Ovarian yolk sac tumour in a 9-year-old girl

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SUMMARY

Yolk sac tumour (YST) is a rare tumour that frequently arises in the gonads as a type of germ cell tumour. It is rare in children, and it is the second most common histologic type of malignant germ cell tumour of the ovary. The patient will come to the hospital with various clinical presentations, like a mass in the abdomen, pain, and sometimes high-grade fever. The alpha-fetoprotein (AFT) is a hallmark of a tumour marker because its level will be elevated in >90% of YST. In this study, a 9-year-2-months-old girl presented with pain and a mass in the suprapubic area. The pre-operative AFT was 107380 IU/ml. Right salpingo-oophorectomy and omentectomy were done, and we started combined chemotherapy. After 9 months of follow-up, the AFT level reduced to a normal level and there was no evidence of disease recurrence. The prognosis of yolk sac tumour is highly stage dependent, but early detection and comprehensive treatment at such a young age has a good prognosis.

INTRODUCTION

Yolk sac tumour (YST), though rare, is the second commonest histopathological subtype of malignant ovarian germ cell tumour after dysgerminoma. This tumour presents as a rapidly growing mass in younger women. YST is usually seen in adolescents and young adults, between 18 and 24 years of age. It is a rare and aggressive malignancy, representing 20% of germinal tumours in the ovary, characterized by its high chemosensitivity. The challenge is to ensure disease control without compromising fertility in adolescent and young women.[1]

CASE REPORT

A 9-year-old girl presented with lower abdominal pain that was localized, persistent, and pricking in nature. The pain gradually worsened till her sleep was disrupted. She was noted to have a suprapubic mass on the second day. Otherwise, she had yet to achieve menarche, no constitutional symptoms, and normal bowel and urinary habits. There is no known cancer in the family history. An initial examination found the abdomen was soft and not distended, with a palpable mass in the suprapubic area extending upwards to the midpoint between the pubic symphysis and umbilicus. The vulva was normal, and the hymen opening was seen.

The ultrasound examination on the next day noted a lobulated heterogenous echogenicity cystic pelvic mass likely from the right adnexa measuring 4.6 × 9.7 × 7.2 cm (AP × W × CC) with the presence of fluid with debris seen in between the mass and the urinary bladder measuring approximately 2.8 × 3.6 × 3.4 cm and increased vascularity within the solid component of the mass. However, no calcification was seen. The feature of a right adnexal mass, most likely a dermoid or teratoma, with the possibility of rupture. The Contrast Enhanced Computed Tomography (CECT) was done the next day and showed a heterogenous enhancing pelvic mass suggestive of a right ovarian yolk sac tumour (YST) with bilateral mild hydronephrosis likely secondary to mass compression. The chest X-ray reported no pleural effusion and no lung metastasis. The pre-operative AFT was 107380 IU/ml, the beta-human chorionic level was 0.2 mIU/ml, and CA-125 was 110 U/ml. The other blood investigations were within normal limits.

The laparotomy was done 7 days after admission, and right salphingo-oophorectomy and omentectomy were performed. There was an intraoperative finding of a pre-operative ruptured tumour with a breached capsule (Figure 1). The histopathological examination was reported as the neoplastic cells exhibit round to oval-shaped vesicular nuclei and inconspicuous nucleoli with eosinophilic to clear cytoplasm. Abundant hyaline globules are present. In some areas, Schiller–Duval bodies are seen (Figure 2). Focal tumour necrosis was also noted. The neoplastic cells are positive for SALL4 and are interpreted as YSTs with no evidence of malignancy in the omentum tissue. The peritoneal fluid for cytology showed scattered lymphocytes and no malignant cells were seen.

She was treated with adjuvant combined chemotherapy of bleomycin, etoposide, and cisplatinum for six cycles, three weekly, completed in 4 months after the operation, and investigation of tumour markers, other blood investigations, and ultrasound of the abdomen and pelvis were done during follow-up. All tumour markers levels showed a reducing trend to reach a normal level and the latest AFP level was 0.93 IU/ml. No local recurrence or distant metastasis was found in CECT, and she was well at home. Her hearing bilaterally was normal prior to the initiation of chemotherapy, and a hearing assessment was done at the Ear-Nose-Throat (ENT) department during follow-up, and her hearing was normal within 9 months of follow-up.

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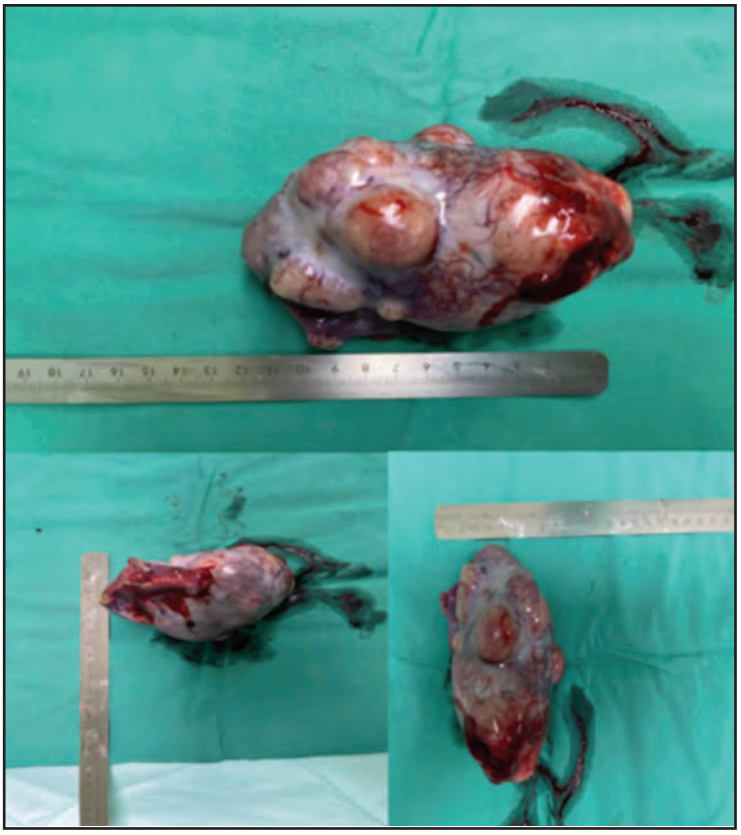

Fig. 1: Intraoperative findings of the right ovarian yolk sac tomour.

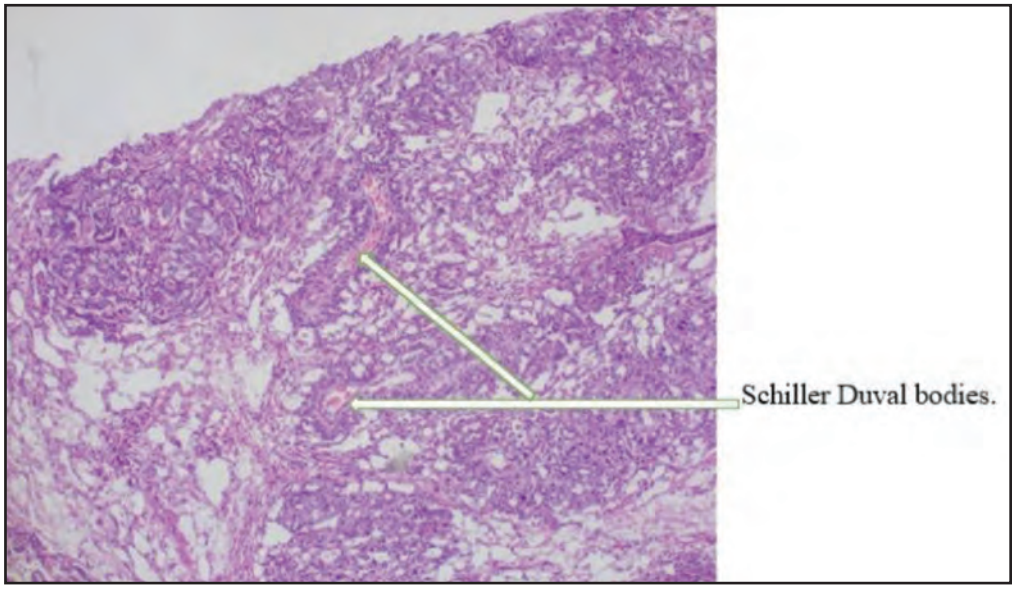

Fig. 2: Histopathological examination finding of Schiller–Duval bodies.

DISCUSSION

The YST is the second most common ovarian germ cell malignancy, following dysgerminoma. Approximately one-third of YSTs are of extra-gonadal origins, such as the vagina, cervix, endometrium, mediastinum, pineal gland, and sacrococcygeal area.[2] At present, the aetiology of YST remains obscure. YST may develop from the false migration of embryonic primitive pluripotent germ cells or oncogenesis of the residual cells when they migrate from the genital ridge of the yolk sac endoderm. Abdominal pain is quite frequently complained of, leading to the discovery of the disease, as seen in our patient. Sometimes, it presents with a mass in the

abdomen, high-grade fever, vaginal bleeding, fever, ascites, or peritonitis secondary to torsion, infection, or rupture of the ovarian tumour. The median age was 18 years (range: 15–22), abdominal pain was the most common presentation (89%), and the mean tumour size was 21 cm (range: 8–30 cm).[3]

An AFP is a specific marker for YST. The association of an adnexal mass and an elevated AFP level is specific to a vitelline component, permitting raised diagnosis with quasi-certainty even before histopathological proof. Imaging investigation classically shows a hyper-vascularized solid-cystic with intratumoral haemorrhagic zones and heterogenous enhancement after administration of contrast product, as seen in our case. The ultrasound examination of the magnetic resonance image is also helpful in showing the hypervascularity and hemorrhagic character of the mass.[1] The typical histopathological features of YST are solid, tubular, and focal papillary patterns with Schiller Duval bodies and sinusoidal structures with fibrovascular core lining formed by tumour cells, frequent mitotic figures, and are cytokeratin positive.[4]

As the tumour was unilateral in our case, the critical management for YST is definitive surgery to eliminate the primary tumour without severe morbidity and fertility-sparing surgery, followed by adjuvant chemotherapy with bleomycin, etoposide, and cisplatin (BEP) regimen chemotherapy. The BEP is considered the gold standard of first-line treatment for germ cell tumours at all stages.[1] Follow-up of these patients includes determining if there is an initially elevated AFP level and repeating it before each cycle of chemotherapy, as in our case. Also, a pelvic ultrasound examination is recommended in cases with conservative surgery to screen for a contralateral recurrence.[5] The survival rate is higher in patients with earlier stages of disease (stages I or II).

CONCLUSION

This is an interesting case of a YST in a young girl at 9 years of age. At a young age, with abdominal pain and a rapidly growing pelvic mass with high AFP levels, the diagnosis of the YST must be kept in mind. The majority of YST is unilateral, and the standard of care is fertility-sparing surgery followed by adjuvant chemotherapy. The follow-up of these patients includes measuring AFP levels before and after the treatment, and annual ultrasound is recommended in cases with conservative surgery for screening of contralateral recurrence.

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CONSENT

Written informed consent was obtained from the patient's father for the publication of this case report and all accompanying images.

CONFLICT OF INTEREST

The authors declare that there was no potential conflict of interest relevant to this article reported.

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