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Moderately differentiated Sertoli-leydig cell tumour in a teenager: Report of an unusual diagnosis in an unilateral ovarian cyst

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Abstract
Mixed type Sertoli – Leydig cell tumours are uncommon ovarian neoplasms of sex – cord stromal origin accounting for less than 0.5% of ovarian neoplasms. Patients commonly present in their second and third decades of life with symptoms of abdominal mass or with hormonal changes, commonly androgenic features like virilization and defeminization. DICER 1 mutations present in younger patients with syndromic association. FOXL-2 have also been documented. Immunohistochemical evaluation show Sertoli cells exhibiting positivity for calretinin, inhibin, FOXL2, SF1 and WT1. The Leydig cells usually exhibit Melan A and inhibin. Prognosis depends on staging (FIGO and TNM) and grading (differentiation). Associated heterologous elements such as cartilaginous or skeletal muscle differentiation confer an unfavorable prognosis also. The mainstay of treatment is surgical, adjuvant chemotherapy may be offered in certain scenarios. We present a case of Mixed type Sertoli – Leydig cell tumour in a young patient, which coupled with the cystic nature of the tumour, posed a diagnostic challenge as these tumors are extremely rare.

Keywords: mixed, sertoli, leydig, tumour, ovary, cyst, young age

Introduction
Mixed type Sertoli – Leydig cell tumours are uncommon ovarian neoplasms, with an incidence of less than 0.5% of ovarian neoplasms. These tumours may be asymptomatic or present with features of virilization and occur among a broad age group of 1 to 85 years with a mean age of 25 years [1, 2]. We present a case of a mixed Sertoli- Leydig cell tumour presenting as an ovarian cyst in a 15-year-old female.

Case Report
A 15-year-old girl presented with mild abdominal pain and no significant virilization or defeminisation symptoms. Serum hormone and tumour marker levels were not elevated. On radiology she was found to have a cystic mass in the left ovary. An exploratory laparotomy was performed, and left ovarian cystectomy was performed. An intact cystic ovary was received, weighing 164 grams, and measuring 74x45x32mm. Cut section shows a multiloculated cyst containing yellowish fluid. The walls of the cyst showed a few firm nodular areas which were ochre yellow and soft. No solid areas, dermoid cyst contents or papillary excrescences were noted. On microscopy, the ovary showed multiple locules with nodular and solid areas with two types of cell populations. One component was composed of trabeculae, cords and tubules of medium sized, darkly staining cells with coarse chromatin, suggestive of Sertoli cells. Interspersed were singly lying, clusters and aggregates of plump cells with granular, eosinophilic cytoplasm suggestive of Leydig cells. An intervening delicate fibrous stroma and edematous areas were also noted. Occasional mitotic activity of around 1 per 10 high power fields was noted. No anaplasia or heterologous elements was noted. The cystic spaces show flattened lining epithelial cells and collagenous stroma. Viable ovarian tissue was noted at the periphery. No capsular breach was noted. Immunohistochemistry was performed for confirmation, with the darkly staining cells strongly positive for calretinin, and alpha inhibin and negative for CK7 and EMA. The Leydig cells were negative for CK7, EMA, calretinin and focally exhibited weak staining for inhibin. Based on the histology and the immunohistochemistry findings a diagnosis of Moderately differentiated Sertoli- Leydig cell tumour of the ovary was proffered. The patient was kept on close follow up and was disease free on last follow up.

Discussion
Sertoli – Leydig cell tumours are rare tumours of sex – cord stromal origin accounting for less than 0.5% of ovarian neoplasms. These tumours have been reported in a wide age range of patients from 1 to 84 years and are commoner in the second and third decades [1, 2, 3]. Patients commonly present with symptoms of abdominal mass or with hormonal changes, commonly androgenic features like virilization and defeminization [4]. In younger patients DICER 1 mutations have been recorded with syndromic association [5]. FOXL-2 has also been documented [6]. These tumours are predominantly solid and unilateral, presenting as stage T1a [7]. On histology, depending on the degree of anaplasia and differentiation, the tumours are subdivided into well differentiated, moderately differentiated, and poorly differentiated subtypes [1, 2, 3, 8]. Associated heterologous elements and anaplasia may be associated and need to be examined as they confer an unfavorable prognosis, commonly associated with moderate to poorly differentiated tumours [9, 10].
Immunohistochemical evaluation of these tumours is performed to confirm sex cord stromal origin, with the Sertoli cells exhibiting positivity for calretinin, inhibin, FOXL2, SF1 and WT1. The Leydig cells usually exhibit

Melan A and inhibin [11, 12]. Staging of the tumours is performed according to the TNM and FIGO systems, and while the mainstay of treatment is surgical, adjuvant chemotherapy may be offered in certain scenarios [13]. Prognosis is favourable, with a survival rate of 100% for well differentiated tumours. 10% of Moderately differentiated and 60% of poorly differentiated tumours exhibit malignant behaviour and may recur in the peritoneal cavity within 2 years [1, 2, 14]. Unfavourable factors include advanced stage at presentation, anaplasia, poorly differentiated tumours, and cartilaginous or skeletal muscle differentiation [1, 2, 15, 16]. In our case scenario, the young age of the patient coupled with the cystic nature of the tumour, posed a diagnostic challenge as these tumors are extremely rare. However characteristic morphology and immunprofile assisted in a definitive diagnosis.

Conclusion

Sertoli Leydig cell tumours are rare sex cord stromal tumors, with well differentiated variants exhibiting an excellent prognosis. Extensive sampling of the tumour is essential to evaluate areas of poorer differentiation and heterologous elements which can alter the prognosis. Close follow up and regular monitoring is required as these tumours are rare and newer methods and therapies for their treatment and diagnosis along with association with mutations are under study.

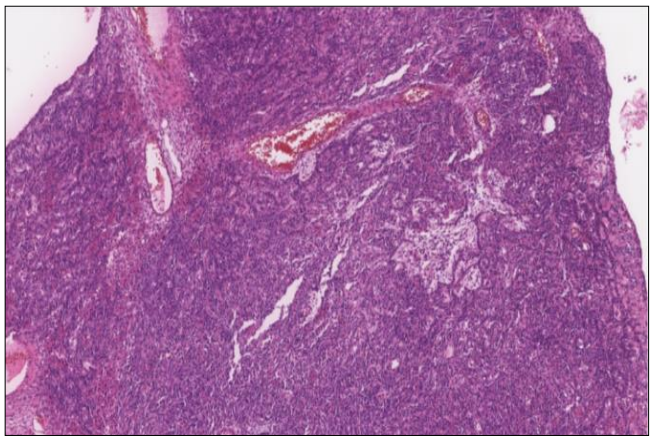

Fig 1: H&E stained section of nodule in wall of cyst with darkly staining Sertoli cells (H&E 1.5X)

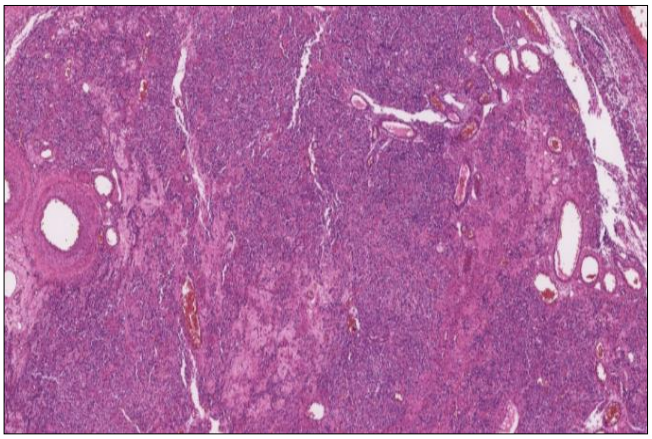

Fig 2: H&E stained section of nodule in wall of cyst with darkly staining Sertoli cells in trabeculae and tubular arrangement and intervening paler cells (H&E 1.7X)

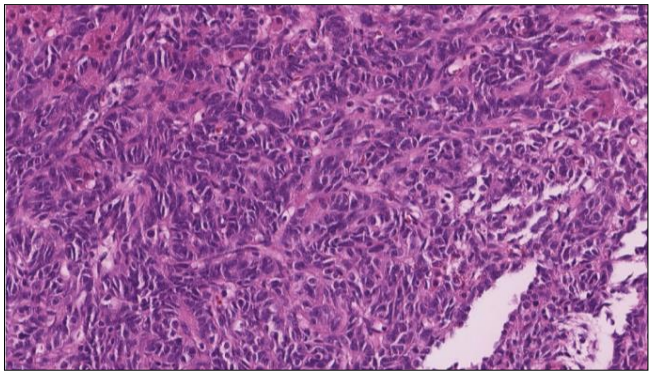

Fig 3: H&E stained section with Sertoli cells in trabeculae and focally retiform appearance. No mitoses or anaplasia is noted (H&E 10X)

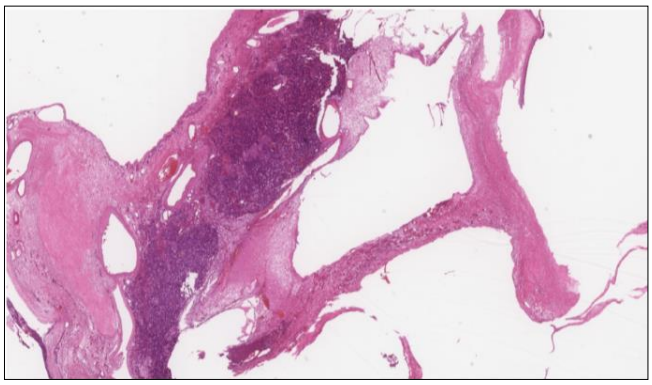

Fig 4: H&E stained section of nodule in wall of cyst with darkly staining Sertoli cells (H&E 1.1X)

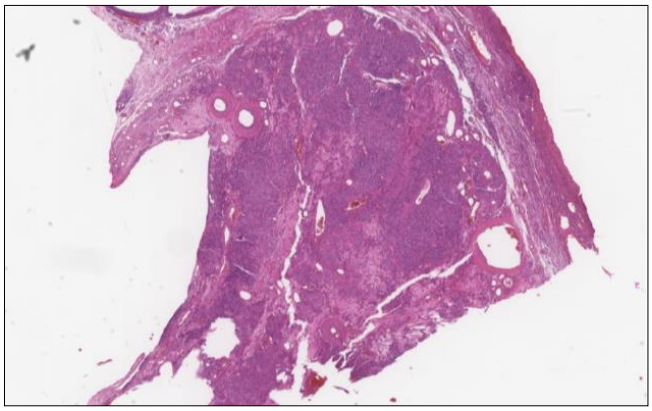

Fig 5: H&E stained section of of wall of cyst with Sertoli cells and intervening paler Leydig cells (H&E 1.1X)

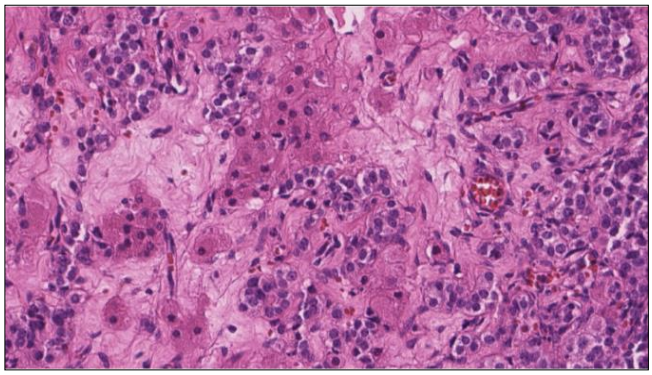

Fig 6: Nodules of Leydig cells in between darkly staining Sertoli cells, no mitoses or anaplasia noted (H&E 15 X)

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Competing Interests
Competing interests: None declared]

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