CASE REPORT

Metachronous Ovarian Steroid Cell Tumour, Not Otherwise Specified in a previously diagnosed Invasive Breast Carcinoma Patient – A rare Case report

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ABSTRACT

Steroid cell tumours, Not otherwise Specified (SCT, NOS) are extremely rare ovarian tumours with an incidence of 0.1% of all the ovarian tumours. They are pure stromal tumours of the ovarian parenchyma composed of steroid cells. These tumours were previously called as *Lipoid cell tumour* or *Lipid cell tumour,* but as it was found later that 25% of these tumours did not contain intracellular fat, the designation *Steroid cell tumour* was proposed. Mean age of incidence of this tumour is 43 years and is usually unilateral and rarely bilateral and patients usually present with endocrine manifestations. In this study we present a case of 50-year-old female with Right ovarian mass measuring 5.5 x 5 x 4cm which on gross examination revealed a solid lesion with yellowish, brownish red haemorrhagic and necrotic areas. Microscopically the lesion was composed of sheets and cords of polygonal tumour cells in lipid rich form predominantly along with little lipid poor form. This case is presented here for its rarity of occurrence.

Keywords: Steroid cell tumor, Ovarian tumor, Stromal tumor

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INTRODUCTION

Steroid cell tumours, Not otherwise Specified are extremely rare ovarian tumours with an incidence of 0.1%. They are pure stromal tumours of the ovarian parenchyma composed of steroid cells. These tumours were previously called as Lipoid cell tumour or Lipid cell tumour, but as it was found later that 25% of these tumours did not contain intracellular fat, the designation Steroid cell tumour was proposed.1 It is usually unilateral and rarely bilateral and patients usually present with endocrine manifestations.2 Incidence of steroid cell tumour is highest in the child bearing age group mainly in the third and fourth decade of life with a mean age of 43 years (premenopausal women) and rarely occurs in post-menopausal women and children.2,3

Clinical manifestations of SCT, NOS depends on the type of hormone elaborated by the tumour, out of which 56% to 77% present with virilisation due to testosterone production, 6% to 23% with hyperestrogenemia presenting as menorrhagia and 6% to 10% with hypercortisolism leading to Cushing syndrome and 25% of SCT, NOS are asymptomatic.4

The following is a case report of a post-menopausal asymptomatic left breast carcinoma – Stage IIIA woman, who was found to have Right ovarian mass -? Likely malignant on follow up PET CT.

CASE DETAILS:

A 50-year-old, post-menopausal asymptomatic woman with history of Left carcinoma breast cT2N2M0 – Stage – III A, status post Left Modified Radical Mastectomy (which was done a year ago) / Post chemotherapy and post radiotherapy, underwent routine PET CT for status post MRM follow up and was found to have a Right ovarian mass that was radiologically suspected to be malignant. Her preoperative blood work up and physical examination appeared to be within normal limits. Hence bilateral salphingo-oophorectomy was done and the sample was sent for histopathological examination.

MORPHOLOGY:

Gross Examination:

Bilateral salphingo – oophorectomy specimen with Right ovary measuring 5.5 x 5 x 4cm with attached fallopian tube measuring 5cm long and Left ovary measuring 2.5 x 1.5 x 1cm with attached fallopian tube measuring 5cm long.

Cut surface:

Right ovary cut surface revealed solid lesion with yellowish, brownish red haemorrhagic and necrotic areas found

replacing the entire Right ovarian parenchyma. Cut surface of the Left ovary and bilateral fallopian tubes were unremarkable.

Microscopy:

Sections studied from the right ovary showed a well circumscribed tumor comprising of tumor cells in irregular cords and large nests and sheets. The tumor cells were round to polygonal and uniform in appearance with few lipid poor forms with abundant eosinophilic cytoplasm and predominantly lipid rich forms with variable sized clear cytoplasmic vacuoles, both types of cells had centrally placed round nuclei with prominent nucleoli. Also seen was extensive hemorrhage. Sections studied from the left ovary and fallopian tubes appeared unremarkable. Hence diagnosed as, **Steroid cell tumor, not otherwise specified** – Right ovary

Figure 1: Gross appearance of right ovarian mass showing yellowish areas with hemorrhage and necrosis.

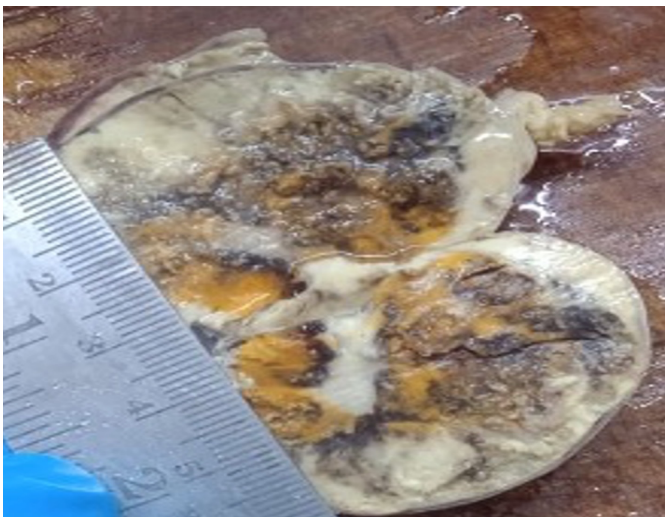


Figure 2: Sheets and cords of polygonal tumor cells with lipid-rich and lipid-poor cytoplasm (H&E).

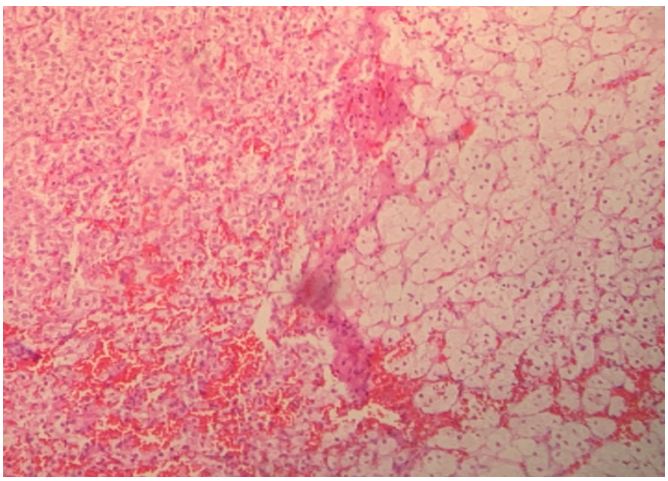
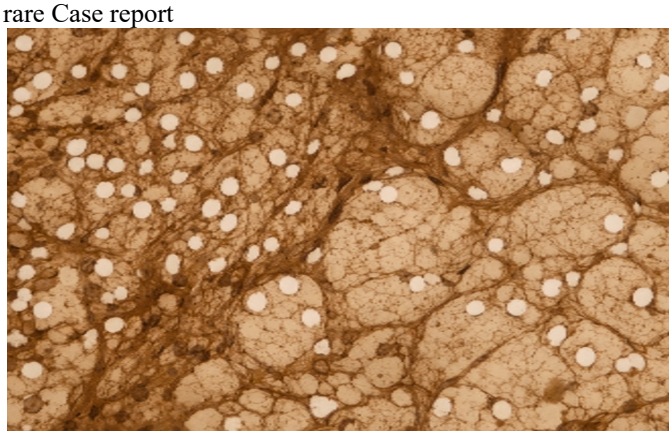


Figure 3: Tumor cells showing strong cytoplasmic positivity for inhibin (IHC).



DISCUSSION:

Steroid cell tumors are a rare subgroup of sex cord stromal tumors of the ovary with an incidence rate of only 0.1% of all ovarian tumors. There are three subtypes of steroid cell tumors according to their cellular origin: 1. Leydig cell tumor arising from Leydig cells in the hilus, 2. stromal luteoma arising from ovarian stroma and 3. steroid cell tumors (not otherwise specified, NOS) when the lineage cannot be identified hence can neither be categorized under stromal luteomas nor under Leydig cell tumors.[5,6] Steroid cell tumors (NOS) constitute about 56% of all steroid cell tumors and these tumors can be functional and produce testosterone, leading to virilization, hyper androgenism, and amenorrhea.[7]

Steroid cell tumor is defined as ovarian parenchymal tumor of sex cord – stromal origin composed of steroid cells with an average age of presentation at 43 years old. Grossly these tumors reach up to an average size of 8.4cm diameter and on cut section typically appear yellow to orangish in color and depending on the presence of necrosis and hemorrhage may also show areas of brown and red to reddish brown respectively. Microscopically similar to the case presented in this study, tumors will be composed of polygonal cells arranged in nests, cords, psuedoglandular pattern and follicle like arrangements. Tumor stroma appears scant to prominent with fibrous tissue and sometimes resembling fibroma. Individual tumor cells have abundant cytoplasm that either appear eosinophilic (lipid poor) or pale and vacuolated (lipid rich) cytoplasm with round nuclei and centrally placed nucleoli.[2]

As per a study by Hayes and Scully, pathological characteristics that indicate malignancy are i) ≥2 mitotic figures per 10 high-power fields is associated with a 92% risk of malignancy; ii) Necrosis with an 86% risk of malignancy; iii) Diameter of >7 cm with a 78% risk of malignancy; iv) Hemorrhage with a 77% risk of malignancy; and v) Grade 2 or 3 nuclear atypia with a 64% risk of malignancy.[8]

Immunohistochemical markers, such as inhibin, calretinin, and steroidogenic factor-1, are positive for these ovarian sex cord stromal tumors.[9] Of all the markers used to identify these tumors, inhibin has proven to be the most helpful to date, as most steroid cell tumors express this marker.[10]

Maissa Ben Thayer et al have also presented a case of co-existence of infiltrating breast cancer with ovarian steroid cell tumor and they have said that there are no available data on the occurrence of ovarian steroid cell tumor and its influence on the prognosis of infiltrating breast cancer.[11]

CONCLUSION:

This case report was presented here not only for its rarity but also intended to strengthen the reader's knowledge on Steroid cell tumours, Not otherwise Specified, for early diagnosis, its importance to distinguish benign from malignancy and for post operative follow up for recurrence or metastasis. Also to explore if there are any correlation between Steroid cell tumours, Not otherwise Specified in a background of carcinoma breast.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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