A RARE CASE REPORT OF SCELOROSING STROMAL CELL OVARIAN TUMOR IN ADOLESCENT AGE GROUP

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ABSTRACT Introduction: Sclerosing stromal cell tumour (SST) is an extremely rare ovarian tumour, a subtype of ovarian stromal tumour of sex cord-stromal origin. So far, about 208 cases throughout the world have been recorded. These tumours occur predominantly in the second and third decades of life. Case report: We report a case of sclerosing cell stromal tumour of the ovary in a 14-year-old girl for its rarity.

KEYWORDS Sclerosing stromal tumour, adolescent, ovarian neoplasm

Introduction

Ovarian sex cord tumours are rare ovarian tumours accounting for approximately 8% of all primary ovarian tumours. Sclerosing stromal tumour cell tumours were first described by Chalvardjian and Scully in 1973 is usually unilateral and well-circumscribed, and its recurrence has not been reported. Histologically it is characterized by a network of thin-walled vessels, sclerosis, heterogeneity of the cellular areas, and ill-defined cellular pseudodolobules separated by a densely hyalinised or markedly edematous stroma. Histopathological and immunohistochemical (IHC) examinations confirm the diagnosis. Herein, the clinical findings and histopathological features of SST are described in a 14-year-old girl. This case is presented for its rarity.

Case Report

Date of admission : 15/11/20
Date of surgery : 18/12/20
Date of discharge : 28/12/20

A 14-year-old girl from Cuddalore was admitted for abdominal distension of 3 months duration, which was sudden in onset, rapid in progression and increased in size, which was not associated with pain. The patient attained menarche 2 years back and gave a history of frequent cycles for the past 3 months (once in 20 days)

No history of loss of appetite or loss of weight.

On examination: General condition: Good
Not anaemic
No clinical evidence of hyperandrogenism (acne, hirsutism)
Height : 154 cm Weight 42 kg
BMI : 17.72kg/m²

Abdominal examination: An abdominopelvic mass of size 24 weeks of gravid uterus was palpable which was variable in consistency, mobile, non tender. No ascites. Basic investigations done: Hb -10.1 gm/dl. O+ve blood group. Ultrasound Examination: A large abdominopelvic mass measuring 14.3*10*11cm with solid and cystic areas. No evidence of significant internal vascularity on colour doppler. Only peripheral vascularity was noted on colour doppler. Left ovary not seen separately. Right ovary seen separately measuring 2.6*1.4cm. Uterus measures 6*3*4.2cm. CECT Abdomen: Large oval solid mass lesion with internal clear cystic areas is seen occupying umbilical, left lumbar and left iliac fossa region of abdomen, mass measures (15.7*11.1*9cm). Right grade 1 hydroureteronephrosis and left grade 2 hydroureteronephrosis seen with compression of left ureter by dilated veins around the mass lesion. Features suggestive of left ovarian solid mass lesion with internal clear cystic areas.

Tumor markers:
LDH : 215U/L
CA 125-5.21 units/ml
ALPHA FETO PROTIEN : 0.68 IU/ml
BETA HCG : 0.20mIU/ml
INHIBIN B : 177 pg/ml
As Preoperative Diagnosis was made as a Granulosa cell tumour.

Planned for Laparotomy and proceeding with conservative fertility-sparing surgery as she is 14 years old.

Under GA, Laparotomy was done.

Intraoperative findings were: Free fluid of about 100ml of fluid present. Tumour arising from left ovary entirely replacing the ovary measuring 10*8cm in size with intact capsule mostly solid in consistency with little cystic spaces, no papillary excrescences, freely mobile, not adherent to adjacent structures. Left fallopian tube stretched over left ovarian mass. Right ovary and tubes normal, uterus normal in size for her age. Left side salphingo-oophorectomy was performed, and the specimen was sent for histopathological examination (HPE).

HPE report showed: The gross and histopathological features are suggestive of SCLEROSING TUMOR OF OVARY.

Ascitic fluid: Negative for malignancy

The postoperative period was uneventful. Suture removal is done. Follow-up was done. The patient remained asymptomatic and stable.

**Figure 1** Gross specimen showing left sided encapsulated ovary with fallopian tube stretched over it.

**Figure 2** Gross specimen of cut section of left ovary which is encapsulated showing solid and cystic spaces with yellowish-white discoloration.

**Figure 3** Microscopy showing cellular areas separated by edematous, collagenous, and hypocellular areas

**Figure 4** Microscopy showing well-encapsulated tumour with a pseudo-lobular pattern

**Figure 5** Microscopy showing spindle and polygonal cells with blood vessels

**Discussion**

Ovarian sex cord tumours are rare tumours that account for approximately 8% of all primary ovarian tumour. Sclerosing tumours account for 2% to 6% of ovarian tumours originating from the stroma of the ovary. Sclerosing Stromal-cell Tumor (SST) develop from a population of actin positive muscle elements arising from the theca externa, namely the perifollicular myoid stromal. Their size varies from 1.5 cm to 20 cm in diameter.

The diagnosis of SST is primarily surgical and histopathological. In contrast, SST is delineated by ill-defined hypercellu-
lar or hypocellular pseudo-lobules with prominent interlobular fibrosis. A definitive diagnosis of SST is made only by pathologic evaluation. The lobules contain heterogeneous cells such as collagen-producing spindle cells and separated by densely hyalinized or markedly oedematous hypocellular stroma. A prominent network of thin-walled vessels with hemangio pericytomatous pattern is noted with areas of cellular heterogeneity in the luteinized theca-like cells. Combination of clinical, morphological, hormonal, immunohistochemical, molecular, tumour markers and radiological studies are required to differentiate and exclude other tumours such as fibroma/fibrosarcoma and lipoid cells.

As a rare ovarian neoplasm, it is impossible to predict the presence of this tumour preoperatively. Thus, it is often misdiagnosed as a malignant tumour that manifests with androgenic features in some cases. SST remains a diagnosis of exclusion and points to suspicion in young patients with an ovarian mass. Post-laparotomy workup employing various modalities has improved the diagnosis of SSTS. Conservative fertility-sparing surgery is the treatment of choice for adolescent girls.

Conclusion
SST is a rare ovarian tumour. The diagnosis is by exclusion. It should be suspected in a young girl who presents with a unilateral ovarian mass. Clinical symptoms and radiological investigations will not conclude a tumour to be sclerosing ovarian tumour. Diagnosis is by histopathological and immunohistochemistry. It is a benign tumor with good prognosis.

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Conflict of interest
There are no conflicts of interest to declare by any of the authors of this study.

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