Sclerosing Stromal Tumor of Ovary: A Rare Case Report

Shweta[1], Kuladeepa Ananda Vaidya[2], & Sukesh[3]

[1]Post Graduate Student, Department of Pathology, Srinivas Institute of Medical Science and Research Centre, Mukka, Mangalore-575021

[2]Associate Professor, Department of Pathology, Srinivas Institute of Medical Science and Research Centre, Mukka, Mangalore-575021

[3]Professor and HOD, Department of Pathology, Srinivas Institute of Medical Science and Research Centre, Mukka, Mangalore-575021

E-mail: shweta992@gmail.com

Type of the Paper: Medical Case Study.
Type of Review: Peer Reviewed.
Indexed In: OpenAIRE.
DOI: http://dx.doi.org/10.5281/zenodo.836195.
Google Scholar Citation: IJHSP

How to Cite this Paper:
Shweta., Ananda., Kuladeepa Ananda, Sukesh. (2017). Sclerosing Stromal Tumor of Ovary: A Rare Case Report. *International Journal of Health Sciences and Pharmacy (IJHSP)*, 1(2), 11-15.
DOI: http://dx.doi.org/10.5281/zenodo.836195.

International Journal of Health Sciences and Pharmacy (IJHSP)
A Refereed International Journal

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Shweta[1], Kuladeepa Ananda Vaidya[2], & Sukesh[3]

[1]Post Graduate Student, Department of Pathology, Srinivas Institute of Medical Science and Research Centre, Mukka, Mangalore-575021

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[3]Professor and HOD, Department of Pathology, Srinivas Institute of Medical Science and Research Centre, Mukka, Mangalore-575021

E-mail: shweta992@gmail.com

ABSTRACT

Sclerosing stromal tumour (SST) is a rare benign ovarian tumor of the sex cord stromal type, occurring predominantly in the second and third decades of life. This tumour is characterized microscopically by having a pseudolobular pattern with cellular areas separated by hypocellular densely hyalinised, edematous stroma interspersed with few prominent blood vessels. Histopathological features which differentiate it from other stromal tumors. We are presenting this case because of its rarity and its simulation with various other ovarian tumours; here we have made an effort to highlight the histomorphological features and microscopic differential diagnoses of this rare tumour.

Keywords: Sclerosing stromal tumour, ovarian tumour, benign, sex cord stromal tumour.

1. INTRODUCTION :

Sclerosing stromal tumours (SST) are rare benignovarian neoplasms of the sex cord stromal category [1]. This tumour was first described by Chalvaridjian and Scully in1973 [1, 2].

2. CASE REPORT :

A 19 year old lady presented with a dull aching abdominal pain, andirregular menstruation since 4 months. On clinical examination, anabdomino-pelvic mass was felt in left adnexal area, and pelvic ultrasonography showed a well-defined solid and cystic mass measuring around 7x6x6cm over the left adnexa. There was no evidence of ascites.Under the impression of a benign ovarian tumour, the patient underwent laparotomy. On laparotomy left ovarian tumour seen with smooth surface, there were no other tumours in the peritoneal cavity or no evidence of ascites. The uterus, the right ovary and bilateral fallopian tubes were normal in appearance. Tumour was removed, with no intraoperative pathologic diagnosis, later fixed in 10% formalin and sent for histopathological examination. Gross examination showed a soft to firm ovarian mass measuring 7x7x6 cm with smooth and intact external surface [FIG 1a], cut section is showing mixed solid and cystic areas with gelatinous material [FIG 1b]. Histopathological examination showed tumor composed of cellular pseudolobules separated by hypocellular densely hyalinized and edematous stroma with few prominent thin walled vasculatures [FIG 2, 3]. Areas with two-cell population seen: rounded polyhedral cells with eosinophilic cytoplasm and spindle shaped fibroblasts. Tumor was focally showing areas of calcification and nests of vacuolated cells. No evidence of necrosis, atypia or malignant changes.Periodic Acid - Schiff and mucicarmine stains were negative. At the periphery of the mass, residual ovarian tissue with developing follicles are seen. Based on these features final diagnosis of sclerosing stromal tumor of ovary was made.

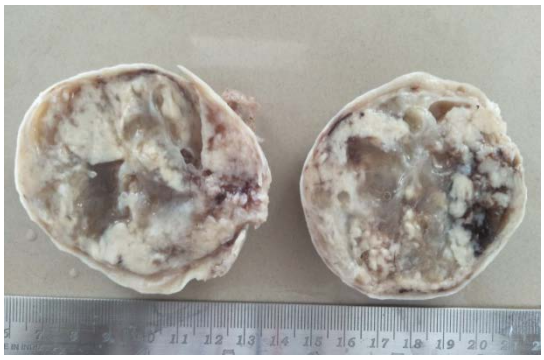
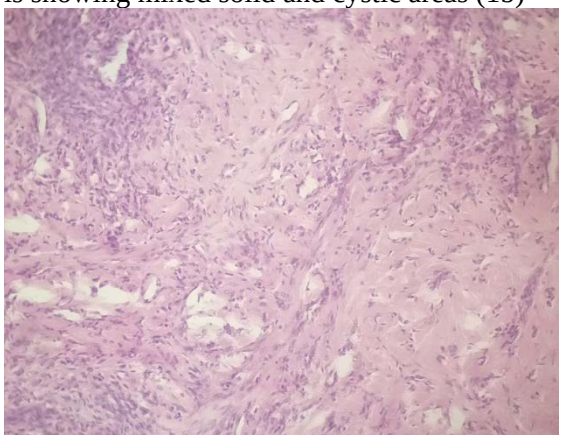


FIG 1a &1b: Gross examination- tumour is with smooth external surface(1a), cut section is showing mixed solid and cystic areas (1b)



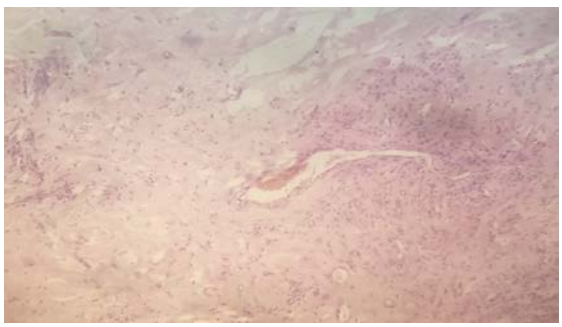


FIG. 2, 3: Histopathology showing tumor composed of cellular pseudolobules separated by hypocellular densely hyalinised, edematous stroma interspersed with few small thin walled blood vessels.

3. DISCUSSION :

Sex cord stromal tumours represent approximately 8% of ovarian neoplasms and SST comprises less than 5% of sex cord stromal tumors. [3,4]. More than 80% of SSTs occur in patients below the age of 30 years [5]. The most common presenting clinical symptoms include menstrual irregularity, pelvic pain and non-specific symptoms related to the ovarian mass and our patient complained of dull aching pain abdomen and menstrual irregularity [6,7]. Some patients have presented with anovulation or masculinization, which resolved spontaneously after removal of tumour.Masculinization or anovulation are occasionally associated with estrogen and rarely androgen secretion [7,8]. In our case there were no clinical features of virilization and hormone levels were normal.

Histologically, SST is characterized by cellular heterogeneity, prominent vasculature, and a pseudolobular appearance composed of both cellular and hypocellular areas and ill-defined cellular pseudolobules separated by a densely hyalinised or markedly edematous stroma. The descriptive term of "sclerosing stromal tumor" was proposed because the cellular areas of this tumor tend to undergo collagenous sclerosis [1, 9, 10].

The main differential diagnoses of SST of ovary include other sex cord-stromal tumors like fibroma and thecoma. Other sex cord-stromal tumors, including fibroma, may be differentiated from SST on the basis of histologic findings of lobulation, hemangiopericytoma-like pattern and positivity for actin in SSTs [11,12].

Calretinin, inhibin, CD34 andalpha glutathione S-transferase positivity (a-GST) was reported to be useful to differentiate STT from thecoma, fibroma and other sex cord stromal tumors [13,14,15]. CD 34 stains the endothelium of often dilated and branching vascular architecture, and clearly distinguishes SSTs from thecoma and fibromas, a-GST positivity within scattered cells appears to be useful in the distinction of SST from diffuse staining the comas and no staining fibromas [13,15].

Vascular tumors are also included in differential diagnosis due to prominent vascularity, but inhibin positivity suggests the diagnosis of SST. Sometimes “massive

ovarian edema'' may be confused with SST, but preserved ovarian tissue within the edematous stroma and absence of heterogeneity favours the diagnosis of massive ovarian edema [15, 16]. The vascular sclerotic and edematous stromal changes are constant features of sclerosing stromal tumors and relate to the local elaboration of some vascular permeability and growth factors (VPF/VEGF) [4, 9]. Sclerosing stromal tumor of ovary can show extensive dystrophic calcification in unusual cases, such a phenomenon in this rare tumor should be kept in mind when confronted by a paucicellular hyalinized ovarian tumour, it may represent a late calcifying stage of SST and needs to be recognized to avoid misdiagnosis and inappropriate management [12], in our case also tumour showedmultiple foci of calcification.

Due to the rarity of this tumour it is not always possible to predict the presence of this tumour preoperatively on the basis of clinical and sonographic findings. But this entity should be kept in mind in young patients with ovarian mass, as all of the sclerosing stromal tumours of the ovary reported in the literature were benign and were treated successfully by enucleation or unilateral ovariotomy [4].

4. CONCLUSION :

Sclerosing stromal tumour (SST) is a rare benign ovarian tumour of the sex cord stromal type. Due to the rarity of this tumour it is not always possible to predict the presence of this tumour preoperatively on the basis of clinical and sonographic findings. Thorough histopathological examination is required for confirmation and to differentiate this tumour from other tumours of ovary.

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