Case Report

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Steroid Cell Tumor: A Rare Ovarian Tumor

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ABSTRACT

Ovarian steroid cell tumors are sex-cord stromal tumors that arise from lutein cells or leydig cells. These are very rare- accounting for <0.1% of all ovarian tumors.. Some of these tumors may be functional and secrete hormones. Nearly 56-77% of the cases present with hyperandrogenism. Rarely steroid cell tumors may arise in the adrenal cortex from the adrenal rest cells

We report a case of a 60-year old lady who presented with gradual onset abdominal distension for two years. She had no other complains and had undergone hysterectomy 20 years back. Her abdomen was over distended due to a large, tense cystic, non-tender mass. Imaging studies revealed a large, mainly cystic abdominopelvic mass; however its origin could not be made out. The left adrenal gland was found to be bulky but morphologically normal. Serum DHEAS levels were normal thus ruling out adrenal pathology. Serum levels of CA 125 and CEA were also normal. Exploratory laparotomy was done. A large thin walled cyst occupied the pelvis and abdomen. . It measured 25x20x16cm and contained 4.2 litre of straw colored fluid. The fluid was drained and the entire cyst wall was excised. Ovaries were not visualized. Frozen section report of the cyst wall was suggestive of a neuroendocrine tumor. Final histopathology report was of benign steroid cell tumor not otherwise specified. In view of the benign nature of ovarian pathology, no further intervention was required. The patient had an uneventful postoperative recovery

Keywords: *Steroid Cell Tumor, Ovarian Tumor, Stromal Tumor, Hyperandrogenism.*

Introduction

Ovarian tumors are usually of epithelial type, occurring in 50-60 years of age group. At times rare varieties of ovarian neoplasms may be diagnosed on histopathology. Steroid cell tumor is one of these rare tumors accounting for less than 0.1% of all ovarian tumors. [1] We report a case of a steroid cell tumor NOS diagnosed in a women presenting with large abdominal mass.

Case Report

A 60-year old postmenopausal lady, para 5, presented to us with complaint of gradual onset abdominal distension for two years. She had no other complains and had undergone hysterectomy 20 years back for fibroid uterus. Records of surgery were not available. She was a known case of hypertension on irregular treatment . She had no other medical illness and was not on any other treatment.

Her blood pressure was high on admission which was controlled on medication. Her other vital signs were normal and general physical examination was unremarkable. Abdominal examination revealed a large, tense cystic, non-tender mass occupying whole of the abdomen almost reaching till xiphisternum. On pelvic examination, the mass was felt high up in abdomen.

Contrast enhanced computed tomography revealed a large, mainly cystic abdominopelvic mass; however its origin could not be made out. Uterus and ovaries were not visualised.The left adrenal gland was found to be mildly bulky but morphologically normal. Routine and biochemical investigations were normal. Serum CA 125 was 36U/ml and CEA was 5.6ng/ml. Serum DHEAS was checked in view of bulky adrenal gland and was found to be normal (30.8ug/dl).

Patient was taken up for exploratory laparotomy . Intraoperatively, a large thin walled cyst measuring 25 x 20 x 16 cm occupied the abdomen. It had a narrow attachment to the retroperitoneum in pelvis. Ovaries were not visualized and there was no free fluid in peritoneum. The cyst contained 4.2 litres of straw coloured fluid which was drained and sent for cytology. The cyst wall was excised completely. It was smooth, brown coloured and showed a 3x2cm solid area . Frozen section taken from the solid part of the cyst wall was suggestive of neuroendocrine tumor. Peritoneal cytology was negative for malignant cells . The final histopathology report from the solid area was of benign steroid cell tumor not otherwise specified and rest of the cyst wall was lined by mesothelial cells.. On immunohistochemistry , tumor cells were positive for inhibin, melan A, CD10 and calretinin and faintly positive for synaptophysin and chromagranin suggesting steroid cell tumor. No significant nuclear anaplasia , atypical mitosis or necrosis was identified. In view of the benign nature of ovarian pathology, no further intervention was required. The patient had an uneventful postoperative recovery and is in follow up with us.

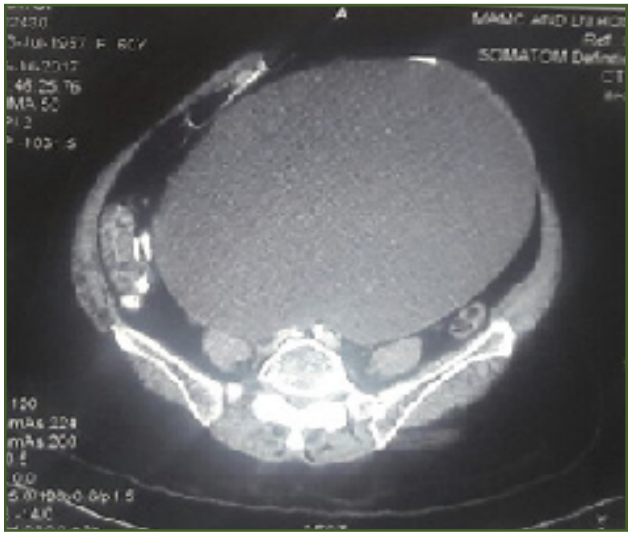

Fig.1: CT scan: Large abdominopelvic mass.

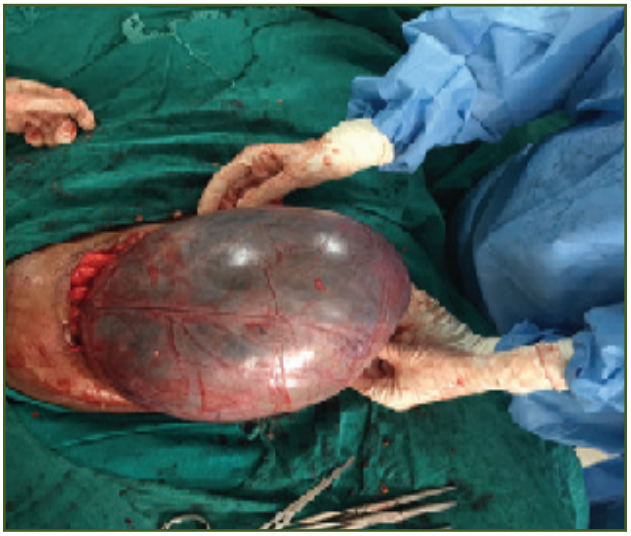

Fig. 2: Intraoperative smooth cyst occupting whole of abdomen.

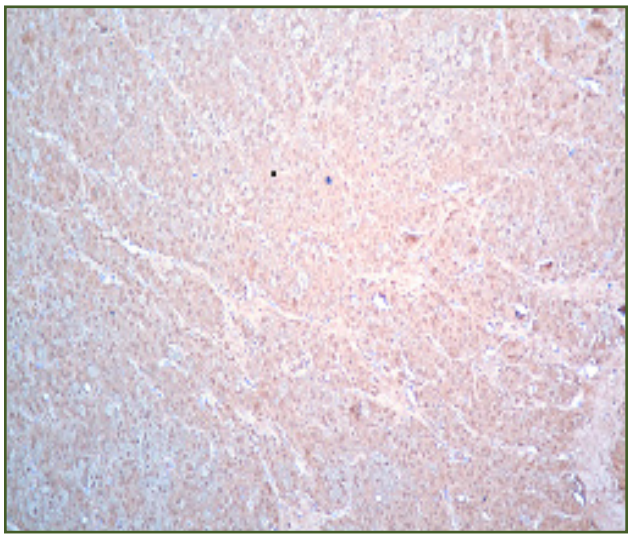

Fig. 3: Positive for calretinin.

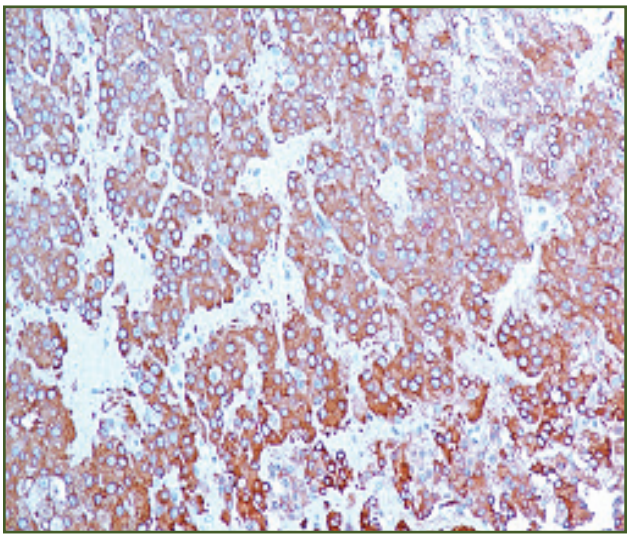

FIg. 4: Positive for inhibin.

Discussion:

This article describes a rare ovarian tumor which presented as a large cystic abdominopelvic mass in a 60 year old woman. There was a diagnostic dilemma concerning origin of the mass. The patient had undergone hysterectomy in the past and ovaries were not identified during the present surgery. Though the adrenal was bulky, serum DHEAS level was normal, thus ruling out adrenal origin. The mass was thus presumed to arise from some residual ovarian tissue left behind at the time of hysterectomy

Steroid cell tumor of the ovary was first described by Scully in 1987. These tumors account for less than 0.1% of all ovarian neoplasms.1 There are three subtypes of steroid cell tumors according to the cell of origin: a) stromal luteomas arising from ovarian stroma(20%), b) leydig cell tumor arising from leydig cells (20%) and c) steroid cell tumor not otherwise specified when the lineage of tumor is unkwown(60%).[2] Our patient had steroid cell tumors NOS. The incidence of steroid cell tumor, NOS is highest in women of child bearing age group, but in rare cases children and postmenopausal women may be affected. These tumors are mostly unilateral but can involve both the ovaries in about 6% of the patients .

56-77% of these are hormone secreting tumors, mostly testosterone and present with features of hyperandrogenism[3] Some of these may produce estrogen or cortisol and present with irregular bleeding or with features of cushing syndrome respectively. Our patient

presented with non specific symptoms and had no features suggestive of hormone producing tumor.

Grossly, tumors are commonly solid, however a combination of solid and cystic or predominantly cystic form may also be seen . . In the presented case steroid cell tumor presented as a solid nodule encased within a large benign cyst. Inhibin and calretinin are sensitive immunohistochemical markers. Calretinin is positive in 60-90% of tumor cells whereas inhibin reactivity ranges from 5-90%.[3] In our case tumor was strongly positive for inhibin and calretinin and weakly positive for chromagranin.

25-40% of steroid cell tumor NOS are clinically malignant.Five correlates of malignant behaviour are i) two or more mitosis per 10 high power fields, ii) tumor diameter of more than 7cm iii) necrosis iv) haemorrhage v) grade 2 to 3 atypia. Histologically, our patient's tumor did not have any of the above criteria and was thus labelled as a benign tumor

Treatment of steroid cell tumor NOS is primarily surgical. A benign tumor limited to ovary is managed by cystectomy or unilateral salpingoopherectomy. For patients with unilateral malignant tumor, but desirous of future fertility, unilateral salpingoopherectomy with preservation of contralateral ovary and uterus is done but they need postoperative surveillance. Metastatic malignant disease requires surgical cytoreduction followed by adjuvant chemotherapy with bleomycin, etoposide and cisplatin. In our case the tumor was benign and was excised completely. No adjuvant therapy was required.

Conclusion

Apparently benign looking ovarian cyst may turn out to be uncommon ovarian neoplasm. A thorough pathological examination with use of immunohistochemical staining technique is vital for correct diagnosis and prognostication.

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