Struma Ovarii

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Citation

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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.
The present case concerns a 75 year old woman that underwent elective open cholocystectomy because of symptomatic cholelithiasis. During the intraoperative investigation of the abdominal cavity, a left adnexal tumor was found incidentally and, a left salpingoophorectomy was performed complementary to cholecystectomy. The histological examination demonstrated typical elements of mature thyroid tissue and the positive immunohistochemical staining for thyreoglobulin confirmed the diagnosis of struma ovarii.

INTRODUCTION

Struma ovarii is a rare ovarian tumor characterized by the presence-entirely or predominantly-of mature thyroid tissue, presenting the same as the thyroid gland, with physiological and pathological changes. The simple presence of thyroid tissue with coexistence and predominance of other cell types does not confirm the diagnosis of struma ovarii[1].

Typically, struma ovarii occurs as a part of benign cystic teratomas, but may occasionally be encountered with other ovarian tumors, either germinal as desmoid cysts and carcinoid tumors or nongerminal as serous or mucinous cystoadenomas and Brenner tumors[2].

OUR CASE

A 75 year old woman underwent elective open cholecystectomy, through a right subcostal incision, because of symptomatic cholelithiasis, estimated by ultrasonography. During the intraoperative exploration of the abdominal cavity, a multilobular, elastic tumor of the left adnexal was incidentally found and a left salpingoophorectomy, through a median subumbilical incision, was performed complementary to cholecystectomy.

The sized 9x9x6cm tumor, on gross examination, was mainly solid and partly cystic filled with a yellowgraywish gelatinous material (Fig. 1, 2).

Figure 1

Figure 1: Multilobular tumor , sized 9x9x6cm (pathological specimen)

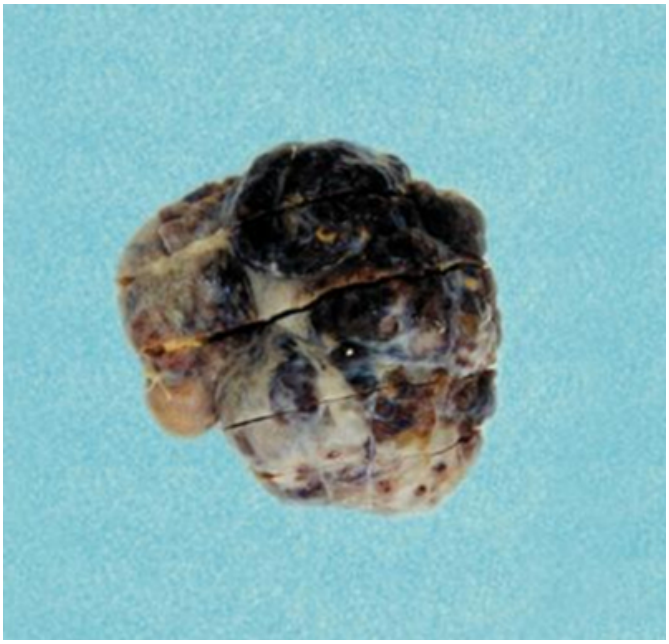
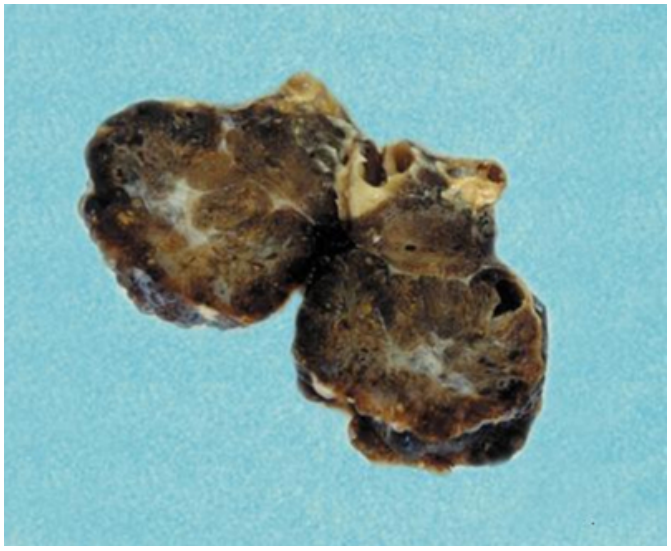


Figure 2

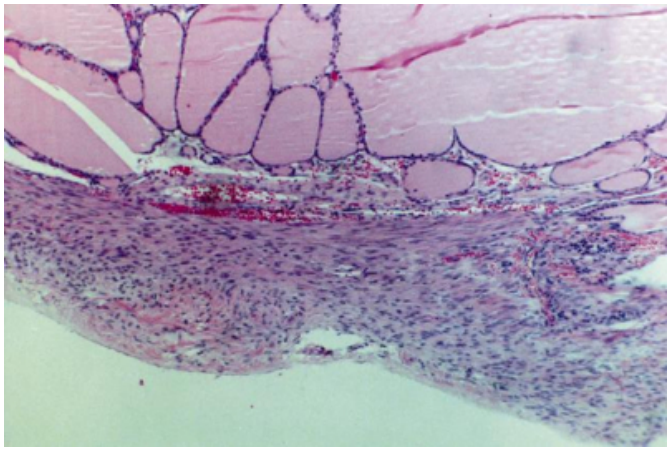
Figure 2: The tumor, on gross examination, was mainly solid and partly cystic filled with a yellowgraywish gelatinous material (pathological specimen)



The histological examination demonstrated the presence of mature thyroid tissue in multiple specimen sections, although there were some areas with follicular dilatations enclosed by ovarian stroma (Fig. 3).

Figure 3

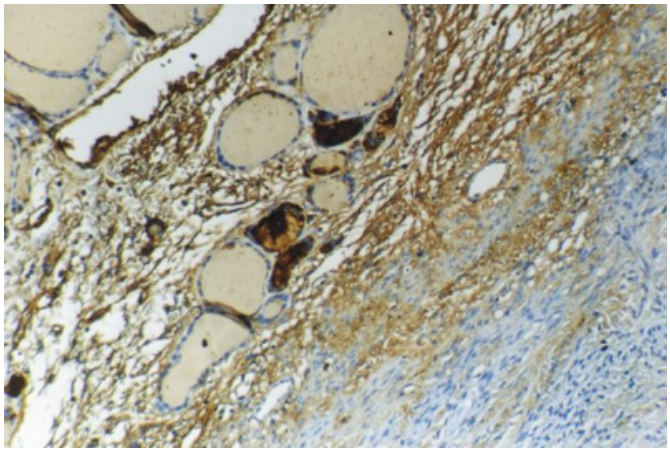
Figure 3: Presence of mature thyroid tissue in multiple specimen sections



The positive immunohistological staining for thyroglobulin confirmed the diagnosis of struma ovarii.(Fig.4).

Figure 4

Figure 4: Positive immunohistological staining for thyroglobulin



The patient's postoperative course was uneventful, with no changes in thyroid function.

DISCUSSION

Struma ovarii was first described in 1899 by Boettlin[3]. Its pathogenesis remains controversial. Today, it is considered that struma ovarii is composed by 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[4].

Its incidence varies in different studies. A Japanese 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%)[5].

Struma ovarii is usually presented during reproductive life and rarely before puberty.

No special correlation exists between tocous and struma ovarii growth. Cases are equally reported amongst childbearing and not childbearing women.

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, as in our case.

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 I^{131} 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[6].

In addition to symptoms and signs caused by the presence of a pelvic mass, struma ovarii may be associated with a number of unusual clinical manifestations (Tabl. 1).

In about 5% of cases, struma ovarii is associated with pleural effusion and ascitis (Pseudo-Meigs' syndrome)[7].

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[8]. The surgical removal of struma ovarii in such cases usually results in resolution of symptoms, although in rare cases may lead to first appearance or exacerbation of hyperthyroidism. Today it is considered that TSH receptors stimulating antibodies release is implicated in postoperative hyperthyroidism pathogenesis[9].

In a recent Brazilian study, a case of clinical hypothyroidism following struma ovarii resection, of a previously asymptomatic woman, is reported for first time[10].

Malignant transformation of struma ovarii is rare (5-10%)[2], [11], [12].

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 histologic criteria alone, with only about 20 cases presenting clinically appreciable metastatic disease[11], [12]. The diagnosis of malignancy 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[11], [13].

Metastatic spread, following pattern of ovarian cancer, occurs in about 5% of malignant cases[8]. 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[8], [14].

Struma ovarii generally appears as a multilobular, encapsulated mass, solid and/or cystic on gross examination. The microscopic examination reveals typical rounded thyroid follicles filled with pinkstaining, homogenous, gelatinous colloid, lined with monoptychial cuboid or columnar epithelium and separated with internal septications. In some cases, microfollicles of fetal adenomas type may be found[15].

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 possitive immunohistochemical staining for thyroglobulin, T3 and T4 confirms the diagnosis of struma ovarii.

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 salpingooophororectomy is the therapy of choice for the vast majority of patients, since most cases are unilateral and benign. Total hysterectomy with bilateral salpingooophorectomy is indicated for bilateral tumors or in postmenopausal patients. In patients with thyroid involvement, concomitant thyroidectomy has been advocated.

In cases of malignant transformation, a combination of complete tumor resection, total thyroidectomy and adjuvant I^{131} ablation is usually mandatory, since there is evidence that struma ovarii behaves like its thyroid counterparts[11]. If evidence of peritoneal metastases is present, appropriate debulking is indicated[15]. Fertility-sparing surgery should be considered in patients who desire preservation of fertility, if disease is confined to the ovary[11], [16]. In these cases, the initial approach must be followed, after completion of chilgbearing, by definitive surgery.

Sequential I^{131} whole body scans and thyroglobulin level measurements are used postoperatively for recurrence and active metastases detection[11], [17].Repeat treatment is still therapeutic if recurrence is detected. Repetitive doses of I^{131} ablation can lead to extended disease free survival. Recombinant human TSH (rhTSH) is occasionally used before I^{131} administration to achieve a concentration of I^{131} activity in the tumour high enough for a significant cytotoxic effect, because I^{131} uptake by most of thyroid carcinomas is quite low in absence of high levels of endogenous TSH[18].

In their vast majority, cases of malignant transformation concern well differentiated carcinomas, with long survival

(15-20 years). Postoperative adjuvant external radiotherapy and chemotherapy are reserved only for the very rare cases of anaplastic or Hürthle cell carcinomas[14].

Figure 5

Table 1: Clinical Presentation Of Struma Ovarii

■ Incidental finding
(In the majority of cases)
■ Symptomatology of pelvic mass
☐ Distension
☐ Abdominal pain
☐ Symptoms caused by neighbouring organs pressure
☐ Irregular menses
■ Associated with pleural effusion and ascites (5%)
(Pseudo-Meigs' syndrome)
■ Clinical hyperthyroidism (8%)
(Approximately 15% of patients present enlargement of thyroid gland, and 25-33% associated and significant thyroid function abnormalities without clinical manifestations)

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