



**Steroid cell tumour of the ovary (Not Otherwise Specified subtype) -
A rare case report**

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Abstract :

Ovarian steroid cell tumours are very rare sex cord stromal tumours comprising 0.1 percent of all ovarian tumours. These tumours are sub classified as stromal luteomas, leydig cell tumours and steroid cell tumour not otherwise specified (NOS). These tumours are interesting as they produce various hormones like estrogen, testosterone, and cortisol. Occasionally these tumours develop in childhood causing precocious puberty but they principally occur in perimenopausal and postmenopausal women with varying degrees of androgenic and estrogenic features. We present one such case of steroid cell tumour of ovary for its rarity. A 42 year old perimenopausal woman presented with complaints of increased body hair growth and balding past 2 years. On examination there was increased hair growth over the face, chest, abdomen and limbs with temporal balding and clitoromegaly.

Examination and radiological imaging revealed a 5.5cm mass confined to Left ovary with no extension to other pelvic organs or abdomen. Her free testosterone was elevated to 6.0ng/ml with normal CA-125. A provisional diagnosis of a virilizing ovarian tumour was made. Subsequently staging laparotomy was done and HPE report revealed Steroid cell tumour of ovary (NOS). Stromal luteomas and Leydig cell tumors are almost invariably unilateral and benign. Steroid cell tumour of ovary (NOS) accounting for 60 of the three tumours have malignant potential. Hence thorough staging laparotomy and periodic follow up with serial hormonal assays are required.

Keyword : steroid cell tumor, ovary, hormone

Introduction:

Ovarian steroid cell tumours are very rare sex cord stromal

tumours comprising < 0.1% (1) of all ovarian neoplasms. These tumours are interesting as they produce various steroid hormones like oestrogen, testosterone and cortisol. Occasionally these tumours develop in childhood causing precocious puberty but they predominantly occur in perimenopausal and post-menopausal women with varying degrees of androgenic and estrogenic features (2). The subtype, *not otherwise specified (NOS)* typically present in premenopausal women with symptoms of androgen excess in more than 50% of the patients and extremely elevated testosterone levels (3). So far only about 74 cases of steroid cell tumours have been reported in the literature (4). We report one such case here for its rarity and discuss its diagnosis, classification, and treatment options.

Case report: History & Evaluation:

A 42 year old perimenopausal woman presented with complaints of increased body hair growth and frontal and temporal balding for 2 years duration. She had no history of any chronic drug intake. Her menstrual history was normal. She has 2 live children in good health.

On examination:

Patient was moderately built and nourished. No generalized lymphadenopathy. There was

figure 2 showing increased body hair

Breasts were normal. Genital examination revealed clitoromegaly. Per abdomen was soft. Per vaginal examination revealed a normal sized uterus and a solid mass 5×4 cm palpable in the left adnexa which was mobile and non-tender.

Investigations:

Treatment:

We made a provisional diagnosis of masculinizing ovarian tumour and proceeded with staging laparotomy and histopathological analysis. consistency. Right ovary was normal. No obvious peritoneal or omental deposits

We did a Total abdominal hysterectomy and bilateral salpingo oophorectomy and partial omentectomy.

Histological analysis:

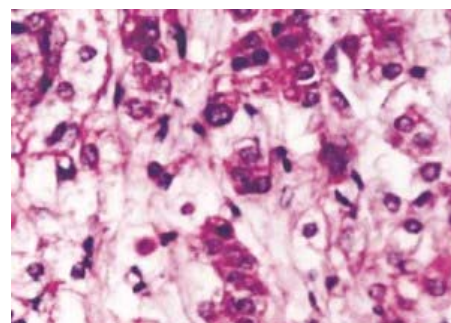


figure 3: Histological section of the tumour showing characteristic eosinophilic and vacuolated appearance

Cut section of the tumour mass revealed solid yellow areas. Microscopically, the specimen showed eosinophilic and vacuolated appearance of the cellular structure which are characteristic of steroid cell of the ovary, not otherwise specified subtype. (fig 3) Post operatively serum testosterone of the patient has fallen within normal range 1.01 pg/ml (Normal 0.02-3.09 pg/ml) in 6 weeks period. The masculinizing features had started to regress and the patient is still on follow up.

Discussion:

Steroid cell tumours are very rare sex cord stromal tumours comprising only less than 0.1 percent of all ovarian neoplasms (1). In the early literature, the distinct group of ovarian neoplasia composed of cells resembling typical steroid hormone-secreting cells (lutein cells, Leydig cells, and adrenal cortical cells)

increased hair growth all over the body with frontal and temporal hair loss.(fig1, 2)



figure 1 showing Masculanizing facial features - frontal and temporal baldism and facial hair



figure 2 showing increased body hair

Breasts were normal. Genital examination revealed clitoromegaly. Per abdomen was soft. Per vaginal examination revealed a normal sized uterus and a solid mass 5×4 cm palpable in the left adnexa which was mobile and non-tender.

Investigations:

CA- 125 was 10U/ml; Serum testosterone- 6 pg/ml (Normal 0.02-3.09 pg/ml)

Ultra sonogram and CT of the abdomen and pelvis revealed a 5× 5 cm solid mass confined to left ovary with no extension to other pelvic organs or abdomen. No pelvic or Para-aortic lymph node enlargement was present.

has been called with different names, such as adrenal rest tumours, adrenal-like tumours, masculinovoblastoma, lipid cell tumour and lipid cell tumour (5). The first three terms have not been in vogue in the more recent literature, as most of these tumours never exhibit any demonstrable fat (6,7), also the last two terms are clearly inaccurate to illustrate this group of neoplasia. The term steroid cell tumour was proposed in the 1964 by Scully (8), to more accurately describe both the morphological features of the tumour cells and their intrinsic property of synthesizing steroid hormones.

Together with some other important tumours (most notably granulosa cell tumours, Sertoli-Leydig cell tumours, adrenal tumours, and unclassified sex cord tumours) the steroid cell tumours belong to the group of virilizing ovarian tumours. In the most recent WHO classification, they are classified based on their histopathology as stromal luteomas (20%), leydig cell tumours (15 to 20%) and steroid cell tumour -not otherwise specified (60%). They are histologically identified based on their absence of pathognomonic features, such as Reinke Crystals,)

Call-Exner bodies, and prominent nucleoli, O,p'-DDD could be considered as an alternative. For both schedules there is no high level of evidence.(10)

The typical presentation of steroid cell, NOS, occurs in premenopausal women with a mean age of 43 and frequently manifests with virilization. More than 80% of the patients have androgenic features and 25% will have hyperoestrogenic symptoms. Hyperoestrogenic features can be due to either direct oestrogen production by the tumour or due to the peripheral aromatisation of androstenedione and testosterone to estradiol in adipose tissue. A comprehensive workup, therefore, includes the evaluation of an adrenal and ovarian source of pathology for the hyperandrogenism. Elevated testosterone levels with normal DHEA, DHEA-S, LH, FSH, and 17-OHP levels warrants abdominal imaging with ultrasound, CT, or MRI of the pelvis to look for an ovarian virilizing tumour(9). Primary management for steroid cell tumour (NOS subtype) is surgery. Staging laparotomy is done and treated according to the stage. Stage Ia who desire for future fertility are done unilateral salpingo-oophorectomy as only 6% of cases are bilateral and all are mostly benign in nature. Stage Ia with completed family, Stage Ib to IV are treated with TAH with BSO with thorough staging. Serial hormone assays are required as 40% of NOS type are malignant. Adjuvant chemotherapy is indicated for higher stages and those with features suggestive of malignancy. Features that were strongly associated with malignancy are an older age (mean age 54 versus 38 years in those with benign disease), tumour size greater than 7 cm in diameter, a mitotic rate of >2 mitosis per 10 high-power fields, and the presence of necrosis (10). GnRH agonists and chemotherapy comprising of a combination of bleomycin, etoposide and cisplatin (BEP) could be used as first-line treatment in malignant steroid cell tumours.

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