

Cystic Struma Ovarii – A pathological rarity and diagnostic enigma

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Abstract

Struma ovarii, a rare ovarian neoplasm, is a monophyletic teratoma composed predominantly of thyroid tissue. It accounts for less than 5% of mature teratomas. Cystic struma ovarii is a rare variant wherein the thyroid component could be minimal in contrast to struma ovarii which has more than 50% of thyroid tissue. Diagnostic difficulties may arise if the Struma ovarii is either cystic or co-exists with any other cystic ovarian tumor. The dilemma gets worse when the tumor reveals only a few typical thyroid follicles and the gross examination shows a multi-loculated cyst with mucoid content. Extensive tissue sampling becomes mandatory in such cases to confirm cystic Struma ovarii and its co-existence with another cystic ovarian neoplasm.

We report one such rare occurrence of an ovarian tumor with co-existent cystic Struma ovarii and Mucinous cystadenoma. The case is reported for its rarity and for the diagnostic challenge encountered.

Keywords: Cystic Struma ovarii, Mucinous cystadenoma, Germ cell tumor, Thyroid tissue

Introduction

Struma ovarii or specialized monodermal teratoma is an ovarian neoplasm of germ cell origin composed predominantly of mature thyroid tissue. It is a rare tumor which comprises 1% of all ovarian tumors and 2.9% of mature teratomas.⁽¹⁾ Cystic type of Struma ovarii is a distinctive variant and may create diagnostic dilemmas because of its rarity and also because of presence of minimal quantity of thyroid tissue, thus resulting in confusion with other cystic ovarian tumors. It may be all the more difficult to identify a coexisting neoplasm unless extensive sampling is undertaken. Co-existence of Cystic struma ovarii and Mucinous cystadenoma, despite being rare, has been reported in literature.^(2,3) The present case report highlights a rare co-existence of Cystic struma ovarii and Mucinous cystadenoma in a unilateral ovarian neoplasm. The co-existence of two tumors with different histogenesis in a neoplasm as in the present case is a pathological entity rarely encountered. In the present case, Mucinous cystadenoma which is of surface epithelial origin is seen co existing with Cytic struma ovarii which is of germ cell origin.

Case Report

A 48-year old female patient presented to the Gynaecological department with menorrhagia of 4months' duration. Per vaginal examination revealed a polypoidal, firm, mobile mass which was felt in the right fornix and measured 8x7cms. Multi-Dimensional Computed Tomography revealed a well-defined, right ovarian multi-loculated lesion with an enhancing mural nodule and calcification suggestive of a Serous cystadenoma. Following this, total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed and the specimen submitted for histopathological examination.(Fig. 1)



Fig. 1: Gross findings uterus with cervix, normal left adnexa and right sided cystic ovarian mass

Gross examination findings: Uterus was enlarged and on sectioning, showed an intramural fibroid measuring 1cm.across. Right ovary was enlarged and cystic and on sectioning showed multiloculations and oozed out mucinous material. (Fig. 2)

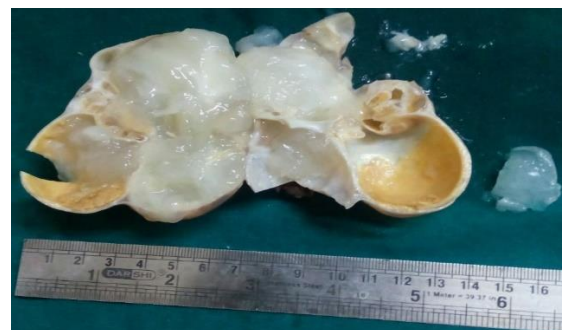


Fig. 2: Cut section of ovarian cystic mass showing multiloculations with inspissated mucoid material

Microscopic findings: Myometrium showed an intramural leiomyoma. Two sections from the right ovarian cystic mass showed a cyst with thickened wall

consisting of thyroid follicles filled with colloid. Sections from the other areas showed a cyst lined by tall, columnar, mucin-secreting epithelium. (Fig. 3 & 4)

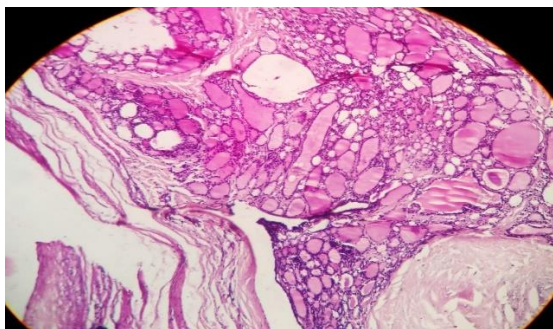


Fig. 3: Photomicrograph showing thickened cystic wall composed of thyroid follicles of varying sizes filled with variable amounts of colloid (10x H&E)

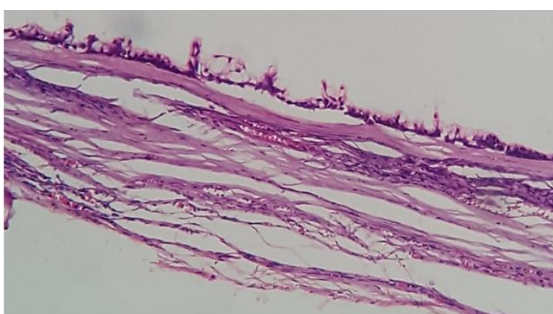


Fig. 4: Photomicrograph showing cyst wall lined by mucin secreting tall columnar epithelium (10x H&E)

Final Diagnosis: Cystic Struma Ovarii associated with Mucinous Cystadenoma in right ovarian cyst.

Discussion

Struma ovarii is a specialized type of monodermal teratoma of germ cell origin.⁽⁴⁾ It is the expression of dominant growth of thyroid tissue in teratoma sometimes to the exclusion of other components.⁽⁵⁾ It is composed predominantly of thyroid tissue and accounts for less than 5% of mature teratomas.⁽⁶⁾

Cystic variant of struma ovarii is an extremely rare lesion and difficult to identify since the quantity of thyroid follicles is minimal and the cystic changes may predominate and mislead the diagnosis.^(7,8) Review of literature revealed that exact incidence of cystic struma ovarii has not been mentioned in any of the earlier studies. One may easily miss the entity totally unless extensive sampling of the entire specimen is undertaken.⁽⁹⁾ On the contrary, in the present case, struma ovarii was diagnosed in the initial tissue sections though the thyroid tissue was minimal. Since the gross examination showed a multiloculated cyst with insistent mucoid material, extensive tissue sampling was undertaken which unveiled a co-existing mucinous cystadenoma. Though the discovery of two co-existing benign neoplasms in ovary did not significantly alter the

management of the patient in anyway, it definitely was a histological delight.

Struma ovarii occurs at a higher age than common mature teratomas. The peak age incidence is reported in the 5th decade but a few cases have also been reported in pre-pubertal and post-menopausal women.^(10,11,12) It usually presents as a palpable abdominal mass, vaginal bleeding and lower abdominal pain as in the present case.^(13,14,15) In literature, 5% to 8% of cases with struma ovarii have features of thyroid hyperfunction which was not seen in our study.^(6,16,17)

They are usually unilateral as in the present case.⁽¹⁾ They range from very small to large lesions growing upto 10cm diameter.⁽¹⁾ Bilateral struma ovarii is very rare and is seen in only 5% of cases.⁽¹⁸⁾

The differential diagnosis for the mucinous cystadenoma component which was considered in the present case was, predominant endodermal differentiation in a mature teratoma associated with struma ovarii. But since the thyroid tissue component was minimal and the rest of the tissue sections showed classic features of mucinous cystadenoma with supportive gross diagnostic features and the diagnostic features of teratoma were absent, this possibility was ruled out.

Szyfelben in their report of 20 cases observed that cystic struma ovarii is a frequently unrecognized tumor owing to the minimal thyroid component which may be missed. They also opined that the reported cases of struma ovarii associated with cystadenoma probably represented an exclusively thyroid type cyst.⁽¹⁹⁾

Some studies have confirmed the thyroid nature of the tissue within the cystic ovarian mass by immunohistochemical positivity for TTF-1 and thyroid hormones.⁽²⁰⁾ The thyroid tissue may show normal thyroid architecture or features of nodular hyperplasia associated with hyperparathyroidism,⁽²¹⁾ Thyroiditis or even Papillary carcinoma⁽²²⁾ and Follicular carcinoma sometimes associated with peritoneal spread referred to as peritoneal strumatosis^(23,24) and Malignant Lymphoma.^(25,26) However, the present case did not show any such association. Some mucinous tumors appear to be derived from teratomas.⁽²⁷⁾ Struma ovarii can be associated with mucinous cystadenoma, Brenner tumor or carcinoid tumor.⁽²⁶⁾

Very occasionally, struma ovarii may present as a mixed tumour containing both carcinoid and strumal(thyroid) elements in which case it is referred to as strumal carcinoid.⁽²⁷⁾

Any type of mucinous tumor can be associated with a teratoma. Two large study series showed 41 mucinous cystadenomas, 44 borderline mucinous tumors and 11 mucinous carcinomas.^(2,3)

We opine that, macroscopic examination alone may not provide any clues to the presence of thyroid tissue which is a purely histological feature whereas, mucinous nature of the neoplasm may be suspected on gross

examination though the absolute confirmation is based on histopathological features.

Conclusion

Cystic struma ovarii being a rare entity with minimal thyroid component may be easily missed unless extensive tissue sampling is undertaken to quantify it and also to look for any co-existing neoplasms. We recommend that Cystic struma ovarii should be considered in the differential diagnosis of all ovarian cystic lesions.

This report highlights the importance of histopathological examination which is the gold standard in the diagnosis of Cystic struma ovarii with minimal thyroid tissue.

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