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Struma Ovarii Masquerading as a Mucinous Ovarian Cyst

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ABSTRACT

Struma ovarii is an uncommon ovarian teratoma distinguished by the unusual presence of thyroid tissue. The vast majorities are benign, and ~5% are malignant. Clinical symptoms, laboratory investigations, imaging modalities are nonspecific. Even histopathologic features may be misleading in a few cases. It is defined as a teratoma composed exclusively of or containing more than 50% thyroid tissue. Common differentials include serous and mucinous cystadenoma. Awareness of existence of such entities may help in utilization of available histologic and imunohistochemical tools for correct diagnosis. Oophorectomy is the treatment of choice for benign struma.

Key words: Struma ovarii, thyroid, teratoma, cystadenoma

INTRODUCTION

Struma ovarii is an uncommon ovarian teratoma distinguished by the unusual presence of thyroid tissue. It accounts for 2.7% of all ovarian teratomas and 1% of all ovarian tumors. [1,2] It is defined as a teratoma composed exclusively of or containing more than 50% thyroid tissue. The vast majorities are benign, and ~5% are malignant. [3] Clinical symptoms, if present, are mostly nonspecific. Laboratory investigations including CA-125 provide little value in diagnosis. The different imaging modalities including ultrasound, CT and magnetic resonance imaging are nonspecific and often suspect of malignancy. Usual radiological features include a large complex multilobulated mass with thick septa and diagnosis is usually made after surgery, based on histopathological findings. [4] Mucinous neoplasms of the ovary account for 10%–15% of ovarian neoplasms. The large majority are benign or borderline, accounting for 80% and 16%–17%, respectively. Mucinous neoplasms of the ovary most commonly affect women in their 20s to 40s and manifest as large unilateral pelvic masses. The imaging usually shows a unilateral multiloculated pelvic mass. [5]

CASE PRESENTATION

40 years old woman from Maharashtra, an Indian state presented in the casualty department with pain in abdomen increasing in intensity since one week. Ultrasonography revealed a partially twisted huge ovarian cyst. An emergency laparotomy with salpingo-oophorectomy was performed. The specimen was sent to a nearby Pathology laboratory from where it was transferred to our centre for histopathologic examination.

The gross examination revealed a 1.3 kg ovarian cyst measuring 16 cms in greatest diameter. The capsule was intact with smooth mildly congested external surface. The cut surface revealed a multiloculated cyst filled with thick almost solidified greenish white material which was assumed to be inspissated mucin by the prosector. After removing this

material thoroughly variably sized locules ranging in size from 1 cm to 8 cms were seen separated by thin walled septa. No solid or papillary areas were present. The fallopian tube was mildly congested. Extensive sampling was performed.

The microscopic examination showed multiple locules with a very thin fibrocollagenous cyst wall lined by flattened to low cuboidal epithelium. An occasional focus lined by mucin containing tall columnar epithelium was noted. A single nodule measuring 6 x 4 mms in size revealed closely packed small cysts. Few ectatic small vessels were present. No other elements were found. The fallopian tube had a few hemorrhagic areas.

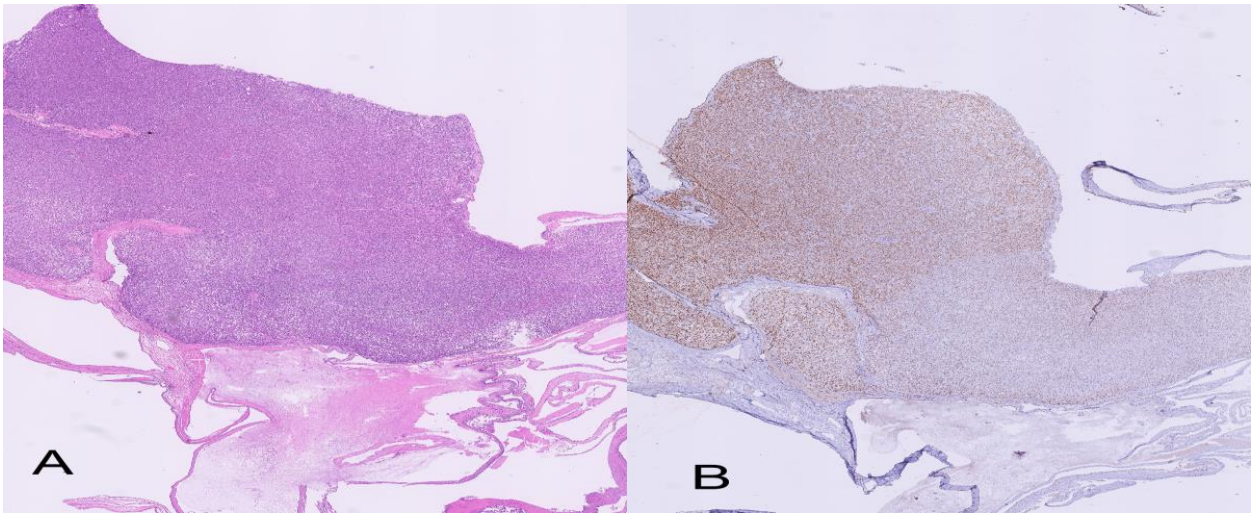

Figure 1: The nodule showing closely packed thyroid follicles with adjacent cystic areas A: H and E stain, B: Positive TTF-1 immunohistochemistry in both nodule and flattened lining of adjacent cysts. 5X

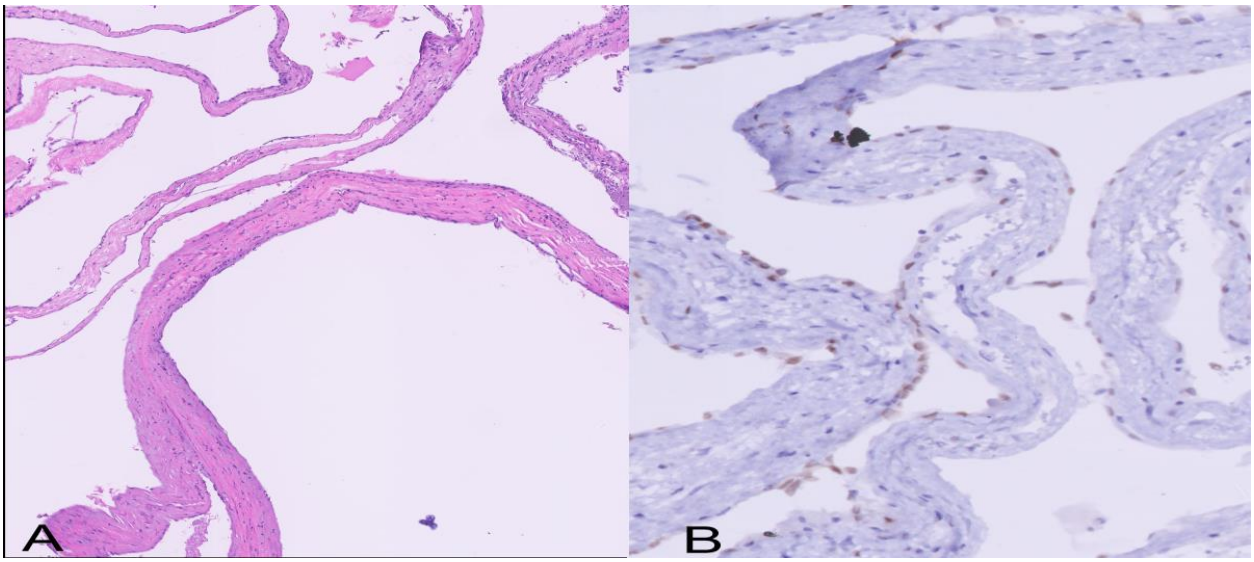

Figure 2: Cystic areas lined by flattened epithelium A: H and E stain, B: Positive TTF-1 immunohistochemistry. 20X

With such a picture, the diagnosis of a benign mucinous cystadenoma with features of torsion was considered most likely. The flattening of lining, lack of intracellular mucin in majority of the cyst and lumen filled with thick fluid resembling colloid aroused suspicion of thyroid tissue. The single nodule having small closely packed cysts was further investigated

with immunohistochemistry (IHC) to rule out possibility of associated component of Sertoli cell tumor or thyroid tissue. Inhibin IHC was negative, while TTF-1 was positive in not only the small nodule but also in the flattened to cuboidal lining of surrounding cysts. Based on IHC, the whole cyst being of thyroidal nature was concluded with a tiny component of mucinous cystadenoma. The proteinaceous fluid initially misunderstood to be mucin was in reality colloid. The final diagnosis of a benign Struma Ovarii was made.

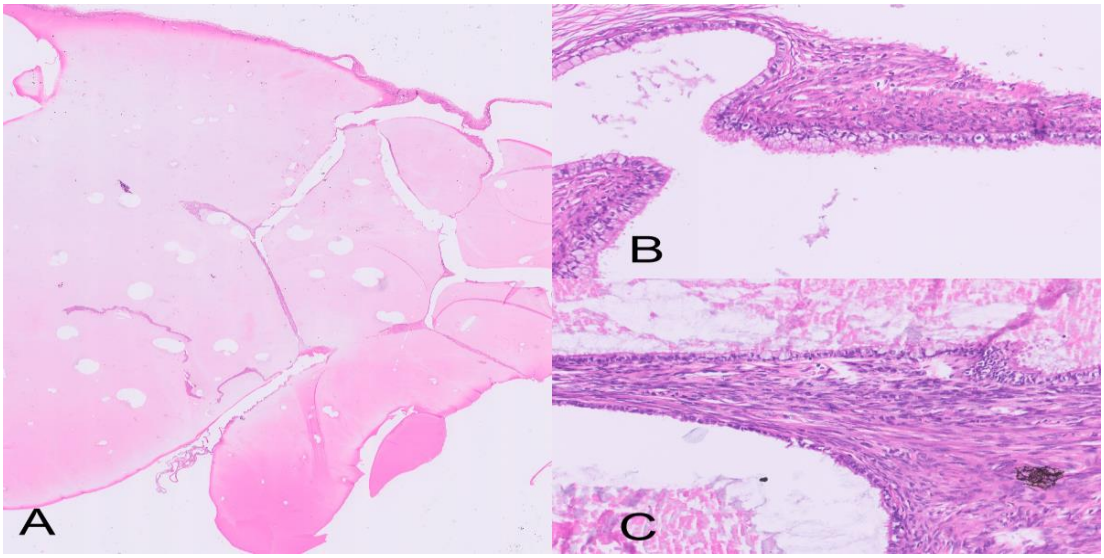

Figure3: A: low power view of cystic areas lined by flattened epithelium, H and E stain 2X. B and C: Cystic areas lined by mucin secreting columnar epithelium. H and E stain 20X.

DISCUSSION

Struma ovarii is a rare histological diagnosis. In a radiology study Saveli et al mention that none of 31 patients of Struma ovaii in their study were diagnosed in ultrasound examination. [6] It is also called a monodermal ovarian teratoma where thyroid tissue predominates and constitutes >50% of the component. This tumor was first described in 1889 by Boettlin. [4] Thyroidal tissue in teratomas is usually present with other components derived from all three germ layers. Approximately 5-15 % of mature cystic teratomas show variable proportion of thyroid tissue; however, proportion of thyroid should be more than 50 % to classify it as struma ovarii. Struma ovarii has been reported to coexist with non-germinal epithelial tumors such as mucinous cystadenoma, Brenner's tumor, and serous cystadenoma. [7]

Although the typical presentation is that of an asymptomatic pelvic mass, some may have lower abdominal pain. Unusual clinical manifestations such as hyperthyroidism, ascites, and Meigs' syndrome have been recognized. Mean age at presentation is 45 years. Most women are premenopausal. However it can occur at any age. CA-125 levels do not rise in most cases. [2] The preoperative radiology usually nonspecific and shows cyst with thick septa. Sometimes struma pearl or intracystic high attenuation lesions, indicating the presence of viscid gelatinous colloid material may be noted. [8] Uncommon macroscopic and especially histological patterns in struma can cause difficulties in diagnosis.

The usual size of these cystic struma ovarii is less than 10 cm however can vary from 3 cm to 20 cms. [9] They have smooth mildly congested capsule. Cut surface shows

multiloculated cyst with thick septa. Few solid areas may be seen. The lumen is filled with greenish brown colloid. Our case had similar findings.

Common differentials include serous cystadenoma which can have similar low cuboidal lining and mucinous cystadenoma from which these are distinguished by a mucin containing tall columnar lining. More so the struma ovarii can co-occur with these cysts, Brenner tumor etc. [10] both differentials were excluded in our patient by a positive TTF-1 Immunohistochemistry. Focal mucinous epithelium was present in our case. Most cases are benign however malignant transformation commonly papillary carcinoma and follicular carcinoma are reported.

Surgical resection remains the main primary treatment. In the case of benign struma ovarii, no further treatment other than the unilateral oophorectomy is necessary. Conservative surgery (cystectomy or oophorectomy) is recommended. Most patients with benign struma do not require extended periods of follow-up or postoperative investigations. [11] Our patient had uneventful recovery no post-operative complications on one month follow up.

To conclude, cystic struma ovarii are uncommon tumors of ovary which are easily misdiagnosed clinico-radiologically. Even histopathologic features may be misleading in a few cases. Immunohistochemistry may be helpful in such cases. Most of the cases are benign and oophorectomy is treatment of choice.

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