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CASE STUDY

A CASE OF YOLKSAC TUMOR IN ADOLESCENT GIRL

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ABSTRACT

Endodermal Sinus Tumour is one of the histological varieties of germ cell tumours. EST have also been referred to as yolk sac carcinoma because, they derived from primitive yolk sac, In the first 2 decades of life, almost 70% of ovarian tumours are of germ cell origin and one third of them are malignant. yolk sac tumour is the third most common malignant germ cell tumour of the ovary, comprising 10 to 15% overall and reaching a higher proportion among children. The EST (Yolk sac tumour) primarily affects the adolescent girls of age group 16 to 18 years. The gross appearance of an EST is soft greyish-brown which is usually well encapsulated and solid. Areas of necrosis and haemorrhage and small cystic spacesare often seen. A rate case of endodermal sinus tumour (Yolk sac tumour) of adolescence is presented. A 16 year old postmenar girl presented with lump abdomen and 3 months amenorrhea. Serum alphafeto protein was elevated. A fertility preserving surgery was done followed by 3 months amenorrhea. Serum. Alphafeto protein level was reduced that refletcs the response to chemotherephy.

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INTRODUCTION

The endodermal sinus tumor also called as yolk sac tumor, is very rare tumor that primarily affects the adolescent girls of age group between 16 to 18 yrs. However it ranks the third among the germcell tumor. It is almost always unilateral tumor and also one of the most rapidly growing tumor. As bilaterality is not seen in these cases, biopsy of other uterus avoided and the normal uterus and ovary preserved. In this article one case of yolk sac tumor successfully treated with fertility preserving surgery followed by postoperative chemotherapy has largely replaced the old treatment of total abdominal hysterectomy and bilateral salphingo oopherectomy with radiotherapy in these young girls.

Case report

A 16 yr old girl presented to us with lower abdominal pain, lump and 3 months of amenorrhoea. The mass was 20X20 CM in size. The abdominal pain and distention was rapidly increasing in size for the past 3 months. Urine pregnancy test was negative. Serum beta HCG was not detected. Her height was normal, secondary sexual characters-breast TANNER STAGE II, Sparse axillary hair and pubic hair. Regular cycle since menarche expect for this 3 month of amenorrhoea. O/E patient was conscious, afebrile, moderately built and nourished with vitals stable. Mass corresponding to 22 weeks uterine size. It was firm to solid irregular mass of 20X20 cm and ascites. Other abdominal organs are normal. Metastatic workup was done. X ray shows B/L pleural effusion without metastasis. karyotyping was not done as she has attained menarche & on regular menstrual cycle since 3 years. Hence the possibility of dysgenetic gonod excluded. Being a postmenarcheal girl, a search for tumor markers of germ cell malignancy carried out. There was increase in the serum level of AFP-450 ng/ml (normal level is <10 ng/ml). Serum lactate dehydrogenase and placental alkaline phosphatase were within normal limits. Under general anaesthesia, laparotomy was done. 2 litres of clear ascetic fluid drained. Left ovary was the site is the site of tumor and left salphingo oopherectomy done. Uterus and right side of the ovary is normal. No obvious areas of secondaries. Biopsy of the ovary was not taken as the other ovary was clinically absolutely healthy and also it was proved preoperatively as an endodermal sinus type in which it was 100% unilateral. Biopsy from omentum, peritoneum, pouch of douglas sent for HPE along tumor and ascitic fluid. She had uneventful postoperative recovery. The girl was followed up postoperatively for 6 months with medical oncologist and started on BEP (bleomycin, etoposide, carboplatin) regimen. Since such platinum based chemotherapy is the preferred regimen. The serum level of alpha beta protein level was measured to monitor response to chemotherapy.post chemotherapy (3 cycles), AFP levels of serum has decreased to negligible levels (3ng/ml). Ascitic fluid was negative for malignant cells.

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The histopathological findings were suggestive of yolk sac tumor. It was encapsulated 20X20 cm tumor. Tumor was predominantly multicystic with necrotic areas (due to rapidity of growth of this tumor). The fallopian tubes and other tissues are unremarkable. Microscopy showed schiller duval bodies which are the most characteristic microscopic picture of yolksac tumors. The tumor showed positivity of AFP on immune peroxidase stain. According to FIGO staging of GCT, This tumor belongs to stageIA Cancer.

DISCUSSION

Germ cell tumors of the ovary are rare tumors but incidence is 80% in adolescent girls <20 years.EST tumor otherwise known as yolk sac tumor rank 3rd amongst all germ cell tumors. Karyotyping is essential in premenarcheal girls as this tumor has preponderance to develop in dysgenetic gonods. karyotyping was not done in our patients because she was a post menarcheal girl with regular menses which rules out genetic gonods. But serum LDH and placental alkaline phosphatase were within normal limits which were well known to be secreted by dysgerminoma. Majority of the girls present in stage I cancer (FIGO staging).the presence of ascites and the presence of pleural effusion explains transdiaphragmatic fluid flow literature showed that children after treatment will have normal menses and had successful pregnancies. In our patient postoperatively she had normal senses. EST tumors are unilateral in 100% cases. Hence biopsy of the opposite ovary in such young young patients is contraindicated. Microscopically the characteristic feature is schiller duval bodies. In mixed types the common occurrence is combination of EST Tumors with dysgerminoma. The tumor secretes alphafeto protein. AFP can be demonstrated in the tumor cells by immune peroxidase staining. There is some correlation between levels of AFP and the extent of disease but not always. The serum levels of this marker is very useful in monitoring the patients' response to treatment. In our patient, after 3 cycles of postoperative chemotherapy, AFP dropped to negligible levels. All patients with EST are treated with either adjuvant or therapeutic chemotherapy. Though several regimens are there, the cisplatin based preferably BEP regimen or POMB-ACE regimen should be used as primary chemotherapy for EST. Our patient was given BEP regimen at our medical college oncology department. Drugs were administered in 3 cycles at 4 weekly intervals after RFT & LFT hematological monitoring.3 to 4 cycles are to be given every 4 weeks in stage 1 completely resected disease and 2 further cycles can be given after negative tumor marker status for patients with Macroscopic residual disease after chemotherapy.

Conclusion

EST is one of the histological varieties og germ cell tumors. EST have also been referred to as yolk sac carcinoma because, they are derived from primitive yolk sac. In the first 2 decades of life, almost 70% of ovarian tumors are of germ cell origin and one third of them are malignant. Yolk sac tumor is the third most common malignant tumor of the ovary, comprising 10 to 15% overall and reaching a higher proportion among children. The EST (yolk sac tumor) primarily affects the adolescent girls of age group between 16 to 18 years. These tumors are unilateral in 100% of cases. Karyotyping is essential in premenarcheal girls as this tumor is preponderance to develop in dysgenetic gonods. Some cases present with ascites (but without peritoneal dissemination) and the presence of pleural effusion explains the microscopic process of transdiaphragmatic fluid flow. Microscopically, the most characteristic feature is the endodermal sinus, or schiller duval bodies.the cystic space is lined with a layer of fattened or irregular endothelium into which projects a glomerulus like tuft with central vascular core. Conservative treatment is recommended for preserving fertility. The treatment of EST consists of surgical exploration, unilateral salphingo oopherectomy and frozen section for diagnosis. The absence of HCG and elevated levels of AFP which was done pre operatively pin pointed the diagnosis of pure form of yolksac tumor. In our case, during surgery a thorough exploration and unilateral salphingo oopherectomy was done. All patients with EST are treated with either adjuvant or therapeutic chemotherapy. Though several regimens are available, the cisplatin based BEP regimen or POMB-ACE regimen should be used as primary chemotherapy for EST. Fertility preserving surgery followed by postoperative chemotherapy has largely replaced the older treatment of total abdominal hysterectomy with bilateral salphingo oopherectomy with radiotherapy in these young girls.

The reasons for administrating post operative chemotherapy include

1. To improve the 2 years survival rate
2. Most of these tumors are chemo sensitive
3. With a combination of conservative surgery and adjuvant chemotherapy, fertility can be preserved.

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