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SCLEROSING STROMAL TUMORS OF OVARY – AN UNUSUAL TUMOR IN A YOUNG FEMALE

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ABSTRACT

Sclerosing stromal tumors are rare benign ovarian neoplasms of the sex cord stromal category. They are frequently seen in second and third decades of life. Herein, we are reporting a unique case of sclerosing stromal tumor of ovary in a 24-year-old female which was clinically suspected as malignancy and diagnosis was confirmed on histopathology. Wherein frozen section played a vital role preventing untoward morbidity of patient due to extensive surgery. Other ovarian stromal tumors include fibroma and thecoma. They tend to occur in the fifth and sixth decades of life. So in such circumstances clinician should keep in mind the rare possibility of a sclerosing stromal tumor in a young female.

Keywords: Benign tumor, ovary, Sex-cord stromal tumor.

INTRODUCTION

Sclerosing stromal tumor (SST) is a rare benign ovarian neoplasm of the sex cord stromal category which occurs predominantly in the second and third decades of life. The tumor is characterized by cellular pseudolobules, prominent interlobular fibrosis, frequently marked vascularity and a dual cell population: collagen-producing spindle cells and lipid-containing round or ovoid cells [1]. The important differential diagnoses are other sex cord stromal tumors including fibroma, thecoma, and lipoid cell tumors [2].

CASE REPORT

A 24-year-old woman was referred for pelvic pain starting six months earlier with no menstrual complaints. She had noticed abdominal distention three months before presentation. On clinical examination, a hypogastric mass was palpable. Abdominal ultrasonography showed a heterogeneous predominantly solid pelvic mass with some cystic foci measuring 9×6 cm.

The patient was hospitalized with the diagnosis of malignant left ovarian tumor. All laboratory tests including tumor markers and serum hormonal assays were normal. Intraoperatively (Fig 1) mass was solid well encapsulated with glistening external surface. Intraoperative frozen section was performed and reported as benign spindle cell tumor of left ovary. In view of this diagnosis patient underwent left salpingo-oophorectomy. Right ovary showed corpus luteal cyst. The cyst was removed and right ovary was preserved.

Gross examination of left ovary showed a well-encapsulated 9×6×5 cm mass weighing 1100 g. The cut surface revealed predominantly solid, with focal cystic and edematous areas (Fig 2). No areas of haemorrhage or necrosis were observed. Capsule was intact. The right ovarian cyst was removed.

Microscopic examination of right ovarian mass showed marked sclerosis, prominent vascularity, and amongst it are seen foci of tumor cells with vacuolated cytoplasm and round to oval nuclei (Figures 3 – 4). Immunohistochemistry was

performed which showed focal positivity for inhibin and CD 34. So the final diagnosis of sclerosing stromal tumor of left ovary was done. On follow up patient doing well. As right ovary was preserved patient conceived and she is now 3 months pregnant.

DISCUSSION

SST of the ovary is a rare tumor derived from the sex cord stroma. This tumor was first described by Chalvaridjian and Scully (1973) and occurs most frequently in the second and third decades of life. The tumor is usually hormonally inactive although some cases with irregular menses and genital bleeding have been reported Peng et al found 114 cases reported until 2003 [3]. The tumor is characterized by cellular pseudolobules, prominent interlobular fibrosis, frequently marked vascularity and a dual cell population: Collagen producing spindle cells and lipidcontaining round or ovoid cells. The heterogeneity due to the variation in cellular size and shape are helpful features in the differential diagnosis of STT, and contrasts with the relative homogeneity of thecoma and fibromas [1, 3]. Also SSTs do not have hyalinized plaques, as do fibromas and thecomas. The finding of a thick rim of compressed residual ovarian tissue at the periphery of the mass suggests a slow growing benign tumor. On the other hand, thecomas and fibromas generally occur in the fifth or sixth decades of life when the ovaries are atrophic, so it is hard to identify residual ovarian tissue at the periphery of the tumor [4].

Our patient presented with pelvic pain with no menstrual irregularities while others reported menstrual irregularity, pelvic pain and non-specific symptoms related to the ovarian mass [3, 5], anovulatory cycles or masculinizing symptoms [6]. The differential diagnoses of SSTs are other sex cord stromal tumors including fibroma, thecoma, and lipid cell tumors. Most of the SSTs occur during second and third decades while fibroma and thecoma are rarely

encountered in the first three decades of life. Thecoma is typically an estrogenic tumor with peak incidence in the sixth decade and lutein cells are distinct. There clinical manifestations are like infertility and irregular menses because of hormone production. Fibroma is a non-functioning tumor, which may have diffuse edema. [5]. SSTs may have a potential for hormone production which is not always manifest or may be of a subclinical nature [7].

Other tumors included in differential diagnosis are vascular tumors due to prominent vascularity of SST, Massive ovarian edema and Krukenberg tumors. Inhibin positivity suggests the diagnosis of SST over vascular tumors. Absence of heterogeneity and preserved ovarian tissue within the edematous stroma favors massive ovarian edema [4]. Krukenberg tumors are the malignant tumors occur in the sixth and seventh decades, are mostly bilateral, and lack the pseudolobulated pattern of sclerosing stromal tumor on cut surfaces. Signet-ring cells of Krukenberg tumors may be confused with vacuolated cells of SST but signet ring cell contain mucin rather than lipid. Mitotic activity and nuclear atypia may be seen in krukenberg tumor.[1]

The etiology of SSTs is unknown. Based on the ultrastructural features, SSTs were thought to arise from pluripotent immature stromal cells of the ovarian. In the literature calretinin, inhibin, CD34 and alpha glutathione S-transferase positivity (a-GST) was reported to be useful to differentiate STT from thecoma, fibroma and other sex cord stromal tumors [7, 8]. Similarly our case showed focal positivity for inhibin and CD34.

It is difficult to distinguish SSTs consisting of solid and cystic areas from ovarian malignancies on the basis of radiological and macroscopic examination. Radiologically, especially on sonograms the appearance of SSTs may be suspected to be malignant ovarian tumors because they show a mixed pattern, with cystic and solid components [9]. Malignant ovarian

tumors usually occur in older women and often show high values of serum tumor markers.

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CONCLUSION

The present case suggests that it is necessary to rule out Sclerosing stromal tumor, a rare tumor in young patients and to confirm the diagnosis by expert intraoperative frozen section and careful histopathological examination in such cases. It will help to prevent untoward morbidity of patient due to extensive surgery and selecting the definitive treatment.

CONSENT

Written informed consent of the patient was taken

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Figure 1: Intraoperative photograph of left ovarian mass

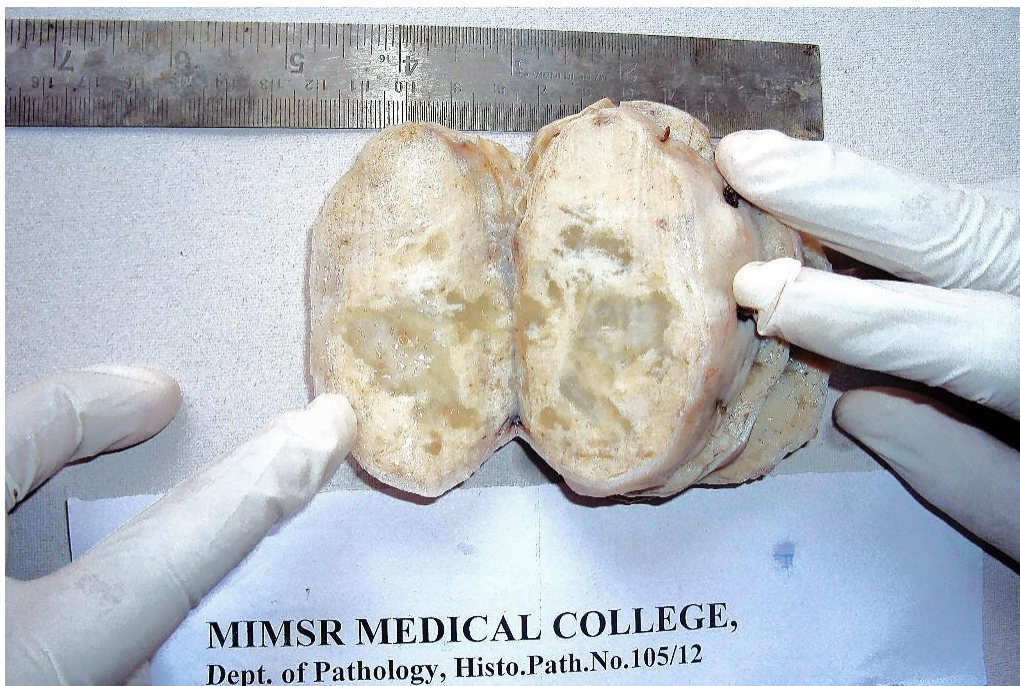


Figure 2: The cut surface of left ovarian mass with predominantly solid with focal cystic and mucoid areas

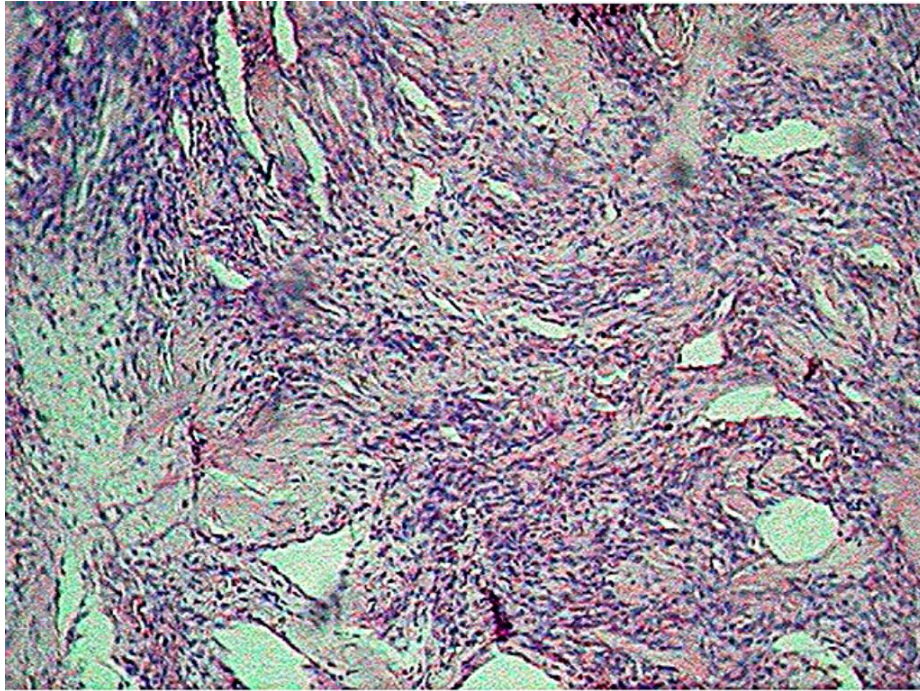


Figure 3: Microscopy showing large sclerotic areas and dilated thin-walled blood vessels in sclerosing stromal tumors (H&E, x400)

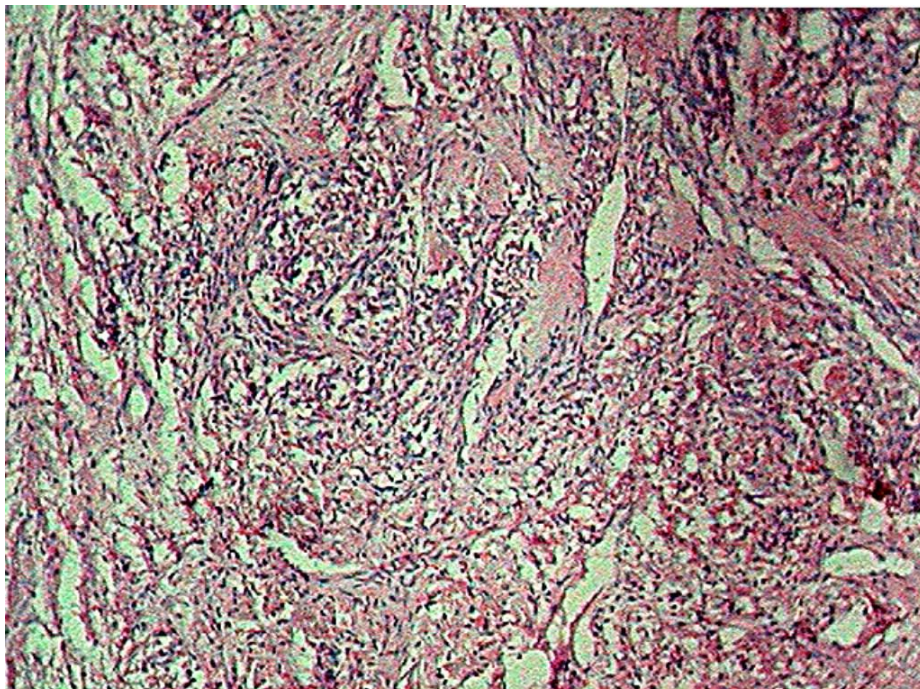


Figure 4: Prominent pseudo lobule formation of spindle cells separated by dense fibrous stroma (H&E, x400)