Case Report

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Pathology Section

Struma Ovarii- A Rare Tumour with an Unusual Presentation

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ABSTRACT

A teratoma composed predominantly or exclusively of a single type of tissue is called monodermal teratoma. Struma Ovarii (SO), a type of monodermal teratoma, is a rare variant of ovarian teratoma composed mainly of mature thyroid tissue. It is usually benign in histology. This is a case of an unusually large cystic ovarian mass reported as SO in a 43-year-old female patient who reported with complaint of acute retention of urine for one day, associated with pain in lower abdomen and distension.

Keywords: Benign, Cystic ovarian mass, Monodermal, Teratoma

CASE REPORT

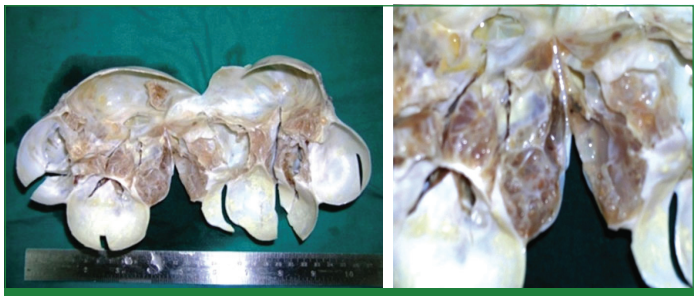
A 43-year-old female presented to the Gynaecology Outpatient Department (OPD), Kasturba Medical College, Manipal, Karnataka, India, with complaint of acute retention of urine for one day, associated with pain in lower abdomen and distention. Patient complained of difficulty in passing urine and incomplete voiding since one year.

On clinical examination, a firm mass measuring 20×20 cm was palpable in the lower abdomen extending up to the umbilicus. It had well defined borders and was mobile. Per vaginally, cervix was pushed to left side, and a firm mobile mass not separate from the uterus was felt extending up to the umbilicus.

Thyroid function tests were within normal range. Tumour markers, CA-125 was mildly raised, 77.3 kU/L, but serum Carcinoembryonic Antigen (CEA) and Alpha-fetoprotein were within normal limits. Paracentesis demonstrated benign peritoneal effusion with reactive mesothelial cells. Radiological investigations, transvaginal ultrasonography and Computed Tomography (CT) scan of the abdominopelvic region showed an abdominopelvic complex cyst measuring 12.7×10.8×11 cm with thin septations and solid components suggestive of ovarian malignancy.

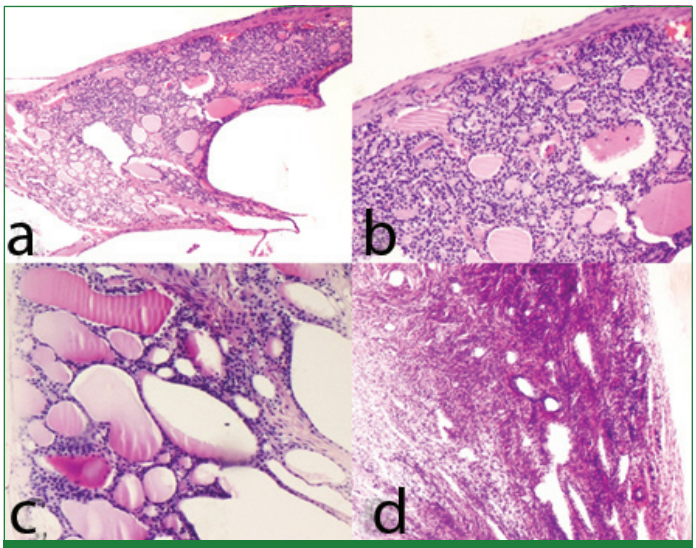
The patient underwent total abdominal hysterectomy with bilateral salpingo-ophorectomy. Peroperatively, an irregular, multilocular cyst measuring 20×15 cm was seen arising from the right ovary. Omentum was free with no bowel adhesions and minimal free fluid was noted in the abdomen.

Grossly, right ovarian cyst, measuring 16.5×11.5×7.5 cm was seen. On cut section the cyst was multiloculated with seromucinous brown material [Table/Fig-1,2]. The inner wall of the cyst appeared smooth with focal brown areas.



[Table/Fig-1]: Multiloculated cyst with seromucinous brown material.
[Table/Fig-2]: Colloid like material. (Images from left to right)

Microscopically, a cyst wall lined by flattened epithelium with bland flattened nuclei and foamy macrophages overlying fibro collagenous stroma with islands of thyroid tissue composed of micro and macro follicles lined by cuboidal epithelium with luminal colloid and colloid laden macrophages, cholesterol clefts surrounded by ovarian stroma was seen [Table/Fig-3a-d]. The ovarian stroma resembled fibroblasts arranged in a storiform pattern and had elongated vesicular nuclei. No other components of teratoma were noted. Sections from the other ovary, tubes, uterus and cervix were unremarkable.



[Table/Fig-3]: a, b and c) Islands of thyroid tissue in the cyst wall (Haematoxylin and Eosin, X40, X100 and X200, respectively); d) Ovarian stroma in the cyst wall (Haematoxylin and Eosin X40).

A diagnosis of SO was given based on the above mentioned histological features. The patient was lost to follow-up following the surgery.

DISCUSSION

Mature cystic teratomas (dermoid cysts) constitute 20% of the ovarian neoplasms [1]. Struma ovarii is a rare variant of ovarian teratoma (3%), composed predominantly of mature thyroid tissue. SO is defined by the presence of more than 50% of mature thyroid tissue [2]. It is predominantly benign on histological examination, with rare malignant transformation in 0.3-5% cases [2]. Metastasis is found in 5-23% of patients with malignant SO [1].

The SO, a monodermal variant of ovarian teratoma, although described earlier, was first recognised in the early part of twentieth century to be composed of thyroid tissue by Ludwig Pick [3]. It is defined as an ovarian teratoma that is composed predominantly (over 50%) or entirely of thyroid tissue and constitutes about 2.5-5% of ovarian teratomas [4,5]. The peak age incidence of struma is

in the fifth decade, however cases have been reported in older postmenopausal women and rarely in prepubertal girls [6]. The common clinical manifestation is abdominal pain or pelvic mass.

A preoperative diagnosis can be suspected in cases with hyperthyroidism, by thyroglobulin measurement or scanning, seen only in 5-8% cases [7]. On radiology, a well vascularised solid component on colour doppler ultrasound and a strongly enhancing solid component in a multilocular tumour of the ovary on CT or Magnetic Resonance Imaging (MRI) can raise the possibility of SO in a solid and cystic teratoma like ovarian tumour.

Grossly, the size of the cyst varies but usually it is <10 cm [8]. The cut surface of the tumour may show light tan glistening thyroid tissue along with haemorrhage, necrosis and foci of fibrin. Microscopically, the tumour is composed of mature thyroid tissue with acini of variable sizes, lined by columnar or cuboidal epithelium and filled with eosinophilic, Periodic Acid Schiff (PAS) stain positive colloid. On Immunohistochemistry, lining epithelium cells show positivity for thyroglobulin. Sometimes, it may resemble adenomatous goitre or adenoma like lesions. The SO has also been reported with appearance of Hashimoto's Thyroiditis.

It is postulated that SO is an autonomous hormone-secreting tumour or that the ovarian thyroid tissue is stimulated by Thyroid Stimulating Hormone (TSH) receptor antibody [5]. Matsuda K et al., reported a case of a 48-year-old woman with symptoms of hyperthyroidism and an ovarian mass [9]. On examination, thyroid gland appeared normal in size and texture, however hormonal findings revealed increased thyroid function which was due to a preoperatively diagnosed hormone producing malignant SO, further confirmed by histopathological examination and immunohistochemistry. The findings of hyperthyroidism disappeared over several weeks after surgery. Malignancy of this entity is rare, has been reported in 5-10% [10,11]. The malignant SO was first described by Wetteland in 1956 [5]. Approximately, 94% of the thyroid type carcinomas arising in SO are unilateral and more commonly involve the left ovary whereas in cases of metastasis from primary thyroid carcinoma, ovarian masses are bilateral and have no teratomatous features [7]. Papillary carcinoma (44%), followed by follicular carcinoma (30%) are the most frequent types of malignancy [2,12]. Some literature also categorises SO as 'biologically malignant SO', which has no evidence of histologically malignant change but shows extra-ovarian and distant metastasis [2]. Because of disambiguate exact percentage of malignancy; the term malignant SO has been replaced by a more appropriate term 'thyroid type carcinoma' originating in SO (specifying the type). Papillary carcinoma, being the most common thyroid type carcinoma in SO, can only be diagnosed if it meets the criteria for conventional papillary carcinoma, that is, a true papillary architecture with a central fibrovascular core and lined by crowded, overlapping, round or oval, enlarged, clear nuclei having ground glass appearance, often showing intranuclear inclusions of cytoplasm. The presence of psammoma bodies increases the likelihood of malignancy.

The second most common type of carcinoma arising in SO is follicular carcinoma. Due to lack of capsule in the ovarian lesion, and the inability to demonstrate capsular invasion, the diagnosis becomes more difficult, however the identification of invasion into the surrounding ovarian tissue and vascular invasion or metastasis can lead to the diagnosis.

The evidence of peritoneal implants in the absence of an identifiable thyroid type carcinoma has been termed as 'peritoneal strumosis', but the use of this term is still controversial because some authors use this term interchangeably with 'malignant SO' and studies also show that they can recur. A distinctive form of ovarian teratoma, 'struma carcinoid' with a mixture of thyroid tissue and carcinoid has also been reported. To come to a diagnosis, use of immunohistochemistry, such as Thyroid Transcription Factor-1 (TTF-1), thyroglobulin and neuroendocrine markers such as chromogranin and synaptophysin can be employed. Metastasis, being uncommon in malignant SO, is seen in 5-23% cases [5].

The recent literatures have suggested a higher rate of recurrence. Salman WD et al., studied 54 cases of Struma, 41 of them were 'proliferative struma' and 13 were 'malignant struma' [7]. Makani S et al., reviewed a total of 39 cases of malignant SO, which have been reported till 2004, and they found nine cases with metastasis (23%) and six cases with recurrence (15%) after an average time of four years. They recommend surveillance of thyroglobin levels during treatment as well as follow-up for at least 10 years because it is a sensitive marker for monitoring both benign and malignant cases of SO [12]. Well differentiated thyroid-type follicular carcinoma can be confused with benign SO, the follicular variant of Papillary Thyroid Carcinoma (PTC), and highly differentiated follicular carcinoma of ovarian origin (HDFCO). They can be differentiated by recognition of surrounding ovarian tissue, vascular invasions, and metastasis which suggest a malignancy rather than benign SO. The follicular variant of PTC other than follicular carcinoma can be diagnosed by the identification of typical nuclear features of PTC with follicle formation but presence of minimal papillary structures. The diagnosis of HDFCO cannot be made until along with benign appearing thyroid follicles extra ovarian dissemination is also detected.

Metastasis from thyroid carcinoma to the ovary can also simulate primary thyroid type carcinoma however; a detailed clinical history and examination can be of help in differentiating the two. Patients having a primary tumour <2 cm in size, confined to the ovary without any aggressive histological or biological features were defined as ' Low risk patients and were advised unilateral salpingo-ophorectomy and thyroxin suppression therapy whereas, the high-risk patients were indicated for additional total thyroidectomy and radioiodine ablation to eradicate extra ovarian metastasis and tumour burden.

The prognosis for low risk patients is reported to be quite good, with a low recurrence rate of about 7.5% over 25 years in a case series of 57 patients [3]. The average time for recurrence was found to be around six years in a review of 59 case series of malignant so by Jean S et al., [13]. Serum thyroglobulin is a good marker to evaluate treatment effects and recurrence of thyroid carcinoma. Therefore, serum thyroglobulin is regarded as a tumour marker to follow for atleast 10 years.

CONCLUSION(S)

The current case unusually presented with a large sized mass along with raised CA-125 levels which raises the suspicion of epithelial tumours. Accuracy of diagnosis will improve with better recognition and knowledge of the wide range of macroscopic and histological features of struma. More precise description of the spectrum of histological changes using terminology applicable to the thyroid gland as well as accurate diagnosis and (although this case was lost to follow-up) a long term follow-up accompanied by serum thyroglobulin estimation will improve the outcome of the disease and aid in diagnosing recurrence.

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