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Case Report

Pathology Section

Steroid Cell Tumor of the Ovary in An Adolescent: A Rare Case Report

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ABSTRACT

Steroid cell tumors are rare sex-cord stromal tumors. Even though benign, these tumors have a risk of malignant transformation. Pathologic evaluation is necessary to rule out malignancy. Clinical, surgical and microscopic correlation along with a regular follow-up is necessary.

Keywords: Amenorrhoea, Sex-cord stromal tumor, Virilisation

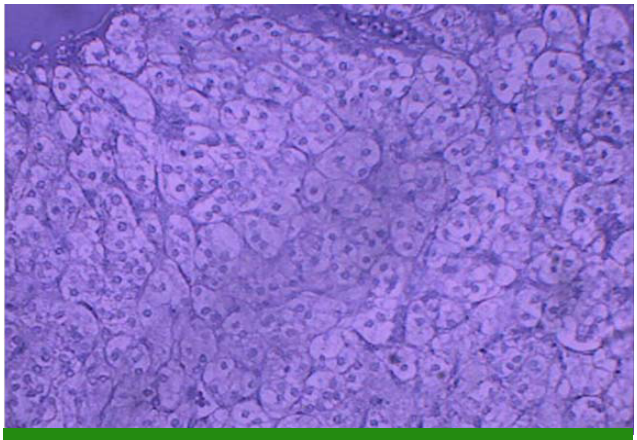
CASE REPORT

A 20 year-old girl was admitted to our hospital with symptoms of hoarseness of voice, abnormal hair growth, weight gain and amenorrhoea. She attained menarche at the age of 14 years and gives the history of amenorrhoea for past 7 months. Physical examination revealed a male pattern of hair growth in the face, legs, arms and abdomen; her breast development was normal. Abdominal examination revealed the presence of a vague mass in the right iliac fossa.

Ultrasonography showed a hyperechoic mass lesion measuring 6.6x5.3cm in the right ovary with irregular hypoechoic component and minimal ascites. Computerized tomography revealed a heterogeneously enhancing well defined lobulated solid soft tissue mass lesion in the right adnexa measuring 5.9x5.5x5.2cm and suggested the possibility of a germ cell tumor. Adrenals were normal and there was no other organomegaly. Haematological and biochemical investigations were within normal limits. Her beta hCG level was negative, alpha-fetoprotein was within normal limits. CA 125, LDH and testosterone levels were 38.14 IU, 214 U/L and 94 ng/l respectively; all three of them were minimally elevated. A clinical diagnosis of ovarian tumor was made.

An informed consent was taken from the patient for the surgery and the histopathological evaluation. The patient underwent an exploratory laparotomy, because of the possibility of malignancy; right oopherectomy was performed. The specimens were sent to the Department of Pathology. Macroscopic examination showed a well circumscribed tumor measuring 6.5x4.2x3cm. The neoplasm had a lobulated, solid, irregular, yellowish cut surface [Table/Fig-1]. Microscopic examination showed a well circumscribed tumor comprising of oval to round cells arranged diffusely and in nests separated by vascular stroma. The tumor cells have well defined borders, round bland nucleus and granular to vacuolated cytoplasm.

[Table/Fig-1]: Macroscopic image - Well circumscribed solid lobulated mass with yellowish cut surface



[Table/Fig-2]: Microscopic image showing oval to round cells with well defined borders, round bland nucleus and granular to vacuolated cytoplasm (H & E X 400)

The stroma at places showed mucoid material [Table/Fig-2]. Histopathological diagnosis was steroid cell tumor of right ovary.

Postoperatively, her complaints regressed and she reattained regular menstrual cycles by 4th month. The patient is followed up closely and regularly with measurement of hormone levels and pelvic ultrasound.

DISCUSSION

Steroid cell tumors are rare sex-cord stromal tumors and they account for only 7% of all primary ovarian tumors [1]. The incidence as per literature is 0.2%. These patients are usually of younger age group and presents with a rapid and sudden history of menstrual irregularity and virilisation symptoms. These tumors develop from sex cord and stromal components of the gonads [2].They include tumors composed of granulosa cells, theca cells, sertoli cells, leydig cells and fibroblasts of stromal origin, singly or in various combinations [3]. These tumors have been classified in three subtypes: NOS, leydig cell and stromal leuteoma [3].These are functional tumors which produce hormones that can induce development of male features (virilisation) in girls [4]. These tumors should be considered in the differential diagnosis of isosexual precocious puberty in children and virilisation in adults.

Steroid cell tumors are generally unilateral and benign and are composed of tumor cells with round to polygonal eosinophilic or vacuolated cytoplasm and centrally located nuclei that may contain prominent nucleoli. The stroma is typically sparse, consisting of delicate connective tissue containing a rich vascular network [5]. These tumors are generally benign but there is a risk of malignant transformation in one quarter of these tumors [2]. Hayes and Scully have described five pathologic features to asess malignancy in these tumors which include size greater than 7 cm in diameter, areas of haemorrhage or necrosis, moderate to marked nuclear atypia, and a mitotic index of 2 or more per 10 HPF [5,6].

Testosterone and DHEA-S levels should be measured for the evaluation of androgen excess. Testosterone levels above 200 ng/dl are the diagnostic threshold for discrimination of androgen secreting tumors and non neoplastic lesions. CA-125 is a useful biomarker and a value more than 35 IU/ml is significant [7]. To rule out an adrenal tumor magnetic resonance imaging is useful. Surgery is the most important and hallmark treatment. Excision of the tumor results in regression of symptoms and disappearance of virilising effects. This case merits presentation because of its rarity and symptoms becoming significant in a 7-month time, in contrast to literature.

CONCLUSION

Steroid cell tumors are very rare and the incidence as per literature is 0.2%. When a patient presents with androgenic symptoms and associated increased testosterone levels it should raise the suspicion of a functional ovarian tumor. Since, there is a risk for malignancy in these tumors, pathologic evaluation is warranted.

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