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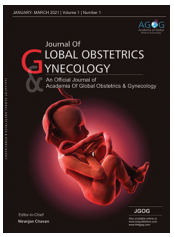
Case Report



Unveiling the Enigma: A Rare Case of Sclerosing Stromal Tumor

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ABSTRACT

Sclerosing stromal tumor is a distinctive sex cord-stromal tumor which is exceptionally rare. It occurs more often in the second or third decades of life. It presents with pelvic or abdominal pain and tenderness, mass, and/or abnormal menses. These tumors are hormonally inactive. This is a case report of rare presentation of a sclerosing stromal tumor in a 45-year female COVID recovered with chronic kidney disease and hypothyroidism. It posed a diagnostic dilemma due to advanced age of presentation and patient underwent exploratory laparotomy with total abdominal hysterectomy and bilateral salpingo-oophorectomy. Diagnosis was confirmed by histology and immunohistochemistry and patient had an uneventful postoperative period with follow-up of 1 year. Due to the rarity of this tumor, it is not always possible to predict its presence preoperatively based on clinical and radiological findings alone. A possibility of sclerosing stromal tumor must be kept in mind for patients with ovarian mass as cases have been reported in older age groups of more than 40 years as well.

Key words: Benign, Ovarian tumor, Rare, Sex cord-stromal tumor

INTRODUCTION

Sclerosing stromal tumor is a distinctive sex cord stromal tumor which is exceptionally rare. It occurs more often in the second or third decades of life.[1] Approximately 8% of all primary ovarian neoplasm are sex cord stromal tumors.[2] Sex cord-stromal tumors of the ovary include granulosa cell tumor, fibrothecomas, Sertoli-Leydig cell tumors, steroid cell tumors, and sclerosing cell tumors. Out of all sex cord stromal tumors, sclerosing cell tumors account for only 2–6%.[3] They are extremely rare benign ovarian neoplasm with close to 200 cases being reported in the literature so far. Sclerosing stromal tumor occur predominantly in the second or third decade of life.[4] Menstrual irregularities, pelvic pain, and abdominal mass are the typical presenting symptoms. Sclerosing stromal tumors are usually hormonally inactive. If hormonally active, they are androgenic and occur mostly during pregnancy. Since these neoplasms are extremely rare, it is not always possible to predict the presence of this tumor preoperatively on the basis of clinical and sonographic findings alone. Diagnosis is confirmed with histopathology and immunohistochemistry. CA-125 is often within normal limits.[5]

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CASE REPORT

A 45-year-old female P4L4 was referred for neoplastic mass lesion in the left adnexa, bilateral renal parenchymal lesion, minimal ascites, and minimal left pleural effusion. On admission, patient had complaints of pain in the abdomen for 8–10 days, fever and breathlessness for 4 days, loose stools, and decreased urine output for 2 days. Patient was diagnosed to be COVID positive and was conservatively managed for the same for 10 days. There was no history of any menstrual irregularities. On per abdomen examination, there was guarding and tenderness on deep palpation, abdominal distension with moderate ascites with no palpable mass. On per vaginal examination, left posterolateral mass of 6 × 8 cm single, firm to hard, regular non-tender, mobile felt in left fornix. CA-125 levels and other tumor markers such as Ca 19.9, lactate dehydrogenase, alpha feto protein, carcinoembryonic antigen, inhibin B, and beta human chorionic gonadotropin were within normal limits. Ultrasonography (USG) abdomen and pelvis was suggestive of 6.5*5.6*6.2 cm sized well-defined smooth

marginated with lobulated surface oval lesion with solid content and few hypoechoic cystic areas within noted in the left adnexa predominately in rectouterine pouch of ovarian origin ORADS 5 with no signs of torsion. Computed tomography plain abdomen and pelvis showed 5.1*6.8*5.1 cm sized large well-defined irregularly thick walled peripherally enhancing heterogeneous solid lesion with few cystic areas in left adnexa with left ovary not visualized separately. Moderate ascites were seen. No obvious lymphadenopathy and rest of abdominal organs within normal limits. Fluid cytology showed proteinaceous material with no malignant cells. Due to her advanced age and a mixed picture of unilateral left adnexal solid lesion with few cystic areas, moderate ascites with no lymphadenopathy, normal tumor markers, and normal fluid cytology, this case posed to be a diagnostic dilemma between a benign and malignant ovarian tumor. Patient was hence posted for exploratory laparotomy and intraoperatively showed moderate ascites with the left ovarian mass of 6*6 cm sized greyish white solid cystic mass, firm in consistency, well-circumscribed not adherent to any surrounding structures. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. Omentum and bowel examined and were found to be normal. Postoperatively, histopathology examination showed left ovarian mass well encapsulated and circumscribed tumor tissue comprising of hypocellular and hypercellular areas. Hypocellular area is composed of lobules of round cells with clear cytoplasm and uniform nuclei. At places signet ring-like cells are seen, stroma is markedly hyalinised and at places shows edema. Stroma shows many staghorn like vessels. No evidence of malignancy or atypia suggestive of sclerosing stromal tumor of ovary. Immunohistochemistry was done and found to be positive for smooth muscle actin and vimentin. Postoperatively, patient was vitally stable with lab parameters within normal limits and patient was counseled regarding the benign nature of the tumor and need for regular follow-up for observation. Patient was followed up for 1 year with routine examination and was found to have no complications.

DISCUSSION

Sclerosing stromal tumor is an extremely rare benign ovarian sex cord-stromal tumor occurring predominantly in the 2nd and 3rd decade of life with only around 200 cases being reported so far. Other ovarian stromal tumors present mostly in 5th or 6th decade of life. However, some cases of sclerosing stromal tumor have been reported after 40 years of age as well.[6,7] It represents around 2–6% of primary ovarian stromal tumors. Sclerosing stromal tumor commonly presents with menstrual irregularities, pelvic pain (which was present in our case), non-specific symptoms of ovarian mass. Sclerosing stromal tumors are usually hormonally inactive; however, masculinization and anovulation may be present in some cases due to estrogen and androgen secretion.[8]

On histopathology, sclerosing stromal tumor is different from other sex cord stromal tumours. It shows heterogeneity with dual cell population, collagen-producing spindle cells, and round or ovoid cells that contain lipid. Cellular pseudo lobules, prominent interlobular fibrosis, and marked vascularity is also observed. Other stromal tumors like thecoma and fibroma have relatively homogenous appearance.[9] Vascular sclerotic and edematous stromal changes are a constant feature of this tumor due to local production of vascular permeability and growth factors. Vascular tumors may be included in the differential diagnosis due to prominent vascularity of these tumors. Sometimes, the edematous stroma of these tumors contains vacuolated cells and signet ring cells which can be confused with Krukenberg tumor of ovary. In contrast to sclerosing stromal tumors, Krukenberg tumors are usually bilateral and seen in 5th or 6th decade of life. To differentiate between the two on microscopy, Krukenberg tumor lack pseudolobular pattern, may show nuclear atypia or mitotic activity, and contain mucin rather than lipid.[10]

Diagnosis is confirmed for sclerosing stromal tumors with immunohistochemistry which shows positivity for smooth muscle actin, inhibin, and vimentin suggesting its stromal origin. CD34 stains the endothelium of dilated and branching vascular architecture seen in sclerosing stromal tumor as opposed to fibroma and thecoma. Alpha glutathione S-transferase positivity within scattered cells also appears to be useful in distinguishing from a diffusely staining thecoma and no staining fibromas.[11]

Sclerosing stromal tumors show a mixed pattern of solid and cystic components on USG which may cause confusion with other malignant tumors.[12] Color Doppler shows prominent peripheral vascularity and vascularity in central inter-cystic spaces.[13] On Magnetic resonance imaging (MRI), heterogeneous solid mass of intermediate to high signal intensity on T2-weighted images is seen or a large mass with hyperintense cystic components is seen. MRI is better to differentiate from other malignant ovarian tumors. Dynamic contrast-enhanced images show early peripheral enhancement reflecting cellular areas with prominent vascular networks and an area of prolonged enhancement in inner portion of the mass representing collagenous hypocellular areas. These findings are not seen in thecomas and fibromas.[14]

CONCLUSION

Due to the rarity of this tumor, it is not always possible to predict its presence preoperatively based on clinical and radiological findings alone. Possibility of sclerosing stromal tumor must be kept in mind for young patients with ovarian masses, with a few cases reported in older age groups of more than 40 years as well. Diagnosis is confirmed with histology and immunohistochemistry. All the reported cases of sclerosing stromal tumor have been benign and have been treated successfully with only enucleation or unilateral oophorectomy with no incidence of recurrence.

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