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Case Report

Cytomorphology of Sex Cord Stromal Tumor with Annular Tubules with Histopathological Correlation: A Rare Case Report

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Abstract

Sex Cord Tumor with Annular Tubules (SCTAT) is a rare type of ovarian sex cord-stromal tumor which accounts for <1% of all ovarian sex cord stromal tumors. Cytologic features of this tumor have been described very rarely in the literature. Here we present a rare case of SCTAT on fine needle aspiration cytology with histological correlation.

Keywords: Sex cord tumor with annular tubules; Cytology; Ovary; Fine needle aspiration

Abbreviations

SCTAT: Sex Cord Stromal Tumor with Annular Tubules; FNAC: Fine Needle Aspiration Cytology

Introduction

FNAC is less often used for the diagnosis of ovarian tumours by clinicians [1]. Sex Cord Tumor with Annular Tubules (SCTAT) is a rare type of ovarian sex cord-stromal tumor that differs from other sex cord-stromal tumors in its distinctive histomorphology, epidemiology, and clinical course. This tumor was first reported by Scully. It accounts for <1% of all sex cords stromal tumors and typically occurs in patients in their third or fourth decades of life, but wider age ranges have been reported (4 years to 74 years) [2-4]. Cytologic features of this tumor have been described very rarely in the literature. Herein, we report a case of SCTAT on FNAC along with cyto-histologic correlation.

Case Presentation

A 10 year old girl presented with complaints of abdominal distension and bleeding PV since 3 months. On physical examination, a large abdominopelvic mass was palpated on the left side. She had precocious puberty because of her increased estrogen levels. She has no other significant medical or family history. All tumor markers were within normal limits. On ultrasound a large well defined multicystic mass measuring 10.6 cm × 7.9 cm × 6.5 cm seen in left adnexa (Figure 1). On MRI a well-defined large left ovarian multicystic lesion with multiple enhancements with thick & thin internal septations suggestive of malignant sex cord tumor-likely granulosa cell tumor was given.

USG guided FNAC was done and smears were highly cellular consisting of sheets and clusters of tissue fragments composed of numerous well-formed tubular structures, comprised of approximately 25-50 cells containing well-delimited central glassy pink material, which were seen in nearly all tissue fragments. The cells were uniform with indistinct cytoplasmic borders and amphophilic, somewhat granular cytoplasm. Nuclei were round to oval and tended to overlap, producing three dimensional areas. Most nuclei contained finely granular chromatin (Figures 2 and 3). Occasional nucleoli were also noted. Nuclear grooves were absent. No mitosis or necrosis was seen. Based on these findings a differential diagnosis of granulosa cell tumor and sex cord stromal tumor with annular tubules was given.

Left sided Salpingectomy was performed. On gross examination of the surgical specimen showed multicystic ovarian mass measuring 13 cm × 9 cm × 4 cm with thin and thick septations and serous fluid was drained. No solid areas were seen. On histopathologic examination the tumor showed the characteristic features of SCTAT, with simple and complex annular tubules containing cells displaying the typical nuclear antipodal arrangement and surrounding basement membrane-like hyaline material, within a hyalinized stroma (Figures 4 and 5). No necrosis or lymphovascular invasion was identified. No mitoses were identified.

Discussion

Sex Cord Tumor with Annular Tubules (SCTAT) is a rare, distinctive ovarian neoplasm. Scully named this tumor SCTAT to distinguish it from other sex cord tumors because of its distinct morphology and association with Peutz-Jeghers syndrome but only rarely occurring in the general population [2]. There are two clinical forms: one occurs in association with PJS which are usually bilateral, multifocal tumors, less than 3 cm, invariably benign and not associated with hyperestrogenism. The other form not occurring in association with PJS differ in that they are usually unilateral, unifocal, and larger than 3 cm, and sometimes behave in a malignant manner, with lethal metastases or recurrences [4]. In our case the tumor was unilateral, multifocal, and greater than 10 cm with features of hyperestrogenism

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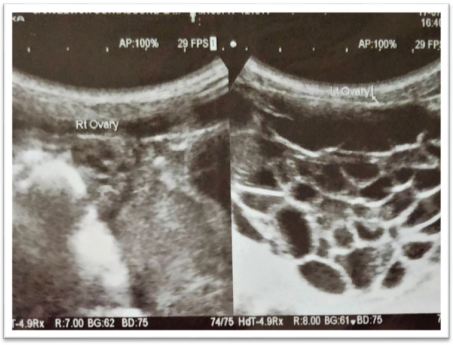

Figure 1: USG showing a large well defined multicystic mass measuring 10.6 cm × 7.9 cm × 6.5 cm seen in left ovary.

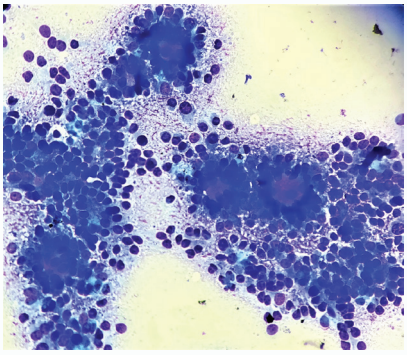

Figure 2: Smears show many tubules with central pink hyaline material. Cells show moderate amount of cytoplasm with round nucleus and fine granular chromatin. (× 40).

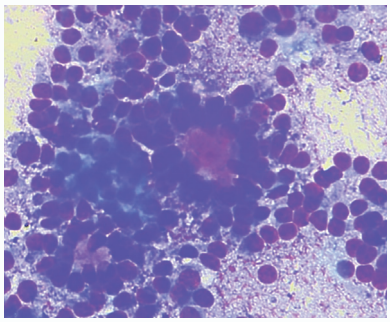

Figure 3: Giemsa smears show tubules with approximately 25-50 cells and central pink hyaline material. Cells are uniform with round to oval nuclei and indistinct amphophilic cytoplasm. Nuclear overlapping is seen (× 40).

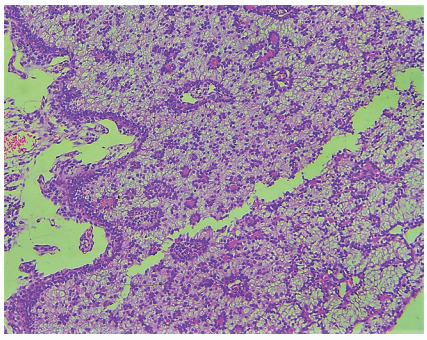

Figure 4: H&E section showing ring like annular tubules showing antipodal nuclear arrangement with eosinophilic basement membrane like material with intervening stromal cells (× 10).

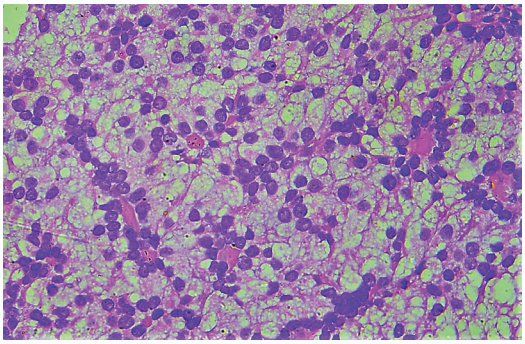

Figure 5: H&E section showing tubules with basement membrane like material. The cells are round with moderate amount of cytoplasm and fine chromatin. Few of the cells show prominent nucleoli (× 40).

and without Peutz- Jeghers syndrome. Most SCTAT are benign; however, malignancy has been reported in 20% of sporadic tumors and malignant PJS-associated SCTAT also have been reported [3,5].

SCTAT has morphologic features intermediate between those of granulosa cell tumors and Sertoli cell tumors; therefore, it has been interpreted as a Sertoli cell tumor by some observers and a granulosa cell tumor by others [6]. The major differential diagnosis on cytology is granulosa cell tumor from which it can be differentiated by poorly-formed Call-Exner body-like rosettes composed of 6 cells to 15 cells [7,8], whereas the tubules in this case were well formed with 25-50 cells.

In our case of SCTAT, in the center of each tubular structure contained well-delimited glassy pink material. In cases of GCT, however, the central area of Call-Exner body-like rosettes contains homogeneous eosinophilic or amorphous pink substance [7]. This material is less dense and less well-delimited than that observed in this SCTAT. The cells seen in cytologic preparations of GCT typically contain scant amounts of cytoplasm, and their nuclei show occasional or frequent nuclear grooves [9]. In our case the cells had moderate amount of cytoplasm and nuclear grooves were absent.

The cytologic features of Sertoli-cell tumors have been reported as virtually indistinguishable from those of GCT. Fine-needle aspirates of Brenner (transitional cell) tumors may show rosettes with luminal hyaline bodies that may suggest the diagnosis of GCT. However, cells of Brenner tumors are typically larger than those of GCT, with more cytoplasm and more prominent nuclear grooves [9]. Brenner tumors are positive with immunocytochemical stains for cytokeratin and negative for inhibin, while SCTAT are negative for cytokeratin and positive for inhibin.

Histologically, SCTAT are characterized by sharply circumscribed, rounded epithelial nests creating an appearance of ring-shaped tubules encircling hyaline cores. The nests show two patterns: simple and complex tubule structures, with and without hyaline cores. The neoplastic nuclei are round, usually with small, single nucleoli, and a characteristic feature is palisading of the neoplastic cells in an antipodal arrangement along the periphery of the nests and around the hyaline cores which was similar in our case [10].

Conclusion

SCTAT is a rare, distinct subtype of ovarian sex cord-stromal tumors with well-described histomorphologic features. Cytologic features have been much less frequently described in the literature.

Recognition of this characteristic morphology observed in this case may prove helpful in diagnosing this tumor on fine needle aspiration and reminds us to keep this tumor in differential diagnosis of sex cord stromal tumors. With the advent of accurate imaging techniques like USG and CT scan, guided FNAC has assumed a definite role in diagnosis and management of these rare entities.

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