

Struma ovarii with literatures review, a Case Report

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Abstract

Struma ovarii is a highly specialized form of ovarian teratoma, characterized by the presence- entirely or predominantly- of mature thyroid tissue. Its most important complications, although rare, are malignant transformation and thyrotoxicosis. In the present 'case report' we describe a case of a 25 year old married woman that seeks medical advice for recurrent lower abdominal discomfort. The diagnosis of complex ovarian cyst confirmed by ultrasound examination of

the abdomen, the operative findings were multilocular ovarian cyst treated by oophrectomy. The histological examination demonstrated typical elements of mature thyroid tissue that confirmed the diagnosis of struma ovarii.

Keywords: ovarian teratoma, struma ovarii, thyroid tissue

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Introduction

Teratomas are tumors with more than one cell type which originate from more than one germ layer. These cells may differentiate into any tissue of the body including hair, teeth, fat, skin, muscle and others. These bizarre tumors are usually located at the midline and paraxial regions of the body. One of the most common locations is the ovary⁽¹⁾.

Ovarian teratomas make up about one – fifth of all ovarian tumors. Most are cystic and benign; 10 % are bilateral. They occur mostly during the childbearing years, typically in the mid – 30s. About 10% are diagnosed during pregnancy⁽²⁾.

Ovarian teratomas include mature cystic teratomas (dermoid cysts), immature teratomas, and monodermal teratomas eg, struma ovarii, carcinoid tumors, and neural tumors . About 1% to 2% of teratomas

may undergo malignant transformation and When malignant transformation occurs, it usually in women older than 40 years⁽³⁾.

Struma ovarii (literally: goiter of the ovary)

Is a rare form of monodermal ovarian teratoma characterized by the presence – entirely or predominantly – of mature thyroid tissue, presenting the same as thyroid gland, with physiological and pathological changes. Simply presence of thyroid tissue with coexistence and predominance of other cell types does not confirm the diagnosis of Struma ovarii⁽⁴⁾.

Typically, struma ovarii occurs as a part of benign cystic teratomas, but may occasionally be encountered with other ovarian tumors, either germinal as dermoid cysts and carcinoid tumors or nongerminial as serous or mucinous cystoadenomas and Brenner tumors⁽⁵⁾.

History and presentation of the case

25 years old married woman had 2 children seeks medical advice in May 2009 for lower abdominal pain and discomfort. There was no history of fever, weight loss, jaundice or bleeding per vagina and there were no symptoms of hyperthyroidism.

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On physical examination there was no pallor, icterus or lymphadenopathy.

Investigations revealed normal haemogram and serum biochemistry. Chest x ray was normal and diagnosis of multilocular cystic mass confirmed by ultrasonography in AL Karama Teaching Hospital/ Wassit Governorate.

During the intraoperative exploration of the abdominal cavity, a multilocular ovarian cyst of the right ovary was found and a right

oophorectomy was performed and biopsy of the left ovary done.

Pathology

Gross examination: right ovary; multilocular cystic mass measured 12* 10 * 9 cm grey smooth slightly thickened outer surface. (Figure 1, Figure 2). Cut surface was multilocular cyst. The cyst filled with grey shiny gelatinous material; the lining of the cyst was smooth. There were no solids areas or intramural nodules within the cyst.



Figure 1&2: multilocular ovarian cystic mass filled with shiny materials.

Microscopical examinations: patient record no. 2622.....May 2009

Compressed ovarian architectures in few sections with replacement of the rest of the ovarian parenchyma by benign colloid filled thyroid follicles (Figure 3&4). No cytological feature

of malignancy was seen and no cartilage or skin or neural tissue was seen. Diagnosis in favor of mature cystic teratomas with benign thyroid follicles contain colloid materials (mnodermal teratma; struma ovarii)

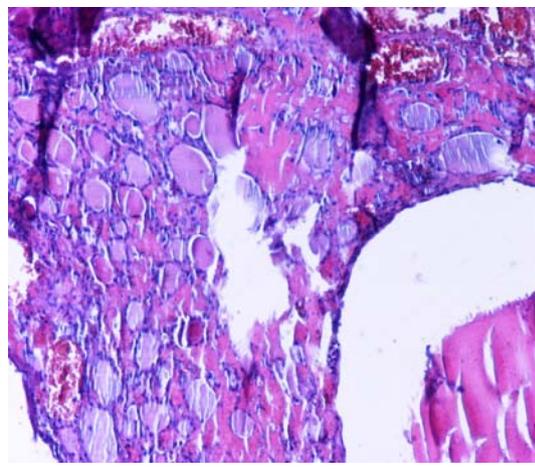
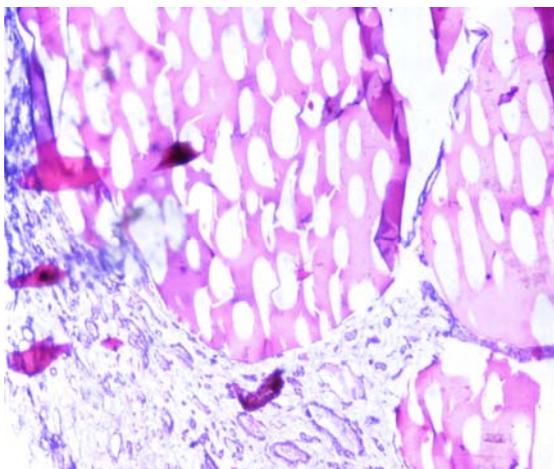


Figure 3&4: Microscopical examinations; thyroid follicles filled with colloid

Discussion

Struma ovarii is a variant of ovarian teratoma in which thyroid tissue components is the major constituent. Thyroid tissue is observed not uncommonly in 5-15% of teratoma, but to qualify as a struma ovarii tumor the thyroid proportion must comprise more than 50% of the overall tissue⁽⁶⁾. Struma ovarii is a relatively rare tumor which comprises less than 1% of all ovarian tumors and 2.7% of all dermoid tumors⁽⁷⁾.

This tumor was first described in 1889 by Boettlin, who observed the presence of thyroid follicular tissue in ovaries, and further reports thereafter were published by Gottschalk⁽⁸⁾. However, due to rarity of this type of tumor there has been a paucity of data in the past literature pertaining to diagnosis and treatment of this tumor. Its pathogenesis remains controversial. Today, it is considered that struma ovarii is composed of mature thyroid tissue growing within ovarian teratomas. Although approximately 15% of ovarian teratomas contain a small, non-significant focus of thyroid tissue, only 0.8-3% are characterized by the presence of functional thyroid tissue or thyroid tissue occupying most of the mass, classified as struma ovarii⁽⁹⁾.

Its incidence varies in different studies. In study of Higuchi et al, published in 1960, reports 3 cases among 1000 solid ovarian tumors (0.3%). In a recent review of 282 ovarian tumors, 2 cases of struma ovarii have been reported (0.7%)⁽¹⁰⁾. Struma ovarii is usually presented during reproductive life and rarely before puberty. The tumor always occurs as a pelvic mass, which may be palpable on physical examination, depending on size and location. Most cases are incidentally found during clinical and imaging examination or laparotomy. Preoperative diagnosis of

struma ovarii is reported rarely, usually in patients with symptoms of hyperthyroidism. The diagnosis can be made by radiological work-up, including CT scan, MRI and I¹³¹ sintigraphy. At this point must be underlined that struma ovarii presents some characteristic MRI findings of a multilobulated complex mass with thickened septa, multiple cysts of variable signal intensities and enhancing solid components⁽¹¹⁾. Struma ovarii is a rare tumor in its pure form, but its true incidence is hard to estimate because of all the variation in the diagnostic criteria due to the fact that some authors reported it as within a teratoma and others only in its pure form. In general, it is an asymptomatic tumor, "benign-like" in most of the cases, and the diagnostic is based only on the histopatological findings⁽¹²⁾. Struma ovarii in our patient was benign and unilateral involving the right ovary and the thyroid hormonal status was in normal range after operation. The tumor is usually nonfunctional and only 8% of patients' present symptoms and signs of hyperthyroidism, as a result of autonomous activation of its thyroid tissue. The surgical removal of struma ovarii in such cases usually results in disappearance of symptoms, although in rare cases may lead to exacerbation of hyperthyroidism because of the release of TSH receptors stimulating antibodies postoperatively⁽¹³⁾. Malignant transformation of struma ovarii is rare (5-10%). Malignancy is defined by various criteria in different studies, principally differing on classifying struma ovarii as either an ovarian or as a thyroid tumor. Most cases of malignant struma ovarii have been diagnosed on the basis of histological criteria alone, with only about 20 cases presenting clinically appreciable metastatic disease. The

diagnosis of malignancy relies on the basis of cytologic atypia, vascular or capsular invasion, or metastases, like in other ovarian neoplasms, has not been universally accepted, since most authors advocate that malignancy should follow the same guidelines as those for thyroid cancer.

Metastatic spread, following pattern of ovarian cancer, occurs in about 5% of malignant cases. In these patients, there may be local implantations, lymphatic metastases to the omentum, liver or mesentery, as well as distal blood metastases to bones, brain or lungs⁽¹⁴⁾.

Struma ovarii generally appears as a multilocular, encapsulated mass, solid and/or cystic on gross examination. The microscopic examination reveals typical rounded thyroid follicles filled with pink staining, homogenous, gelatinous colloid, lined with monoptychial cuboid or columnar epithelium and separated with internal septations.

In some cases, microfollicles of fetal adenomas type may be found. Malignant transformation of the thyroid tissue may be follicular, papillary, or mixed in pattern, and in rare cases can include elements of cystadenocarcinoma, Brenner tumor, carcinoid or melanoma. The positive immunohistochemical staining for thyroglobulin, T3 and T4 confirms the diagnosis of struma ovarii⁽¹⁵⁾.

Because of its rarity, there is no consensus on struma ovarii treatment. Each case must be managed individually. Definitive therapy depends on the extent of the disease and the future childbearing wishes of the patient.

Simple salpingoophorectomy is the therapy of choice for the vast majority of patients, since most cases are unilateral and benign. Total hysterectomy with bilateral salpingoophorectomy is indicated for

bilateral tumors or in postmenopausal patients. In cases of malignant transformation, a combination of complete tumor resection, total thyroidectomy and adjuvant I¹³¹ ablation is usually mandatory; since there is evidence that struma ovarii behaves like its thyroid counterparts⁽¹⁴⁾.

If evidence of peritoneal metastases is present, appropriate debulking is indicated.

Fertility-sparing surgery should be considered in patients who desire preservation of fertility, if disease is confined to the ovary. In these cases, the initial approach must be followed, after completion of childbearing, by definitive surgery.⁽¹⁵⁾

It was concluded that Struma ovarii is a rare mature ovarian cystic teratoma and the preoperative diagnosis of which is difficult. To our knowledge it is the first reported case in our territory.

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