AN ATYPICAL AGGRESSIVE PRESENTATION OF A MALIGNANT ENDODERMAL SINUS TUMOUR IN A YOUNG GIRL - A CASE REPORT

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Abstract

Malignant ovarian germ cell tumours (OGCT) comprise only 2-5% of all ovarian malignant cancers. Endodermal sinus tumour (Yolk sac tumour-YST) is the second most common malignant germ cell tumour of the ovary after dysgerminoma and comprises approximately 20–25% of malignant germ cell tumours and 2 - 5% of all the ovarian malignancies. YST is almost always unilateral and large. YST are highly aggressive and because of the early metastatic or invasive behaviour, their prognosis has been poor. Here, we report a case of endodermal sinus tumour in a 19-year-old girl with lower abdominal pain for one month. A pelvic mass was found and being investigated but emergency laparotomy was performed for severe acute abdominal pain. An intraoperative diagnosis of Stage IIIC ovarian tumour with bilaterality and intraperitoneal rupture was made. She required extensive debulking surgery, later confirmed as yolk sac tumour.

Keywords: Yolk sac tumour, endodermal sinus tumour, ovarian germ cell tumour

Introduction

Endodermal sinus tumour, also known as yolk sac tumour, is a rare malignant ovarian tumour that usually occurs in the second decade of life. It is most commonly unilateral and large. The most common presenting symptoms for patients with endodermal sinus tumours are, a rapidly enlarging mass and pain. The OGCT should be considered as a diagnosis in any girl or young woman with a pelvic mass. Prior to menarche, 90% of ovarian tumours are OGCTs, between the ages of 13 and 20, 60% are of germ cell origin, with 65% being malignant. In the embryo, germ cells appear in the wall of the yolk sac, migrate to the genital ridge and become incorporated into the developing gonad. In common with testicular GCTs, ovarian GCTs are derived from the differentiation of these germ cells into somatic tissues (teratoma), germinal epithelium (dysgerminoma), extra-embryonic trophoblast (choriocarcinoma) and yolk sac (yolk sac tumour).

Because of the aggressive nature and occurrence of YST in young adolescent period, greater concern about future fertility is needed. The feasibility of fertility sparing surgery has been discussed at length for decades. The recommended treatment is unilateral salpino-oophorectomy followed by combination chemotherapy. Affected patients can be diagnosed by elevated serum α-fetoprotein (AFP) and lactate dehydrogenase (LDH) levels. However, reports of pre-operative diagnosis of endodermal sinus tumour in an adolescent by combining images from ultrasound and magnetic resonance imaging (MRI) as well as AFP determination are rare. We report a case of endodermal sinus tumour of a very virulent nature.

Case Report

A 19-year-old girl presented to the emergency with lower abdominal pain for the past one month. Pain was crampy and increased with urination, defecation and eating.

Patient had been taking NSAIDS with minimal relief. She also complained of constipation on and off and increased frequency of micturition. Constipation was not relieved by taking oral laxatives. There was history of loss of appetite and loss of weight. Patient’s menarche started at the age of 13, and she had normal menstrual cycles but associated with spasmodic dysmenorrhoea.

The physical examination revealed an alert, conscious, oriented female in distress. Her pulse rate was 96 beats per min with bilateral oedema of lower limbs. Her pulse rate was 96 beats per minute. The abdomen was distended. The bowel sounds were present.

Ultrasound revealed presence of 17X 19X18.5 cm large right ovarian solid mass with few cystic areas occupying pelvis and lower abdomen with bilateral hydronephrosis and hydroureter with minimal ascites. Left ovary was not visible. Contrast-enhanced computed tomography (CECT) showed presence of malignant ovarian mass 9.5 X 15.4 X 20 cm with possible invasion of rectum and sigmoid colon, minimal malignant ascites, peritoneal deposits and retroperitoneal lymphadenopathy with bilateral hydroureteronephrosis (Figure 1).
Figure 1. Photograph of the coronal view of CECT whole abdomen showing the ovarian mass.

Her karyotype was normal (46XX). At this stage routine tumour markers were sent for investigation. Her case was evaluated by multidisciplinary team. Bilateral ureteric stenting was done. Despite being on antibiotics and intravenous analgesics, her condition deteriorated with severe acute pain and tachycardia, necessitating an urgent laparotomy with the diagnosis of torsion. Tumour marker reported as follows - CA 125 - 54.5, LDH 1639, AFP >400, CEA 0.96 and HCG <2.39, suggestive of ovarian malignancy.

A midline laparotomy was performed with the help of oncology team. Peroperative findings were large right polypoidal necrotic mass with ruptured capsule involving left ovary (Figure 2).

Figure 2. Photograph of the intra-peritoneal ruptured ovarian tumour exhibiting polypoidal and extremely fragile mass.

Posterior wall of uterus, sigmoid colon, hepatic flexure of transverse colon and omentum with enlarged para-aortic lymph nodes and omental caking was present (Figure 3).

Figure 3. Photograph of the uterus (circled) and adhered involved colon (yellow arrow).

A 2x2 cm firm nodule was felt in the right lobe of the liver. Surgical Staging was done and found to be of stage IVB ovarian cancer. Frozen section revealed malignant germ cell tumour. A pelvic clearance including total abdominal hysterectomy, bilateral salpingo-oophorectomy, right hemicolecctomy, infracolic omentectomy, colostomy, and peritoneal washings were done in an attempt to achieve optimal debulking and better chemotherapeutic effect. Peritoneal washings revealed malignant cytology.

Histopathological examination revealed endodermal sinus tumour containing areas of tumour necrosis and haemorrhage. There were scattered eosinophilic globules and Schiller-Duval bodies throughout the specimen (Figure 4), involvement of 40% of myometrium, bilateral parametrium, contralateral ovary, posterior lip of the cervix and adjacent transverse colon and omentum.

Figure 4. Microphotograph of the endodermal sinus tumour showing myometrial invasion (yellow arrow) and the Schiller’s Duval Bodies (inbox Red Arrow). H & E stain 10x (Inbox 40x).
No normal ovarian tissue was found. Vaginal flap was free of the tumour. The patient was started on bleomycin, etoposide and cisplatin regimen immediately after confirmation with biopsy report on postoperative day 10 after removal of stiches. Patient is on regular follow up.

Discussion

Yolk sac tumours are rare ovarian malignancies. Their incidence increases sharply around puberty and decreases in older age groups. Germ cell tumours frequently occur in girls with dysgenetic gonads with Y chromosome component. The most frequent presentation is that of abdominal pain and large pelvic mass. The average size of the tumour is usually 14 cm at the time of presentation and are frequently unilateral. If complications such as hydronephrosis or ovarian torsion are present, more acute symptoms can occur as seen in the present case. Before the introduction of effective chemotherapy, the prognosis of the patient diagnosed with yolk sac tumour involving surgical treatment was poor. After combination chemotherapy was introduced, the survival rate improved dramatically. A recent study reported that an elevation of the serum markers β-human chorionic gonadotropin and AFP are independent poor prognostic factors in malignant ovarian germ cell tumours. However, serum AFP was not an independent prognostic factor in YST.

Microscopic examination of the endodermal sinus tumour revealed multiple patterns. The most common appearance is a reticular, honeycombed structure of communicating spaces lined with primitive cells. Schiller-Duval bodies, which resemble the endodermal sinus of the rodent placenta, are also present. The Schiller-Duval body in our patient consisted of a cystic space, lined with a layer of flattened endothelium, into which projected a glomerulus-like tuft with a central vascular core.

In terms of management, there may be arguments for conservative surgery in younger women if tumours are diagnosed early or unilateral. However, preservation of fertility is difficult in cases of bilateral involvement as reported by Kurman et al. It has also been well established that staging and tumour reductive surgery strongly affects the prognosis of the disease and so tumour reductive surgery is advisable when ascites is minimal. If the disease is extensive, pelvic clearance is indicated.

Conclusion

We report an atypical presentation of acute abdomen as a case of highly aggressive endodermal sinus tumour in a young girl. Dilemma about the optimal or suboptimal debulking was present during surgery but in view of patient’s poor socio economic status and aggressive nature of the tumour, maximum debulking was planned to have a better effect of post operative chemotherapy and longevity of the patient.

Editor’s comments

Yolk sac tumours are highly malignant occurring in children and young adults. Prognosis is stage-dependent and an early diagnosis can result in a drastic difference in the final outcome of the treatment of this highly aggressive disease. Further, the prognosis is improved with the use of new drugs like bleomycin, etoposide and cisplatin.

References