



Case Report

Role of Histopathological Analysis in Diagnosing Steroid Cell Tumour: A Case Report

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ABSTRACT

Steroid cell tumour of the ovary is an extremely rare tumour. They are classified into three types with the commonest, the steroid cell tumour not otherwise specified (NOS) consisting approximately 60% of them. They occur mostly in middle age, are mostly benign and mostly cause virilising symptoms though a small proportion can present with oestrogenic effects. Diagnosis is based on presence of symptoms, presence of specific histopathological features with absence of certain histopathological features and positive specific immunostaining results. Here the author reports a case of a 42-year old woman with abdominal mass but without virilising symptoms but on whom a diagnosis of ovarian steroid cell tumour was reached by histopathological analysis which revealed presence of specific histopathological features with absence of certain histopathological features.

Keywords: *Histopathological features, Ovary, Steroid cell tumour, Virilisation.*

INTRODUCTION

Ovarian steroid cell tumours belong to the class of ovarian tumours called the sex cord-stromal tumours. They are very rare constituting about >0.1% of all ovarian tumours¹ but form more than half of the sex-cord stromal tumours. These tumours were formerly called lipid or lipoid cell tumours. They are characteristically benign and unilateral.^{2,3} The tumours are classified into 3 according to cell of origin namely stromal luteoma from ovarian stroma, Leydig cell tumor from Leydig cells, and steroid cell tumour, not otherwise specified (NOS).¹ The last constitutes approximately 60 % of all steroid cell tumour with about 1/3 of them being malignant.¹ Because these tumours are extremely rare, the preponderance of literature on them is case reports. Here the author reports a case of a 42-year old woman with abdominal mass, without virilising symptoms but on whom a diagnosis of ovarian steroid cell tumour was reached by histopathological analysis which revealed presence of specific histopathological features with

absence of certain histopathological features. A review of literature is also presented.

CASE PRESENTATION

A 42-year-old multiparous female presented in the Gynaecology clinic of our hospital, with a 2-month history of abdominal distension and associated pain and loss of appetite for fifteen days. The patient reported no other associated symptoms. Abdominal examination revealed distension, a mass in the left lumbar region and associated mild ascites. Examination of the vagina revealed no pathology. Abdominal ultrasound revealed the mass as having solid and cystic parts. The uterus was of normal uterine size but had two small subserosal fibroid in the anterior wall. There was free fluid in the peritoneal cavity. An exploratory laparotomy was performed alongside total abdominal hysterectomy (TAH), and bilateral salpingo-oophorectomy, (BSO). Pelvic and mesenteric

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lymph nodes were also harvested. The specimens were submitted to the histopathology laboratory of our hospital for analysis.

Gross examination by histopathologist revealed a firm yellow-coloured tumour on the left ovary which measured $10.4 \times 5.8 \times 3.2$ cm in size with a lobulated surface. Cut sections revealed yellow surface with solid and cystic parts. Haemorrhage and necrosis were not seen nor was there capsular invasion (Figure 1).



Figure 1: Gross picture of tumour showing yellowish, lobulated surface

Carefully sampled blocks of tissue were taken from both the solid and cystic parts for processing. Apart from two small subserosal fibroids on the uterus, the rest of the uterus, the fallopian tubes and the right ovary were grossly normal. Harvested lymph nodes were also processed. Light microscopy revealed sheets, nests or cords of medium to large round to polygonal cells with abundant granular, eosinophilic cytoplasm with a small eccentric nucleus and intervening thin fibrovascular stroma (Figure 2).

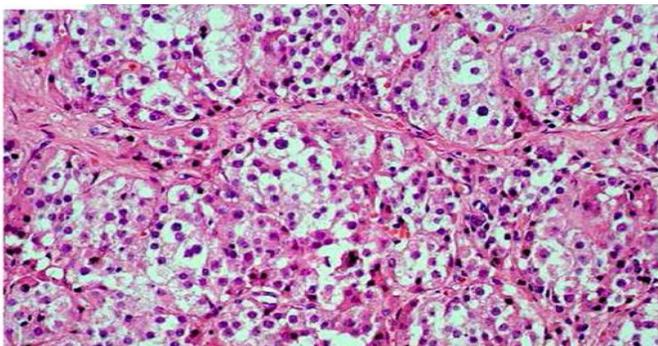


Figure 2: Photomicrography of sheets of benign tumour cells with abundant eosinophilic cytoplasm, central nuclei. Intervening stroma is also evident.

Neither Reinke crystals nor Call-Exner bodies nor prominent nucleoli were seen. Mitosis, haemorrhage and necrosis were also absent. Periodic acid-Schiff reaction was negative excluding glycogen. The sampled lymph nodes showed reactive changes.

Histopathological features observed supported a diagnosis of benign ovarian steroid cell tumour not otherwise specified (NOS).

DISCUSSION

Historically, tumours now called steroid cell tumours were called lipid cell or lipid tumours but this was changed after Hayes and Scully¹ proposed the name steroid cell tumours after finding that >25% of the tumour did not contain lipid. Steroid cell tumours typically occur in the 5th (steroid cell tumours NOS) and 6th (stromal luteoma and Leydig cell tumour) decades of life average age being 43 years and 58 years respectively. All three types may rarely, occur in prepubertal period and while steroid cell tumours NOS is rare in the postmenopausal period. The tumour varies widely in size ranging between 1cm and 45cm while 8.4cm is the average.^{3,4} The patient on whom this report is made was aged 42 years while the mass measured 10.4cm in the widest diameter.

These tumours are usually grossly well circumscribed with yellow to orange surface when sectioned which signifies the high lipid content features also seen in the case here reported. When they are lipid-poor, steroid cell tumours may range from red to dark brown in colour. They are characteristically solid but may contain cystic, necrotic and haemorrhagic areas.⁵ The present tumour has similar microscopic features with the benign subtype namely sheets, nests or cords of medium to large round to polygonal cells with abundant granular, eosinophilic cytoplasm and a small eccentric nucleus with intervening thin fibrovascular stroma. In malignant types, the cells show nuclear atypia and there is mitosis.

Generally, the diagnosis of steroid cell tumours NOS should depend on finding virilising signs, appropriate histologic feature and immunoreactivity to specific markers including inhibin and calretinin which are the ones most useful in differentiating between sex cord stromal tumour from virilising

non-sex cord stromal tumours.⁶ Other tumours to be differentiated from steroid cell tumours NOS include Leydig cell tumours, luteinized granulosa cell tumours, clear cell carcinomas, and metastatic renal cell carcinomas. Absence of pathognomonic features seen in other virilising ovarian tumour including Reinke Crystals, Call-Exner bodies, and prominent nucleoli differentiates steroid cell tumours NOS from such tumours.⁷ This was useful in diagnosing the tumour herein reported as those features were not seen. Periodic acid-Schiff which demonstrates presence of glycogen is positive in clear cell carcinomas and metastatic renal cell carcinomas but not in steroid cell tumours as was the case in this tumour.

A very important aspect of the diagnosis of steroid cell tumours is to determine if a given tumour is malignant. According to Hayes and Scully,¹ histopathologic features that highly correlate with malignancy include tumour size ≥ 7 cm in diameter (78%), presence of necrosis (86%), haemorrhage (77%), grade 2 to 3 nuclear atypia (64%) and ≥ 2 mitotic figures/10 high power field (92%). However, tumours with pathologically benign features have been known to exhibit malignant behaviour.^{8,9} It is therefore accepted that the only definite proof of malignancy is the presence of metastasis.¹⁰ The case here presented showed benign features both pathologically and biologically.

Mostly the tumours secrete hormones with androgenic effects but on occasions secrete those with oestrogenic effects or may not cause endocrine symptoms at all. About 50% of steroid cell tumours NOS tumours present with excessive androgenic symptoms which include hirsutism, virilisation, temporal alopecia, anovulation, clitoriomegaly, obesity, hypertension and impaired glucose tolerance etc.^{1,2,6} In approximately 8% of all steroid cell tumours, estrogenic effects are seen namely isosexual precocity and abnormal uterine bleeding in prepubertal girls and irregular vaginal bleeding and non-endocrine symptoms namely abdominal swelling and abdominal pains in reproductive and postmenopausal women.¹¹ The case presented in this report did not show any virilising features a feature similar to that reported by Medhi *et al.*¹²

Surgery is the accepted mode of treatment for steroid cell tumours with a range of as minimal as unilateral oophrectomy for stage I disease to as aggressive as TAH with BSO especially in women that have completed their family.^{13,7} Because of the rarity of the tumour, the low incidence of

malignancy, metastasis and recurrence, a number of issues about them are at best uncertain including clinical outcomes, the place of and guidelines for postoperative adjuvant chemotherapy are not established.^{13,14} As such regular follow-up should be an important component of the management of this condition.

CONCLUSION

Steroid cell tumours are rare and difficult to diagnose especially if classical virilising symptoms are absent. Careful histopathological analysis of submitted specimens can be very useful in diagnosis especially in resource limited settings.

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