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Case Report

Sclerosing stromal cell tumor of ovary

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Introduction

Sclerosing Stromal-Cell Tumor (SST) is a rare, benign sex-cord stromal ovarian neoplasm. Most patients present with menstrual irregularities, abdominal or pelvic mass and associated discomfort or pain. Some cases also present with hormonal activity, anovulation, infertility, virilization, ascites and Meig's syndrome.^{1–3} First described in 1973, SSTs constitute 6% of stroma-derived tumors and more than 80% are observed in young women in 2nd and 3rd decades of life.^{4,5} SSTs are hormonally inactive, although some cases related to pregnancy and androgenic symptoms have been described.⁵ This case report describes one such case, where histopathology proved essential in clinching the diagnosis.

Case report

A 28-year old female patient, primipara presented with complaints of menstrual irregularity, pelvic pain and low backache for 4 months. There was no history of dysmenorrhoea, urinary or bladder symptoms. On clinical examination there was no abdominal mass, hirsutism or palpable lymph nodes. Vaginal examination revealed a minimal pericervical ectropion, anteverted uterus and a 6 cm × 6 cm firm, non-tender mass with cephalo-caudal mobility palpable through Pouch of Douglas with no forniceal tenderness. Ultrasonography revealed a 6.5 cm × 5.5 cm × 5 cm iso hyperechoic lesion anterior to uterus. A differential diagnosis of endometrioma and broad ligament myoma was given. A contrast-enhanced computed tomography scan of abdomen and pelvis revealed a homogeneously enhancing solid mass of 7 cm × 6.5 cm × 6 cm with areas of necrosis involving right adnexa (Fig. 1). All hematological, biochemical, hormonal and serological markers including tumor markers such as CA 125 (13.7 U/ml), alpha fetoprotein (1.6 ng/ml), carcino-embryonic antigen (1.10 ng/ml), beta HCG (2.0 mIU/ml), CA 19.9 (16.0 U/ml), LDH (293 IU/ml) and inhibin (0.22 ng/ml) were normal.

Laparotomy revealed no free fluid, peritoneal deposits or any lymph node involvement. Left tube and ovary were normal. A 7 cm × 7 cm × 7 cm solid encapsulated mass was seen arising from right ovary without any extension into fallopian tubes or uterus. Suspecting benign ovarian mass, right oophorectomy was done. The right ovary appeared gray with bossellated appearance. Cut section revealed a homo-

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Fig. 1 – Contrast-enhanced computed tomography scan of abdomen and pelvis reveals a homogeneously enhancing solid mass of 7 cm × 6.5 cm × 6 cm with areas of necrosis involving right adnexa.

genously solid mass with yellowish discoloration (Fig. 2). Histopathology with hematoxylin and eosin showed pseudo lobular pattern of growth with cellular and hypo cellular areas of oedematous fibro-collagenous tissue. There was increased vascularity and thin walled blood vessels with stag horn/hemangio pericytomatous appearance, tumor cells comprised of disorganized array of fibroblasts, rounded and vacuolated cells with foci of edema and hyalinization. No mitotic figures, necrosis or atypia was seen. Peritoneal wash cytology was negative for malignant cells. Residual ovarian tissue with primordial follicles and follicle cysts were seen at periphery. Tumor was negative for periodic acid schiff and mucicarmine stains. Immunohistochemical staining of tissue blocks revealed vimentin positive, and desmin and inhibin negative status (Fig. 3) establishing the diagnosis of SST. Post-operative recovery was uneventful and the patient resumed menses in the next month. Two years follow up revealed normal menstrual cycles with no features of recurrence (Fig. 4).

Discussion

Sclerosing Stromal-cell Tumor (SST) develop from a population of actin positive muscle specific elements arising from the theca externa, namely the perifollicular myoid stromal cells.^{1,5} Their size varies from 1.5 cm to 20 cm in diameter.⁶ The

diagnosis of SST is primarily surgical and histopathological where in SST is delineated by ill-defined hypercellular or hypocellular pseudo-lobules with prominent interlobular fibrosis. Definitive diagnosis of SST is made only by pathologic evaluation however intraoperatively a frozen section analysis showing background of pseudo lobular pattern, heterogeneity of cellular areas and densely hyalinized or marked edematous stroma indicates a benign pathology like SST. The lobules contain heterogenous 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, tumor marker and radiological studies are required to differentiate and exclude mimicking tumors such as thecoma, fibroma/fibrosarcoma, lipoid and Krukenberg's tumor. Further, many of these tumors can present with Meig's syndrome. Fibroma and thecoma are rarely encountered in the first three decades. Thecoma show a peak incidence in the sixth decade and are estrogenic in nature with distinct lutein cells. Fibroma may have diffuse edema and are usually non functional.^{7,8}

Immunohistochemistry plays a diagnostic role with SSTs showing positivity for vimentin, CD 99 and smooth muscle actin; positive or negative for inhibin, desmin, calretinin and S-100 protein; and negative for pancytokeratin. Calcitonin, CD34, inhibin, and alpha glutathione S-transferase help to differentiate SST from thecomas, fibromas and other sex cord stromal tumors.⁵ In this case, the tumor was positive for vimentin and was negative for desmin and inhibin.

A mixed pattern of solid and cystic components on sonography may lead to the SSTs being suspected as a malignancy.⁹ Dynamic contrast Magnetic Resonance Imaging (MRI) findings such as early peripheral enhancement and prolonged enhancement in inner mass representing collagenous hypocellular area are more specific in distinguishing this benign neoplasm from other malignant ovarian tumors.^{1,5,10} In turn, it may avoid excessive surgical intervention if diagnosed timely in preoperative period. Increased peripheral and slight central vascularization with intercytic spaces on color doppler may strongly hint toward the diagnosis of SST. On contrast enhanced computed tomography SST usually

Fig. 2 – Gross specimen of right ovary measuring 7 cm × 7 cm × 7 cm with gray-white areas and bossellated appearance. Cut section revealed a homogeneously solid mass with yellowish discoloration.

Fig. 3 – (a) H&E image showing typical HPC pattern of vessels. (b) H&E pseudo-lobular pattern of growth with cellular and hypocellular areas of oedematous fibro-collagenous tissue.

Fig. 4 – Immunohistochemical staining of tumor with (a) Vimentin, (b) Desmin, (c) Inhibin.

shows a well-defined round mass with strong peripheral enhancement and centripetal progression, although it was absent in this case.

Cytogenetic analysis in 13–21% of all examined cases revealed Trisomy of Chromosome 21 using FISH.⁷ One case has been reported showing Monosomy of Chromosome 16.⁹

As a rare ovarian neoplasm, it is not always possible to predict the presence of this tumor preoperatively and thus it is often misdiagnosed as an aggressive neoplasm. SST remains a diagnosis of exclusion and thereby warrants a high index of suspicion in young patients with ovarian mass. A detailed post-laparotomy work up employing various modalities has improved diagnosis of SSTs. Surgical treatment for a benign

neoplasm remains non aggressive with ovariectomy, enucleation, oophorectomy and salpingo-oophorectomy optimized for age and fertility of the patient. Till date no cases of malignancy have been noted in the 200 cases of SSTs reported.¹

Conclusion

Sclerosing Stromal-cell Tumor (SST) demands a high index of clinical, radiological, histopathological and histochemical suspicion to be diagnosed amongst mimicking entities such as thecoma, fibroma/fibrosarcoma, lipoid and Krukenberg's

tumor. Further, with a correct diagnosis and follow up, a good prognosis and reproductive life can be assured to a hitherto young patient presenting with apprehension of malignancy.

Conflicts of interest

The authors have none to declare.

REFERENCES

1. Lee HC, Kim BR, Lee JS, et al. A case of ovarian sclerosing stromal tumor with massive ascites and elevated CA 125. *Korean J Obstet Gynecol*. 2009;52:120–124.
2. Jung NH, Kim T, Kim HJ, Lee KW, Lee NW, Lee ES. Ovarian sclerosing stromal tumor presenting as Meigs' syndrome with elevated CA-125. *J Obstet Gynaecol Res*. 2006;32(6):619–622.
3. Liou JH, Su TC, Hsu JC. Meigs' syndrome with elevated serum cancer antigen 125 levels in a case of ovarian sclerosing stromal tumor. *Taiwan J Obstet Gynecol*. 2011;50(2):196–200.
4. Chalvardjian A, Scully RE. Sclerosing stromal tumors of the ovary. *Cancer*. 1973;31:664–670.
5. Akbulut M, Colakoglu N, Soysal ME, Duzcan SE. Sclerosing stromal tumor of the ovary: report of a case and review of the literature. *Aegean Pathol J*. 2004;1:84–89.
6. Saitch A, Tsutsumi Y, Osamura Y, Watanabe Y. Sclerosing stromal tumor of the ovary. Immunohistochemical and electron-microscopic demonstration of smooth-muscle differentiation. *Arch Pathol Lab Med*. 1989;113:372–376.
7. Kawauchi S, Tsuji T, Kaku T, Kamura T, Nakano H, Tsuneyoshi M. Sclerosing stromal tumor of the ovary: a clinicopathologic, immunohistochemical, ultrastructural, and cytogenetic analysis with special reference to its vasculature. *Am J Surg Pathol*. 1998;22:83–92.
8. Damajanov I, Drobnjak P, Grizelj V, Longhino N. Sclerosing stromal tumor of the ovary: a hormonal and ultrastructural analysis. *Obstet Gynecol*. 1975;45:675–679.
9. Lopes JM, Seruca R, Hall AP, Branco P, Castedo SM. Cytogenetic study of a sclerosing stromal tumor of the ovary. *Cancer Genet Cytogenet*. 1990;49:103–106.
10. Yerli H, Agildere AM, Bilezikci B, Karadeli E. Sclerosing stromal tumor of the ovary with torsion: MRI features. *Acta Radiol*. 2003;44(6):612–615.