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The Value of Frozen Section in Sclerosing Stromal Cell Tumor of the Ovary – Two Case Reports

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Introduction

When an ovarian tumor is encountered in a teenager, the chance of it being a germ cell tumor is about 60 to 70% and the incidence of malignancy is high especially in premenarchal girls. The role of conservative surgery need not be overemphasized in these patients and therefore a frozen section diagnosis is of utmost importance for ensuring the correct treatment.

Case No. 1

An 18 year old nulligravida married for six months complained of pain in the lower abdomen of two years duration. She attained menarche at 16 years of age and was suffering from polymenorrhea since then. She weighed 45 kgs and had no lymphadenopathy. Cardiovascular examination revealed a systolic ejection murmur confirmed by echocardiography and dignosed as pulmonary stenosis. Abdominal examination and other systemic examinations were normal. Speculum examination revealed the cervix and vagina to be healthy. On vaginal examination, the uterus was normal in size and a hard mass of 10x8 cm was palpable in the posterior and right lateral fornices.

Transabdominal USG showed the uterus to be normal and a mass arising from the right ovary. It was echogenic with few small cystic areas and measured 6x8 cm. Left ovary was cystic. Both kidneys and liver were normal. All other investigations including LFT and RFT were within normal limits. Laparotomy undertaken with a provisional diagnosis of germ cell tumor revealed straw-colored ascitic fluid of about 150 ml in the pelvis and a normal uterus. The right ovary was replaced by a solid grey white tumor of 6 cm x 8 cm. The left ovary showed normal ovarian tissue with a clear cyst of 3x4 cm. Both tubes were normal. There was enlargement of numerous mesenteric lymphnodes and also paraaortic nodes measuring 2x1x1 cm. All other intraabdominal viscera were normal. Intraoperatively the features were suggestive of dysgerminoma. Frozen section was done. The report showed a cellular tumor with areas of necrosis and collaginisation where categorization was not possible but most probably there was a malignant germ cell tumor.

Right salphingo-oopherectomy with left ovarian cystectomy and partial omentectomy with biopsy of the paraaortic and mesentric lymphnodes were performed. Histopathological examination after special stains showed features of sclerosing stromal cell tumor of right ovary and serous cyst of left ovary. Paraaortic and mesenteric lymphnodes showed reactive lymphadenitis and omentum showed only fibrofatty tissue.

The patient is being followed up for the past 3 years and is having regular menstrual flow .

Case No. 2

A 14 year old girl who attained menarche one year back presented with complaints of irregular cycles with increased duration of menstrual flow and passage of clots. Her general and systemic examinations were normal. Abdominal examination revealed a firm freely mobile mass of 8x6 cm in right iliac fossa. There was no other mass or organomegaly or free fluid. On vaginal and rectal examinations, the uterus was felt to be smaller than normal size and a firm nodular mass of 8x6 cm was felt through the right fornix.

Transabdominal USG showed uterus measuring 4x4x3.5 cm with endometrial thickness of 4.5 mm. A complex echogenic mass of 7x5x6 cm was seen superior and to the right of the uterus (Photograph 1). All other abdominal organs were normal. A provisional diagnosis of germ cell tumor of the right ovary was made.

At laparotomy, a 8 x 6 cm solid mass with few cystic areas and subcapsular focal hemorrhages was seen arising from right ovary. There was one complete torsion of the pedicle in clock-wise direction. The left ovary, both tubes and the uterus were normal. Paraaortic nodes were not palpable. Right salphingo-ovariotomy was performed and frozen section done which reported malignant germ

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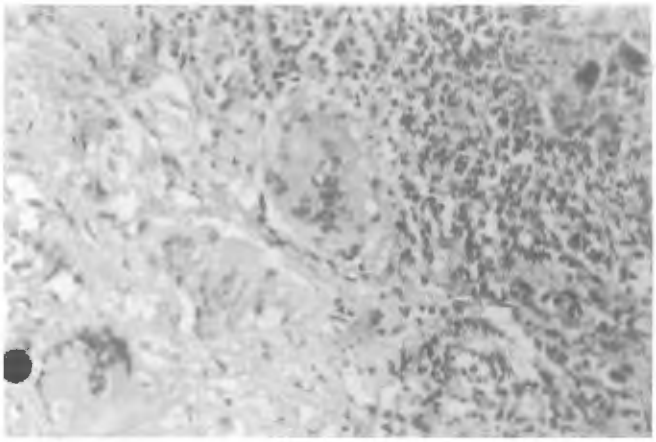
cell tumor with dysgerminoma like areas. Hence, left ovarian wedge biopsy and omental biopsy were taken. Pelvic and abdominal CT scan performed on the 10th postoperative day did not reveal any positive findings. Peritoneal fluid cytology showed reactive mesothelial cells and polymorphs. The final histopathological report was as follows :-

The cut section of the tumor shows fleshy nodular areas, the largest of which measures 3 cm and there are necrotic areas at the periphery. On microscopic examination, there are extensive areas of hemorrhagic necrosis and many vessels with fibrin thrombi. Focal areas show faint positive stain for fat and some areas show sclerosis with thick walled blood vessels (Photograph 2). There is no increase in mitotic activity of the tumor cells and the tumor is confined to the ovary. Features suggest sclerosing stormal cell tumor. The girl is being followed for one year and is having regular menstrual cycles.

Photograph 1 : Transabdominal sonogram showing complex solid mass with cystic areas superiorly towards left. A part of uterus is seen in the middle towards right below the enechoeic st cture-the bladder (Case No.2)



Photograph 2 : The histopathological features of sclerosing stromal cell tumour showing pseudolobular pattern and hyperchromatic nuclel (Case No.2).

Discussion

Sclerosing stomal cell tumor of ovary is considered to be a rare benign neoplasm occurring mostly in young women of reproductive age but cytogenetic studies reveal its low grade malignant features with monosomy of chromosome 16[1]. Its association with endometrial adenocarcinoma was reported though the tumor was inactive[2]. However, no association was found between the stromal tumors and ovarian cancer in a Norwegian study involving 41 cases of stromal tumors encountered during pregnancy[3].

The origin of the tumor from stromal cells was confirmed by electron microscopic studies[4] but Shaw and Dabbs[5] proposed that the tumor may originate from muscle specific action positive elements existing in the theca externa and the perifollicular and myoid stomal cells[5]. Clomiphene citrate therapy was implicated in the pathogenesis of these tumors occurring in cases of infertility[6].

The clinical symptoms of sclerosing tumor of the ovary depend on the degree of differentiation reached by the tumor cells. The cases with steroid secreting cells had polymenorrhea disappearing after its removal whereas the cases with fibroblastic morphology were asymptomatic[7,8]. Disappearance of menstrual abnormalities after tumor removal was observed in our cases also. Masculanising features were reported in two cases diagnosed during pregnancy with documented increase in androgen secretion[9,10].

Eventhough CT scan can differentiate malignant neoplasms from benign, sclerosing stromal cell tumor could not be differentiated because of its complex appearance with solid components and thick septas in the presence of free fluid[8]. Same applies to ultrasonographic appearance as in both of our cases the tumor appeared more echogenic with few small cystic areas. Ismail and Walker[9] observed calcifications and ascites on USG. At laparotomy it is difficult to come to a conclusion because of the features described in Case No.1. Ascites was reported in 14.2% of benign stromal tumors and 40% of the fibromas measuring

more than 10 cm^3. Presence of ascites in sclerosing stromal tumors was reported in three cases[8,9] and was also present in our Case No.1.

Frozen section is of utmost importance for proper management of these cases as most of them occur in the younger age group. It is better to adopt a conservative primary surgery in the young even in the presence of ascites though as in our cases the frozen section report is that of malignancy. The accuracy of frozen section in differentiating benign from malignant neoplasms of ovary was reported to be 93.8% by Obiakor et al[11]. They have recommended careful examination of the omentum and peritoneum and biopsies of suspicious lesions when a benign frozen section is reported so as to avoid a second surgical intervention for staging .

As sclerosing stromal cell tumors require special immunohistochemical studies (by vimentin, desmin and muscle specific acting stains)[10] for definitive diagnosis, at frozen section, a pseudolobular pattern with focal areas of sclerosis and presence of spindle and polygonal cells along with collagen[9,10] should be carefully looked in to.

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