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CASE REPORT

SCLEROSING SEX CORD STROMAL TUMOR A RARE CASE REPORT

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ABSTRACT

Sclerosing stromal tumor is a rare benign neoplasm that accounts for 2–6% of ovarian stromal tumors. It is generally found in patients who are young, 80% are < 30 years of age. Patients often present with menstrual irregularities or symptoms related to the presence of a pelvic mass. The tumors are almost always unilateral and range from 1.5 to 20 cm in largest dimension. We present a case of a 29 year old female, who presented with complaints of lump in abdomen since three months. Her clinical and radiological examination raised suspicion towards an adnexal tumor? malignant. It was only after histopathological examination that the diagnosis of a benign sclerosing sex cord stromal tumor was confirmed.

INTRODUCTION

These neoplasms account for approximately 6% of all ovarian tumors and for most clinically functioning ovarian tumors. They contain elements of sex cord and stromal derivation in varying combinations and with differing amounts of cytological atypia and mitotic activity. The most common subtype of sex cord–stromal tumor is the endocrinologically inactive fibroma[1].The great array of patterns that may be encountered within tumors in the sex cord–stromal family lead to numerous issues in differential diagnosis.

Case Report

We present a case of a 29 year old female, who presented with complaints of lump in abdomen since three months. The patient complained of abdominal discomfort. Patient had menorrhagia and her menstrual cycles were regular. Patient's blood investigations were within normal limits. CA-125 was slightly raised. USG abdomen (Figure 1 A) revealed 25 x 18 x12cms sized cystic lesion seen in the left ovary with thick internal sepatations measuring 5mm with few low lying echoes within few of the septae. The left ovary was not visualized separately. The lesion was separate from the uterus and was diagnosed as mucinous cystadenoma.

CT abdomen (Figure 1 B) revealed- a large complex mass arising from the pelvis (presumably left ovary) extending into the abdomen upto the level of the body of the pancreas superiorly. The mass was smooth in outline, showed cystic and solid enhancing components and septae with peripheral vascularity supplied by the uterine artery. Final diagnosis of large ovarian tumor presumably mucinous cystadenoma was given.

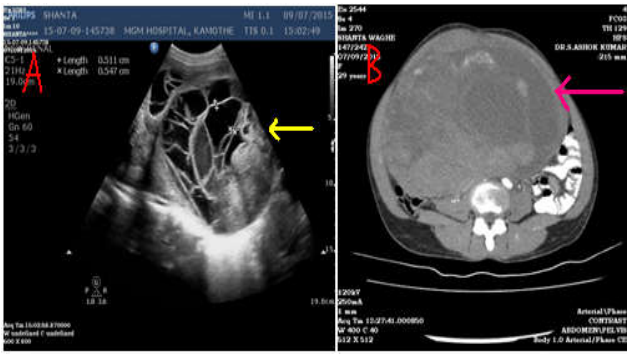

Figure 1 A – Ultrasonography image (yellow arrow- tumor),
B – CT – Abdomen image (pink arrow –tumor)

Gross findings revealed a well encapsulated, bosselated, irregular, glistening mass, weighing 2.1kilograms, measuring 28 x 20 x 15 cms. On cut-section, solid and cystic areas were identified, largest cyst measured 9 x 6 cm. Multi-loculated cystic areas containing blood mixed serous fluid were identified. Solid component appeared multi nodular and yellowish-white. (Figure 2 A) Microscopic findings: Sections

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showed a well encapsulated tumor comprising of cellular pseudolobules made of round to oval cells, few with vacuolated cytoplasm along with dense collagenous stroma.(Figure 2 B, Figure 3 A). Also seen were thecoma fibroma like areas, few areas showing hemangiopericytoma like pattern. Stroma also contained few cells with vacuolated cytoplasm along with dense collagenous stroma. Immunohistochemistry .showed uniform positivity for vimentin (Figure 3 B), and uniformly negative for cytokeratin. Also the sections were negative for alcian blue.

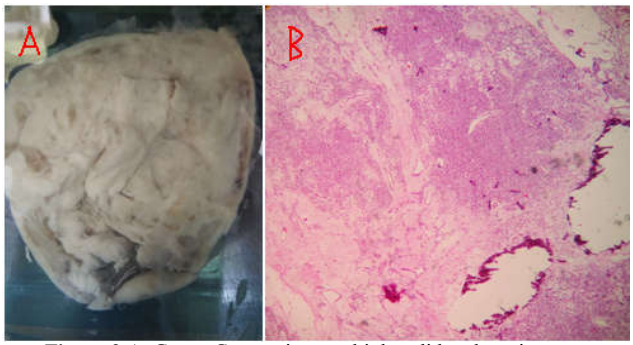

Figure 2 A–Gross :Cut-section- multiple solid and cystic areas, B – Microscopy – 40x magnification showing pseudolobules.

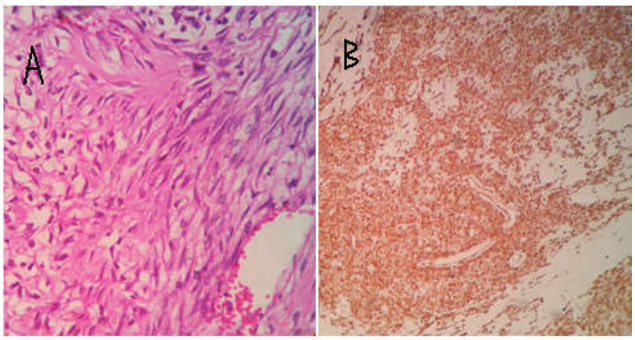

Figure 3 A – Microscopy – 400x magnification; B – Vimentin-Positive - 100x magnification.

DISCUSSION

Sex cord stromal tumors account for approximately 6% of all ovarian tumors and for most clinically functioning ovarian tumors.[1] Sclerosing stromal tumor is a rare benign neoplasm that accounts for 2–6% of ovarian stromal tumors. It is generally found in patients who are younger than those with either thecomas or fibromas, as 80% are < 30 years of age. Patients often present with menstrual irregularities or symptoms related to the presence of a pelvic mass. [2] Our patient presented with complaints of lump in the abdomen since three months. The patient had complaints of menorrhagia. These ovarian tumors differentiate in the direction of sex cord and/or the specialized ovarian stroma.[3]This includes female-type cells (granulosa and theca cells), male-type cells (Sertoli and Leydig cells), and indifferent elements.[4,5,6]. These various elements can occur in combination[7] and exhibit a wide range of differentiation, which often seems to recapitulate the patterns produced during embryogenesis of both the ovary and testis, as proposed many years ago by Gunnar Teilum.[8] The tumors are almost always unilateral and range from 1.5 to 20 cm in largest dimension. They have a well defined border that, in some cases, has led to enucleation of the tumor at surgery. They are white to yellow and typically solid, or solid with multiple cystic spaces. [2] Our patient had a tumor measuring 28 x 20 x 15 cms, weighing 2.1 kilograms. On cut-section, solid and cystic areas were identified. Solid component appeared multi nodular and yellowish white. Sclerosing stromal tumor is a benign ovarian neoplasm that shares many features with fibroma and thecoma. However, it occurs in a younger age group, has a less homogeneous gross appearance, and is characterized microscopically by a lobular pattern of growth, interlobular fibrosis, marked vascularity, and the presence of a dual cell population: collagen-producing spindle cells and lipid-containing round or oval cells. Some of the latter may have a signet ring appearance and thus simulate a Krukenberg tumor[9]. The sections from our patient's tumor showed cellular pseudolobules made of round to oval cells, few with vacuolated cytoplasm along with dense collagenous stroma. Immunohistochemistry showed diffuse positivity for vimentin and uniform negativity for cytokeratin thus Krukenberg tumor was ruled out.

Calcitonin, inhibin, CD34, and α-glutathione S-transferase (αGST) positivity has been reported to be useful to differentiate sclerosing stromal tumors from the comas, fibromas and other sex cord stromal tumors[10]. The heterogeneous appearance of the SST contrasts with the relative homogeneity of fibromas and thecomas. Although fibromas may be edematous and vascular, the edema is generally diffuse rather than focal, and pseudolobules and lutein cells are absent or rare. Hyaline plaques, a conspicuous feature of many fibromas and the comas, are rare in SSTs.[1]

CONCLUSION

Sclerosing stromal tumor is a very rare benign tumor. Though benign, the tumor can be very large occurring upto 20cms causing lot of agony and anxiety to the patient. Also radiological imaging can be misguiding towards a nonspecific ?malignant neoplasm. The benign nature of the tumor can be confirmed only after histopathological examination. The patient generally does not have any recurrence and any menstrual abnormalities if present are generally resolve post surgery. [11]

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