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Ruptured sex cord stromal tumor: A rare case report

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Abstract
Background: Sex cord tumor with annular tubules is a rare ovarian neoplasm that accounts for <6% sex cord stromal tumors [1]. It is seen mostly in reproductive period and less commonly in pediatric population [2]. One-third of cases are associated with hyperestrogenism and Peutz- Jegher's syndrome.
Case report: A 26 years old unmarried girl came in gynaecology OPD with pain abdomen and abdominal lump since 4 months. After necessary investigations, she was planned for elective surgery but was lost to follow-up then. After 2 months, she presented in emergency with acute pain abdomen with shock. On examination, abdominopelvic mass was felt palpable corresponding to 20 weeks. Exploratory laparotomy with left sided salpingo-oopherectomy was performed. Hemoperitoneum of around 2 litres and a left-sided ovarian tumor 15x10cm in size with necrosis, hemorrhage and ruptured capsule was found. On histopathology report it was confirmed to be as sex cord tumor with annular tubules (SCTAT).
Conclusion: We describe for the first time to the best of our literature search the incidental detection of SCTAT in a patient previously thought to be case of germ cell tumor. Meticulous histological staining and awareness is required for detection of such unusual incidental lesions.

Keywords: Sex cord tumor, peutz- jegher's syndrome

Introduction
SCTAT is a rare ovarian tumor which was first described by Scully in 1970 [1]. It accounts for only 8% by overall ovarian neoplasms and only 6% of sex cord stromal tumors [1]. It is strongly associated with Peutz- Jegher's syndrome, also with adenoma malignum of cervix, turner's syndrome, dysgerminoma, endometrial carcinoma etc [3-7]. It is estrogen-progesterone secreting tumor with low malignant potential.

Case History
A 21 year's old unmarried girl presented in OPD with pain abdomen and abdominal lump since 4 months. On examination a left-sided abdominopelvic mass was palpable which was confirmed as ovarian mass on ultrasound (suggestive of left adnexal non-defined solid echogenic lesion with internal cystic wall 12*9*10 cm with internal vascularity) and CT scan as 8.5x16.1x17.4 cm predominantly solid mass suggestive of germ cell tumor with secondary necrotic adenopathy. She was planned for elective surgery but was lost to follow-up. Her tumor markers profile has been like:-
CA- 125 19.86 U/ml
CEA - 1.03 ng/ml
AFP - 3.0 ng/ml
LDH - 542 U/l (Raised)

After 2 months, she presented in emergency with acute pain abdomen with nausea, vomiting and shock. On abdominal ultrasound, hemorrhagic ascitic fluid was observed. Emergency exploratory laparotomy was performed followed by left-sided salpingo-oopherectomy. Per operatively hemoperitoneum around 2 litres and left sided ovarian tumor of 15x10cm with necrosis, hemorrhage and ruptured capsule was found. On histopathology, it was found to be SCTAT with inhibin, calretinin, CD99 and vimentin markers positive on immunohistochemistry. It's Ki 67 labelling index was 14%. No evidence of extraovarian extension/ metastasis was noted.

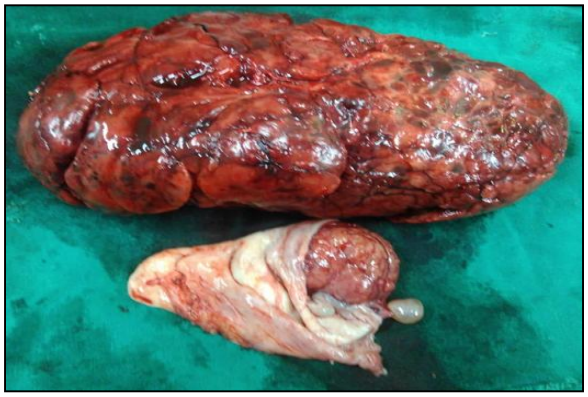

Fig 1.a: Gross specimen showing (Above) ruptured ovarian tumor with left tube and ovary and (below) left ovary shown with ruptured capsule.

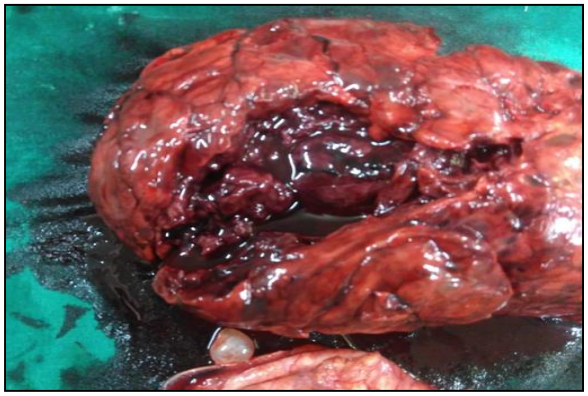

Fig 1.b: Gross specimen showing ovarian tumor with areas of necrosis and haemorrhage

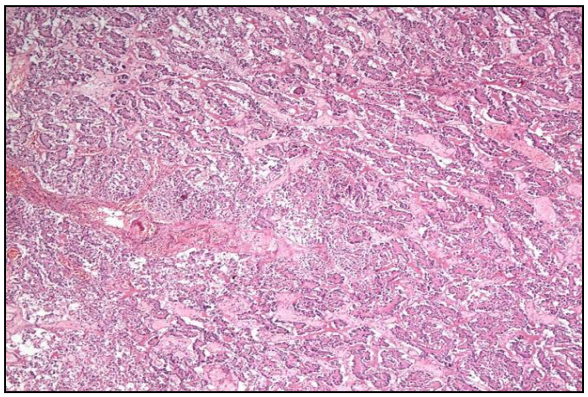

Fig 2.a: histopathology slide showing sex cord stromal tumor with annular tubules.

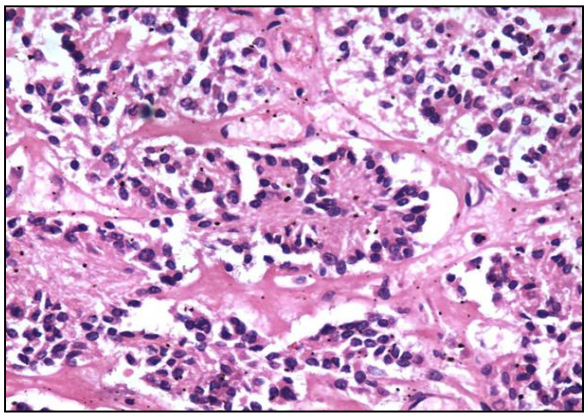

Fig 2.b: Histopathology slide showing annular tubules (Hematoxylin and eosin)

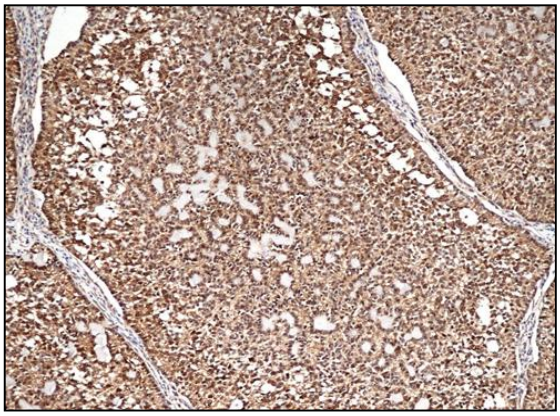

Fig 3.a: Calretinin reactivity in tumor cells

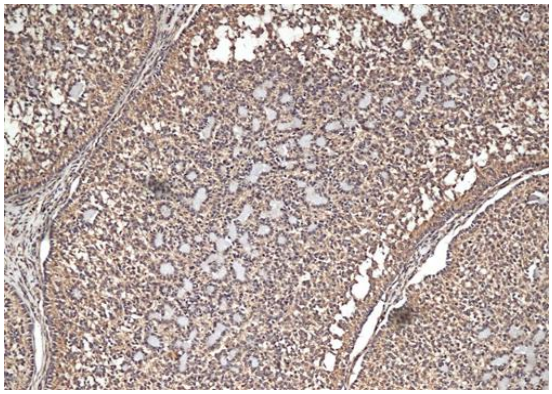

Fig 3.b: CD99 reactivity in tumor cells

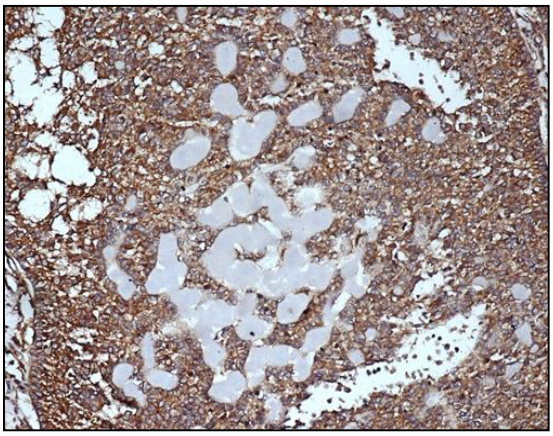

Fig 3.c: Alpha- Inhibin reactivity in tumor cells

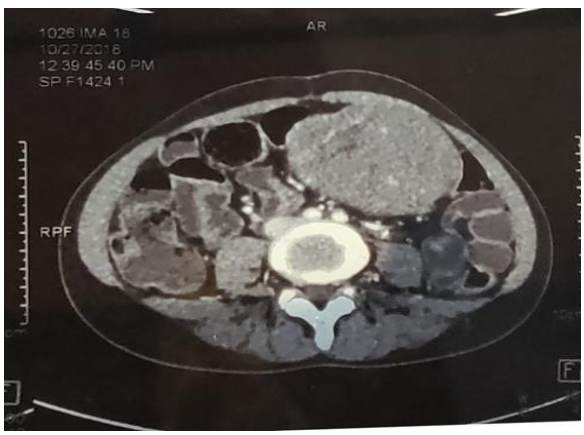

Fig 4-a: CT abdomen film showing adnexal mass

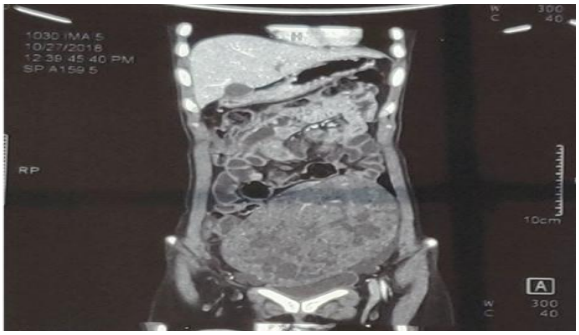

Fig 4-b: Another film showing adnexal mass

Discussion

SCTAT is a unusual ovarian neoplasm with its predominant component having morphologic features intermediate between granulosa cell tumor and sertoli cell tumor. Clinical manifestations of SCTAT are due to its estrogen-progesterone secreting activity like menorrhagia, precocious puberty, postmenopausal bleeding etc [3]. In this case, patient didn't have any clinical/ biochemical features of hyperestrogenism. SCTATs are strongly associated with Peutz-Jegher's syndrome however present case was not associated with it clinically. Those associated with Peutz- jegher's syndrome are usually benign, multifocal, bilateral and small whereas those without PJ syndrome are usually unilateral and larger in size.

Complex tubules are not seen in sertoli cell tumors. In micro follicular granulosa cell tumors, hyaline bodies in lumens of tubular structures are not very dominant, a feature important for differential diagnosis.

Few cases have been reported for acute abdomen that eventually resulted in exploratory laparotomy. Selma sengiz erhan *et al*. reported a case in which a 12 years old girl presented with acute abdomen which was later found out to be ruptured SCTAT on histopathology. It was positive for calretinin, vimentin and inhibin. The patient was lost to follow-up postoperatively but as per telephonic call she had no complaint in six years after surgery. In case reported by kanika *et al.* in 2017, a 12 years old girl presented with acute abdomen due to spontaneous rupture of tumor resulting in hem peritoneum. On exploratory laparotomy, left salpingo-oopherectomy and total omentectomy was performed and it revealed out yolk sac tumor on histopathology. She was started on BEP regimen but poor general condition and disseminated carcinoma resulted in poor outcome. Sunita Sudhir *et al*. reported a case of immature teratoma of the ovary in a 14-year-old girl who presented with acute pain abdomen. On laparotomy, it was a ruptured malignant germ cell ovarian tumor and unilateral salpingo-oophorectomy was performed. It was stage 1, grade 3. The patient was put on BEP regimen and responded well to chemotherapy. Saha *et al*. reported a case of a 14-year-old girl who presented with acute abdominal pain with distension of abdomen. On laparotomy, it was a suspected malignant ovarian tumor stage IC and ipsilateral salpingo-oophorectomy was done, and on histopathological examination, it was a juvenile granulosa cell tumor. It was not followed up with chemotherapy and uptill the time of reporting, the patient was healthy without any recurrence. In yet another case reported by Daher *et al*., rupture of mixed germ cell tumor was detected in a 12-year-old girl who presented with acute pain abdomen, vomiting and distension. Emergency laparotomy revealed a ruptured ovarian tumor with omental invasion and ipsilateral salpingo-oophorectomy was done with omentectomy. It was then followed by cisplatin-based chemotherapy and the patient responded well.

In our case, she presented with acute pain abdomen, where exploratory laparotomy followed by left sided salpingo-opherectomy was performed and patient responded well to it.

Thus, in the above mentioned case, the patient had all her tumor markers negative which has been found as per scanty literature available. However, histopathology showed annular tubular pattern with immunohistochemistry markers positive for calretinin, CD99 and inhibin. Her postop period went uneventful. She followed up both in oncology and gynaecology department and doesn't require chemotherapy but was lost to follow up thereafter.

Based on scanty literature available, the surgery alone with fertility preservation should be attempted in these patients. To summarize, a rare case of sporadic SCTAT is presented and its diagnosis and management have been highlighted.

Therefore, any case presenting in the OPD should be timely taken up for elective surgery to avoid any unfortunate emergency laparotomies in future resulting into better prognosis of the patient.

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