

Steroid Cell Tumour: NOS of The Ovary in A Young Female

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ABSTRACT

Sex cord stromal cell tumours constitute 5-8 % of all ovarian neoplasms. Steroid cell tumours are a type of sex cord stromal cell tumours. The name “Steroid cell” stands for the morphology and the functionality of these tumours. Steroid cell tumours- Not otherwise specified constitute about 56% of all steroid cell tumours and presents more commonly with androgenic manifestations in third to fourth decade.

We present a case, 22 year old, who presented with virilizing symptoms. MRI was suggestive of an right ovarian mass for which right salpingo-oophorectomy was done. Histopathology revealed features classical of Steroid cell tumour- Not otherwise specified. We present the case for a relatively younger age at presentation and the classical histomorphology.

Keywords: Virilizing, Steroid Cell, Young

Introduction

Steroid cell tumour- NOS is a type of sex cord stromal cell tumour of the ovary. It presents usually in the third or the fourth decade of life with virilizing symptoms. We present a case of a relatively younger female, 22 year old, who presented with virilizing symptoms. Microscopy of the ovarian tumour resected showed features classical of Steroid cell tumour- NOS type. We present this case to emphasize on the consideration of this entity in the clinical scenario of rapid onset of virilizing symptoms in a female and also to document it's occurrence at a younger age than the classical description in the literature.

Case Report

The patient, 22 year old, married since 3 years presented with history of primary infertility and excessive hair growth with change in the voice. She had history of oligomenorrhea and noted decrease in the size of the breast. There was no other significant past medical or surgical history. On general examination, vital parameters were within normal limits. Hirsutism was present. Genital examination revealed the presence of Clitoromegaly. Laboratory investigations revealed S. Androstenedione levels of more than 10ng/ml (Range: 0.3-2 ng/ml) and the level of S. Testosterone was 6.79 ng/ml (Range: 0.15-0.7 ng/ml). S. FSH level was 6.8 mIU/ml (Range: 2.3-18.5 mIU/ml), S. LH level was 7.95 mIU/ml (Range: 0.4-105 mIU/ml) while S. ACTH level was 21.2 pg/ml (Range: 5-27 pg/ml). MRI Pelvis revealed right ovary measuring 4.6 X 4.4 X 3.4 cm which showed a focal lesion in the ovarian stroma. The left ovary was normal. Patient underwent right salpingo-oophorectomy with left ovarian drilling.

On Gross examination, the right ovary measured 5 X 4 X 2 cm. Externally, the ovarian surface was congested at places. Cut section revealed a bright yellow mass measuring 3.5 X 2 cm, which was soft to feel. Normal ovarian parenchyma could be appreciated in the periphery. (Figure 1) The right fallopian tube measuring 3.5 cm in length was also received which was normal.

Microscopy of the yellow mass revealed a well circumscribed tumour which predominantly showed medium to large sized polygonal cells in sheets and clusters separated by fibrous septae which gave a lobulated appearance. (Figure 2) Individual cells showed small nuclei, inconspicuous nucleoli and abundant pale vacuolated cytoplasm. (Figure 3). No reinke crystals could be identified. There was no evidence of atypical mitosis and necrosis in the sections studied. Normal ovarian parenchyma could be identified at the periphery.

On the basis of the classical histomorphology and the supportive hormonal studies, a diagnosis of steroid cell tumour- Not otherwise specified was made.

Post operatively, the serum testosterone levels returned back to normal levels. S. Testosterone level was 0.54 ng/ml. (Range: 0.15-0.7 ng/ml) and the patient improved symptomatically. This confirmed the source of excess androgen to be the ovarian tumour.

Discussion

Steroid cell tumours are rare ovarian neoplasms. They account for approximately 0.1 % of all ovarian neoplasms. They are classified into three types- stromal luteoma, Leydig cell tumour and Steroid cell tumour- Not Otherwise

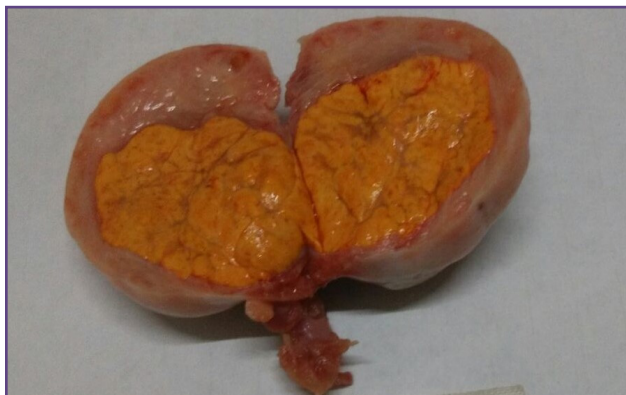


Fig. 1: A well circumscribed yellowish solid tumour and the surrounding ovarian parenchyma seen.

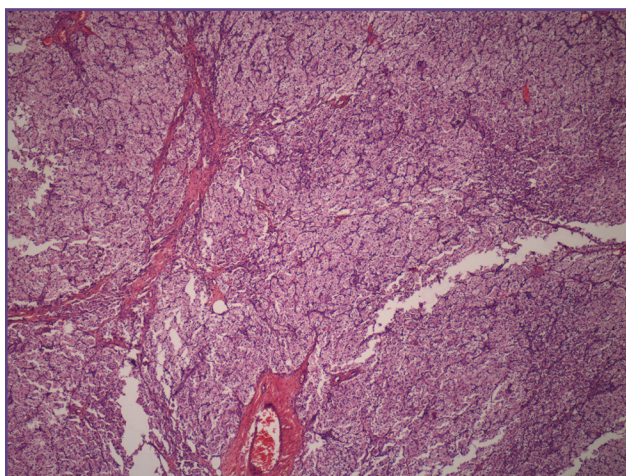


Fig. 2: Tumour cells are arranged in sheets, clusters in a lobulated fashion, separated by thin fibrovascular septae (HE X 40).

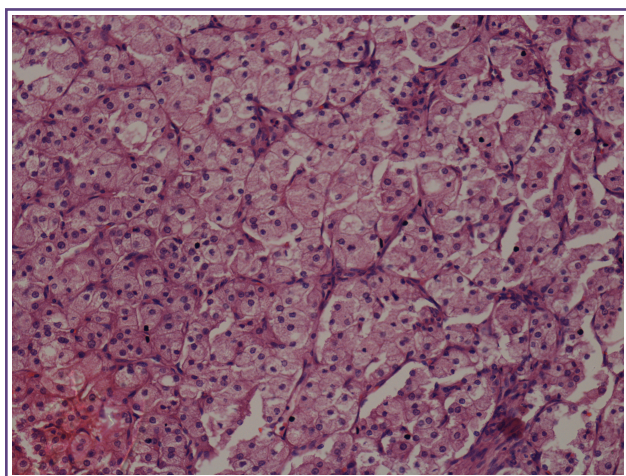


Fig. 3: Individual tumour cells are round to polygonal with abundant clear cytoplasm and small round nuclei. (HE X 400).

Specified(NOS). Steroid cell tumour- NOS accounts for about 56% of all steroid cell tumours [1]. They commonly present with virilizing symptoms in the third or the fourth decade of life. Androgenic manifestations associated with this tumours are attributable to hormones like androstenedione, α -hydroxyprogesterone, and testosterone secreted by the tumour cells [2-3]. However, rarely the diagnosis of such tumours can be incidental when the patients do not develop any of the above symptoms. Our case also presented with virilizing symptoms. However, the age of presentation was slightly younger than the age classically described.

Most of these tumours are unilateral and can vary in sizes from 1.2 to 45 cm [4]. They are usually solid and yellow to brown in colour. Rarely, the tumours may show cystic areas. Our case showed a bright yellow tumour with solid consistency. Histomorphology shows polygonal cells arranged in sheets and clusters which show small nuclei, inconspicuous nucleoli and abundant vacuolated cytoplasm. Classical histomorphology was seen in our case on the basis of which diagnosis was made.

Steroid cell tumours are usually positive for inhibin and stain variably with anti- cytokeratin antibodies and vimentin. However, Immunohistochemistry does not help to differentiate between the various types of sex cord stromal cell tumours and the distinction is mainly made on the basis of the histomorphology. As our case showed classical histology of Steroid cell tumour- NOS, Immunohistochemistry was not required.[5]

Steroid cell tumours are usually benign in behavior. However, large tumours with hemorrhage and necrosis, tumours showing considerable nuclear atypia and mitosis are more commonly malignant in behavior. [5]

Differential diagnosis of this tumour includes leydig cell tumour and stromal luteoma. Tumour cells in Leydig cell tumour show dense eosinophilic cytoplasm [6] while stromal luteoma is associated with stromal hyperthecosis and shows degenerated vascular spaces [7]. Our case did not show the presence of reinke crystals nor was there evidence of stromal hyperthecosis and degenerated vascular spaces. Hence, the two other possibilities were also ruled out and a diagnosis of Steroid cell tumour- NOS was made.

Post operatively, S. Testosterone levels decreased and the patient improved symptomatically, confirming the source of excess androgen to be the ovarian tumour.

Treatment of these tumours is decided on the basis of the staging, the extra- ovarian spread of the tumour and the desire to preserve fertility [8,9]. Unilateral salpingoophorectomy is the treatment of choice in young patients, as they are

desirous of preserving fertility^[10]. The same was done in our case.

We present this case for the rarity of this type of ovarian tumour and also its presentation at a relatively younger age.

Conclusion

Steroid cell tumours- Not otherwise specified is a rare ovarian tumour which presents with signs and symptoms of androgen excess. A diagnosis of Steroid cell tumour – NOS should be considered apart from other virilizing ovarian and adrenal tumours in young patients presenting with a rapid onset of virilization.

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