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Case Report

Ovarian Sclerosing Stromal Tumor, Incidental Discovery in a 26-Year-Old Female with a History of Amenorrhea

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1. ABSTRACT

1.1. Introduction: Sclerosing Stromal Tumor (SST) is a rare form of benign pure ovarian stromal tumour, derived from the ovarian sex cord stroma. It usually occurs unilaterally in young women and can be clinically, radiologically and morphologically eluting. Accurate diagnosis requires thorough pathological studies, both by morphology and immunohistochemistry. The main modality of treatment is surgical resection and clinical follow-up.

1.2. Case Report: We report a case of a right ovarian SST in a 26 years old lady, whom presented to the gynaecology clinic with a 45 days history of amenorrhea. Neither associated pain nor virilisation signs or symptoms at the time of presentation were found. Pelvic MRI revealed a 4.9 cm x 3.9 cm, mildly enlarged right ovary with a well-defined 4.1 cm x 3.4 cm x 4.0 cm area of abnormal signal intensity. On complimentary ultrasound, this ovarian lesion was isoechoic with internal arterial vascularity. Tumour markers (Ca 125, Ca 19-9, Ca 15-3) were all within the normal range. The patient underwent laparoscopic salpingo-oopherectomy and the detailed pathological studies were that of SST.

1.3. Conclusion: SSTs are generally considered to be rare. However, they should be thought of as a differential diagnosis when young patients present with menstrual irregularities, pelvic pain and when symptoms are refractory to conventional management.

2. Keywords: Sclerosing stromal tumor; Ovary; Stromal-sex cord; Presentation; Adult patients

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3. CASE PRESENTATION

A 26-year-old lady, Para 0/Abortion 2, presented in July 2021 with a 45 days history of amenorrhea. No reported associated pain at the time of presentation and the home pregnancy test was negative. She reached menarche at age of 12 years and her medical history revealed initial 3 days of severe dysmenorrhea every menstrual cycle, relieved by intravenous analgesics. This pain has been present since she got married two years back. Her period is characterized by heavy prolonged bleeding for almost ten days, however, her cycles are regular and about 30 days in length. She is a known case of chronic anaemia, but denied having any virilisation symptoms, such as more than usual body hair, acne, smaller breasts or a deeper voice. She had two previous uncomplicated spontaneous abortions at around six weeks of gestation and two uneventful pregnancies with normal spontaneous vaginal delivaries.

3.1. Laboratory workup investigations

The CBC was normal, except an elevated platelets count, reduced mean corpuscular volume (MCV) and Mean Corpuscular Hemoglobin (MCH). White blood cell count: 6.55 x 103/uL, red blood cell count: 5.46 x 106/uL, haemoglobin: 10.3 g/dL, platelet Count: 580 x 103/uL, MCV: 62.8 fL, MCH 18.9 pg. The Serum Beta HCG: 0.0 and Tumour markers (CA 125: 5.5, CA 15-3: 17.3, CA 19-9: 15.7) were all within the normal limits. Other hormonal profile included FSH = 5.91mlU/ml, LH =19.73mlU/ml, Prolactin =26.7ng/mL and TSH = 2.59ulU/ml.

3.2. Radiological work-up investigations

Pelvic ultrasound revealed a right ovarian complex isoechoic lesion with internal arterial vascularity. No cystic component, fat density or calcification is noted.

Pelvic MRI (Figure 1 and Figure 2) revealed a 4.9 cm x 3.9 cm, mildly enlarged right ovary with a well-defined 4.1 cm x 3.4 cm x 4.0 cm abnormal signal intensity area. This lesion was heterogenous, predominantly hyperintense on T2-weighted images, hypointense on T1-weighted images, and showed peripheral diffusion restriction and heterogenous post-contrast enhancement with central necrosis. The left ovary measured 2.7 hyperintense x 2.6 cm and appeared normal. Cervix and vagina were unremarkable. The uterus was normal in size with no myometrium mass. The endometrial thickness was normal, about 0.8cm. Mild free fluid identified in the pelvis, and few sub-centimetre bilateral pelvic and inguinal lymph nodes identified, up to 0.5 cm. However, no significant lymphadenopathy noted. The overall radiological appearances were suggestive of a neoplastic lesion, most likely a Germ Cell Tumour.

The patient underwent an elective laparoscopic right Salpingo-Oopherectomy and peritoneal fluid was collected for cytologic evaluation.

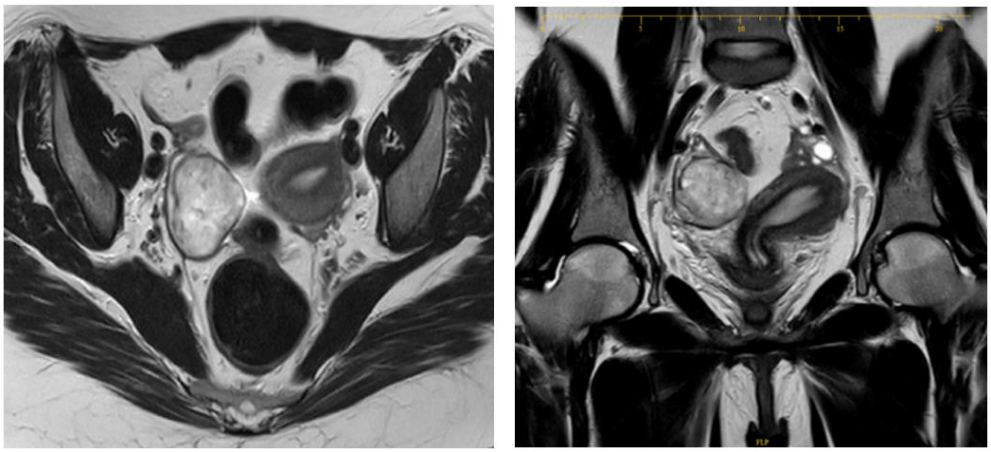

Figure 1 and 2: MRI Pelvis: Both Coronoal & Axial Views show an enlarged right heterogenous ovary with? Central necrosis.

3.3. Intraoperative findings

The right ovary was enlarged, with a 5 cm firm complex mass (Figure 3). Omental adhesions to the anterior abdominal wall, with further extension to the right pelvic side were found along with an abnormal thickening of the mesosalpinx was. The left adnexa and uterus were normal and no intraperitoneal, hepatic or diaphragmatic deposits found.

Omental adhesiolysis was performed and omental biopsy was taken. The right adnexa has been dissected, inserted into an endobag and removed through the umbilical port.

The right adnexa, omental biopsy and the collected peritoneal fluid were sent for pathological evaluation. The patient has been discharged after day one of her surgery, and the post-operative stage was uneventful.

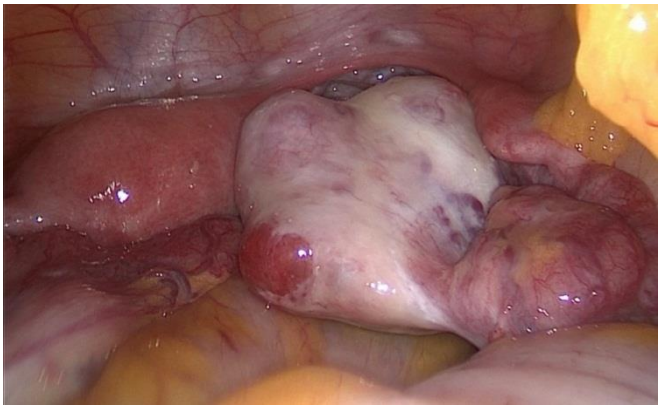

Figure 3: Intra-operative image revealing an enlarged, firm and complex ovarian mass.

3.4. Pathology findings

Macroscopic examination included a 55 mm x 50 mm x 37 mm greyish white right ovary with a cut surface medium-sized by a firm well-demarcated, nodular golden brown solid mass with central gelatinous zone. Foci of congestion and microcysts were noted but no necrosis or calcification was found. The ovarian cortical remnant included several subscapular thin-walled simple cysts. The right fallopian tube including its fimbrial end was normal and measured 75 mm x 18 mm. The Omental biopsy measured 22 mm x 20 mm x 10 mm, formed by a grossly normal piece of fatty tissue and weighed 1.68gram.

The peritoneal fluid was serous in nature and tannish brown in color.

Mrphologically, the right ovarian tumour was formed by a pseudo-lobular growth of mixed, predominantly clear cells with intervening areas of hypocellular, edematous and/or collagenous stroma. The neoplastic tumour cells were irregularly dispersed in moderately cellular, partly coalescent pseudolobules (Figure 4) formed by dual cell types; a dominant luteinized theca-lick cells with clear or vacuolized cytoplasm and bland roundish eccentrically located vesicular nuclei, with or without apparent nuclei (Figure 5). The less dominant other cell type was rather fusiform or spindly in shape with a fibroblast-like, appearance and mainly seen within the hypocellular areas or around dilated capillary channels (Figure 6). The stroma within the neoplastic nodules had a rich thin sinusoidal vascular capillary sized meshwork and include random areas of hypocellular oedema and/or hyalinization with occasional scattered plasma cells.

Ectatic small to medium sized, thin-walled blood vessels were also present and most of which had a characteristic hemangiopericytomatous/Staghorn pattern (Figure 7).

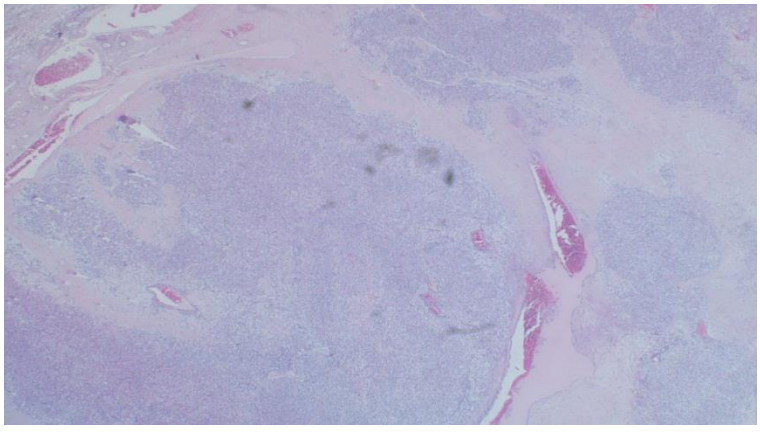

Figure 4: The ovarian tumour shows a nodular growth pattern with mixed cellular components, dilated blood channels and sclerosed stroma. [Photomicrograph
H&E stain, LPF].

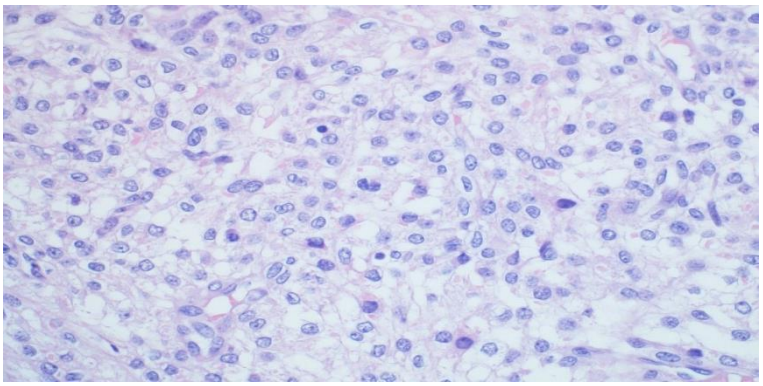

Figure 5: The ovarian tumour has a domienet large Theca-Like Cells with clear or vaculated cytoplasm. [Photomicrograph H&E stain, HPF].

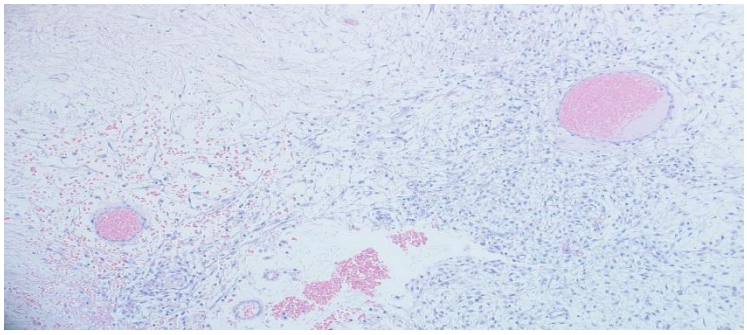

Figure 6: The ovarian tumour with a hypocellular edematous area formed by the less domienet spindle cell elements. [Photomicrograph H&E stain, HPF].

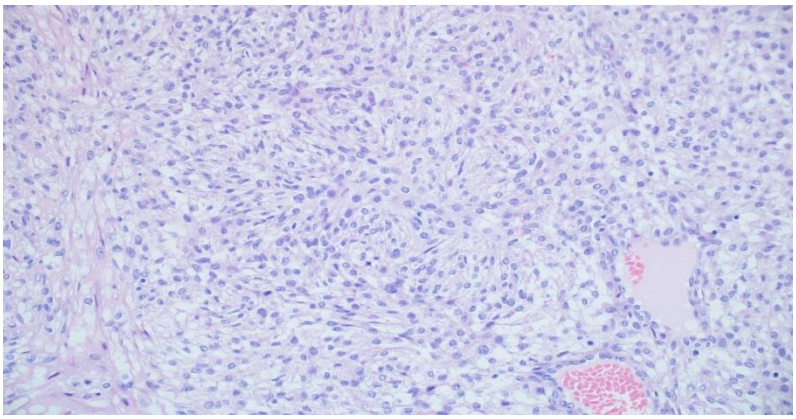

Figure 7: The ovarian tumour with both large clear Theca-like and spindle cell elements. [Photomicrograph H&E stain, HPF].

Only minor foci of ischemic necrosis and RBCs extravasation were noted. No sex cord component, significant cellular atypia or apparent mitosis is seen. The tumour was surrounded by a variably thickened fibrous tissue capsule, showed no ovarian capsular breach and it was completely excised.

Histochemical & immunohistochemistry results: Reticulin stain highlighted the well-delineated nodular tumour pattern with variable pericellular Reticulin framework.

Vimentin & CD10 stains were diffusely (3+) immunoreactivities within both neoplastic theca and fibroblast-like cellular components; CD99 was diffusely (3+) positive with a perinuclear dot-like immunoreactivity. SMA was also diffusely (3+) immunoreactive within the fibroblast-like spindle cells only-no theca cells positivity.

CD56 showed a multifocal (3+) immunoreactivity with the theca-like cells, likewise, variable multifocal immunoreactivities were noted with Calretinin (3+), Inhibin (3+), S-100 protein (3+), Melan-A (1-2+), ER (1+), PR (1+) and WT-1 (1+).

Tumour proliferative index (by Ki-67) : <1%

The tumour cells were negative for Desmin and CD34 (CD34 only highlighted the stromal thin capillary sinusoidal meshwork). All Germ cell tumour associated (OCT ¾, B-hCG, AFP & Alkaline phosphatase) markers and epithelial (CK AE1/AE3, CK7 & CK20) markers were diffusely negative.

The above detailed morphological and special studies were indeed are those of a benign primary ovarian tumour, and much supportive of an ovarian SST.

The hosting residual ovarian tissue was not peripherally compressed by the tumour and harboured many follicles at different stages of maturation, including several closely adjacent cystic forms and a single Corpus luteum (Figure 8).

The right fallopian tube and the omental biopsy were normal and peritoneal fluid was negative for tumour cells.

The patient has been on a regular follow up for a period of six months following her surgery. The operative wound has healed and the ultrasound scans were all unremarkable.

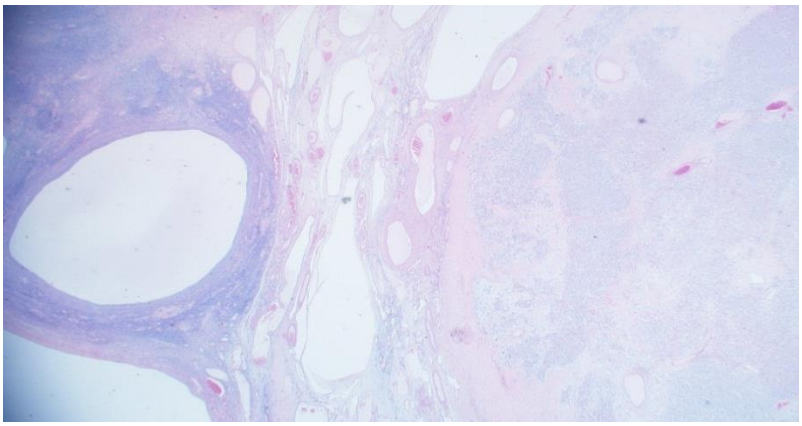

Figure 8: The ovarian tumour interface with hosting ovarian tissue; it is associated with an irregular peripheral fibrous tissue pseudocapsule. Also note the non-affected ovarian cortex with cystic follicles. [Photomicrograph H&E stain, LPF].

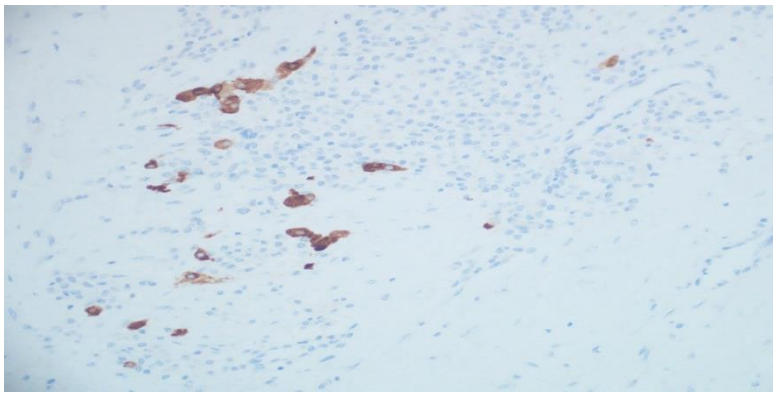

Figure 9: The ovarian tumour with Inhibin multifocal Theca-like cell Staining. [Photomicrograph, Inhibin IHC stain, MPF].

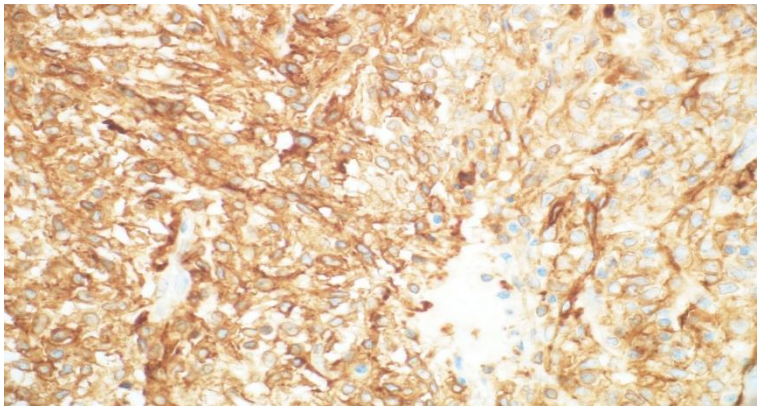

Figure 10: The ovarian tumour with diffuse CD-10 staining within both the Theca-like and spinle cell components. [Photomicrograph, Inhibin IHC stain, MPF].

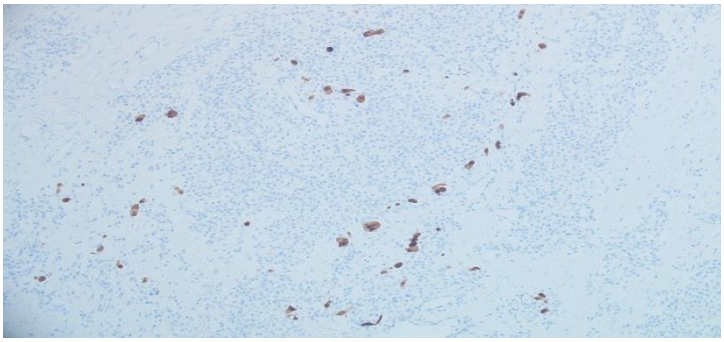

Figure 11: The ovarian tumour with Calretinin multifocal Theca-like cell Staining. [Photomicrograph, Inhibin IHC stain, LPF].

4. DISCUSSION

Ovarian neoplasms are rare in adolescents and mainly of a germ cell origin. Ovarain Sex cord-stromal tumors (SCSTs) and according to the WHO Classification are divided in to three groups; A) Pure stromal tumors (Fibroma, Cellular fibroma, Thecoma, Luteinized thecoma associated with sclerosing peritonitis, Fibrosarcoma, Sclerosing stromal tumor, Signet-ring stromal tumor, Microcystic stromal tumor, Leydig cell tumor & Steroid cell tumor and Steroid cell tumor, malignant), B) Pure sex cord tumors (Adult granulosa cell tumor, Juvenile granulosa cell tumor, Sertoli cell tumor & Sex cord tumor with annular tubules) and C) Mixed sex cord-stromal tumors (Sertoli-Leydig cell tumor, Well-differentiated, Moderately differentiated, with heterologous elements, Poorly differentiated with heterologous elements, Retiform with heterologous elements & Sex cord-stromal tumors, NOS).[1]

Some ovarian SCSTs produce steroid hormones, particularly androgens or estrogens, and thus may present with signs of virilization or estrogen excess.[2] Ovarian SCSTs are less common than tumors of epithelial cell and germ cell origin. Benign ovarian SCSTs account for <4 percent of benign ovarian neoplasms, and malignant ovarian SCSTs account for <8 percent of malignant ovarian neoplasms. In contrast to the more common epithelial ovarian malignant neoplasms, most patients with malignant SCSTs are diagnosed with early-stage disease and have no association with BRCA germline mutations, neither a genetic predisposition to breast cancer. An exception is granulosa cell tumors, which appear to be more common in women who have a family history of breast and/or ovarian cancer.[3]

4.1. Clinical presentations and diagnostic consederations

Patients with SCSTs generally present with a non-specific signs and symptoms, they somehow follow the same pattern of presentation of patients with epithelial or germ cell ovarian neoplasms. For example patients could present with abdominal or pelvic mass related symptoms or as an incidental finding of adnexal mass on examination or imaging. Though Some SCSTs can have characteristic clinical presentations such as estrogenic and androgenic effects with abnormal uterine bleeding, endometrial neoplasm, or, in a child, precocious puberty. The differential diagnosis in patients who present with both an adnexal mass and abnormal uterine bleeding includes SCSTs as well as an ovarian metastasis from a primary uterine cancer, an endometrial metastasis from a primary ovarian malignant neoplasm, and separate primary ovarian and endometrial carcinomas.

Likewise, signs of androgen excess (virilization) should be sought, including hirsutism, acne, alopecia (male pattern baldness), menstrual abnormalities (oligomenorrhea, amenorrhea), clitoromegaly, and deepening of the voice.[4] Ascites and/or pleural effusion, with or without other abdominal symptoms and elevated serum cancer antigen 125 (CA 125) levels have been reported in some cases. However, it should be bared in mind that, neither ascites/pleural effusion, nor an elevated CA 125, is necessarily indicative of an advanced epithelial ovarian carcinoma in a patient with a pelvic mass.[5]

Pseudo-Meigs syndrome (a clinical syndrome of pleural effusion, ascites, and an ovarian mass that is not a fibroma or fibroma-like mass/tumor) has been reported from a number of sources, such as leiomyomas, struma ovarii,

mucinous cystadenoma, teratoma, and malignancies that are metastatic to the ovary (particularly colorectal cancer). [6]

Luteinized thecoma associated with sclerosing peritonitis commonly have massive ascites, which cause acute abdominal distension and pain. Some patients may have bowel obstruction along with it.

Grossly, some tumours can adopt a solid appearance, while others can be solid and cystic, however, all are limited by the overlying capsule. Ovarian SCSTs arise from the dividing cells that would typically give rise to a specialized gonadal stroma surrounding the oocytes, namely, granulosa cells, theca cells, Sertoli cells, Leydig cells, and fibroblasts. The neoplastic cells show bimodal differentiation; fusiform fibroblast-like cells and theca-like cells with vacuolised cytoplasm. SCSTs display at least some areas with unequivocal gonadal stromal differentiation, such as granulosa, theca, Sertoli, or Leydig cell differentiation. They may display indifferent morphologic features, such as fibroblastic differentiation, and may contain areas of other types of stromal differentiation (e.g., cartilage or skeletal muscle) or areas of epithelial differentiation ("heterologous elements"). This may happen because the dividing cell population from which these neoplasms arise may still have capacity for various lines of differentiation. Tiltman and Haffeyee suggested that SSTs and thecoma are probably closely related entities as they share some morphological features and antigenic determinants such as smooth muscle actin and vimentin.[5,6-14] Immunohistochemical studies are mandatory for tumour triaging and ideally, Vimentin, Smooth Muscle Actin, Desmine, Progesterone Receptor, Calretinin, Inhibin are positive. CD-10, C-kit and Melan-A can be positive, while S-100, Pancytokeratin, Cytokeratin7, Estrogen Receptor are negative in all the cases.

Generally, SCSTs are low grade tumours with rare lymph node metastases and good prognosis, however, some neoplasms can be aggressive and have a lethal outcome.[7,15]

Ovarian SST is a rare neoplasm of sex-cord stromal origin, usually occure in young patients in the 2nd or 3rd decades, with average age of 18 to 28 years.[8] Clinically, it is a unilateral and asymptomatic, though can be associated with menstrual irregularities, abdomino-pelvic pain, or discovered during pregnancy.[9]

Most of the reported patients had no signs or symptoms of hormonal imbalance, as SST is typically hormonally inactive. However, and according to the literature, if the tumor is hormonally active, it usually shows androgenic activity with virilization, cliteromegaly and amenorrhea.[10] SSTs can also cause anovulation and infertility due to an increased estrogenic activity, typically, gets rectified following tumor excision.[11]

Tumour markers such as CA 125, CA 15-3, CA 19-9 and patient's hormonal profile including FSH, LH, Prolactin & TSH are always within normal limits.[12]

The management of SSTs is either via Laparoscopic oopherectomy, or laparotomical adnexal mass excisions. Post-operative patients follow up for a period of 1 to 5 years is almost always uneventful.[13]

5. CONCLUSION

The significance of these tumors is that it is necessary to consider there rare existence and to be able at least to distinguish the histomorphological appearances, especially during intra-operative frozen section examination, despite the alarming clinical and radiological features, in order to protect the other adnexa, particularly at early ages

(2nd and 3rd decades) presentation. Our findings support the conclusion that SCSTs are benign-character tumors that stem from ovarian stroma.

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