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Sclerosing Stromal Tumor of Ovary: A Rare Case Report with Review of Literature

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Abstract: ***Sclerosing stromal tumor of ovary is an extremely rare benign tumor of ovary of the sex cord stromal category, and most commonly occurs in young women. These tumors are found to be inactive hormonally and present with non-specific symptoms. As it often mimics a malignant neoplasm of ovary, pre-operative diagnosis of the tumor is challenging. Many young women undergo oophorectomy following which Sclerosing stromal tumor is diagnosed on histopathological examination. Hereby, we report one such rare case in a 17-year-old female who presented with 6 months of menorrhagia and pelvic pain. She underwent unilateral oophorectomy along with salpingectomy for right ovarian mass. Histopathological examination showed features of Sclerosing Stromal tumor of ovary. It is important for the pathologists to be aware of the morphological features of this rare benign entity which mimics malignant neoplasm clinically and radiologically, to avoid unnecessary extensive surgery and mental distress to the patients.***

Keywords: Sclerosing stromal tumor, Ovary, benign, sex cord stromal tumor, Rare tumor

1. Introduction

Sclerosing stromal tumors (SST) of the ovary have been described as a pathologically and clinically distinct entity of benign ovarian sex-cord stromal tumor category [1].

These tumors resemble malignant tumor of ovary because of its radiological and gross appearance.

It is a rare ovarian disease with prevalence of approximately 1.5 to 6 % of ovarian stromal tumors occurring predominantly in young women [1].

It is distinguished from other ovarian tumors on histopathology by cellular heterogeneity, prominent vascularization of blood vessels, and a predominant hypercellular and hypocellular areas separated by oedematous and collagenous area [2], [3].

All SST's reported in literature till date are benign with no evidence of recurrence following conservative surgery [3], [4]. We report a case of 17-year-old female who developed an SST of the right ovary.

2. Case Report

A 17-year-old unmarried girl reported to the gynaecology outpatient department with complaints of menorrhagia and pelvic pain for the last 6 months. There was no other significant personal or family history.

Ultrasonography revealed a 10×8 cm large, solid cystic lesion in the right pelvic area extending up to the midline suggestive of right ovarian mature cystic teratoma.

Contrast enhanced computed tomography (CECT) showed well defined solid heterogeneously enhancing lesion with mild free fluid in the pouch of Douglas. Findings were suggestive of neoplastic etiology. The left ovary was normal in shape and echotexture.

All routine, haematological, biochemical test were normal except for an elevated CA-125 level of 92 IU/ ml.

With the preoperative diagnosis suggestive of right ovarian tumor, the patient underwent right oophorectomy and salpingectomy.

Pre operatively, the left ovary and uterus were normal and there were no deposits on the bowel wall, omentum or other viscera.

Peritoneal wash was done and sent for fluid cytology, which showed only reactive mesothelial cells and no evidence of cancer.

Grossly, the right ovarian mass measured 10x8x5 cm with attached fallopian tube. The mass was well encapsulated, smooth, congested and bosselated at places, grey white. On cut section, the mass was solid with few cystic areas, yellow to grey white with focal mucinous areas. No necro-haemorrhagic areas were observed. Normal ovarian tissue was not seen.

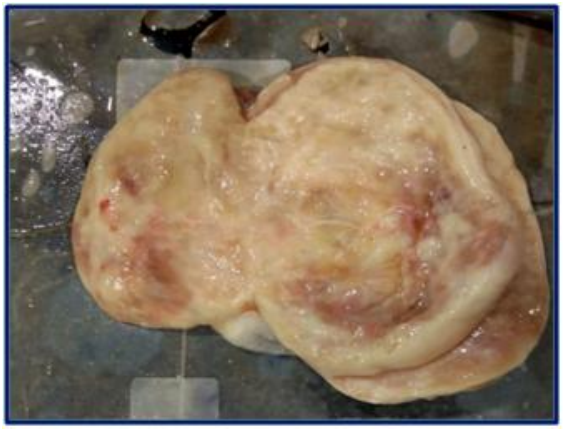

Figure 1.1: Gross image of the cut surface of the ovarian tumor showing solid area, grey-white with focal cystic and mucinous change

Microscopic examination showed ovarian tumor comprising of ill- defined pseudo lobules of hypocellular and hypercellular areas along with numerous hemangiopericytoma like vascular spaces. Dual cell population was seen comprising of cells with spindle nuclei, vesicular chromatin with moderate cytoplasm and other cell population showed lipidized cells having nucleus pushed towards the periphery.

Intervening stroma showed zonal edema and collagenous areas along with few foci of hyalinization.

Sections from attached fallopian tube were unremarkable.

The differential diagnosis considered were sex cord stromal tumours like thecoma, fibroma and lipoid cell tumours, massive oedema of the ovary. As the histopathological feature in the present case was without malignant features, Classical of SST, we arrived at the final diagnosis of Right Ovarian Sclerosing Stromal tumor.

3. Discussion

SST is a rare benign stromal ovarian neoplasm. It was first described by Chalvardjian and Scully in the year 1973 [3], [4].

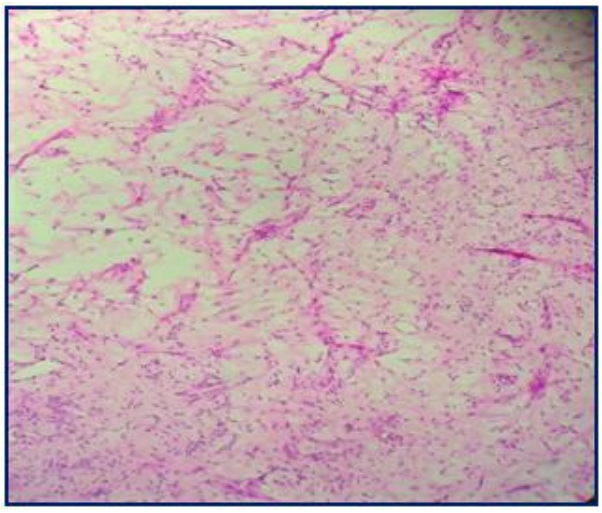

Figure 1.2: Microscopy of the ovarian tumor showing hypercellular and hypocellular areas with lipidized cells

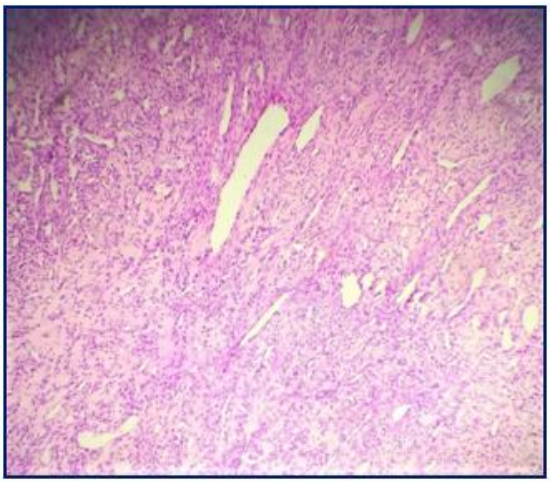

Figure 1.3: Microscopy of the ovarian tumor showing hemangiopericytoma like blood vessels in the stroma

The SST's are unilateral tumours commonly affecting females in their second and third decades [5], [6].Our patient was a 17-year-old female with a unilateral right-sided ovarian mass.

The clinical symptoms of SST are vague and non-specific. The patients usually present with pelvic or abdominal pain accompanied by tenderness in the lower abdomen, menstrual irregularities, dysmenorrhoea/hypermenorrhoea and/or abdominal mass. Most of the times SST's reported in literature are hormonally inactive [3], [5], [6]. In a few documented cases in literature hormonal activity has been reported. Lam and Geittmann[7] proved that sclerosing stromal tumors synthesized dehydroepiandrosterone. Damajanov et al. [8] have documented that elevated levels of both estrogenic and androgenic hormones were responsible for menstrual irregularity, amenorrhoea and infertility. Our patient presented with 6 months history of menorrhagia along with pelvic pain.

On imaging studies most commonly, they are solid or complex cystic adnexal mass with marked vascularity and they are often confused with other solid cystic tumors including teratoma and malignant ones [9].

On CT Scan our patient showed right ovarian mass lesion with heterogeneously enhancing solid component.

Reportedly, SST's can appear as small solid masses to large multi-cystic lesions. In our case, the lesion was well encapsulated, solid with few cystic areas having focal mucinous changes.[3] For arriving at the definitive diagnosis histopathological examination is needed [3], [5]Microscopically, these tumours have a pseudo-lobular appearance with alternating hypercellular and hypocellular areas with areas of hyalinization. The hypercellular areas comprise of spindle cells with dense collagenous tissue. In the hypocellular areas, there are clusters of lutein cells which are round to ovoid cells with perinuclear vacuolation. Also seen are areas of sclerosis with prominent blood vessels giving it a hemangiopericytoma like appearance. Similar features were seen in our case.[3], [5], [6].

Immunohistochemical staining is required only when the histopathological features are inconclusive or when unusual changes are present. All SST's typically show positive staining for inhibin, alpha-glutathione S-transferase (αGST). The endothelium of the branching blood vessels is positive for CD34[6].

Sex cord-stromal tumors like fibroma and thecoma occur in the fifth or sixth decades of life. They are differentiated from SST by the presence of cellular pseudolobules, prominent interlobular fibrosis, marked vascularity and a dual cell population – collagen producing spindle cells and lipid containing round to ovoid cells. This heterogeneity in the cell size and shape helps to differentiate SST from the relative homogenous tumors like thecomas and fibromas. Also, SSTs lack hyalinized plaques which are present in thecomas and fibromas.[10]

Massive edema of ovary may be confused with SST. The preservation of ovarian tissue within the edematous stroma and absence of heterogeneity favours the diagnosis of massive edema of the ovary. Also, in SST the edema if present, is zonal as opposed to that seen in massive edema of ovary or edematous fibroma.

The presence of vacuolated cells and signet-ring like cells may lead to the mistaken diagnosis of Krukenberg tumor of the ovary. But these malignant tumors are known to occur in the sixth or seventh decades of life, are most commonly bilateral and lack the pseudolobulated pattern which is characteristic of SST. In addition to the above features, the signet-ring cells of Krukenberg tumor contain mucin while similar cells in SST contain lipid, and Krukenberg tumor being malignant will exhibit cytologic atypia [10].

Vascular tumors are also included in the differential diagnosis due to prominent vascularity, in such cases, inhibin positivity suggests the diagnosis of SST [10].

The patient in our case had an elevated CA-125 level. This is an unusual finding and is believed to be a consequence of physical irritation and inflammation. A few cases of SST with an elevated CA-125 levels have been reported in literature, which reinstates that CA-125 is not a specific marker of ovarian malignancy [11].

It is difficult to diagnose SST by only clinical and radiological examination as these tumours are rare and difficult to diagnose. But in a young patient presenting with an ovarian mass, SST should be considered as one of the differential diagnosis. SST is a benign tumor and can be treated successfully with fertility preserving surgeries[3].

4. Conclusion

This uncommon case of SST of the ovary in a young female emphasizes the importance of histopathological examination as the Gold Standard for the diagnosis and even though rare, it should be considered in the differential diagnosis specially in a young woman.

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