MAR Pathology & Clinical Research (2023) 2:6

Case Report

Ovarian Sex Cord Tumour with Annular Tubules in a 52-Year-Old Female: A Case Report

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Received: 12 October 2023

Published: 01 November 2023

Abstract

Sex cord tumor with annular tubules (SCTATs) is a relatively rare ovarian neoplasm often having a syndromic association with Peutz-Jeghers syndrome (PJS). Other associations described with this rare neoplasm include adenoma malignum of cervix, Turners syndrome, dysgerminoma, gonadoblastoma, endometrial carcinoma, and endometriosis of fallopian tube. We describe the incidental detection of SCTAT. Non-PJS SCTATs tend to be larger and could be more prone to distant metastasis, warranting subsequent follow-up.

Introduction

Sex cord tumor with annular tubules (SCTATs) is a rare ovarian tumor first described in 1970 by Scully [1]. It accounts for only six % of sex cord stromal tumors, which in turn account for only 8% of the overall ovarian neoplasms [1]. Since Scully described it, numerous case reports and a few series have contributed to the better understanding of this rare morphological entity. There is a strong association of ovarian SCTAT and Peutz-Jeghers syndrome (PJS). In a series by Young et al., of 74 cases one third of patients with SCTAT, 27, had PJS [2]. SCTAT has been documented to be an estrogen-progesterone-secreting tumor with low malignant potential. Other associations reported with SCTAT include adenoma malignum of cervix, Turners syndrome, dysgerminoma, gonadoblastoma, endometrial carcinoma, and endometriosis of fallopian tube [2–6].

Case History

A 52-year-old lady presented with abdominal pain and DUB for the past one year. On examination a left sided pelvic lump was palpable which was confirmed as fibroid on ultrasound. On USG scan a well circumscribed mass was identified measuring 10 × 10 cm . No lymph nodes or pelvic extension was noted suggestive of benign etiology.

Panhysterectomy was performed for fibroid to relieve symptoms., with preoperative assessment. Grossly the specimen showed 8 cm intramural fibroid with ovaries of 1x1x0.5cm. Grossly the ovaries were unremarkable.

Ovarian tumor markers, including carcinoembryonic antigen, carbohydrate antigen 19-9, carbohydrate antigen 125, alpha fetoprotein, and human chorionic gonadotropin, were within the normal range. Serum estradiol levels were within normal limits done after the tumor detection.

The patient was reexamined and reinvestigated to discard Peutz-Jeghers syndrome. General physical examination and gastrointestinal endoscopy ruled out syndromic SCTAT. Thus a final diagnosis of unilateral sporadic sex cord stromal tumor with annular tubules was given. No evidence of extraovarian extension/metastasis was noted.

On microscopy, the tumour had ring-shaped tubules which were separated by stroma. The cells had small single nuclei and pale cytoplasm. Within the tubules, the cells were arranged in interconnecting configuration, and at the centre of each of these, there was a core of eosinophilic materials (Fig. 1). The nuclei of the cells were palisading both around the hyaline cores and at the periphery of the tubules. There was no necrosis and the capsule was free from the tumour. Other ovary was unremarkable. On immunohistochemistry calretinin (Fig. 2) WT1 (Fig. 3) and inhibin (Fig. 4) were strongly positive. The diagnosis of SCTAT was confirmed.

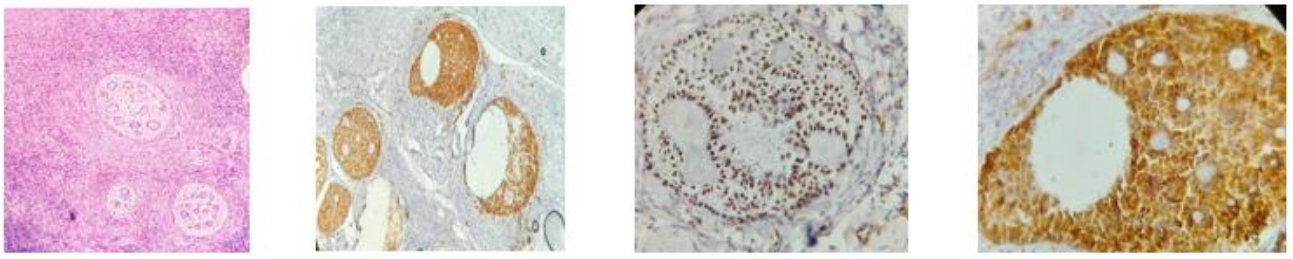

Figure 1

(a) The tumor had ring shaped tubules which were separated by stroma. The cells had small single nuclei and pale cytoplasm. Within the tubules, the cells were arranged in interconnecting configurations ans central eosinophillic material. [HE, 40x]. (b) Immunohistochemistry showing tumor cells positive for calretinin [immunohistochemistry with DAB as chromogen, 40x]. (c) Immunohistochemistry showing tumor cells positive for WT1 [immunohistochemistry with DAB as chromogen, 40x].(d) Immunohistochemistry showing tumor cells positive for inhibin [immunohistochemistry with DAB as chromogen, 40x].

Discussion

The sex cord tumor with annular tubules (SCTATs) is a distinctive ovarian neoplasm, the predominant component of which has morphologic features intermediate between those of the granulosa cell tumor and those of the Sertoli cell tumor; focal differentiation into either granulosa cell or Sertoli cell tumor may occur. Ultrastructurally, Charcot-Bottcher filament has been noted in some tumors supporting Sertoli cell origin while being absent in some. It was then suggested that SCTAT associated with PJS is a hamartoma while that without is granulosa cell tumor [7].

The majority of patients reported in various series with SCTAT have been in the reproductive age group. Rare pediatric cases have been reported [8]. The clinical manifestations of SCTAT are mainly due to estrogen-progesterone secretion like: menorrhagia, postmenopausal bleeding, precocious puberty, and sterility thus detected usually earlier than ovarian epithelial neoplasms [2, 8–10]. In the present case the patient was in middle age group and though did not have any clinical or biochemical features of high estrogen production; she had symptoms related to the fibroid.

SCTATs have a strong association with PJS and thus can be broadly classified as those with and those without PJS [8]. In the present case no features of PJS were identified. Those associated with PJS tumors are usually benign, multifocal, bilateral, very small, or even microscopic in size and calcified [2]. The ones without PJS are usually unilateral and larger in size [2, 8].

In PJS-associated SCTAT, the serum tumor marker inhibin and immunohistochemical markers inhibin, estrogen receptor, progesterone receptor, and androgen receptor have been studied as diagnostic tools, though morphology is the gold standard [8]. Non-PJS-associated SCTATs also commonly secrete estrogen and progesterone, although the sensitivity is low. In the present case inhibin was strongly positive in the tumor nests along withWT1 and calretinin; serum estrogen levels were normal in our case.

In addition to its association with PJS, many other coexisting conditions have been reported, mostly with single case reports. These include adenoma malignum of cervix, Turners syndrome, dysgerminoma, gonadoblastoma, endometrial carcinoma, and endometriosis of fallopian tube [2–6]. The latter two conditions can be explained by estrogen production by many of these tumors.

Though malignancy and distant metastasis are known to occur in 20% of non-PJS SCTAT, these tumors are considered to be a tumor with low malignant potential. PJS-associated SCTATs are usually benign [2]. The exact behavior and long-term prognosis in these patients are almost unknown and thus the present

management guidelines are based on a scanty literature available, which suggests that surgery alone with preservation of fertility (if possible) should be attempted in these patients. A school of thought suggests that management strategies in these patients can be generalized as per the management of granulosa cell tumors [8]. In the present case, panhysterectomy was already done and tumor was incidental microscopic finding, follow up will be done.

To summarize, a rare case of microscopic SCTAT is presented and its diagnosis and management have been highlighted.

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