Case Report

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Pathology Section

Struma Ovarii: A Rare Presentation in a Paediatric Patient

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ABSTRACT

Struma Ovarii (SO) is a rare form of teratoma of ovary containing predominantly thyroid tissue, usually comprising more than 50% of the overall mass. It accounts for 1% of all ovarian tumours in adults. Due to its rare presentation in paediatric age group, the exact incidence is not known.The association of SO with hyperthyroidism has been noted in approximately 8% of cases. Radiological evaluation helps in preliminary diagnosis of a complex ovarian cyst, however, a definitive diagnosis of SO can only be made upon extensive histopathological evaluation. Present report is of SO in a 12-year-old female patient, who initially presented with pain abdomen, vomiting and hyperthyroidism with mildly elevated thyroid hormones Triiodothyronine (T3) and Tetraiodothyronine (T4). Magnetic Resonance Imaging (MRI) whole abdomen showed evidence of right-sided dermoid with mild ascites. An exploratory laparotomy was done followed by right ovarian cystectomy and histopathology revealed a mature teratoma predominantly comprising of thyroid tissue (>90% of total ovarian mass), thereby confirming the diagnosis of SO. After one month of surgical resection, the patient's thyroid profile showed decrease in fT3 and fT4 levels. The index case highlights the unusual occurrence of SO in a paediatric patient along with the role of an extensive histopathological evaluation in diagnosis of the same.

Keywords: Histopathology, Ovarian teratoma, Thyroid tissue

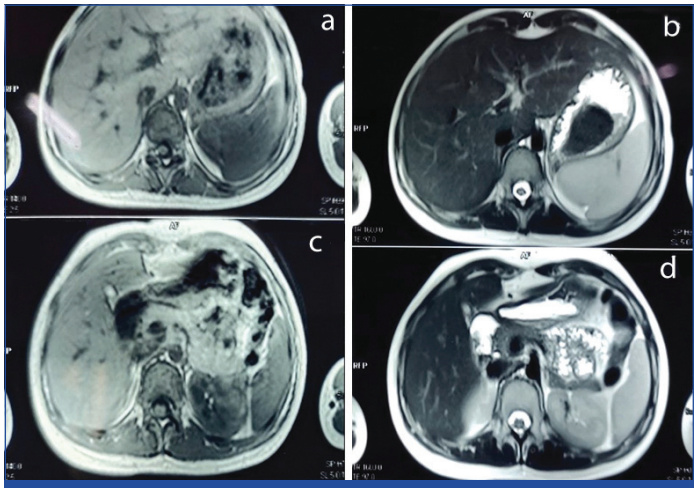
CASE REPORT

A 12-year-old female patient presented to hospital with complaints of pain abdomen and vomiting for last five days. There was no history of tuberculosis and any other chronic or major illness. The patient had attained menarche one year back. Her routine haematological parameters (Complete Blood Count) were within normal limits. However, thyroid function tests revealed slightly elevated T3 and T4 levels with normal Thyroid Stimulating Hormone (TSH) levels. Rest of the biochemical tests, i.e., liver and kidney function tests, lipid profile and blood sugar levels were within normal reference range.

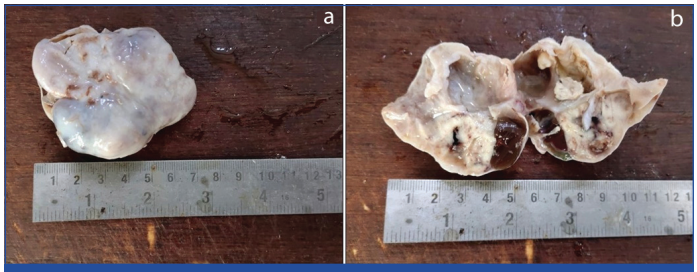
The patient's ultrasound abdomen revealed a right, solid-cystic ovarian mass measuring 4.2×6 cm. Physical examination revealed mild abdominal distension with no palpable pelvic mass or any sign of ascites. Urine pregnancy test was negative. Her serum tumour markers, i.e., Cancer Antigen 125 (CA-125) Carcinoembryonic Antigen (CEA), human Chorionic Gonadotropin (hCG) were within normal reference range. Clinically, a provisional diagnosis of twisted ovarian cyst was made, along with germ cell tumour of ovary (teratoma). The patient was advised for surgical intervention. MRI whole abdomen was done and revealed a large heterogenous signal intensity lesion with fat signal and fluid signal intensity with free fluid level/haemorrhagic foci in pouch of douglas inseparable from right ovary-likely right dermoid ovarian cyst with mild ascites [Table/Fig-1a-d].

Exploratory laparotomy by a Pfannenstiel incision found a multiloculated complex right-ovarian cystic mass. The left ovary, uterus and the rest of the abdomen appeared free from disease. A right ovarian cystectomy was performed. The postoperative course was uneventful, and the patient was discharged on Day 4. The surgical specimen was sent for detailed histopathological examination.

Grossly, a specimen labelled as right ovarian cyst, measuring 5.5×5.0×2.5 cm was received. The external surface appeared asymmetrical with grey-white glistening areas. Cut section showed multiloculated cyst filled with grey-brown material (colloid-like), haemorrhagic areas, grey-white areas, focal cartilaginous areas [Table/Fig-2a,b]. Few cystic spaces were noted filled with mucoid material



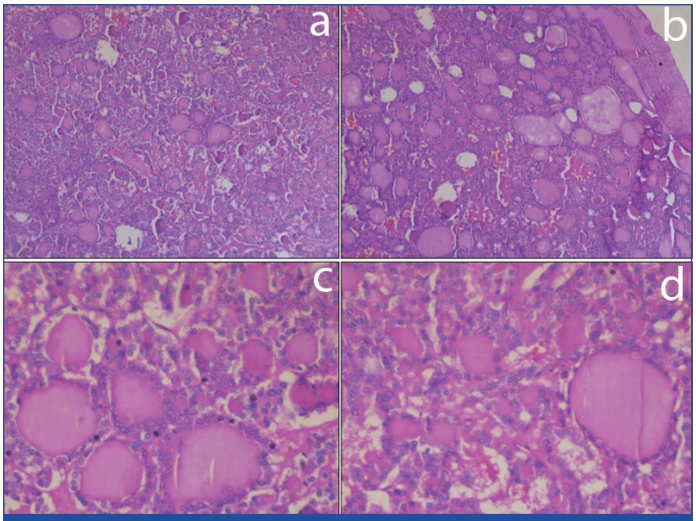
[Table/Fig-1]: MRI whole abdomen showing a large heterogenous lesion with variable signal intensity, inseparable from right ovary- likely right dermoid ovarian cyst with mild ascites.



[Table/Fig-2]: a) Image showing an asymmetrical cystic lesion, measuring 5.5×5.0×2.5 cm with grey-white glistening areas. b) Cut section revealing a multiloculated cyst filled with colloid-like material, grey-white areas and few cystic spaces filled with pultaceous material along with a tooth embedded in the wall of cyst.

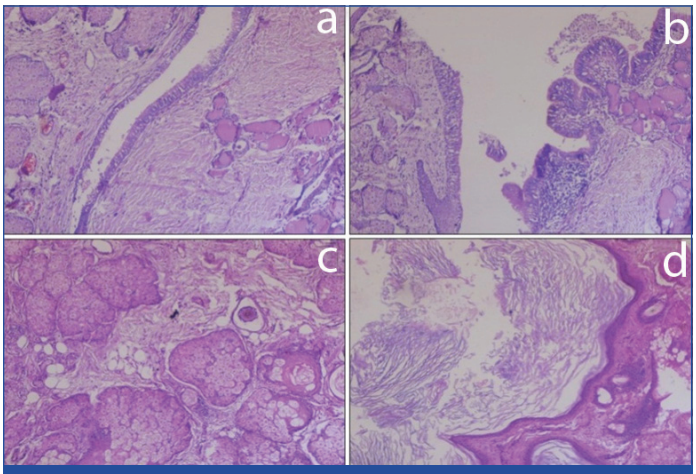
and few filled with pultaceous material. On serial sectioning single tooth was found embedded in the wall of the cyst [Table/Fig-2b].

Multiple sections were taken from the right ovarian cyst specimen. Sections showed partly encapsulated lesion showing extensive areas (>90%) comprising of benign thyroid follicles arranged in small groups, microfollicles and macrofollicles with luminal colloid. Intervening areas showed fibrocollagenous septa with delicate vessels [Table/Fig-3a-d].



[Table/Fig-3]: a,b) Low power view showing partly encapsulated lesion with extensive areas (>90%) comprising of benign thyroid follicles arranged in microfollicles and macrofollicles with luminal colloid a,b (H&E,10X, 20X). c,d) Intervening areas showed fibrocollagenous septa with delicate vessels c,d (H&E,40X).

Sections from the compressed ovarian parenchyma showed histological evidence of ruptured epidermal cyst, sheets of sebaceous glands underneath squamous lined epithelium, focal cartilaginous areas and focal ovarian stroma lined by respiratory epithelium [Table/Fig-4a-d]. No neural tissue or any atypical cells were noted after extensive sectioning.



[Table/Fig-4]: a,b) Histological images showing focal ovarian stroma lined by respiratory epithelium and surrounded by aggregates of thyroid follicles a,b (H&E,40X); c) Foci of sebaceous glands underneath squamous lined epithelium H&E,40X; d) Evidence of epidermal cyst (H&E,40X).

Based on clinical, radiological and a detailed histopathological evaluation, a diagnosis of SO (mature ovarian teratoma-right ovary) was made. Post surgery, her thyroid profile was also found to be within normal reference range. Till the writing of the present report and follow-up of one year with ultrasound, thyroid profile and CA-125 at regular intervals, no evidence of recurrence was found.

DISCUSSION

Teratoma is a tumour of germ cell origin and usually consists of more than one germ cell layer structures [1-3]. Based on cellular components, it is divided into mature, immature and monodermal variants. The two forms of monodermal ovarian teratomas are SO and carcinoid. SO is a rare ovarian tumour, categorised as a monodermal and highly specialised teratoma with presence of predominantly thyroid tissue (>50%) and accounting for approximately 5% of all ovarian teratomas. Usually SO does not secrete thyroid hormones, however, 8% of patients show clinical and biochemical features of hyperthyroidism [2-4]. The definitive diagnosis of SO is only made based on histopathological findings.

This entity was described for the first time in the year 1889 by Boettlin, who happened to see the presence of thyroid tissue in ovaries and subsequently few cases were reported by Von Kalden in 1895, Gottschalk in 1899 and Mayer in 1903 [2-4]. However, as SO comprises of <1% of all ovarian tumours, there is limited data pertaining to SO in literature. It consists of 5% of all ovarian teratomas. This is supported by few series in literature, Ghartimagar D et al., [3], Yoo SC et al., [4], and Khediri Z et al., [5] reported SO cases to be 5%, 4.8%, 8.6%, respectively.

The SO is usually noted after the age of 40 years, the peak incidence being in the 5th decade [3-5]. However, few cases have been reported in prepubertal and postmenopausal women. In series by Ghartimagar D et al., of total seven cases, three were seen in >40 years, three cases noted in 30-40 years and only one was seen in 26-year-old woman [3]. Khediri Z et al., also reported three cases of SO in 17 years, 31 years and 40 years old female patients [5]. Index case was much rarer in the fact that the patient was a 12-year-old female.

Majority of SO cases present with non specific symptoms. Yoo SC et al., noted 42% of patients of SO with non specific symptoms, just diagnosed during routine ultrasound evaluation. These non specific symptoms could be pain abdomen, palpable abdominal mass, vaginal bleeding, ascites, hydrothorax, elevated thyroid hormones, etc., [4]. It is noted in literature that approximately 8% of SO patients can have simultaneous presence of hyperthyroidism, which was noted in index patient also [5-8]. In a study by Kaur S et al., a case of SO was noted with preoperative high levels of T3 and T4 [6]. The postoperative thyroid profile was normal as noted in index case.

Radiological evaluation serves as an important tool in the diagnosis and classification of ovarian mass, however for a definitive diagnosis of SO, a detailed histopathological evaluation is essential [7,8]. The differential diagnosis given on radiology could be dermoid cyst, benign cyst, endometrioma, malignancy. MRI shows multilocular cystic lesion with variable signal intensity within the loculi. However, no definitive opinion can be given just on the basis of MRI findings in suspected cases of SO.

Usually, the SO cases appear on unilateral side, and the right ovary being affected commonly [6-8]. Bilateral SO is very rare, noted in just 6% of cases. Rana V et al., had reported a case of bilateral SO in an elderly patient with pseudo-Meig's syndrome [8]. Khediri Z et al., also found the occurrence of SO in the right ovary [5] as seen in our case. However, in study by Ghartimagar D et al., all four cases were noted in left ovary [3].

Majority of cases of SO measure 0.5 to 10 cm in their dimension and grossly appear as cystic mass containing brown gelatinous material (colloid like) with some solid areas [3-6]. Microscopically, it consists predominantly of normal looking or hyperplastic thyroid follicles along with other areas like sebaceous glands, squamous epithelium, etc. Malignant form of SO is very rarely noted with occurrence of 5-10%, with histological evidence of papillary, follicular or mixed form [7-9]. The treatment of benign SO is usually the surgical resection of the involved ovary, especially in younger women. However, in case of malignancy or recurrence few factors like size of the tumour, presence of ascites, adhesions during surgery etc are taken into consideration [9-12]. Due to the rarity of this tumour, there is lack of literature pertaining to its diagnosis and treatment, especially in cases of malignant transformation [8-12].

In the present case, surgical resection of the involved right ovary was done, considering the paediatric age group of the patient. Postsurgery, her thyroid levels were within normal range. No recurrence was noted in index patient till the last follow-up of one year.

CONCLUSION(S)

The SO is a rare ovarian tumour with majority behaving in a benign fashion. Though radiology can give a preoperative differential

diagnosis of various cystic lesion involving the ovary, no definitive diagnosis can be given based on imaging. The case highlights the importance of detailed histopathological examination in giving an accurate diagnosis of benign teratoma with predominant thyroid tissue. The index is also rare in the fact that, SO was noted in a paediatric female patient. Index case reiterates the importance of an integrative approach in the diagnosis and early management of a rare tumour.

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