

Endodermal Sinus Tumor in an Adolescent Girl

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Abstract: Endodermal sinus tumor (EST), also known as yolk sac tumor (YST), is a member of the germ cell tumor group. It is the second most frequent malignant ovarian germ cell tumor just after dysgerminoma, with an incidence of approximately 1% of ovarian malignancies. It usually occurs around the second decade of life. In this case report, a 19yrs old unmarried girl presented with complaints of pain and mass abdomen for two months with elevated tumor markers and ultrasound features suggestive of right malignant ovarian tumor, staging laparotomy proceeded with as uterus and the left ovary was found to be normal, right ovariectomy done; Histopathology report came as endodermal sinus tumor and patient is now on chemotherapy. This case is reported for its rarity.

Keywords: yolk sac tumor, alpha fetoprotein, staging laparotomy, fertility sparing surgery, BEP regimen.

INTRODUCTION

Malignant ovarian germ cell tumors are rare but aggressive, accounting for approximately 1% to 2% of all ovarian malignancies [1]. The peak incidence is found in young women or adolescent girls. Yolk sac tumor (YST) is the second most common tumor in ovarian tumors of this group [2]. YST is a highly malignant tumor that metastasizes early and invades all intraabdominal structures. Especially following the addition of cisplatin to combination regimens, survival rates reached excellent values, even for patients with advanced stage tumors. Another important consideration in young females is the impact of chemotherapy on gonadal and reproductive function. Today for early stage disease the aim of chemotherapy is to be efficient and at the same time to minimize toxicity and retain reproductive function. Different studies reported the resume of normal menstrual function and in some patient's pregnancies after chemotherapy.

CASE REPORT

A 19 years old female was admitted on 21/9/2016 in gynaecology department, Government Rajaji Hospital, Madurai, with abdominal pain and mass abdomen for the past two months. On palpation of abdomen a firm irregular mass of size 18*18 cm with lower border not palpable and occupying suprapubic

and right iliac fossa with restricted mobility. Ultrasound examination revealed well defined large lobulated mixed echogenic mass with few cystic areas in pelvis measuring 18*17*10cm with moderate ascites, possibility of right ovarian tumor. Uterus normal, Other ovary could not be visualised separately.

Biochemical and laboratory investigations showed hemoglobin of 10g/dl, platelets of 2,71lakhs; urea-29mg/dl and creatinine-0.7mg/dl. Liver function were normal; Serum human gonadotropin hormone levels -1037 mIU/ml, Sr. LDH – 952 U/L and CA-125 levels-220 IU/L were elevated. There was a marked elevation of serum alpha-fetoprotein (AFP) level to 601 ng/ml. Subsequently, CT of the abdomen was performed. A right ovarian mass measuring 18*18cms with solid and cystic components seen, Planned for staging laparotomy.

Laparotomy done on 30/9/16, Abdomen was opened by subumbilical midline incision. 1000ml of straw coloured ascitic fluid was present and taken for analysis. Right ovarian solid tumor of size 20*18cm with fallopian tube stretched over the tumor and the omentum was adherent to it. Left ovary, left fallopian tube and uterus appeared healthy. No deposits in pouch of douglas. No peritoneal or intraabdominal deposits, Right ovariectomy with omental biopsy taken.



Fig-1: yolk sac tumor intraop picture

Specimens were sent for histopathological examination. The HPE report showed ovarian parenchyma with tumor, composed of pleomorphic large rounded cells with vesicular nucleus seen arranged in sheets, cords and glandular pattern. Tumor

cells seen forming schiller duval bodies and are supported by myxoid necrotic stroma suggesting endodermal sinus tumor. Tumor infiltrating the capsule, Omental tissue shows tumor deposits.

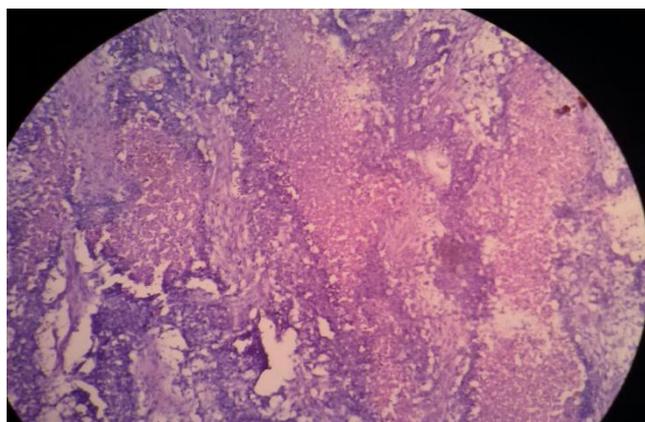


Fig-2: HPE picture showing Schiller Duval body

The postoperative course was normal. Chemotherapy with the BEP regimen (Bleomycin, Etoposide, Cisplatin) for 3 cycles was planned as per protocol. 2 cycles of BEP regimen given, After 2 cycles of chemotherapy serum AFP was 3890 ng/ml and serum beta HCG was 6.9. 2 mIU/ml more cycles of EP has been given. Patient has been asked to review after 2 weeks for repeat tumor markers evaluation.

DISCUSSION

Yolk sac tumor is almost always unilateral and large with a diameter that may vary from 5 to 50 cm (median 15 to 19 cm). The typical neoplasm manifests as a large complex pelvic mass and often characterized by extremely rapid growth and extensive intra-abdominal spreading with poor prognosis. Clinically abdominal pain is the principal symptom leading the discovery of the disease. Other symptoms are the presence of an abdominal or pelvic mass vaginal

bleeding, ascites or peritonitis secondary to torsion, infection or rupture of the ovarian tumor.

Yolk sac tumors can be cystic, with signs of hypervascularization and areas of haemorrhage. If YST appear with a solid portion, multiple small arterioles with lower RI are detectable by color Doppler ultrasound, haemorrhagic spots can be demonstrated by T1 weighted MRI, hypervascularity can be shown on contrast-enhanced T1 weighted scans. The malignant evolution of YST consists in locoregional extension involving uterus, pelvic peritoneum, rectum and bladder. Other authors described the involvement of the omentum, abdominal peritoneum and serosal surfaces of bowel in 30% of the cases. Retroperitoneal lymph nodes and liver parenchyma were also involved in advanced stages.

The standard management of malignant ovarian germ cell tumors is complete surgical excision

[3]. Because most of them are unilateral, it's possible executing a fertility sparing surgical treatment and should be followed by cycles of chemotherapy while monitoring the rate of decline of serum AFP. Currently, initial surgery followed by adjuvant chemotherapy including bleomycin, etoposide and cisplatin (BEP) is considered the standard for the treatment of Yolk Sac Tumor [4]. Patients with YST had a 3-year survival rate of 13%.

Compared with other regimens, BEP appears to be the best active first-line option for primary, metastatic, or recurrent disease. Progressive or recurrent ovarian YST after treatment with BEP chemotherapy is associated with a poor prognosis [5]. It should also be stressed that secondary cytoreductive surgery could play an important role when tumors are limited and resistant to chemotherapy. For follow-up after chemotherapy, the determination of initially elevated markers (AFP) should be repeated before each cycle of therapy, soon after the end of the treatment and during the 2 years after the end of chemotherapy. An annual pelvic ultrasound is necessary in the case of conservative treatment, to screen for a contralateral recurrence.

CONCLUSION

The risk of of infertility following treatment of germ cell tumor is always a concern, even if the majority of patients, especially in the early stage, will maintain their ovarian function and fertility. However when fertility sparing surgery followed by chemotherapy and regular follow up is done most patients will be cured and will be able to give birth.

REFERENCES

1. Berek JS. Practical gynecologic oncology.
2. Levitin A, Haller KD, Cohen HL, Zinn DL, O'Connor MT. Endodermal sinus tumor of the ovary: imaging evaluation. *AJR. American journal of roentgenology*. 1996 Sep; 167(3):791-3.
3. Kaur S, Bodal VK, Bal MS, Bhagat R, Gupta N. Malignant mixed germ cell ovarian tumor in pregnant female.
4. Rogers PC, Olson TA, Cullen JW, Billmire DF, Marina N, Rescorla F, Davis MM, London WB, Lauer SJ, Giller RH, Cushing B. Treatment of children and adolescents with stage II testicular and stages I and II ovarian malignant germ cell tumors: A Pediatric Intergroup Study—Pediatric Oncology Group 9048 and Children's Cancer Group 8891. *Journal of clinical oncology*. 2004 Sep 1; 22(17):3563-9.
5. Rouge TD, Pautier P, Duvillard P, Rey A, Morice P, Haie-Meder C, Kerbrat P, Culine S, Troalen F, Lhommé C. Survival and reproductive function of 52 women treated with surgery and bleomycin, etoposide, cisplatin (BEP) chemotherapy for ovarian yolk sac tumor. *Annals of oncology*. 2008 Aug 1; 19(8):1435-41.