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CASE REPORT

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A large yolk sac malignancy in a girl, an uncommon yet challenging ovarian tumor: A case report

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Key Clinical Message

Yolk sac tumors are rare and malignant germ cell tumors of the ovary occurring in children and young women. Fertility-sparing surgical intervention with adjuvant chemotherapy has shown to improve prognosis.

Abstract

We present a case of a 14-year-old girl who presented with the complaints of lower abdominal pain and distention. Her tumor markers were increased, and radiological investigation suggested the diagnosis of malignant left ovarian mass. Histopathology confirmed the diagnosis of Yolk sac tumor. She was subsequently managed with fertility-sparing surgery and adjuvant chemotherapy.

KEYWORDS

fertility, ovarian neoplasm, yolk sac tumor

1 | INTRODUCTION

Yolk sac tumors (YST), also known as endodermal sinus tumors or primitive endodermal tumors, are those tumors that derive from malignant cells that are differentiating along the extraembryonic yolk sac lineage.[1] They are classified under germ cell tumors by the World Health Organization.[2] Delving into embryology, migration of the germ cells occurs from the midline dorsal mesentery to the gonadal ridge, toward the fourth to sixth week of development. The misplacement of the cells along the line and the malignant transformation of those germ cells is the cause of the primary germ cell tumors. It can arise anywhere along the migration from the mesoderm to the future cranial area.[3] These tumors, accounting for 1% of all malignant ovarian tumors, occur in adolescents and young women.[4] Since the diagnosis of the tumor is at a younger age, fertility preservation is the area of concern. Also, the outcome of fertility preservation surgery is good with promising obstetrics and gynecological outcomes.[5] Fertility-sparing surgical interventions have increased the overall 5-year and event-free survival rates to 84% and 79%.[6]

Herein, we present a case of a 14-year-old girl who was diagnosed with a germ cell tumor, a histologic YST managed with fertility-sparing surgery, and later treated with adjuvant chemotherapy.

2 | CASE PRESENTATION

A 14-year-old girl, who had not attended menarche, presented to our Gynecology Outpatient Department with

chief complaints of lower abdominal pain and distention for 17 days. The abdominal pain was insidious in onset, pricking type, mild intensity, non-radiating, and not severe enough to hamper her daily activities. The abdominal distention was progressive. She lost 3 kgs over 2 months, but the abdominal pain and distention were not associated with bloating, water brash, vomiting, yellowish discoloration of the body, cough, shortness of breath, loss of consciousness, excessive hair growth, and hoarseness of voice. She had no history of a similar illness in the family.

On abdominal inspection, the abdomen was distended above the umbilicus. And, on deep palpation 22 × 14 cms solid cystic mass was felt, which was non-tender, had a smooth surface, and regular margin with side-to-side motility. Tumor markers were increased. Serum alpha-fetoprotein was (AFP) > 2000 ng/mL (Reference Range [Ref.] 0–8.5 ng/mL). *Carcinoembryonic antigen* (CEA) was 22.5 ng/mL (Ref. 0.0–3.0 ng/mL). *Cancer antigen 125* (*CA 125*) was 514.9 U/mL (Ref. <35 U/mL). Lactate dehydrogenase (LDH) was 1081 U/L (Ref. 115–221 U/L). Serum beta-human chorionic gonadotropin (hCG) was 2.91 μIU/Ml (Ref. 0–5 μIU/mL). Contrast Enhanced Computed Tomography (CECT) abdomen-pelvis revealed 18.3 × 15.5 × 10.2 cms well-circumscribed cystosolid lesion in abdominopelvic cavity with solid component showing heterogenous enhancement and without separate visualization of left ovary (Figure 1) which all suggested the diagnosis of malignant left ovarian mass.

Staging laparotomy with left salphingo-ophorectomy with infra-colic omentectomy with appendectomy with peritoneal biopsy with bilateral paracolic gutter biopsy with mesenteric lymph node sampling was done. Gross examination of tumor (Figure 2) revealed a large 25 × 20 cm mass of ovarian tumor with adherent mesentery on the right side with small bossulation on the surface without surface deposits. The cross-section (Figure 3) showed a yellowish fleshy mass with intracystic hemorrhage with areas of necrosis. Histopathology (Figure 4) revealed a germ cell tumor, morphology favoring YST with a maximum tumor dimension of 16.5 cm. Characteristic Schiller–Duval bodies were also identified as characterized by central vessel core lined by tumor cells and followed by cystic spaces. Being the tumor limited to one ovary with capsule intact, no tumor on the ovarian or fallopian tube surface, no malignant cells in peritoneal washings, and no regional lymph node metastasis, the TNM staging is pT1An0 and FIGO staging is IA (AJCC 8th edition/CAP protocol 2021). The patient was referred to the oncologist for treatment beyond. She was started on three cycles of BEP (bleomycin, etoposide, and cisplatin) regimen and completed it. On her 1 year follow-up, she was clinically stable. The repeat imaging was done which did not show signs of tumor.

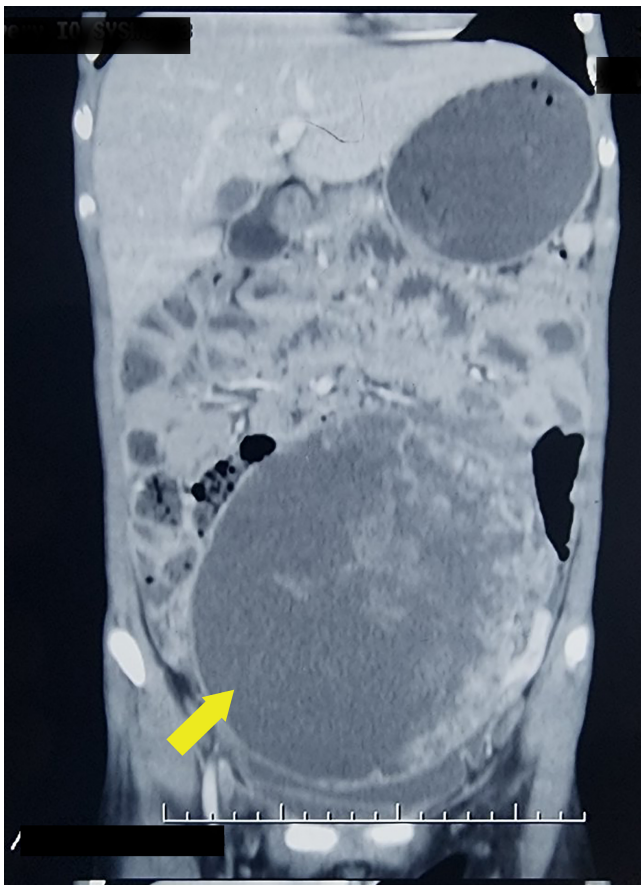

FIGURE 1 Contrast enhanced computed tomography of abdomen and pelvis, axial view, showing large cystosolid lesion extending from L4 to S4, with solid component showing heterogenous enhancement.

3 | DISCUSSION

Germ cell tumors, like their counterparts in the testis, are cancers of germ cells. YST ranks as the second most common histological subtype of malignant ovarian tumor of germ cell origin after dysgerminoma.[7] YST usually arise in the gonads, testes, or ovary; however, approximately one-third of them have extragonadal origins. Most patients are adolescent girls or young women, between the age group 18–30 years,[8] presenting with abdominal pain and a rapidly growing pelvic mass with a short duration of symptoms. Mass metastasizes fast and intrudes intra-abdominal structures and retro-peritoneal lymph nodes, and usually involves a single ovary.[9]

The several Diagnostic tools for YST are Ultrasonography (USG), Computerized Tomography

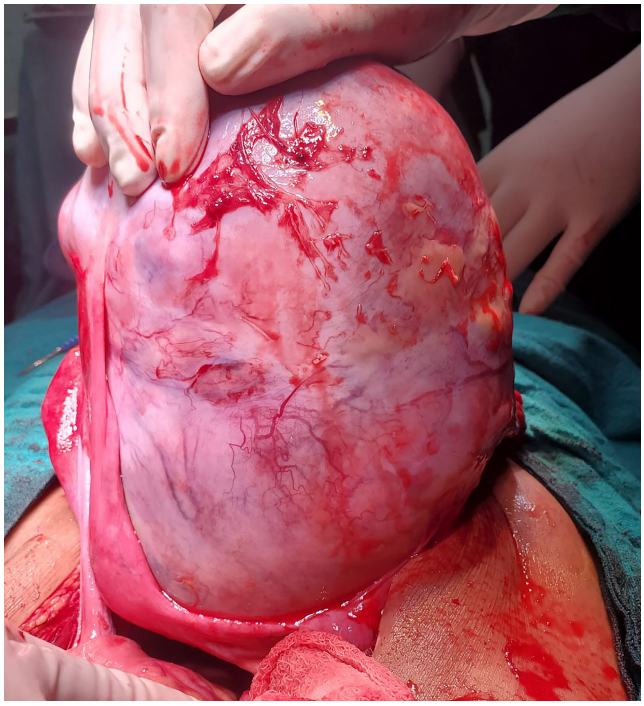

FIGURE 2 Gross examination of tumor.

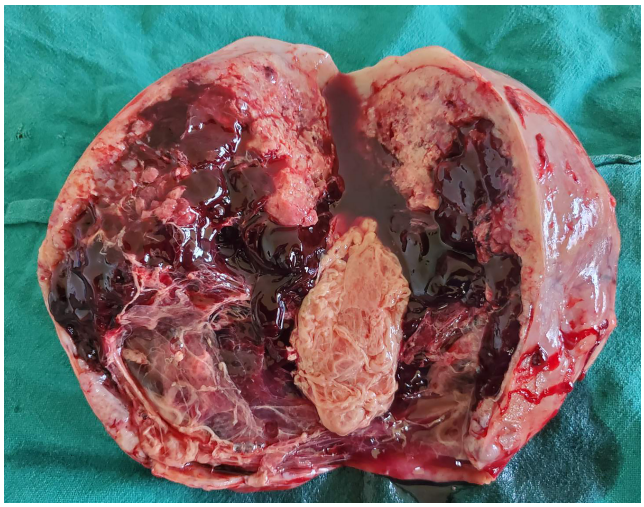

FIGURE 3 Cross-section of the tumor showing yellowish fleshy mass with intracystic hemorrhage with areas of necrosis.

(CT), Magnetic resonance imaging (MRI), and histopathological analysis. USG detects the adnexal mass and shows ascites. CT scan shows adenopathy and carcinosis. The tumor may appear as a solid or cystic structure or can be a combination of both.[10] MRI reveals the hypervascularized and hemorrhagic feature of the mass.[11] For histopathological analysis, a biopsy can be taken by exploratory laparotomy. It is an emerging technique that also detects tumor details.[12] YST has variable histopathological properties. Reticular or microcystic areas formed by a loose meshwork lined by flat or cuboidal

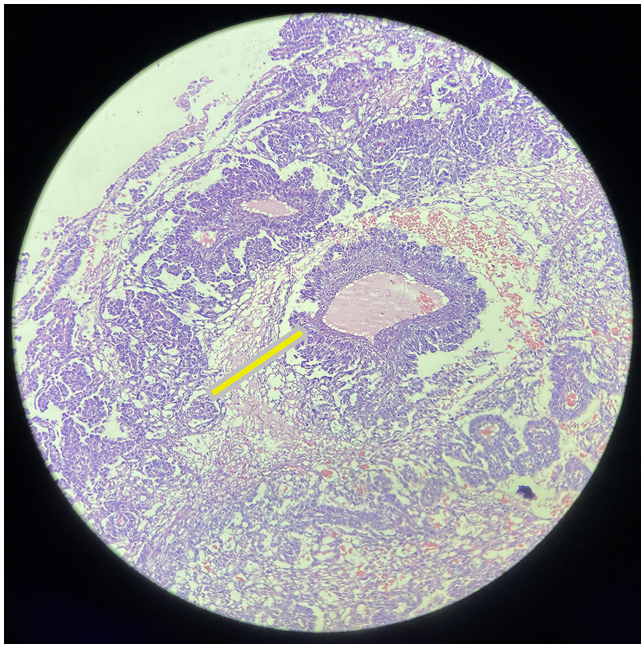

FIGURE 4 (Hematoxylin and eosin, ×100) showing characteristics, Schiller–Duval bodies.

cells are the most typical types.[13] The pathognomic histopathological feature is the Schiller Duval body. They are rounded glomeruloid bodies composed of a central blood vessel enveloped by tumor cells within a space that is also lined by tumor cells.[14]

Though extremely sensitive but not tumor-specific,[15] elevation of AFP is prognostically important and is an important index for stopping chemotherapy in YST.[16] It is a glycoprotein that is normally produced during gestation by the fetal liver and yolk sac. It has been reported that the lowering level of post-operative serum AFP could be a useful marker for determining if residual cancer cells still exist after surgery.[17]

The major concern in these types of tumors is the preservation of reproductive potential but since YST unlike dysgerminomas are confined to one ovary; it permits conservative, fertility-sparing surgical intervention that is unilateral salpingo-oophorectomy with preservation of normal appearing uterus and contralateral ovary.[18] Also, Nishio et al.[19] have shown that the fertility-sparing surgical intervention when added with adjuvant chemotherapy favored good outcome in terms of fertility and survival. With the combined treatment of surgery and adjuvant multi-agent platinum therapy, a survival rate of greater than 90% can be achieved in women with early-stage ovarian germ cell tumors and up to 80 percent of those with advanced disease.[20,21] For chemotherapy, the regimen of choice is BEP regimen. It is significantly superior to non-BEP therapy in 5-year overall survival rate.[22]

4 | CONCLUSION

Yolk sac tumors of ovary are the rare and highly malignant tumors occurring primarily in children and young women. Appropriate radiological investigations and serum alpha-fetoprotein levels in the blood lead to prompt diagnosis. Fertility-sparing surgical interventions added with chemotherapy improve prognosis with the preservation of reproductive and sexual function.

AUTHOR CONTRIBUTIONS

Aashish Poudel: Conceptualization; resources; supervision; writing – original draft; writing – review and editing. **Prajwal Sedain:** Conceptualization; visualization; writing – original draft; writing – review and editing. **Biraj Pokhrel:** Conceptualization; investigation; supervision; validation; writing – original draft; writing – review and editing. **Aakash Sapkota:** Writing – original draft; writing – review and editing. **Anita Chamlagain:** Writing – review and editing. **Nisha Sharma:** Writing – review and editing. **Sanyukta Rajbhandary:** Supervision. **Bishal Khaniya:** Supervision. **Neebha Ojha:** Supervision.

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CONFLICT OF INTEREST STATEMENT

The authors declare that there is no conflict of interest regarding the publication of this paper.

DATA AVAILABILITY STATEMENT

Not applicable.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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