CASE REPORT

A Rare Case of Unilateral Ovarian Sex Cord Tumor with Annular Tubules

Siddhi Gaurish Sinai Khandeparkar, Maithili Kulkarni, Vaishali R. Aphale, Bageshri P. Gogate, Sheetal Gosavi, Ketki Ghanekar

Department of Pathology, Smt. Kashibai Navale Medical College and General Hospital, Pune, Maharashtra, India

ABSTRACT

Sex cord tumor with annular tubules (SCTATs) is a rare ovarian tumor accounting for <1% of sex cord stromal tumors. It is of two types sporadic and syndromic. Syndromic SCTATs are regarded as benign, are usually bilateral, and are associated with germline STK11 mutations on chromosome 19p13.3 characteristic of Peutz–Jeghers syndrome. Sporadic SCTAT cases are unilateral and about 20% of them are regarded to manifest extra ovarian spread. Here, we intend to put on record a rare case of sex cord stromal tumor with annular tubules in a 49-year female presenting with left adnexal mass.

Key words: Ovarian tumor, hyperestrinism, sex cord tumor with annular tubules, inhibin

INTRODUCTION

Sex cord tumor with annular tubules (SCTATs) is a rare ovarian tumor accounting for less than 1% of sex cord stromal tumors. It was first described in 1970 by Scully.[1] It is of two types sporadic and syndromic.[2] Syndromic SCTATs are regarded as benign, are usually bilateral, and are associated with germline STK11 mutations on chromosome 19p13.3 characteristic of Peutz–Jeghers syndrome (PJS).[3] Sporadic SCTAT cases are unilateral and about 20% of them are regarded to manifest show extra ovarian spread.[4] Here, we intend to put on record a rare case of sex cord stromal tumor with annular tubules in a 49-year female presenting with left adnexal mass.

CASE REPORT

A 49-year, Gravida-2-Para-2-living-2, married female presented to gynecologic outpatient department with complaints of abdominal pain for a week and menorrhagia for 3 years. She had a history of death of one fetus at 36 weeks of gestation and lower segment cesarean section with twin pregnancy. Laboratory tests showed CA-125 of 13 U/mL (normal range: <35 U/mL). Ultrasound (USG) showed a well-defined complex solid-cystic lesion measuring 6.5 × 5.4 cm in left adnexa. The patient underwent total abdominal hysterectomy with left salpingo-oophorectomy. Resected specimen was sent for histopathological evaluation.

Grossly, globular soft to firm tissue mass measuring 8 cm in diameter and weighing 750 g was seen. The capsule was intact. Cut surface was solid, yellowish with few tiny cystic areas. Microscopically tumor cells were arranged in sheets and in simple and complex tubular areas. Few tubules were ring shaped with peripherally oriented nuclei around central hyaline material while few showed interconnecting rings revolving around multiple hyaline bodies. Individual tumor cells were medium sized, round to oval with moderate amount of cytoplasm with vesicular chromatin with few nuclei showing grooves. There was minimal atypia and mitosis of 1/10 HPF. There was no lymphovascular invasion noted. Histopathological diagnosis of unilateral sex cord stromal tumor with annular tubules was offered. Immunohistochemistry (IHC) done with inhibin

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Address for correspondence:
Siddhi Gaurish Sinai Khandeparkar, Department of Pathology, Smt. Kashibai Navale Medical College and General Hospital, E-517, The Island, Wakad, Pune - 411 057, Maharashtra, India.
Mobile: +91-9604791261.
E-mail: siddhigsk@yahoo.co.in

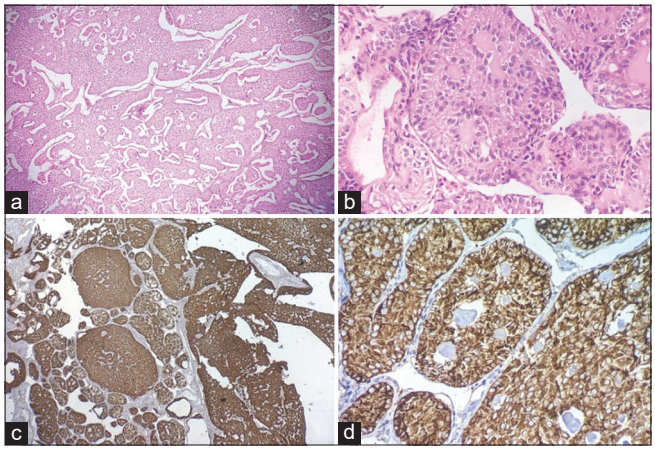

Figure 1: Photomicrograph showing (a) sharply delineated simple and complex tubules containing basement membrane-like material on ×100 and (b) ×400 magnification (c) tumor cells expressing strong cytoplasmic inhibin positivity on ×100 and (d) ×400 magnification

showed strong cytoplasmic immunoreactivity confirming the diagnosis (figure 1). The endometrial study showed disorderedly proliferative endometrium with tiny endometrial polyp. There was seen intramural leiomyoma of 1 cm in diameter. The patient was reexamined and radiologically reinvestigated to rule out PJS. General physical examination and gastrointestinal endoscopy ruled out syndromic SCTAT. On follow-up, the patient is disease free for 1½ years.

DISCUSSION

The SCTAT is a distinctive ovarian neoplasm exhibiting morphologic features intermediate between those of the granulosa cell tumor and those of the sertoli cell tumor.[1] About one third of the patients with SCTAT are associated with PJS characterized by mucocutaneous pigmentation and gastrointestinal polyposis.[2] Sporadic SCTATs occur most commonly among women of reproductive age group.[5] Our patient was 49 years and did not have PJS on clinical and radiological evaluation. Other associations reported with SCTAT include adenoma malignum of cervix, Turner's syndrome, dysgerminoma, gonadoblastoma, endometrial carcinoma, and endometriosis of fallopian tube.[1] On histopathological evaluation, there was seen disorderedly proliferative endometrium, tiny endometrial polyp, and intramural leiomyoma in this case. This can be explained as signs of hyperestrinism in present case. However, serum estradiol levels were not performed in this case.

The characteristic histomorphology of variably size round nests of sharply delineated simple and complex tubules containing basement membrane-like material, which may also be present around the tubules helps arriving at an accurate diagnosis. However, gonadoblastoma (seen in indeterminate gonads, composed of germ cells, and sex cord stromal cells distorted by hyalinization and calcification), adult granulosa cell tumor (Call-Exner bodies, tumor cells with coffee bean nuclei), and sertoli cell tumor (tubules lined by columnar cells) may be regarded as histomorphological differentials.[4,6]

SCTATs are known to show immunoexpression of immunohistochemical markers such as inhibin, calretinin, and WT1 which are also seen in most sex cord stromal tumor. SCTAT do not express placental alkaline phosphatase, CD117, and alpha-fetoprotein.[2,5] Thus, characteristic histomorphological features together with IHC could help in arriving at accurate diagnosis and ruling out differentials.

USG remains the most sensitive and cost-effective imaging modality for initial assessment of adnexal masses.[7] Other imaging modalities such as CT, magnetic resonance imaging, and positron imaging tomography scans can be used for better characterization of ovarian SCTAT, detection of extraovarian disease, and identification of other possible primary neoplasms.[7]

SCTAT has been reported in literature to be treated by surgically (salpingo-oophorectomy) in non-syndromic cases. Complete removal is advised in recurrent cases. Prognosis is overall favorable as documented in literature. Other treatment modalities include adjuvant chemotherapy and radiotherapy or combination of surgical treatment and chemotherapy and/or radiotherapy.[2]

Malignant potential of SCTAT cannot be reliably assessed on histologic evaluation as high-risk features such as mitotic rate, lymphovascular invasion, or ovarian surface involvement are not consistently associated with poor outcomes. Malignant SCTAT may have early or late recurrences decades after initial presentation.[4] Thus, regular follow-up of the cases even if regarded as sporadic is warranted.

CONCLUSION

Our experience with the present case highlights the rarity of the lesion, its association with signs of hyperestrinism, awareness of the characteristic histomorphological appearance along with immuno-histopathological studying in arriving at an accurate diagnosis.

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