features consistent with yolk sac tumor. The final pathological staging was stage 1A. She then underwent 6 cycles of adjuvant chemotherapy (bleomycin, etoposide and cisplatin) and had an uneventful recovery.



Here is a cut section image of the yolk sac tumor showing multiloculated solid -cystic areas, serous to purulent fluid with hemorrhage and necrotic areas in it.

DISCUSSION

MOGCT s which originate from the primitive germ cell constitute 15-20 %of the ovarian tumors.[1] They differentiate to mimic tissues of either the embryonic like ectoderm, endoderm and mesoderm or of extraembryonic tissues like yolk sac and trophoblast.[2,3] As per the scheme by Telium[2,3] :1. If there is no differentiation, a germinoma would develop. 2. With differentiation, embryonal carcinoma would develop. 3. With extraembryonic differentiation, a yolk sac tumor or choriocarcinoma would develop.

They are usually present in adolescent girls with an sudden, unilateral, progressively enlarging abdominopelvic mass due to their rapid growth potential,[2] assosiated with pain. Elevated alphafetoprotein is the hallmark of the tumor,[2] which was lacking in our case. Fall in AFP levels post exploration indicate absence of residual disease and is a valuable prognostic index,[2] Pre-operative radiological investigation modalities like USG and CT scan are extremely useful in establishing diagnosis.[4] Histopathological diagnosis post exploration confirms the diagnosis. It shows presence of specific Schiller Duval bodies.[5] Due to the younger age of presentation of the tumor fertility- sparing surgery is the mainstay of the treatment. The BEP (Bleomycin, Etoposide and cisplatin) chemotherapeutic regime post exploration is proved efficacious in treating MOGCTs.[6] Early metastasis is common and the disease has a poor prognosis . In our patient there was no metastasis and she had complete remission when given chemotherapy after surgery which is treatment of choice in MOGCT.

CONCLUSION

MOGCTS are highly malignant ovarian tumors that occur in adolescent girls that tend to have a poor prognosis owing to its rapid growth rate. Hence a high degree of suspicion, early diagnosis and treatment involving a fertility sparing surgery followed by adjuvant chemotherapy is the cornerstone of treatment.

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