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Extragonadal yolk sac tumour originating from the broad ligament - A rare case report

[1]Dr. Arsha Narayanan, Senior Resident, Department of Pathology, Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana.

[2]Dr. Nisha Marwah, Professor, Department of Pathology, Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana.

[3]Dr. Anku Alisha, Junior Resident, Department of Pathology, Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana.

[4]Dr. Vibhav Goel, Junior resident, Department of Pathology, Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana.

[5]Dr. Priyanka Verma, Junior resident, Department of Pathology, Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana.

[6]Dr. Sunita Singh, Senior Professor and Head, Department of Pathology, Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana.

Corresponding Author: Dr. Arsha Narayanan, Senior Resident, Department of Pathology, Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana.

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Abstract

Extra gonadal yolk sac tumours are extremely rare and the most common sites involved are mediastinum, retro peritoneum, sacrococcygeal region and pineal gland. We report a case of extra gonadal yolk sac tumour, originating from broad ligament of uterus in an 18-year-old girl admitted with complaints of abdominal distension with pain for 3 months and found to have a left adnexal mass per operatively.

The patient was diagnosed with yolk sac tumor originating in broad ligament on histopathology. There were only four reported cases of broad ligament yolk sac tumors in literature prior to this.

It is imperative that the surgeons should be aware of the challenges of accurate diagnosis and its implications on therapy in these cases.

Keywords: AFP, broad ligament, extragonadal, yolk sac tumour.

Introduction

Yolk sac tumor (YST), otherwise known as endodermal sinus tumor, is a malignant germ cell tumor most commonly occurring in the gonads with differentiation into yolk sac structures and is usually accompanied by elevated serum alpha-fetoprotein (AFP) levels. Yolk sac tumor is the third most common malignant germ cell tumor of the ovary and comprises approximately 1% of ovarian malignancies. It occurs principally in children and young women with a median age of 19 years[1]. The most common presenting complaints are abdominal pain, abdominal enlargement or an abdominal mass. About 10% of patients have acute abdominal symptoms due to rupture or torsion of the tumor. Yolk sac tumor appears limited to the ovary at diagnosis in about 50% of patients. High levels of AFP are present in the serum of most patients with yolk sac tumors and some have elevated levels of CA125 as well. Only less than 10% YSTs arise in extra-gonadal sites in female patients; which can be sacrococcygeal region, retro peritoneum, mediastinum, pineal gland, stomach, liver, omentumpelvis and genital system.[2] YSTs originating from the broad ligament are very rare and difficult to distinguish those from ovarian cancer and other pelvic mass preoperatively. Up to now, there have been only 4 case reports of primary broad ligament YST reported before in English literature. Here, we share one case of primary YST of the broad ligament reported in our institution. This study was approved by the ethics committee of University of Health Research and Sciences, Rohtak, Haryana.

Case report

An 18-year-old unmarried female presented in the Gynaecology emergency with complaints of abdominal distension and pain for 3 months. On palpation, a non-mobile well-defined firm to hard mass was identified in the pelvis involving both iliac fossa and reaching up to the umbilicus. On CECT abdomen, a huge solid necrotic enhancing mass lesion of size 221 x 192 x 145 mm was identified in the abdominopelvic region, causing com pression and displacement of bowel loops and abdominal structures. The patient had undergone an exploratory laparotomy and left sided salpingooophorectomy following the investigations. Per operatively, a large friable mass was identified involving the adnexa (broad ligament) and was removed.

The gross examination of specimen (left adnexal mass) sent for histopathology revealed a friable mass in multiple pieces measuring 20 x 14 x 10 cm. Left ovary measuring 6.5 x 4 x 1.5 cm was sent along with fallopian tube of length 6.4 cm. Representative histopathology sections from friable mass revealed a tumor composed of atypical cells arranged in trabeculae, sheets, papillae, reticular and microcystic pattern and forming Glomeruloid perivascular structures (Schiller-Duvall bodies). These cells revealed mild to moderate nuclear pleomorphism and were mitotically active. Tumour was seen arising in the broad ligament invading peri tubal tissues, ovarian tissues and omentum. Immuno his to chemical staining was positive for Alpha-Feto Protein (AFP) and CK (Cytokeratin) and negative for CK7 and CA125, confirming the diagnosis of Yolk Sac Tumor. Post operative chemo therapy with Bleomycin-Etoposide- Cisplatin (BEP) was started and the patient was kept on follow up. After follow-up for 21 months, there is still no evidence of relapse.

Discussion

Yolk sac tumor, also known as endodermal sinus tumor is a malignant germ cell neoplasm in which there is differentiation into yolk sac structures. They are limited

to ovary at the time of diagnosis in 50% of cases. In general, yolk sac tumors are large in size, with an average diameter of 16 cm. The cut surface of tumor is tan, white or gray with small cysts and areas of hemorrhage and necrosis. The tumor can have a honey comb appearance due to the presence of numerous cysts. There are numerous histologic patterns of yolk sac tumor growth. The two most common and distinctive patterns are the reticular, or microcystic pattern and the endo dermal sinus pattern. The reticular pattern consists of a loose meshwork of microcystic spaces lined by a single layer of flattened or cuboidal cells. These have clear or amphophilic cytoplasm and atypical hyper chromatic nuclei. The endo dermal sinus pattern, also known as the festoon or pseudo papillary pattern, consists of labyrinthine anastomosing glands and papillae lined by columnar cells with clear or amphophilic cytoplasm and fusiform, hyper chromatic nuclei[1].

Schiller-Duval bodies, a characteristic finding is papillary or Glomeruloid structures in which fibro vascular cores covered by columnar tumor cells project into glands or cystic spaces lined by cuboidal cells. They are diagnostic of yolk sac tumor. Immuno his to chemistry: The cases of yolk sac tumors are almost invariably positive for cytoplasmic staining with Alpha-Feto- Protein (AFP). Though SALL4 also shows diffuse strong nuclear positivity, they are not specific for YST. Other IHC markers that can be positive are cytokeratin (CK) and Glypican 3. These are negative for OCT4, EMA, CK7, CD30 as well as CD117.[2]

Herein, we share a case series of primary YST originating from the broad ligament. The broad Ligaments are the folds of the parietal peritoneum that reflect over the female genital tract, extending from the lateral aspect of the uterus to the pelvic wall. They are a double-layered sheet of mesothelial cells continuous with each other, and between them is the extraperitoneal tissue, referred to as the parametrium, which comprises the connective tissue, smooth muscles, and neuro vascular elements. Although secondary involvement of the broad ligament by malignant tumors arising elsewhere in the abdomen and pelvis is common, primary tumors in this location are rare. To be considered tumor from the broad ligament, it should not be connected with either the uterus or the ovary[3]. Thus, the imaging approach to establish the differential diagnosis includes excluding an ovarian, uterine, or tubal origin by recognizing these separately.

After review of the English literature, there were only 4 cases of primary broad ligament YST published before. Review of previous reported cases reveal that YSTs primarily occur in the younger population. The median age from theses case is around 19 years. The mechanisms of YST from the broad ligament need to be explored[4].

For cases of female patients with extragonadal YSTs, there is still lack of standard guidelines. According to NCCN guidelines for ovarian YSTs, the most common regimen is BEP regimen, while some patients with recurrent disease can also be treated with high-dose chemo therapy and stem cell transplantation as a salvage therapy[5]. Considering both the ovarian YST and the broad ligament YST occurring outside the uterus and metastasizing easily, the treatment strategies suited for ovarian disease may be also suited for the broad ligament YST and need to be explored in the future[6].

The initial surgical treatment recommended for yolk sac tumor is unilateral salpingo-oophorectomy with limited debulking of extraovarian tumor. It is not necessary to biopsy the contralateral ovary since bilateral involve

Ment is rare in initial stages[7]. Majority of the patients have rapid development of metastases and high mortality, even when the tumor appeared limited to the ovary at operation. Radiotherapy proved ineffective for treating these tumors. Com bination chemotherapy using VAC regimen (vincristine, dactinomycin and cyclo phosphamide) was given previously in these cases and it resulted in a survival rate of about 80% in stage I tumors and about 50% in cases of advanced disease. It is proved that chemo therapy regimens containing platinum provide even better results, and standard therapy is now with a combination of bleomycin, etoposide, and cisplatin (BEP)[8]. The serum AFP level can be used to monitor the response to treatment and to detect tumor recurrence.

Conclusion

YSTs originating from the broad ligament are extremely rare and are usually accompanied by elevated serum AFP. The case of YST from the broad ligament we shared was the 4th case in the English literature. Due to paucity of cases, the treatment strategies are not standardized and need to be explored.

Acknowledgement

All the authors have contributed to concept, literature search, data acquisition, data analysis, Manu script, editing and review.

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Legend Figures

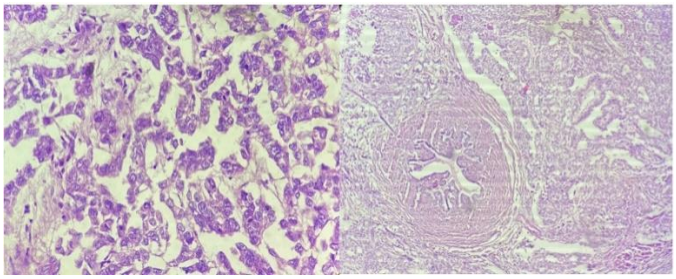

Figure 1: (A) Atypical cells in Extragonadal Yolk sac tumor (H & E, 400X); (B) Tumor involving the par tubal areas (IHC, 100X)

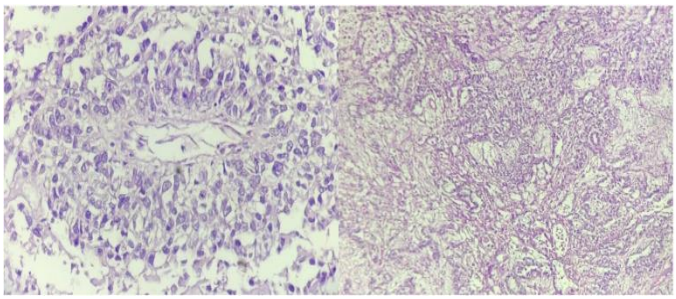

Figure 2: (A) Schiller Duval body (H & E, 400X); (B) Tumor cells arranged in microcystic pattern (IHC, 100 X)[1]

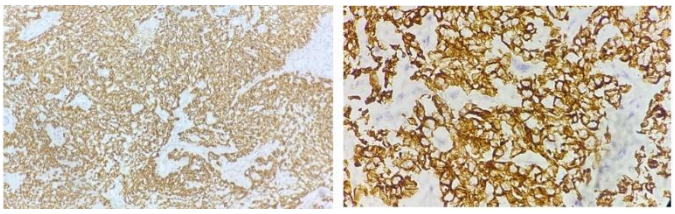

Figure 3: Extragonadal Yolk sac tumor: Cytokeratin (CK) positivity (A) IHC, 100X (B) IHC, 400X

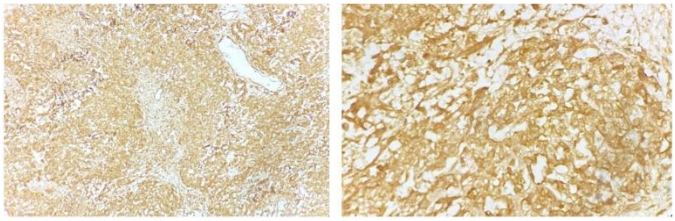

Figure 4: Extragonadal Yolk sac tumor: AFP positivity (A) IHC, 100X (B) IHC, 400X