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Case Report

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Advanced Ovarian Yolk Sac Tumor in Nine-Year-Old Girl: Case Report and Literature Review, 2023

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ABSTRACT

Background: Yolk sac tumor or primary endodermal tumor is a rare and aggressive malignant tumor, representing 15% to 20% of germ cell tumors of the ovaries, characterized by its high sensitivity to chemotherapy, often-present adolescents from 18 to 30 years old.

Case Report: This is a 9 years old female presented with abdominal pain and distention associated with constitutional symptoms and significant weight loss, which she cannot exactly quantify. She had no family history of similar problems and denies any change in bowel habit, vaginal discharge or bleeding. Besides, she had no development of any secondary sex characteristics. On physical examination, she was not in distress with stable vital signs. Chest was clear to auscultation and abdomen was distended with slight deviation to the right side, which is hard, non-tender mobile, with irregular border about 20x18cm size, reaching to the level of umbilicus. There was no sign of fluid collection or inguinal lymph node enlargement. serum tumor markers revealed alpha fetoprotein level >1210 ng/dl, CA 125 of 70.1U/mland CT scan showed right ovarian mass with paraa ortic lymph node enlargement and attachment of the mass to the rectum. Under general anesthesia, midline incision done and about 120mla serous ascetic fluid of was evacuated. Hard, irregular right adnexa mass was identified and a huge retroperitoneal mass fixed to big blood vessels with multiple enlarged para aortic lymph nodes was identified. The omentum was with multiple kicks and the liver surface was irregular with multiple nodules at its lower edges. Peritoneal fluid cytology revealed malignant metastasis and the histopathology analysis was consistent with yolk sac tumor. She was staged surgically as 4B and discharged after clinical improvement to go abroad for chemotherapy treatment.

Conclusion: The histopathology, cytology and intraoperative clinical finding were consistent with serum tumor markers that she had advanced stage yolk sac tumor with poor prognosis. Community and health professionals' awareness for early recognition of the disease for better prognosis and initiation of pediatric chemotherapy in Eritrea is crucial, as these diseases are highly responsive to radio-chemotherapy.

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Introduction

Ovarian germ cell tumors (OGCTs) account for about 15~20% of all ovarian neoplasms. Only 1~2% of OGCTs are malignant called malignant ovarian germ cell tumors (MOGCTs) and constitute around 3–5% of all malignant ovarian neoplasms. Yolk sac tumor (YST) or endodermal primitive tumor accounting the second most common tumor in MOGCTs and is rare and typically occurs in gonads [1-4]. In comparison with epithelial ovarian tumors, the endodermal tumor of the ovary is very malignant and develops rapidly with a very short duration of symptoms and rapid metastases invading all intra-abdominal structures and retroperitoneal lymph nodes [1, 2, 4].

Ovarian yolk sac tumors are often detected at an early stage in young women, usually in the second or third decade of life, presenting with pain due to very rapid growth and markedly elevated serum alpha-fetoprotein (AFP). It has been suggested that AFP can be applied as a feasible tumor marker because its level was elevated in >90% of YST and the prognosis of patients can be monitored by the AFP level after operation [5]. The tumor is almost always unilateral and the median diameter is 15 to 19cm. Often there is rapid growth with extensive intra-abdominal spread leading to poor prognosis [2, 4, 6].

The typical histological appearance is a clear cell proliferation organized in a generally microcystic network with Schiller-Duval bodies which are cellular structures resembling a fetal glomerulus, pathognomonic of endodermal tissue. AFP is a specific tumor marker; the association of an adnexal mass and a high AFP level are specific to a vitelline component of the tumor allowing the diagnosis to be made with almost certainty [2]. Even before histological proof, its rate was high in more than 90% of yolk sac tumors [2, 7, 8].

The standard treatment approach to OYSTs is debulking surgery, followed by adjuvant chemotherapy. It has been found that the overall 5-year disease-free survival rates with stage I/II and stage III/IV OYSTs were 94% and 90%, respectively. Fertility-sparing surgery is often possible, as the tumors are unilateral. Even in patients with bulky metastases, a normal appearing uterus and contralateral ovary can be safely preserved allowing for future fertility [3, 4]. Given the very high chemo-sensitivity of yolk sac tumors, potential nodal metastasis should be targeted by adjuvant chemotherapy, and nodal dissection should be carried out only in the case of nodal abnormality [4, 6]. The BEP protocol (Cisplatin, Etoposide, Bleomycin) extrapolated from the treatment of testicular germinal tumors showed an efficacy equivalent to the PVB protocol (Cisplatin, Vinblastine, Bleomycin) with less toxicity [5]. We report a malignant yolk sac tumor with liver metastasis in a nine-year-old girl [2].

Case Report

This is a 9 years old female child from villages around Asmara, the capital city of Eritrea; presented with abdominal pain and distention of two weeks' duration. She visited with this complaint to her nearby clinic and referred to Orotta National Referral Pediatric hospital on 22/12/2022. After 04 days stay in the pediatric hospital for investigation and further management, she was transferred to Orotta National Referral Maternity Hospital with the diagnosis of ovarian tumor. She had history of non-projectile vomiting of ingested material of 1-2 times/day accompanied by loss of appetite and generalized body weakness. Her mother reported that she had significant weight loss that she can't exactly quantify but there was no history of fever, diarrhea, vaginal discharge or bleeding. She is good in school performance and vaccinated with currently available vaccines for her age. She had no history of any chronic illness and previous exposure to radio chemotherapy. There is no report of similar illness in the family.

On physical examination, she was sick looking, but not in cardiorespiratory distress. Vital signs revealed blood pressure of 90/60mmHg on left arm sitting position, pulse rate of 109 beats per minute, respiratory rate of 30breaths per minute, random blood sugar of 120g/dl and temperature of 36.7oc on right axilla. She had pink conjunctiva and non-icteric sclera without any enlargement of lymph around the neck. Chest was clear to auscultation with good air entry. Abdomen was distended to the level of umbilicus; asymmetrical deviated more to the right side. There was no skin discoloration or collateral veins distention. A hard, non-tender, mobile mass with irregular border of 20x18cm size, reaching to the level of umbilicus was identified. Fluid thrill and shifting dullness were negative and, on per rectal digital examination the mass was palpated more on the right side of adnexa.

She was investigated with complete blood count, β-hCG titter on urine dipstick, renal and liver function test, contrast enhanced CT scan of abdomen and pelvic, ultrasonography and tumor markers. Her hemoglobin was 11.9g/dl, Alpha feto protein level was >1210 ng/dl and CA 125 was 70.1U/ml (Table: 1). She was negative for β-hCG titter on urine dipstick. The CT scan revealed right ovarian mass with para-aortic lymph node enlargement and attachment of the mass to the rectum (Figure: 1).

Table 1: Different investigations of the case

Investigations	Result	Reference value
Hematology		
WBC	10.38x10^3/µL	3.71 - 10.67 x10^3
Hemoglobin	11.9g/dl	12.00 - 16.75
Hematocrit	37.5%	35.1 - 48.7
Platelets	562.8 x10^3/µL	150.5-366.8 x10^3
Serum tumor markers		
CA-125	70.1U/ml	0.0 - 34 U/mL
CA19-9	2.6 U/mL	0.0 - 39 U/mL
Carcinoembryonic Ag	1.08 ng/mL	0 - 3.0 ng/mL
Alpha-fetoprotein (AFP)	>1210 ng/dl	0.0 - 8.1 ng/mL
Renal, liver function test and serum electrolytes		
BUN	8 mg/dL	6 - 20mg/dL
Creatinine	0.6 mg/dL	0.5 - 1.1mg/dL
AST	34 U/L	0 - 31U/L
ALT	11 U/L	0 - 31U/L
Albumin	3.3 g/dL	3.4 - 4.8g/dL
Sodium (Na)	138 mmol/L	135 - 145mmol/L
Chlorine (Cl)	100 mmol/L	101 - 111mmol/L
Potassium (K)	4 mg/dL	8.8-10.2mg/dL
Imaging, Histopathology		
CT scan, with contrast	Right adnexal mass with lymphadenopathy	
Peritoneal cytology	Malignant metastatic cells	
Histopathology	Yolk Sac Tumor	

Figure 1: CT scan with contrast enhanced

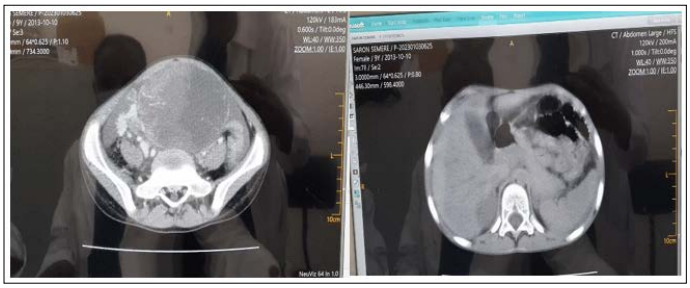

Figure: 1Big right ovarian mass of about 20x18cm size with para-aortic lymphadenopathy and attachment to the posterior intra-abdominal structures

With the diagnosis of advanced ovarian mass, patient was scheduled for laparotomy and family was counseled about the possible interventions and prognosis of the disease. Under general anesthesia, midline incision was done and about 120ml of serous ascetic fluid was evacuated. Hard, irregular right adnexal mass was identified and resected but huge retroperitoneal mass of about 10cm x8cm size, fixed to big blood vessels with multiple enlarged para aortic lymph nodes was identified which was unrespectable and left behind. The para-aortic lymph nodes were markedly enlarged with multiple kickson the omentum and the liver was nodular with irregular edge. Right salpingo-oophorectomy with resection of the adnexal mass and omentectomy was done, and all samples sent cyto-histopathology analysis (Figure: 2). She was staged surgically as 4-B, and abdomen was closed after complete

gauze count and securing hemostasis. The peritoneal fluid cytology revealed malignant cell metastasis and the histopathology revealed yolk sac tumor. She was discharged from hospital after clinical improvement to go abroad for chemotherapy.

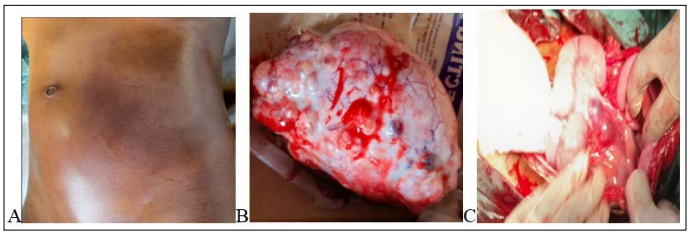

Figure 2: (A) Swollen abdomen that reached to the level of umbilicus (B) Irregular, about 20x18cm right ovarian mass intra operatively after removal (C) enlarged multiple para-aortic lymph nodes with size 4x3cm, 2x3cm and 2x2cm, fixed with the posterior structures.

Histopathology Description

Macroscopic Description: One piece of ovarian tumor with attached tubes measuring 12x20x18cm and on cross section, it shows smooth nodular inner surface with necrosis and hemorrhage.

Microscopic Description: Sections show infiltration encapsulated malignant tumor forming sheath microcytic structure with a papillary structures seen and hyaline globules. The tumor features high mitotic activities and featuring vascular invasion, not hemorrhagic, also schiller Duval body seen. Features are consistent with Yolk Sac Tumor.

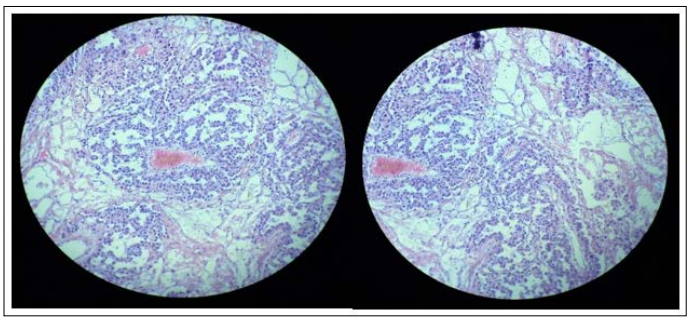

Figure 3: vascular invasions with schiller Duval body

Discussion

Yolk sac tumors are highly malignant, rapidly growing tumors with a very brief duration of symptoms and metastasize rapidly to the peritoneum, lymph nodes, and via the hematogenous route. Malignant pediatric ovarian tumors seem becoming increasing in Eritrea. There are also previous two reported malignant ovarian cancers in the pediatric age group with poor prognosis recently in Eritrea [1, 2, 5]. The pathogenesis is not clear and they had no any identified risk factor for the disease process [9].

The disease progress was so rapid in weeks as the caregivers reported. But the intraoperative finding like liver nodularity doesn't seem to be of such short duration. So, the clinical manifestation of abdominal pain and distension could be seen long after the onset of the problem. This was consistent with literatures that abdominal pain and distension may be the only clinical presentation, which is a common manifestation of the most highly malignant tumor of the ovaries. The abdominal pain could be mainly due to the retroperitoneal mass because it persists, even progressively worsened after surgery and due to the fast-spreading pattern of the disease to nearby intra-abdominal structures and the pressure effect of the mass could have an effect for the clinical presentation [3, 5, 9]. Complete or near complete cyto reduction could not only affects chemotherapy response but also the quality of life is also affected.

Intraoperatively, there was intraperitoneal and retroperitoneal mass without direct continuation. The origin of retroperitoneal mass could be from right side of the para aortic lymph nodes. Although the left ovary and uterus was healthy, left side para aortic lymph nodes were enlarged. The intra-abdominal lymph nodes were so prominent and it was confined to the abdomen not extending to inguinal or cervical lymph nodes. The liver was also severely affected with the bowel and the peritoneal surface look normal. She had about 20x18cm, unilateral ovarian tumor with hard nodular liver and metastasis to the para aortic lymph nodes that suggests advanced level of the disease. This early metastasis is the hallmark of yolk sac tumor, reliable with most literatures. In advanced stages, retroperitoneal lymph nodes and liver parenchyma are involved [1, 4, 6]. The size of the mass and being unilateral is also reported in many studies [2, 6]. This highly malignant and early metastasis of the tumor could be associated with poor prognosis of the disease [4, 6, 8].

It has been suggested that AFP can be applied as a tumor marker as its level is elevated in about 90% of cases [2]. This case had AFP level of >1210 ng/dl with was consistent with the histopathology. Almost all literatures reported high level of AFP in patients with Yolk sac tumor as AFPof 71,300 ng/ml, 1156 UI/ml, 13220 ng/ml and 28 000 ng/ml [1, 5, 7]. This markedly elevated serum AFP level could be used in diagnosis, management and prognosis of yolk sac tumor [2].

Yolk sac tumor have typical histological finding with clear cell proliferation organized in a generally microcytic network with Schiller-Duval bodies, which are pathognomonic of endodermal tissue. This was similar to this case, which showed infiltration encapsulated malignant tumor forming sheath microcytic structure with Schiller Duval body [3, 5]. This indicated that the significance of histopathologic analysis for the diagnosis of yolk sac tumor.

The peritoneal cytological analysis revealed malignant metastasis, which was constant with the intraoperative finding of enlarged para-aortic lymphadenopathy and liver metastasis. The serum tumor markers and histopathology results were also consistent for the advanced stage of the disease. All these findings suggest the general clinical presentation of yolk sac tumor for being highly malignant with rapid progression of the disease.

A debulking surgery with fertility preserving unilateral salpingo-oophorectomywas done and it was surgically classified as stage IV-B of the FIGO 2014 classification. Literatures reported that treatment generally involves debulking surgery of tumors followed by systemic chemotherapy and treating young patients is by using fertility-sparing strategy. The patient age is helpful in many of these considerations [2, 3, 7]. As yolk sac tumors are highly responsive to chemotherapy, early initiation is crucial for the management of the disease process [4, 6].

Even though she had advanced ovarian tumor and poor prognosis, she could have benefit from chemotherapy just within two-four weeks postoperatively. As pediatric chemotherapy is not started in Eritrea, she was sent abroad, and her outcome cannot be determined. It has been found that the overall 5-year disease-free survival rates with stage I/II and stage III/IV OYSTs were 94% and 90%, respectively. In addition, the prognosis has improved considerably, even for patients at an advanced stage, with survival going from 14% to almost 90% [2, 5].

Conclusion

The onset of the disease was acute with malignant metastasis on peritoneal cytology analysis, intraoperative finding with metastasis, high AFP level and histopathology, which revealed advanced stage of the disease consistent with the clinical findings of yolk sac tumor. Health professional's awareness and starting pediatric chemotherapy in Eritrea is highly recommended. Raising community awareness and further research to determine the prevalence of ovarian tumors is crucial for early intervention and decreasing the morbidity and mortality related to the disease process.

Declarations

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Competing of interest: Authors did not have any conflict of interest to disclose

Ethical approval: Written informed consent was obtained from the guardian to present in case report

Author's contribution: All authors have contributed on data analysis, interpretation and writing.

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