

Malignant Yolk Sac Tumor of the Ovary Presenting as Adnexal Torsion in a Child: A Rare Case Report with Review of Literature

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Abstract

Introduction: Ovarian yolk sac tumor is a rare malignant germ cell neoplasm. We report its presentation with adnexal torsion in a nine-year-old child, highlighting the role of imaging in assessment and management.

Case Presentation: A nine-year-old girl presented with acute right lower quadrant pain, nausea and vomiting. Examination revealed abdominal tenderness and guarding. Laboratory investigations showed leukocytosis, raised inflammatory markers and markedly elevated alpha-fetoprotein. Ultrasound demonstrated a heterogeneous right adnexal mass with absent Doppler vascularity, raising suspicion of torsion. CT and MRI characterized the mass and associated pelvic and para-aortic lymphadenopathy. Emergency laparotomy revealed twisted vascular pedicles. Right salpingo-oophorectomy and lymph node dissection were performed. Histopathology confirmed yolk sac tumor with nodal involvement. The postoperative course was favorable, with complete recovery within two weeks.

Conclusions: This case illustrates the rare presentation of ovarian yolk sac tumor with adnexal torsion in a child. Imaging supports assessment of the mass and its complications; suspected torsion requires timely surgical assessment.

Keywords: Malignant Yolk Sac Tumor, Ovarian Germ Cell Tumor, Adnexal Torsion, Pediatric Ovarian Tumors, Multimodality Imaging

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Abbreviations

1. AFP - Alpha-Fetoprotein
2. LDH - Lactate Dehydrogenase
3. HCG - Human Chorionic Gonadotropin
4. CA-125 - Cancer Antigen 125
5. CA-19.9 - Cancer Antigen 19.9
6. CEA - Carcinoembryonic Antigen
7. CECT - Contrast Enhanced Computed Tomography
8. MRI - Magnetic Resonance Imaging
9. US - Ultrasonography
10. RLQ - Right Lower Quadrant
11. BEP - Bleomycin, Etoposide, and Cisplatin (Chemotherapy regimen)
12. OGCT - Ovarian Germ Cell Tumors
13. GCT - Germ Cell Tumors
14. CT - Computed Tomography
15. DWI - Diffusion Weighted Imaging
16. ADC - Apparent Diffusion Coefficient
17. PID - Pelvic Inflammatory Disease
18. UTI - Urinary Tract Infection
19. ESR - Erythrocyte Sedimentation Rate
20. PAS - Periodic Acid-Schiff
21. SALL4 - Spalt-Like Transcription Factor 4

Introduction

Ovarian germ cell tumors arise from the germ cells of the ovary and account for ~ 15-20% of all ovarian neoplasms (1). Malignant germ cell tumors of the ovary are rare with an incidence of ~ 1-2% (2). Yolk sac tumors, also known as endodermal sinus tumors, are highly aggressive with a predilection for gonadal sites (3). The gonads, particularly the ovaries, are the primary sites for these tumors. However, extragonadal sites like broad ligament, retroperitoneum, mediastinum and sacrococcygeal region have also been reported (3-5). These tumors are usually seen in young women and adolescent girls. Occurrence in children less than 10 years of age is extremely uncommon with a few sporadic case reports. Unlike epithelial tumors, malignant yolk sac tumors are locally aggressive and possess high metastatic potential. Due to their aggressive nature, early detection and management are critical for improving survival outcomes.

This highlights the rare presentation of malignant yolk sac tumor complicated by adnexal torsion, emphasizing the role of multimodality imaging in timely management.

Case Presentation

A 9-year-old girl child was brought to the emergency department with acute onset, severe right lower abdominal pain, nausea and few episodes of non-bilious vomiting. No history of preceding trauma, fever, or changes in urinary or bowel habits could be elicited. She had no prior abdominal surgeries or known health issues.

On physical examination, the patient appeared visibly distressed and was clutching her abdomen. She was afebrile with stable vital signs. Abdominal examination revealed marked tenderness in the right lower quadrant with guarding. Laboratory investigations showed a leucocytosis (white blood cell count of 18,600/ μ L) and elevated inflammatory markers (ESR 32 mm/hr). Notably, alpha-fetoprotein (AFP) level was markedly elevated at 32,134 ng/mL, strongly suggestive of a malignant germ cell tumor. Lactate Dehydrogenase (LDH) was also elevated at 635 IU/L. Levels of other tumor markers were within normal limits (Human Chorionic Gonadotropin (HCG)- 0.74 ng/ml; CA-125- 14.6 U/ml; CA-19.9 <0.8 U/ml; Carcinoembryonic Antigen (CEA)- 1.52 ng/ml).

Imaging Findings

Pelvic ultrasound revealed a large, heterogeneous, solid-cystic mass measuring 8 x 7.5 cm in the right adnexa. The mass exhibited mixed echogenicity, with areas of necrosis (Figure 1). Colour Doppler imaging demonstrated no vascularity within the lesion, suspicious of adnexal torsion. Free fluid was noted in the pelvis, raising further concern of a possible adnexal torsion. Few enlarged para aortic lymph nodes also noted.

Contrast enhanced computed tomography (CECT) confirmed the presence of a lobulated heterogeneously enhancing solid pelvic mass measuring approximately 8.0 x 8.0 cm in the right adnexa (Figure 2). It was seen compressing the uterus, displacing it towards the left, and causing a significant mass effect on the urinary bladder and the right ureter, resulting in moderate right-sided hydroureteronephrosis. Minimal ascites was present. Multiple enlarged heterogeneously enhancing para-aortic and pelvic lymph nodes were identified, largest measuring up



Figure 1: Grey-scale ultrasonogram showing a heterogeneous solid mass in the pelvis, predominantly involving the right adnexa.

to 2.5 cm in diameter, consistent with nodal metastasis. No evidence of distant organ involvement was observed.

Magnetic resonance imaging (MRI) was subsequently performed for further characterization. It revealed a lobulated solid lesion in the right hemipelvis, appearing isointense on T1 and heterogeneously hyperintense on T2 with areas of restricted diffusion (Figure 3). Right ovary could not be visualized separately from the lesion. Uterus was displaced to the left. The left ovary was visualized distinctly and measured 1.5 x 1.0 cm. Minimal hemoperitoneum was noted. It was also compressing the urinary bladder and the right ureter, causing moderate hydroureteronephrosis. Additionally, multiple enlarged

heterogeneous pelvic and paraaortic lymph nodes were noted.

Considering the radiological features, malignant ovarian germ cell tumor with multiple metastatic lymph nodes was considered. In view of acute presentation, absent vascularity on colour Doppler and presence of intraperitoneal hemorrhagic fluid, possibility of associated adnexal torsion was also considered.

Management

Considering the possibility of adnexal torsion, emergency laparotomy was performed. Intraoperatively, a solid right adnexal

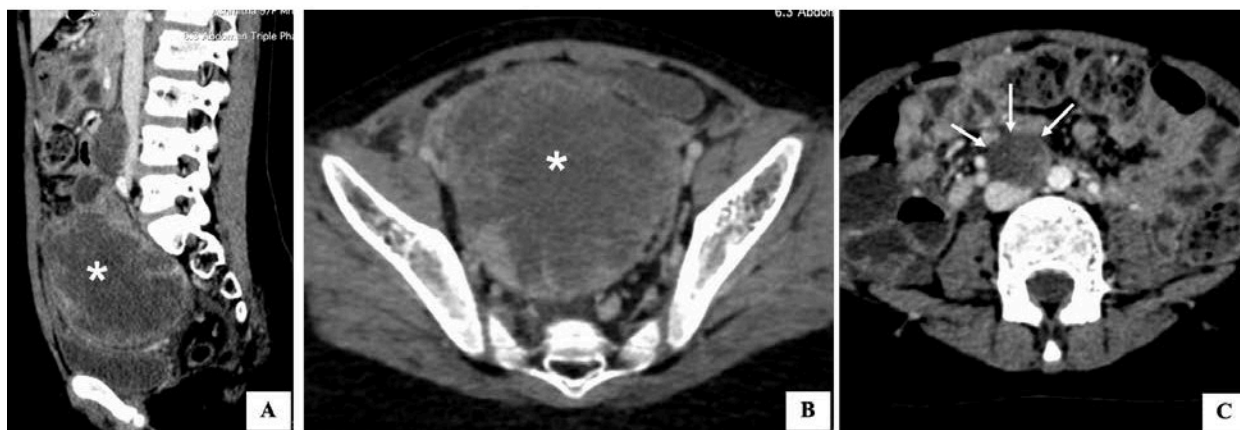


Figure 2: Sagittal reformatted (A) and axial (B and C) contrast enhanced CT images show a heterogeneous lesion in the pelvis (asterisk) with enhancing solid areas, predominantly in the periphery. An enlarged heterogeneously enhancing paraaortic lymph node is also noted (white arrows).

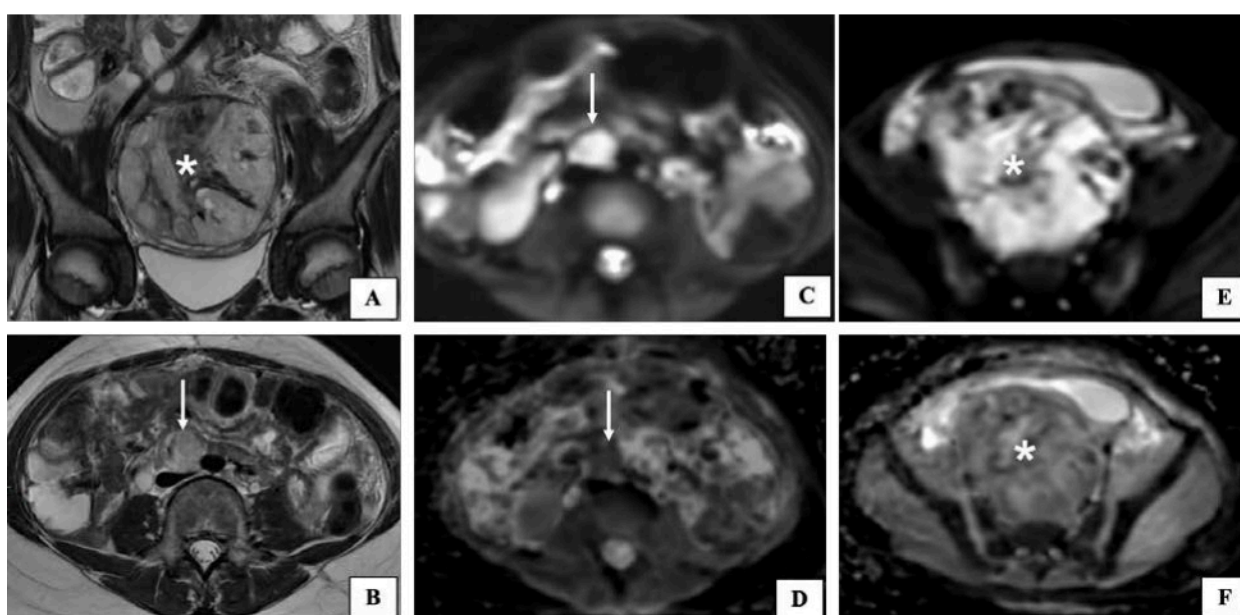


Figure 3: MRI Coronal T2 (A), axial T2 (B), axial DWI (C and E) and axial ADC (D and F) images show a heterogeneous solid lesion (asterisk) in the pelvis appearing hyperintense on T2 with internal hypointense fibrous septae and demonstrating restricted diffusion. An enlarged necrotic paraaortic lymph node (white arrow) is also noted with diffusion restriction.

mass with necrotic lymphadenopathy was observed, complicated by twisted vascular pedicles. Right salpingo-oophorectomy with lymph node dissection was performed. The postoperative course was favorable, with complete recovery within two weeks.

Histopathological Examination

Histopathological examination (Figure 4) revealed a tumor composed of cells arranged in microcystic, festoon, and glandular patterns. The tumor cells exhibited nuclear pleomorphism and atypia. Hyaline globules were observed, along with many areas showing Schiller-Duval body formation. Extensive areas of hemorrhage and necrosis were noted. Dense inflammatory infiltrate composed of neutrophils was present, and

brisk mitotic activity was observed. The ovarian capsule was ruptured. The fallopian tubes were also infiltrated by tumor cells. The tumor cells showed Periodic Acid Schiff (PAS)- positive hyaline globules and strong immunoreactivity to Alpha-FetoProtein (AFP) and Sal-like protein 4 (SALL4). These findings were diagnostic of malignant yolk sac tumor. Para aortic and Pelvic lymph nodes showed evidence of metastatic involvement.

Discussion

One of the most common causes of Right lower quadrant (RLQ) pain in children is acute appendicitis, which typically presents with periumbilical pain migrating to the RLQ,

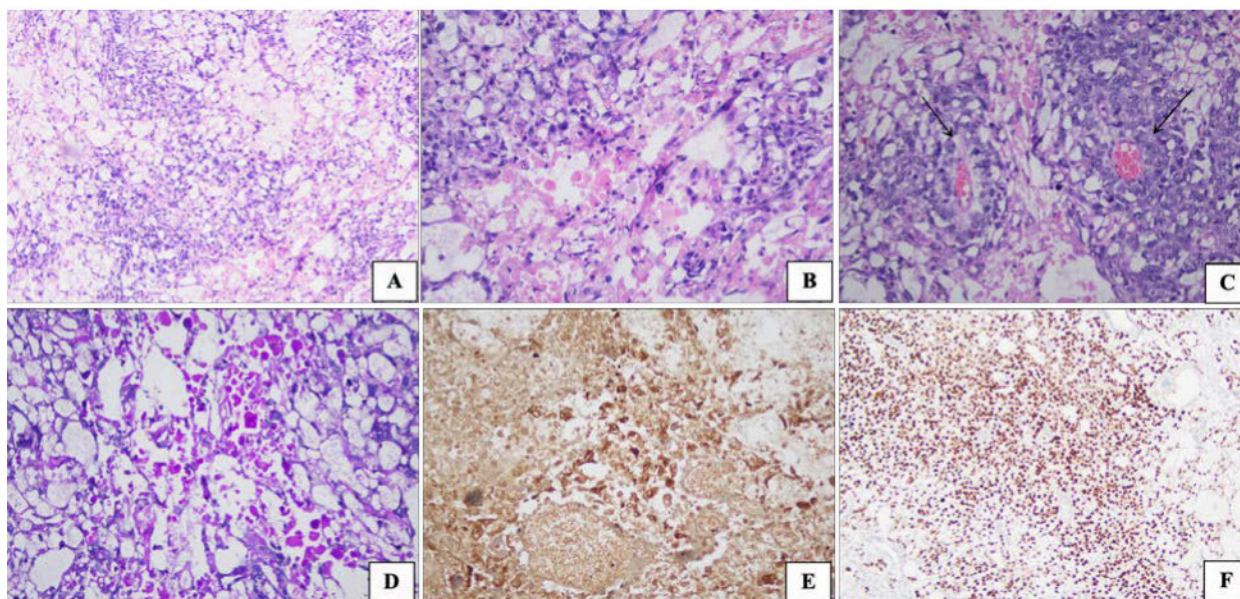


Figure 4: Microscopic images representing the histomorphology and immunophenotyping of the tumor. Histopathological images with hematoxylin and eosin (H&E) stain in 10x (A) and 20x (B and C) magnification show tumor cells arranged in microcystic and glandular configuration, demonstrating an eosinophilic cytoplasm, nuclear pleomorphism and cellular atypia. Multiple Schiller-Duval bodies (black arrows in C) characterized by a central vessel with surrounding sheets of tumor cells are noted. Periodic Acid Schiff (PAS) stain (D) highlights the presence of hyaline globules. The tumor cells show positive immunoreactivity to AFP (E) and SALL4 (F).

accompanied by fever, anorexia and nausea (6). Ultrasound is the initial imaging examination for suspected appendicitis in children. Its performance depends on visualization of the appendix and the interpretation of associated findings (7). Mesenteric adenitis can mimic appendicitis, often follows a viral illness and is usually self-limiting (8). Intussusception is another important consideration in infants and toddlers; ultrasound can establish the diagnosis, and image-guided enema reduction is a treatment option in appropriate cases.

In female children, gynecologic causes must be considered. Ovarian torsion is a surgical emergency, often presenting with sudden abdominal pain, nausea and vomiting. Ultrasound is the initial imaging modality, but imaging alone cannot confirm or exclude torsion. Clinical suspicion requires timely surgical assessment (9).

Another important consideration is ovarian cyst rupture, which may cause acute RLQ pain and free fluid visible on imaging. Additionally, Mittelschmerz, or mid-cycle ovulation pain, may present as self-limited RLQ pain in pubescent girls (10). For sexually active adolescents, pelvic inflammatory disease (PID) and ectopic pregnancy are

critical considerations. Pregnancy testing should be considered when clinically appropriate to assess the possibility of ectopic pregnancy (11). Urinary tract infection may also cause abdominal pain in children; evaluation includes urine testing according to age, symptoms and clinical findings (12).

Ultrasound is the initial modality for assessing pediatric adnexal lesions. Gray-scale imaging evaluates lesion morphology and Doppler assesses vascularity. Reduced or absent Doppler flow may support suspicion of torsion, but preserved flow does not exclude it (9). Functional cysts often have a simple appearance, whereas complex or solid components warrant further characterization. CT and MRI can provide additional information about adnexal lesions and disease extent when clinically indicated (13).

MRI is useful for characterizing masses that remain indeterminate after ultrasound. Its soft-tissue contrast helps assess lesion composition and features associated with malignancy. Pediatric evidence supports MRI assessment of ovarian masses, while risk models developed in adults require evaluation in children (14). CT may be used selectively for staging or other clinical indications, with consideration of radiation exposure (13). Additional imaging should not delay surgical assessment when torsion is suspected (9).

Germ cell tumors are a major category of ovarian neoplasms in children (15). They include benign and malignant histologies. Reported malignancy proportions vary with the population and referral setting: institutional series of ovarian masses or tumors cannot establish a universal malignancy rate for all pediatric ovarian germ cell tumors (16,17).

Mature teratomas are benign and contain differentiated tissues such as skin, hair and fat. They may cause symptoms when complicated by torsion or rupture. Malignant histologies include dysgerminoma, yolk sac tumor and mixed germ cell tumors. Immature teratomas contain immature tissues and require separate consideration according to their histology and stage. Yolk sac tumors are associated with elevated alpha-fetoprotein (AFP), while mixed tumors contain more than one germ cell component (18).

Serum tumor markers contribute to diagnosis, treatment monitoring and follow-up. AFP is characteristically elevated in yolk sac tumors; its serial measurement helps assess treatment response.

Beta-human chorionic gonadotropin (beta-hCG) may be elevated in choriocarcinoma and some mixed germ cell tumors. Dysgerminoma can be associated with elevated lactate dehydrogenase (LDH). CA-125 may be elevated in epithelial ovarian tumors or other conditions. Inhibin and estradiol may be elevated in sex cord-stromal tumors, including granulosa cell tumors, which may cause precocious puberty.

The utility of tumor markers extends beyond diagnosis to guiding management. Preoperative levels of AFP, β -hCG, and LDH can provide insights into the likelihood of malignancy and the tumor subtype, assisting in surgical planning. Postoperative monitoring of these markers is essential to evaluate treatment response and identify early recurrences. Despite their utility, serum tumor markers must be interpreted alongside clinical findings and imaging, as false positives can occur in certain physiological or pathological conditions (19).

Treatment of pediatric germ cell tumors requires multidisciplinary assessment, balancing oncologic control with preservation of unaffected reproductive organs and reduction of treatment-related toxicity. Surgery should follow oncologic staging principles and preserve unaffected ovarian tissue where appropriate. The need for chemotherapy is determined by histology, stage and pediatric treatment protocols (20).

Platinum-based chemotherapy is an important component of treatment for many malignant germ cell tumors. Pediatric and adolescent protocols differ in their drug schedules and risk stratification.

Selected stage I dysgerminomas may be managed with surgery and surveillance. More advanced disease generally requires chemotherapy. Management of immature teratomas differs from that of yolk sac and mixed malignant tumors. Radiotherapy is not a routine treatment for pediatric ovarian germ cell tumors (21).

Treatment response is assessed using the relevant tumor markers and clinical and imaging findings. Persistent marker elevation warrants evaluation for residual disease or recurrence. Long-term follow-up also addresses treatment-related toxicity and reproductive outcomes. Prognosis varies with age, stage, primary site and histology; a single survival estimate should not be applied to all pediatric germ cell tumors (22).

Conclusions

This case illustrates ovarian yolk sac tumor presenting with adnexal torsion in a child. Ultrasound supports initial evaluation of pediatric adnexal masses, with MRI or CT selected according to the clinical question. When torsion is suspected, further imaging should not delay timely surgical assessment.

Declarations

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Competing interests

The authors declare no competing interests.

Ethical considerations

The authors report that procedures conformed to institutional ethical standards and the Declaration of Helsinki.

Consent for publication

The authors report that written informed consent was obtained from the patient's parents for publication of the case and radiological images, with patient anonymity ensured.

Data availability

The clinical findings and relevant imaging are described in this case report. No separate research dataset was generated.

Authors' contributions

KAK: concept and design, literature research, manuscript preparation and editing.

EA: guarantor of integrity, concept and design, literature research, manuscript preparation and editing.

KN: concept and design, literature research, manuscript editing.

PS: concept and design, literature research, manuscript editing.

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