Introduction

Background

Struma ovarii is a rare ovarian tumor defined by the presence of thyroid tissue comprising more than 50% of the overall mass. Most commonly, they occur as part of a teratoma, but may occasionally be encountered with serous or mucinous cystadenomas. Struma ovarii were first described in 1899 and comprise 1% of all ovarian tumors.

Several variants of the tumor exist. Benign strumosis is a rare version of mature thyroid tissue implants throughout the peritoneal cavity. Strumal carcinoid is defined by the presence of carcinoid tissue within a struma and is exceptionally rare. The vast majority of struma ovarii are benign; however, malignant disease is found in a small percentage of cases.

The symptoms of struma ovarii are similar to other ovarian tumors and are nonspecific in nature. The tumor can be characterized by radiological imaging; however, the final diagnosis is made upon pathological and histological examination of the tissue itself. Surgical resection remains the definitive treatment for benign disease, and surgery with adjuvant radiiodine therapy has been shown to be successful in treating metastatic and recurrent disease.

Frequency

United States

Struma ovarii is rare. Approximately 1% of all ovarian tumors and 2.7% of all dermoid tumors are classified as struma ovarii.

Mortality/Morbidity

Malignancy is defined by various criteria in different studies, principally differing on classifying struma as either a thyroid or ovarian cancer. Several other types of tumors, such as Brenner tumor or cystadenoma, may also be found with a struma.

- Malignant change seems to occur in about a third of cases.
- Metastatic spread, which follows the pattern of ovarian cancer, occurs in approximately 5% of malignant cases.

Although the tumor is predominately composed of thyroid tissue, thyrotoxicosis is seen in only 5% of all cases. Only 1 case of thyrotoxicosis resulting from peritoneal strumosis has been reported.

Race

Because of its rarity, no clear racial predilection for struma ovarii has been determined.

Sex

Defined as a tumor of ovarian origin, struma ovarii occurs exclusively in genetic females.

Age

- Struma ovarii typically presents during the reproductive years. The fifth and sixth decades are the ages of peak frequency.
Struma ovarii rarely occurs before puberty.

**Clinical**

**History**
The symptoms of struma ovarii are similar to other ovarian tumors and are nonspecific in nature. They include the following:

- Abdominal pain
- Palpable abdominal mass
- Abnormal vaginal bleeding
- Ascites (Reported in up to one third of cases\textsuperscript{[6]})
- Pseudo-Meigs syndrome (ascites in the setting of hydrothorax) (Reported in fewer than 10 cases)
- Incidental discovery on pelvic imaging or surgery

Rarely, thyroid hyperfunction is the presenting symptom, seen in 5-8% of patients with struma ovarii.\textsuperscript{[2]}

**Physical**
Struma ovarii is difficult to diagnose and physical examination often does not reveal any abnormalities. The tumor may present as a large abdominal mass, which can be palpable on examination depending upon size and location. Patients may also experience expanding abdominal growth and a fluid wave consistent with ascites. Rarely, dyspnea and pulmonary crackles can be indicative of Pseudo-Meigs syndrome.

**Differential Diagnoses**

| All other forms of ovarian neoplasms, both benign and malignant | Hyperthyroidism |
| Ectopic Pregnancy | Metastatic thyroid cancer to the ovary |
| Endometrioma | Physiological ovarian cyst |
| Hydrosalpinx | Tubo-ovarian abscess |

**Workup**

**Laboratory Studies**

- CBC count
- Blood type and screen
- Cancer antigen 125
  - Nonspecific marker elevated in a variety of benign clinical settings, including menstruation, pregnancy, endometriosis
  - Elevated in epithelial ovarian, endometrial, bowel, breast, and lung cancer
  - Elevated in only 8 cases reported in the literature in the setting of struma ovarii\textsuperscript{[6]}
- Thyroid function tests are ordered only in patients with symptomatic hyperthyroidism.

**Imaging Studies**

- Triple-contrast CT scan of the abdomen and pelvis should be performed to evaluate the extent of disease and the involvement of lymph nodes and other adjacent structures (eg, bowel). Typically, struma ovarii appear as a multicystic mass with no or moderate cystic wall enhancement.\textsuperscript{[7]}
If a triple-contrast CT scan is not available and bowel involvement is suspected, sigmoidoscopy or colonoscopy should be performed.

Pelvic sonography is optional if a CT scan has already been performed. Frequently, this is an initial study.

Mammography should be performed in patients with pelvic masses of unknown origin.

Chest radiographs should be obtained in indicated patients.

In select cases, preoperative evaluation with uptake of sodium iodide I-123 has been performed to demonstrate thyroid uptake in pelvic masses.

### Other Tests

- Papanicolaou test
- Iodine-131 scanning (In patients with suspected struma, this will evaluate active thyroid tissue in the pelvis or abdomen.)
- Thoracentesis in patients with pleural effusion (Cytology may reveal adenocarcinoma in the pattern of malignant thyroid cells.)

### Histologic Findings

On gross examination, the struma is brown or green-brown and solid, but it can also be partly or entirely cystic, filled with gelatinous fluid. The struma is rarely bilateral. Most strumal tissue is not functionally active, and cases associated with thyrotoxicosis can be due to autoimmune stimulation of the normal thyroid gland.

Pathological examination reveals thyroid tissue as the major component of the mass, and is most commonly found in a teratoma. Thyroid tissue may be papillary, follicular, or mixed pattern, and it can include elements of mucinous cystadenocarcinoma, Brenner tumor, carcinoid, or melanoma. Birefringent crystals of calcium monohydrate are present in most patients, which is considered specific for tumors of thyroid origin. Immunohistochemical staining for thyroglobulin, triiodothyronine (T3), and thyroxine (T4) can confirm the diagnosis.

Malignancy is defined by histological features of the tumor including cellular atypia and hyperplasia, nuclear pleomorphism, mitotic activity, and invasion into surrounding vessels or the ovarian capsule. Currently, the pathological criteria used in diagnosing thyroid carcinoma are widely accepted as the standard in diagnosing malignant struma ovarii.[8] However, there is still controversy over the defining characteristics of a malignant struma ovarii tumor.

Malignant struma ovarii is divided into 3 different categories by histology.

- Papillary type is the most common and identified by “ground glass” or overlapping nuclei
- The follicular variant of papillary carcinoma shares the same nuclear characteristics as the papillary type but has a follicular architecture.
- Follicular carcinoma is identified by follicles of mitosis around vascular and capsular structures.[8]

### Treatment

#### Surgical Care

For the vast majority of cases, surgical resection of the ovary is sufficient to treat benign, unilateral disease. A paucity of evidence exists in the literature regarding conservative management in cases with evidence of malignancy. In these patients, serum
thyroglobulin levels can be followed as a marker for recurrence following fertility-sparing unilateral salpingo-oophorectomy. In patients who do not desire future fertility, malignant struma ovarii necessitates surgical staging for ovarian cancer with pelvic washings, total abdominal hysterectomy, bilateral salpingo-oophorectomy, lymph node sampling, total thyroidectomy, and radioactive I-131 ablation. The recurrence rate in patients with malignant struma ovarii who undergo surgery without subsequent radioablation has been cited as high as 50%.

Follow-up

Further Inpatient Care

For patients with a benign struma ovarii, standard surgical follow-up is sufficient.

For patients with malignant disease on surgical pathology, postoperative adjuvant therapy with radio-ablative iodine-131 is recommended. After surgical staging, a thyroidectomy is suggested before adjuvant treatment to potentiate the effects of radioablation. As normal thyroid cells preferentially uptake I-131, thyroidectomy would ensure delivery to the malignant cells. Additionally, a thyroidectomy would provide pathological confirmation that the struma is indeed ovarian in origin.

It is crucial for the surgeon to be aware of the intra- and postoperative complications of thyroidectomy (including hypocalcemia, damage to the recurrent laryngeal nerve, and/or need for postoperative thyroid replacement), and to be comfortable with their management. Radioactive I-131 ablation has been shown to treat malignant disease in both its initial presentation and any subsequent recurrence with excellent efficacy, although the rarity of the disease and lack of data surrounding its long-term management prove challenging to clinicians.

Thyroglobulin is the preferred tumor marker followed in patients with malignant struma ovarii and should be followed sequentially after surgery and ablation. Increases in serum thyroglobulin should be followed up with total body I-131 scanning to detect recurrence, which is treated with subsequent radioablation.

Complications

Significant changes in thyroid function may occur in the immediate perioperative period.

Prognosis

For the vast majority of patients, the struma is benign, and the prognosis is excellent. Even in malignant cases, adjuvant iodine-131 ablation with surgical extirpation has proven curative. Recurrences may be detected using iodine-131 scanning, and repeat iodine radioablation can lead to extended disease-free survival.

In an analysis of 88 patients with malignant struma ovarii, several factors were identified as being associated with recurrence or extraovarian spread. These include adhesions, peritoneal fluid of 1 liter or more, ovarian serosal rent, a papillary histology, or a struma component 12 cm or more. The overall survival rate for all patients is 89% at 10 years and 84% at 25 years.

Patient Education

For excellent patient education resources, visit eMedicine's Endocrine System Center. Also, see eMedicine's patient education article Thyroid Problems.

Miscellaneous

Special Concerns

- Current controversies revolve around the diagnosis of malignancy, ie, whether to classify the struma using ovarian or thyroid carcinoma criteria.

- Because struma ovarii is extremely uncommon, there is no consensus on treatment. Each case must be managed individually.
References


Keywords

teratomatous ovarian tumor, hyperthyroidism, cystadenomas, strumosis, teratoma, pelvic mass, oophorectomy, total hysterectomy, bilateral salpingo-oophorectomy, thyroidectomy

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