

Ovarian Sertoli Cell Tumor: A Rare Case of Sex Cord Stromal Tumor

Neelu Gupta¹, Dharm Chand Kothari², Neeraj Arora²

¹Associate Professor, Department of Pathology, Sardar Patel Medical College, Bikaner, Rajasthan, India, ²Post-graduate Student, Department of Pathology, Sardar Patel Medical College, Bikaner, Rajasthan, India

Abstract

Sertoli cell tumors are rarest sex cord-stromal tumors, these occur most often in women of reproductive age, but occasionally arise in children and postmenopausal women. Two-thirds of Sertoli cell tumors secrete steroid hormones. Girls with hormonally active tumors present with precocious pseudopuberty and vaginal bleeding. Older women have irregular bleeding, postmenopausal bleeding, or, rarely, virilization, depending on the type and amount of hormone secreted. Patients with hormonally inactive neoplasms have nonspecific symptoms such as pain or abdominal swelling, or their tumors are incidental findings. Grossly Sertoli cell tumors are unilateral, encapsulated and cut surfaces are grey, tan, brown, or yellow and are predominantly solid, or cystic. Microscopically, they are composed of closely packed tubules separated by fibrous stroma. The tubules are lined by cuboidal to columnar cells with abundant pale eosinophilic cytoplasm, with little atypia or mitotic activity. We report a case of Sertoli cell tumor in a 22-year-old female presenting with complaints hoarseness of voice, hirsutism, flat breast and pelvic mass with ascites underwent unilateral salpingo-oophorectomy. Histopathological examination revealed a diagnosis of Sertoli cell tumor of the ovary.

Keywords: Ovary, Sertoli cell tumor, Sex cord tumor

INTRODUCTION

Sertoli-Leydig cell tumors are uncommon, comprising <0.1% of ovarian neoplasms.¹ Sertoli cell tumors are among the rarest sex cord-stromal tumors.² A Sertoli cell tumor is a sex cord-gonadal stromal tumor of a Sertoli cells. Although Sertoli cells normally occur only in the testis, this type of tumor may also rarely occur in the ovary of females.²

They differ from Sertoli-Leydig cell tumors in that they do not contain Leydig cells or immature gonadal stroma. Tumors with complex annular tubules have been classified as Sertoli cell tumors. Sertoli cell tumors occur most often in women of reproductive age group, but they occasionally arise in children and postmenopausal women. The average patient age is about 30 years.^{2,3}

Girls with hormonally active tumors present with precocious pseudopuberty and vaginal bleeding. Older women have irregular bleeding, postmenopausal bleeding, or, rarely, virilization, depending on the type and amount of hormone secreted. Patients with hormonally inactive neoplasms have nonspecific symptoms such as pain or abdominal swelling, or their tumors are incidental findings. Sertoli cell tumors are unilateral, and most are clinically benign tumors that can be treated by unilateral salpingo-oophorectomy.

Microscopic Pathology

These are tumors composed of Sertoli cells that grow in mature fibrous or hyalinized stroma. A tubular pattern is characteristic.² Sertoli cell tumor include a lipid-rich type, in which the cells have abundant clear, foamy cytoplasm, and an oxyphilic type composed of cells with abundant granular eosinophilic cytoplasm. Most Sertoli cell tumors are well differentiated, and the neoplastic cells have uniform nuclei and few mitotic figures. Sertoli cell tumors that are confined to the ovary at diagnosis and that exhibit minimal or no minimal nuclear atypia and mitotic activity can be viewed as benign.

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Corresponding Author:

Dr. Dharm Chand Kothari, Room No. G, Hostel No. 4, UG Hostel, Sardar Patel Medical College Campus, Bikaner - 334 001, Rajasthan, India. Phone: +91-9509536304. E-mail: drdckothari@gmail.com

CASE REPORT

A 22-year-old nulliparous Hindu female present to the gynecologic clinic with complaints of progressive irregular bleeding (menometrorrhagia) for 1 year duration. She had also noticed a gradual change in her voice for 1 year and excessive hair growth on her face, chest, and limbs for the last 6 months. In addition, she complained of abdominal discomfort due to left abdominal lump. She also complains that her breast was atrophied.

On physical and clinical examination revealed pseudo precocity, menometrorrhagia, hirsutism, breast atrophy, clitoral hypertrophy and hoarseness.

Per abdominal examination revealed a mass in left pelvic area 14 cm × 12 cm and ascites.

Ultrasonography: A cystic mass measuring 13 cm × 11 cm with ascitic fluid.

Contrast-enhanced computed tomography: Abdominal lump measuring 17 cm × 13 cm in size complex cystic mass is abutting anterior abdominal wall and displaces intestinal loops.

Carbohydrate antigen: 125 was 40 µ/ml.

Ultrasound guided fine-needle aspiration cytology, which revealed the presence of benign cystic cells.

Routine Investigation

Hematological investigation: Hemoglobin - 14.5 gm%, total leukocyte count - 9500/mm³, erythrocyte sedimentation rate - 20 mm at the end of 1 hour. Bleeding time and clotting time was within normal limit.

Urine microscopy and biochemistry was normal.

Her chest X-ray was clear and not shows any opacity.

So her general physical examination was normal except for the presence of hirsutism, breast atrophy, clitoromegaly and left pelvic mass.

The patient underwent exploratory laparotomy under general anesthesia because there was a strong suspicion of malignancy. Left salpingo-oopherectomy was done, and sample was sent for histopathological examination.

Pathologic Finding

Gross

Tubo-ovarian mass measuring 15 cm × 13 cm × 5 cm shows well circumscribed tumor with fallopian tube. Outer surface of mass was grey white, smooth and

vascular marking was prominent. The cut surfaces were multilocular multicystic and grey white to brown in color and were predominantly cystic. Cysts are filled with serous, mucinous and hemorrhagic fluid. A few gray, brown solid areas present in between the cystic areas (Figure 1).

Multiple section from cyst and solid areas were taken processed and stain with hematoxyline and eosin and examined under microscope. Histopathological findings confirm the diagnosis.

Microscopy

Section shows hollow, solid, simple or complex tubules (Figures 2 and 3). Hollow tubules are lined by columnar to cuboidal cells with small round to oval nuclei with a moderate amount of eosinophilic cytoplasm (Figure 4). Simple tubules are surrounded by a basement membrane and contain central hyaline bodies (Figure 5). Complex



Figure 1: Cut surfaces of tubo-ovarian mass were multilocular multicystic and grey white to brown in color. A few grey brown solid areas present in between the cystic areas

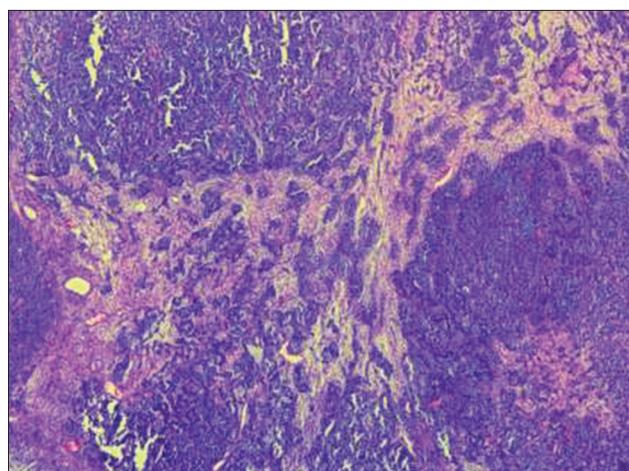


Figure 2: Hollow, solid, simple or complex tubules with hyalinized stroma (4x)

tubules form multiple lumens filled with hyaline bodies and surrounded by a thick basement membrane. Solid tubules filled with pale cells with moderate to abundant cytoplasm containing lipid (Figure 6). In some tumors, cells distended by intracytoplasmic lipid are seen in a pattern known as "folliculomelipidique" (Figure 6). The nucleus is typically oval or spherical with a small nucleolus. The cytoplasm is clear or lightly vacuolated, stains for lipid (Sudan black B) is positive.

On the basis of clinical history, physical examination and histopathological findings a diagnosis of Sertoli cell tumor was made.

DISCUSSION

Sertoli cell tumors are among the rarest sex cord-stromal tumors.² Patients range in age from 2 to 79 years. Mean ages of 21 and 38 years and median ages of 33 and 50 years

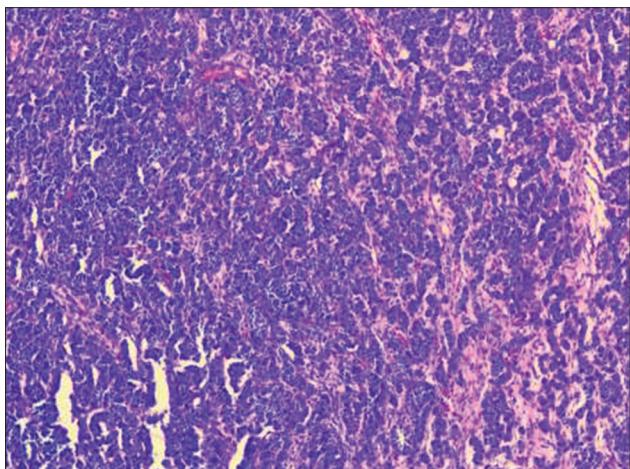


Figure 3: Hollow, solid, simple or complex tubules (H and E, 10x)

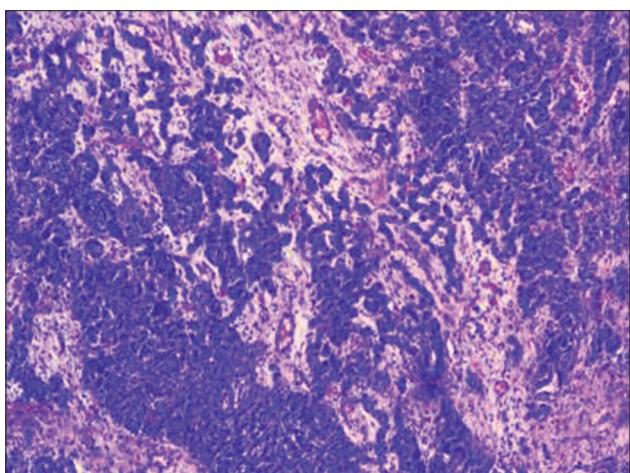


Figure 4: Cords, trabeculae, and diffuse growth pattern of sertoli cells with areas of hyalinized stroma. Leydig cell is not seen (H and E, 10x)

have been reported in the two largest series.⁴ The average patient age is about 30 years.^{2,3}

The tumors are functional in 40-60% of cases, most often estrogenic, but occasionally androgenic or rarely both. Rarely, the tumor produces progestins.⁴

Clinical manifestations include isosexual pseudo precocity, menometrorrhagia, amenorrhea, hirsutism, breast atrophy, clitoral hypertrophy and hoarseness.

Girls with hormonally active tumors present with precocious pseudopuberty and vaginal bleeding. Older women have irregular bleeding, postmenopausal bleeding, or, rarely, virilization, depending on the type and amount of hormone secreted. Patients with hormonally inactive neoplasms have nonspecific symptoms such as pain

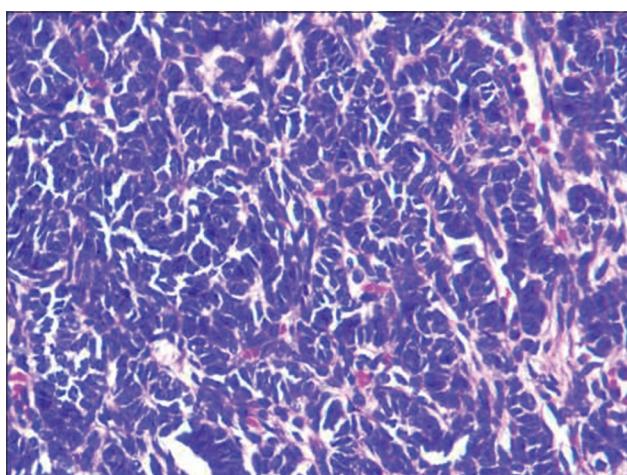


Figure 5: Hollow, solid, simple or complex tubules. Tubules are surrounded by a basement membrane and contain a central hyaline body. Tubules are lined by columnar to cuboidal cells with small round to oval nuclei with a moderate amount of eosinophilic cytoplasm (H and E, 40x)

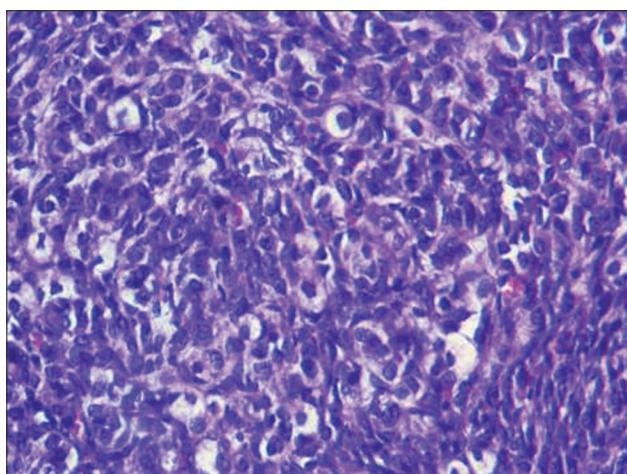


Figure 6: Diffuse sheet of sertoli cells. Some cells shows cytoplasmic clearing (H and E, 40x)

or abdominal swelling, or their tumors are incidental findings.

Cases with menstrual disturbances or postmenopausal bleeding may show hyperplasia or adenocarcinoma of the endometrium. A peritoneal decidual reaction may be seen. Patients with Sertoli cell tumor may have elevated levels of serum estrogen, progesterone and luteinizing hormone.⁴ Occasionally, a Sertoli cell tumor occurs in a patient with Peutz–Jeghers syndrome.^{2,5,6} Sertoli cell tumors are unilateral, and most are clinically benign tumors that can be treated by unilateral salpingo-oophorectomy. Occasional poorly differentiated or invasive sertoli cell tumors recur or metastasize and cause the patient's death.²

Gross Pathology

These are unilateral neoplasms, and the ovaries are involved with equal frequency. They range in size from 1 to 28 cm with an average of 7-9 cm. They are well-circumscribed, solid neoplasms with a smooth or lobulated external surface, a fleshy consistency and a yellow-tan sectioned surface. Areas of hemorrhage and/or cystic degeneration may be seen in larger tumours.⁴

Microscopic Pathology

These are tumors composed of sertoli cells that grow in mature fibrous or hyalinised stroma. A tubular pattern is characteristic.² The tubular pattern tends to be a simple one, consisting of round or oval open glands with a central lumen or long, closed cord-like tubules two or three cells thick. Mixed tubular patterns are often present. In some tumors, cords, trabeculae, or areas of diffuse tumor cell growth are present. Sertoli cells are cuboidal or columnar and have round to oval nuclei, which sometimes contain small nucleoli. Bizarre but regenerative nuclei do not appear to affect the prognosis adversely. The cytoplasm varies from clear to eosinophilic. Uncommon variants of sertoli cell tumor include a lipid-rich type, in which the cells have abundant clear, foamy cytoplasm, and an oxyphilic type composed of cells with abundant granular eosinophilic cytoplasm.⁵ Most Sertoli cell tumors are well differentiated, and the neoplastic cells have uniform bland nuclei and few mitotic figures. Sertoli cell tumors that are confined to the ovary at diagnosis and that exhibit minimal or no nuclear atypia and mitotic activity can be viewed as benign. Less than 10% of sertoli cell tumors are clinically malignant. These have a poorly developed pattern of tubules, the sertoli cells have enlarged atypical nuclei, and five or more mitotic figures are seen per 10 hpf.^{2,7,8} Malignant sertoli cell tumor can bear a resemblance to endometrioid

adenocarcinoma, but can be differentiated from it by the presence of more typical patterns of sertoli cell growth and by differences in immunophenotype. Sertoli cell tumors are variably positive for keratins, vimentin and alpha-inhibin. Steroidogenic factor 1 and Wilm's tumor 1 stain the tumor cell nuclei in virtually all Sertoli cell tumors, and thus are good, be it not specific, immunohistochemical markers for these tumors.^{9,10} CD99 and calretinin are positive in about 50% of cases. The tumors are negative for epithelial membrane antigen.⁴

Differential Diagnosis

Sertoli cell tumors must be distinguished from strumaovarii, carcinoid and endometrioid carcinoma. Phenotypic females with the androgen insensitivity syndrome may be correctly diagnosed as having a Sertoli cell tumor of the ovary if the syndrome has not been diagnosed preoperatively.⁴

CONCLUSION

Sertoli cell tumor should be kept in mind as a differential diagnosis in a young female presenting with the complaints of hirsutism, irregular bleeding, and atrophy of breast with pelvic lump.

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