JOURNAL OF CURRENT TRENDS IN CLINICAL MEDICINE & LABORATORY BIOCHEMISTRY

Case Report

A Case of Yolk Sac Tumour in Adolescent Girl

[1]N.K Mahalakshmi*, [2]C.Shanthi.

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 are 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 18years. 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 rare case of endodermal sinus tumour (yolk sac tumour) of adolescence is presented. A 16 year old postmenarcheal girl presented with lump abdomen and 3 months amenorrhea. Serum alphafeto protein was elevated. A fertility preserving surgery was done followed by 3 cycles of postoperative chemotherapy. The tumour marker Alphafeto protein level was reduced that reflects the response to chemotherapy.

KEY WORDS : Alphafeto protein, BEP regimen, Endodermal sinus tumour, Schiller-Duval bodies.

Introduction

The endodermal sinus tumour , also known as yolk sac tumour, is a very rare tumour that primarily affects the adolescent girls of age group 16-18 years. However, it ranks the third among the germ cell tumours. it is almost always a unilateral ovarian tumour.It is one of the most rapidly growing tumour.

[1]Associate professor, [2]Professor
Department of obstetrics and Gynaecology,
Govt. Rajaji Hospital and Madurai Medical College,
Madurai-625020.

***Corresponding Author**
Dr.N.K.Mahalakshmi
Associate Professor of Obstetrics and Gynaecology
Govt. Rajaji Hospital and Madurai Medical College,
Madurai, Tamilnadu - 625020,
Email id: drnkmaha@gmail.com
Mobile No.+91944579410

As bilaterality is not seen in these cases, biopsy of the other ovary is avoided and the normal ovary and uterus are preserved. In this article, one case of yolk sac tumour successfully treated with fertility preserving surgery is presented. Fertility preserving surgery followed by postoperative chemotherapy has largely replaced the older treatment of Total abdominal hysterectomy with bilateral salphingo oophorectomy with radiotherapy in these young girls.

Case Report

A 16 year old girl presented to us with lower abdomen pain, lump and 3 months amenorrhea. The mass was 20 x 20 cm in size and the tumour was about 28 week uterine size. The abdominal pain and distension 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 cycles since menarche except for this 3 months amenorrhea.

On examination patient was conscious, afebrile, moderately built and nourished with vitals stable. P/A: Mass corresponding to 28 weeks uterine size. It was firm to solid mass arising from the pelvis. The USG/CT showed a solid irregular mass of 20 x 20 cm and ascites. Other abdominal organs were normal. Metastatic work up was done. X-Ray chest shows bilateral pleural effusion without metastasis. Karyotyping was not done as she has attained menarche and on regular menstrual cycles since 3 years. Hence the possibility of dysgenetic gonads is excluded. Being a post menarcheal girl a search for tumour markers of germ cell malignancy was carried out. There was increase in the serum level of AFP which was 450ng/ml(normal level of AFP<10ng/ml). Serum Lactate dehydrogenase(LDH) and Placental alkaline phosphatase were within normal limits which rule out dysgerminoma component. The haematological values & the blood biochemical values were within normal range.

Under general anaesthesia, laparotomy was done. 2 litres of clear ascitic fluid drained. Left ovary was the site of tumour and left salphingo oophorectomy was carried out. Uterus and the ovary was normal. No obvious areas of secondaries. Biopsy of the other ovary was not taken as the other ovary was clinically absolutely healthy and also it was proved preoperatively as a endodermal sinus type in which it was 100% unilateral. Biopsy fromomentum, peritoneum, pouch of Douglas sent for HPE along with tumour and ascitic fluid. She had uneventful postoperative recovery.

The girl was followed up postoperatively with medical oncologist & started on BEP (Bleomycin, Etoposide, Carboplatin) regimen since such platinum based chemotherapy is the preferred regimen. The serum level of AFP was measured to monitor response to chemotherapy. After post chemotherapy (3 cycles), AFP levels of serum has decreased to negligible levels (3 ng/ml). Ascitic fluid was negative for malignant cells.

The histopathological findings were suggestive of yolk sac tumour. It was encapsulated 20 x 20 cm tumour(Figure No. 1). Tumour was predominantly multicystic with necrotic areas(due to the rapidity of growth of these tumours). The fallopian tubes and the other tissues were unremarkable. Microscopy showed schiller duval bodies which are the most characteristic microscopic feature of yolk sac tumours(Figure No.2).The tumour showed positivity for AFP on immune peroxidase stain(Figure No. 3). According to FIGO staging of GCT, this tumour belongs to stage IA cancer.

Discussion

Germ cell tumours of the ovary are rare tumours but incidence is 80% in adolescent girls < 20 years. EST tumours otherwise known as yolk sac tumours rank 3rd among germ cell cancers. Karyotyping is essential in premenarcheal girls as this tumour has preponderance to develop in dysgenetic gonads. Karyotyping was not done in our patient because she was a post menarcheal girl with regular menses which rules out dysgenetic gonads. But serum LDH and Placental alkaline phosphatase were within normal limits which are well known to be secreted by dysgerminoma. Majority of the girls present in stage Icancer (FIGO Staging).The presence of ascites (but without peritoneal dissemination) and the presence of pleural effusion explains the microscopic process of trans diaphragmatic fluid flow[4].

Literature showed that children after treatment have normal menses and had successful pregnancies. In our patient postoperatively she had normal menses.

EST tumours are unilateral in 100% cases. Hence biopsy of the opposite ovary in such young patients is contraindicated. Microscopically the characteristic feature is Schillerduval bodies. In mixed types the common occurrence is combination of EST tumours with dysgerminoma. The tumour secretes alphafeto protein. AFP can be demonstrated in the tumour cells by immune peroxidase staining. There is some correlation between levels of AFP and the extent of the disease but not always. The serum levels of this marker(AFP) is very useful in monitoring the patients' response to treatment. In our patient, after 3 cycles of postoperative chemotherapy, AFP levels 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 oncology department. Drugs were administered three cycles at 4 weekly intervals after RFT and LFT haematological 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 tumour marker status for patients with macroscopic residual disease after chemotherapy.

Conclusion

EST is one of the histological varieties of germ cell tumours. 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 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 18years. These tumours are unilateral in 100% of cases. Karyotyping is essential in premenarcheal girls as this tumour has preponderance to develop in dysgenetic gonads. 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 (Fig.No.2).The cystic space is lined with a layer of flattened 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 oophorectomy and a frozen section for diagnosis. In mixed germ cell tumours. The absence of HCG and the elevated levels of AFP which was done pre operatively pin pointed the diagnosis of a pure form of yolk sac tumour. In our case, during surgery a thorough exploration and unilateral salphingo oophorectomy was done.

All patients with EST are treated with either adjuvant or therapeutic chemotherapy. Though several regimens are available, the cisplatin based preferably BEP regimen or POMB-ACE regimen should be used as primary chemotherapy for EST. Fertility preserving surgery followed by post operative chemotherapy has largely replaced the older treatment of Total abdominal hysterectomy with bilateral salphingo oophorectomy with radiotherapy in these young girls.

The reasons for administering post operative chemotherapy include:

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

Acknowledgements:

We are greatly thankful to

Prof. Dr.Meenakumari, H.O.D, Dept of pathology, Govt. Rajaji Hospital and Madurai Medical College, Madurai, Tamilnadu, India.

Prof. Dr. P.N. Raja Sekaran, H.O.D, Dept of Medical Oncology, Govt. Rajaji Hospital and Madurai Medical College, Madurai, Tamilnadu, India

And the following Junior residents of Dept of O&G;

1. **Dr.Vimala**
2. **Dr.Suhas**
3. **Dr. R.S.Ramya**

Fig.No. 1. Laparotomy photograph of Yolk sac tumour of left ovary.

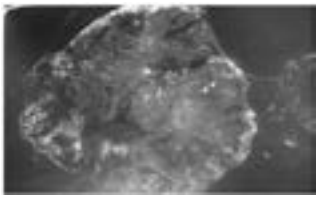


Fig.No. 2.Photograph showing Schiller duval Bodies characteristic of yolk sac tumour

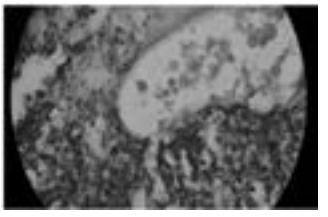
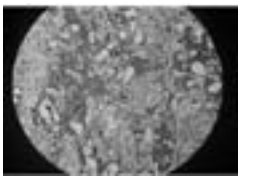


Fig .No.3.Photograph showing Immunopositivity for Alpha Fetoprotein



References

1. Sowmya MS, Siddhartha D. Outcome of germ cell tumours of ovary treated with adjuvant chemotherapy. Int J sci stud. 2014;2(8):136-138
2. Tangirj. Ovarian germ cell malignancies. Obstetgynecol .2003; 101:2
3. Bailey J, churchd. Management of germ cell tumours of the ovary. Rev Gynaecolpract. 2005; 5:201-601.
4. Ai Miyoshi, Takashi, Takeya Hara, Asuka Tanaka, Naoka Komura, Shinnosuke Komiya, serikskanao, Mayoko Mimura, Masaaki Nagamatsu and Takeshi Yokoi. Etiology of ascites and pleural effusion associated with ovarian tumours. Literature review and case reports of three ovarian tumours presenting with massive ascites, but without peritoneal dissemination. Case reports in OG – Hindawi corporation-Articles in press)