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Research Article

SCLEROSING STROMAL TUMOUR OF OVARY– A CASE STUDY AND REVIEW OF LITERATURE

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ABSTRACT

Sclerosing stromal tumour of ovary is a distinct sex cord stromal tumour bearing a morphological resemblance to a variety of tumours. Commonly affecting young females in 2nd and 3rd decades, presenting as lower abdominal mass with pain and menstrual irregularities it takes a benign course with good prognosis. Some are hormone producing and symptomatic. Distinct histomorphology with IHC helps in confirming the diagnosis. One such case is reported here in a young female which raised the suspicion of malignancy during radiological and intraoperative examination.

INTRODUCTION

Sclerosing stromal tumour (SST) is an extremely rare benign sex cord stromal tumour of ovary affecting mainly young women comprising 1.5 to 6% of all sex cord stromal tumours of ovary[1]. In 1973 it was first described by Chalvardjian and Scully and till date fewer than 209 cases have been reported[2,3]. About 80% of cases present below 30 years of age[1,4,5]. Most common presentation is pelvic pain and abdominal mass with or without menstrual irregularities[6]. Majority are hormonally inactive. However, androgen and estrogen excess have been reported in this tumour[4,7].

Sclerosing stromal tumour, a solid cystic mass with rich vascularity is often misdiagnosed as malignancy on ultrasonography as well as intraoperatively[4,5]. So also variation in cell morphology gives a resemblance to other sex cord stromal tumours and even metastatic neoplasms. Distinct histomorphology with immunohistochemistry (IHC) confirms the diagnosis[4].

Case report

A 15 years old female presented with a mass in left pelvic region for last one year and pain abdomen for six months. She had no menstrual irregularities or any other abnormalities. On examination a globular firm mass was palpable in the left lower abdomen.

Her routine haematological and biochemical investigations were within normal limits. Ultrasonography revealed a solid left adnexal mass of size 7x6x5 cm with normal right ovary and uterus.

Fig 1 Gross photo showing inhomogeneous cut surface

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There was no collection of free fluid in the pouch of douglas, no lymphadenopathy or any other mass lesion. Other visceral organs were normal. Resection of the mass was done followed by histopathological examination.

Gross specimen measured 6x5x4 cm in size, a well encapsulated grey white tumour with smooth surface. Cut section appeared mostly solid, firm but heterogeneous in appearancewith focal pale areas (fig-1). Histomorphology revealed cellular and hypocellular areas imparting a pseudolobular appearance (fig-2).

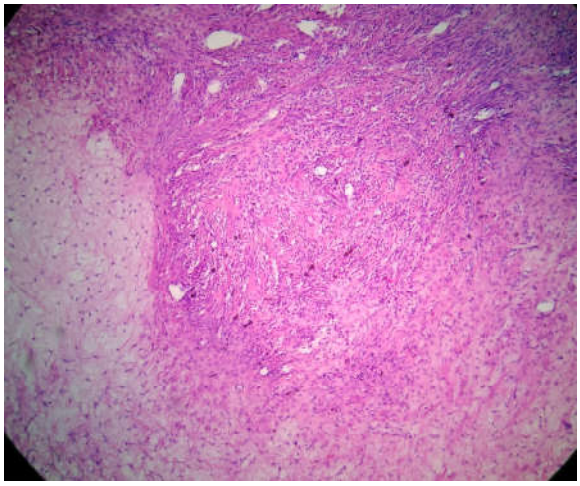

Fig 2 Cellular, hypocellular areas with pseudolobule (HP, 400X)

Dual population of cells were observed in cellular areas comprising spindle shaped fibroblastic cells and round, oval to polygonal cells with clear cytoplasm and round nuclei with fine chromatin. Eccentric nuclei in these clear cells gave a signet ring appearance (fig-3).

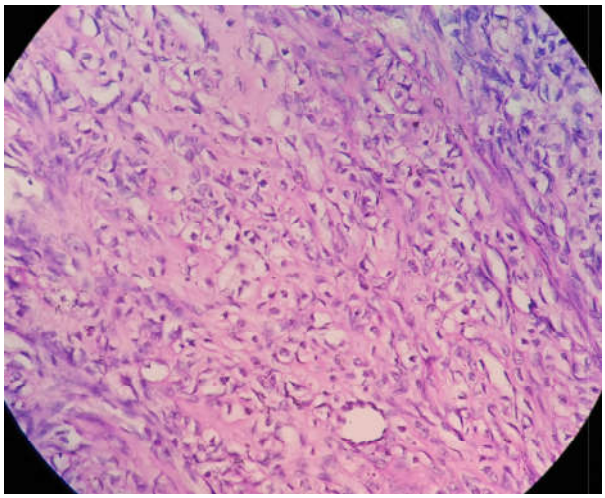

Fig 3 Spindle cells, clear cells & signet ring cells (HP, 400X)

Dense collagenised and oedematous stroma separating the pseudolobules was prominent. Many thin walled blood vessels with branching staghorn appearance was a key feature (fig-4). Normal ovarian stroma was apparent at places with no mitotic figures. Thus possibilities of fibroma, thecoma, vascular tumour, krukenberg tumour were thought of apart from massive ovarian oedema and sclerosing stromal tumour. Special staining and IHC workup were carried out. PAS and mucicarmine stains were both negative.

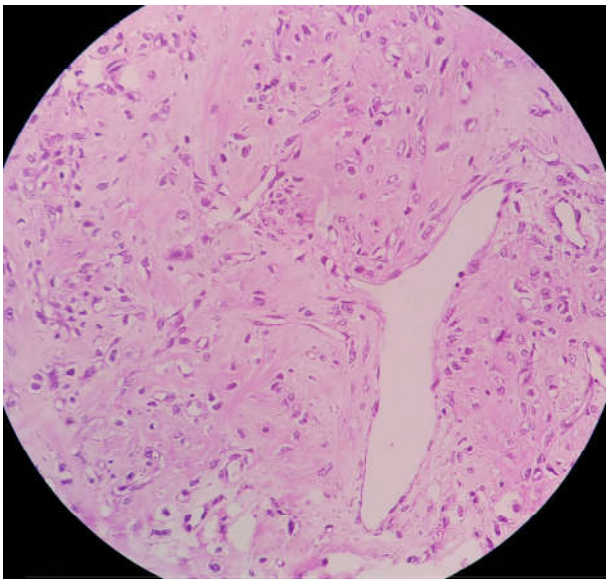

Fig 4 Thin walled BV with staghorn pattern (HP, 400X)

IHC: Tumour cells showed a strong positive reaction to vimentin (fig-5), SMA (fig-6) and focally for Inhibin (fig-7). Cells were negative to CK and desmin. Blood vessels wereintensely highlighted by CD 34 (fig-8).

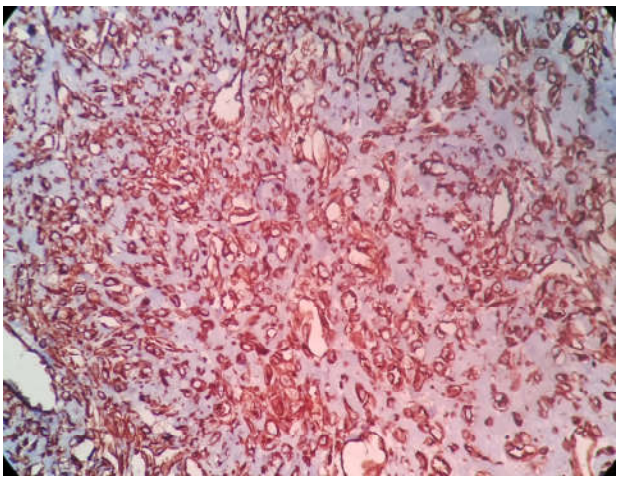

Fig 5 Vimentin positive (IHC)

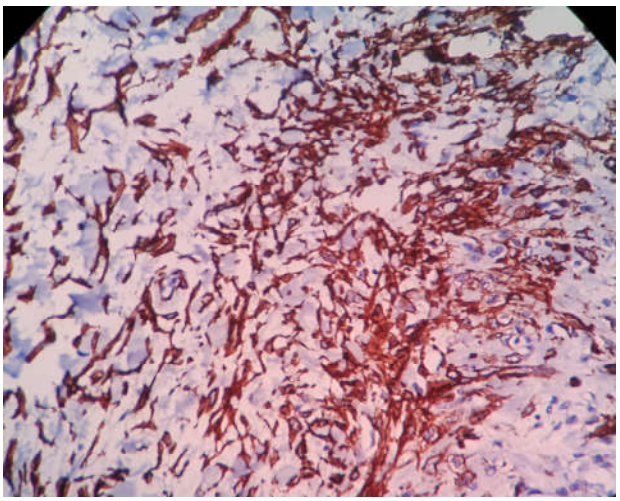

Fig 6 SMA positive (IHC)

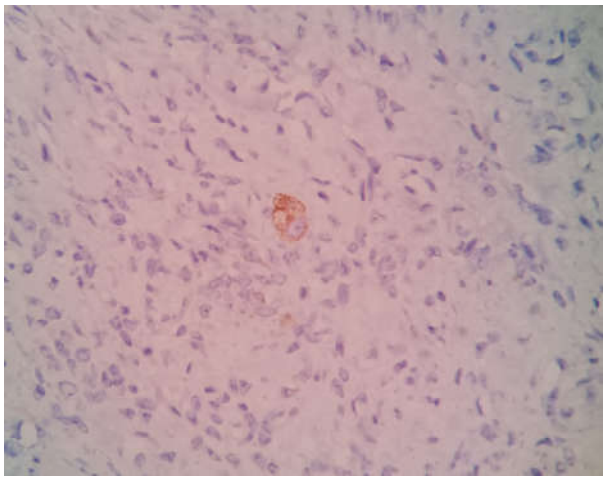

Fig 7 Inhibin focally positive (IHC)

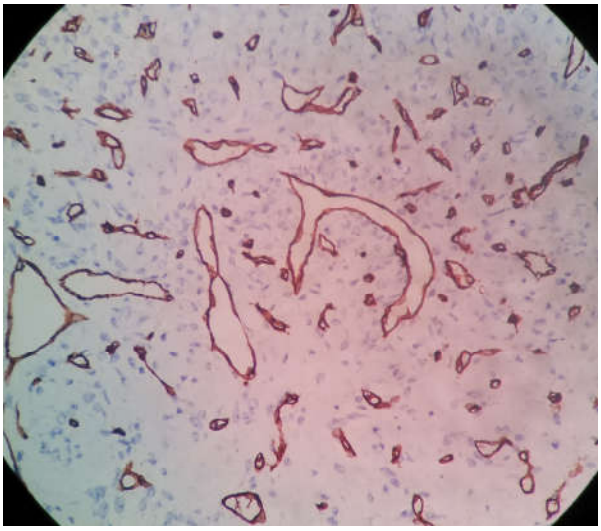

Fig 8 CD 34 positive (IHC)

Distinct histomorphology and IHC picture with clinical and radiological correlation established the diagnosis of sclerosing stromal tumour of ovary.

DISCUSSION

Ovarian tumours are very uncommon in adolescent girls and majority are of germ cell origin. SST of ovary is a distinct subtype of sex cord stromal tumour and is extremely rare[7]. Most common presentation is menstrual irregularities, pain abdomen and palpable abdomino-pelvic mass[4,5,6,7]. Some may present with hirsutism, infertility and an ovulation. Both androgen and estrogen production has been reported[4]. It is mostly unilateral, solid and inhomogeneous on cut surface.

Etiology of this tumour is not very well defined. However, ultrastructural features suggest origin from pluripotent immature myoid stromal cells of ovarian cortex[7,8].

Microscopically cellular areas showing pseudolobule formation separated by hypocellular oedematous stroma is very typical[4,6]. Two types of cells both spindle cells and polygonal clear cells give a resemblance to other sex cord stromal tumours like fibroma, thecoma and juvenile granulosa cell tumour[2,4,5]. Clear cells at times show a morphology of signet ring cells thus raising the suspicion of krukenberg tumour[1,5]. Prominent thin walled blood vessels give an impression of a vascular tumour, with a staghorn appearance almost resembling haemangiopericytoma[5,7]. Interlobular areas show fibrosis, collagen deposition and oedematous stroma[4]. Oedema in SST is focal may resemble massive ovarian oedema where it is extensive[3].

Immunohistochemical analysis in SST reveals positive staining for inhibin, calretinin, SMA, vimentin, VEGF, ER and PR[4,5,7,9]. Epithelial markers CK and EMA are negative[4,5]. CD 34 highlights endothelial linings of proliferated vascular channels thereby differentiating SST from fibroma, thecoma and other sex cord stromal tumours[4]. In our case vimentin, SMA , CD 34 were intensely positive and inhibin was focally positive. CK and desmin were negative. Studies reported IHC can show variable results (inhibin +/-, Calretinin +/-, desmin +/-)[4,10]. However, characteristic gross and histomorphology (solid inhomogeneous cut surface, pseudolobule formation, oedematous stroma, thin walled pericytoma like blood vessels)alongwith positivity for vimentin, SMA and CD 34 favour the diagnosis of sclerosing stromal tumour (SST) of ovary.

There is an entity called signet ring stromal tumour of ovary. This is a distinct variant of sclerosing stromal tumour which is characterised by predominantly signet ring cells, absence of pseudolobule, no vascular proliferation and PAS, oil red O both negative. Signet ring appearance may be due to protrusion of extracellular oedema into the stromal cells like an inclusion or swollen mitochondria.

Cytogenetic studies using FISH technique show trisomy of chromosome 12 in about 13-21 % of cases, while trisomy 16 was reported in one study[6,11].

CONCLUSION

In young females presenting with unilateral solid cystic complex ovarian mass possibility of SST should be considered even if it is rare. It needs differentiation from a variety of mimickers both benign and malignant, like fibroma, thecoma, juvenile granulosa cell tumour, metastatic carcinoma, krukenberg tumour, massive ovarian oedema and vascular tumours. Characteristic histopathology with IHC establishes the diagnosis. It takes a benign course, with less aggressive surgery which is the treatment of choice, carries a good prognosis.

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