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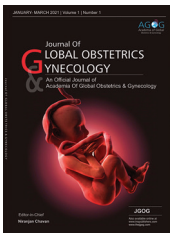
Case Report



A Rare Case of Benign Sclerosing Stromal Tumor Masquerading as Germ Cell Tumor

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ABSTRACT

Aim: To understand the diagnosis of a rare ovarian tumor that masquerades as another tumor. **Background:** Sclerosing stromal tumor (SST) is a rare ovarian tumor that usually occurs in young adults in the second and third decades of life. **Case Description:** A case of a SST of the ovary with a review of literature to study a detailed algorithmic approach toward it. The patient was diagnosed with this tumor only after histopathological examination postoperatively, which masquerading as a dysgerminoma on blood investigations and imaging studies. **Conclusion:** The tumor has distinct histological features and is easily recognizable when a high index of suspicion is maintained in young patients presenting with an ovarian mass. **Clinical Significance:** These tumors are benign and can be treated successfully by enucleation or unilateral oophorectomy, and up to 200 cases have been reported in the literature until now.

Key words: Sclerosing stromal tumor, Ovarian tumor, Dysgerminoma, Tumor markers

BACKGROUND

Ovarian sex cord tumors are infrequent neoplasms that account for approximately 8% of all primary ovarian neoplasms.[1] A sclerosing tumor originates from the stroma of the ovary. Most sclerosing stromal tumors (SSTs) have been observed in young adults in the second and third decades of life.[2] Menstrual irregularity and pelvic pain are the most frequent complaints during presentation.

CASE DESCRIPTION

A 23-year-old married nulligravida patient presented with complaints of dull, aching pain over the right iliac fossa intermittently for 3 months with gradually increasing intensity. The patient had been a known case of an ovarian tumor for 2 years but was lost to follow-up and had currently presented to the outpatient department with aggravation of symptoms. The patient had an ultrasound of the abdomen and pelvis suggestive of a midline cystic mass with a solid component with the right ovary not seen separately from it of size 18 cm × 15 cm × 10 cm with mild compression signs on the right ureter and right kidney, probably a right ovarian cyst adenoma. The patient did not have any history of vomiting episodes. She had irregular menses with polymenorrhea without significant dysmenorrhea or heavy menstrual bleeding. The patient was tested negative for pregnancy on the beta human chorionic gonadotropin rapid kit test.

On general examination, the patient was vitally stable, without signs of pallor or cervical or supraclavicular lymphadenopathy. On abdominal examination, a midline mass corresponding to 26 weeks in size, firm in consistency, and arising from the pelvis with free side-to-side mobility was palpated without any signs of guarding, tenderness, or rigidity. The cervix and vagina were otherwise healthy. On per vaginum examination, a 26-weeks-size mass felt through the right and anterior fornix and uterus was not felt separately from it. The mass did not move with cervical motion. No forniceal tenderness was elicited.

Routine blood investigations were advised, along with a computed tomography (CT) scan and magnetic resonance imaging (MRI) for demarcating the planes and the exact size of the pelvic mass. A CT scan was done primarily to check for lymph node involvement by the tumor, while an MRI was done to study

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the plane and extent of the tumor. CT findings showed a 19.8 cm × 14.7 cm × 9.2 cm well-defined multiloculated cystic lesion in the pelvic-abdominal region in midline, with the right ovary not seen separately [Figure 1]. The solid component of the lesion showed arterial enhancement with progressive enhancement on venous and a delayed phase-suggestive of dysgerminoma. No lymph node involvement was noted. On MRI, it was seen extending from the coccyx inferiorly to the L3 vertebra superiorly, with compression seen on the urinary tract, aorta, inferior vena cava, and iliac vessels posteriorly, the anterior abdominal wall anteriorly, and the urinary bladder inferiorly. There was mild ascites. Tumor marker investigations as reported are shown in Table 1.

Table 1: Tumor marker levels

Tumour Marker	Serum level	Laboratory reference level
Lactate dehydrogenase	298.8 U/L	140–280 U/L
Carcino embryogenic antigen	0.52 ng/mL	0–5 ng/mL
CA 19–9	6.74 U/mL	0–37 U/mL
CA 125	15.1 U/mL	0–35 U/mL
Beta HCG	<1.2 mIU/mL	<5.3 mIU/mL

HCG: Human chorionic gonadotropin

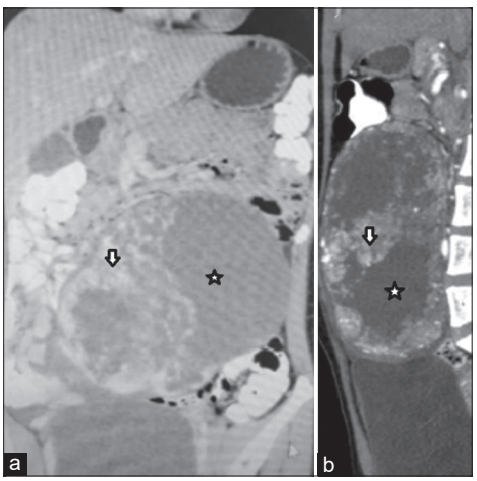

Figure 1: Coronal (a) and Sagittal (b) section view of computed tomography scan of the ovarian tumor with solid (arrow) and cystic (star) areas

All the above findings were suggestive of a germ cell tumor, probably a dysgerminoma. Decision to perform exploratory staging laparotomies with adequate exposure and care sought to prevent spillage. Intraoperatively, a 20 cm × 15 cm × 8 cm mass with cystic consistency and scanty firm areas [Figure 2a] was seen arising from the right ovary [Figure 2b]; however, the right ovary was not seen separate from the mass, and the right fallopian tube was entirely stretched over the mass and was also dilated. The mass was seen to be highly vascular, and it arose from the right ovary and did not invade any surrounding structures. The left-sided ovary was normal in appearance. A right-sided salpingo-oophrectomy by applying a clamp to the infundibulo-pelvic ligament was performed. The specimen was sent for histopathological examination, and the peritoneal and cyst fluid were sent for cytological study.

Histopathology of the specimen was reported as benign SST. Gross examination showed cystic and firm areas [Figure 2c], while microscopic examination showed a pseudo-lobular pattern of cellular zones separated by broad acellular sclerotic tissues with numerous branching vascular spaces of varying size in cellular areas [Figure 2c]. A cytological study of the peritoneal as well as tumor cyst fluid showed the absence of malignant cells.

The patient had an uncomplicated post-operative course and was discharged on day 10 of surgery. Since the literature has no documentation of recurrence of this tumor, the patient and relatives were counseled regarding the same and the effect of the surgery on the patient's subsequent fertility.

CASE DISCUSSION

Sertoli–Leydig cell tumors, fibro-thecomas, steroid cell tumors, and sclerosing stromal tumors are only a few examples of the numerous types of ovarian sex cord-stromal cancers.[3] They are often unilateral and tightly constrained; the literature has not documented their recurrence. According to a theory, SST develops from the theca externa naturally occurring muscle-specific actin-positive elements known as perifollicular myoid stromal cells.[3]

Due to their mixed pattern of cystic and solid components, SST can mimic malignant ovarian tumors on ultrasound; nevertheless,

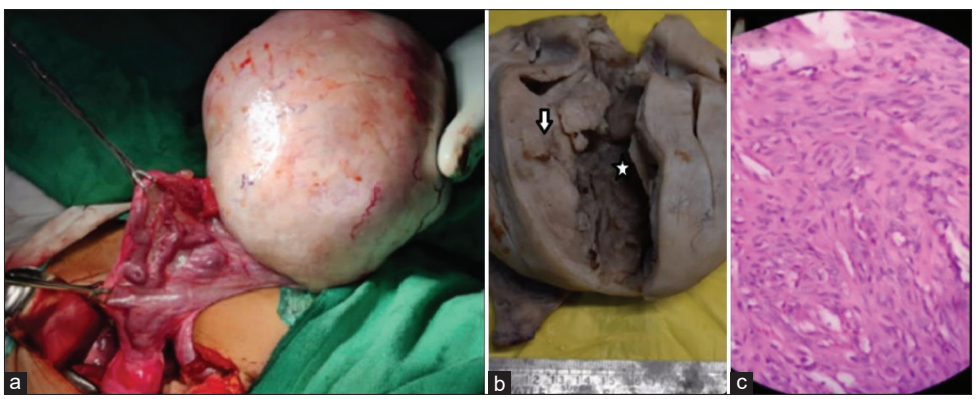

Figure 2: Ovarian mass (a) with gross cut section (b) showing solid (arrow) and cystic (star) areas and ×40 magnification microscopic examination (c)

color Doppler ultrasonography can highlight extensive peripheral vascularity and central inter-cystic gaps. The same characteristics found on ultrasonography may also be seen on CT. SST can be distinguished from malignant ovarian tumors using MRI more effectively than clinical examination, which may reveal a big mass with hyperintense cystic components or a heterogeneous solid mass with varied intermediate-to-high signal intensity. On the basis of radiological and macroscopic examination, it is challenging to distinguish SST composed of solid and cystic areas from ovarian cancers because these tumors additionally seem very vascular, providing the impression of malignant tumors.

CLINICAL SIGNIFICANCE

The diagnosis of SST of the ovary, which is an exceedingly rare and distinctive sex cord-stromal tumor, depends on the patient's young age (often in the second or third decade of life), the unilaterality of the tumor, and the distinctive macroscopic and histological appearance of the tumor. Due to the rarity of this neoplasm, it is not always possible, as was the case in our instance, to predict the presence of this tumor preoperatively based solely on clinical and radiological signs. Young patients with ovarian masses should always have the possibility of a sclerosing stromal tumor considered in the differential diagnosis. These benign tumors can be successfully removed by unilateral oophorectomy or enucleation. Surgical resection of the tumor is curative since, to date, no local or distant recurrences have been reported in the literature.

ACKNOWLEDGMENT

We would like to thank the medical staff of the pathology and radiodiagnosis departments of LTMMC and LTMGH Hospital, Sion, Mumbai, who played an invaluable role in the management of this patient.

Patient's Informed Consent

Taken.

Funding

No funding sources.

Conflict of Interest

None declared.

Ethical Approval

Not required.

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How to cite this article: Nimbkar A, Panchbudhe S, Neeraja G. A rare case of benign sclerosing stromal tumor masquerading as germ cell tumor. J Glob Obstet Gynecol 2022;2(2):45-47.

Source of support: Nil, **Conflict of Interest:** Nil.