

Case Report

Strumma ovarii: a case report and literature review

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ABSTRACT

Struma ovarii is an uncommon monodermic teratoma in which thyroid tissue is the predominant element. Its low prevalence, wide age of presentation, variable clinical manifestations, and the lack of accurate diagnostic methods, contribute to the overall high percentage of misdiagnosis (90%). Surgery is the best available therapeutic option and the prognosis in most cases is good. This article summarizes the most relevant points for the diagnosis, management, and follow-up of this pathology.

Keywords: Strumma ovarii, Ovarian tumor, Teratoma, Papillary thyroid cancer

INTRODUCTION

Among malignant ovarian tumors, there is a group of germ cell tumors, which are associated with great heterogeneity and low prevalence.¹ Ovarian teratomas are the most common of this group and are subdivided into mature, immature, and monodermal teratomas.² Struma ovarii (SO) is classified as a monodermal ovarian teratoma, and 50% of its composition is thyroid tissue. It represents 2.7% of ovarian teratomas and 0.5% of malignant ovarian tumors.²⁻⁴

The etiology of ovarian teratomas is still under investigation, among the most accepted mechanisms are the failure of meiosis I or the fusion of the primary oocyte; failure of meiosis II or fusion of the secondary oocyte and endoreduplication of the haploid genome of a mature ovum, where the origin of the OS is due to errors in meiosis I or II and its ability to spread.

Clinical presentation

SO is a teratoma that is present from birth, but the age of presentation is from the second to the seventh decade of

life. Most patients are asymptomatic, in some cases, it presents as a left unilateral tumor smaller than 10 cm, which is accompanied by lower abdominal pain and abnormal vaginal bleeding.⁵⁻⁶ Less frequently, isolated ascites or as part of a pseudo-Meigs syndrome, symptoms of urinary or intestinal obstruction, infertility, hot flashes, abdominal distension, local pain, thyroid hyperfunction, or hypofunction have been described in the literature.⁴⁻¹²

Diagnosis

Imaging studies are usually the initial diagnostic tool for the evaluation of pelvic masses.⁹ Addley et al reported that in the cases of OS analyzed, the most used imaging study is ultrasound, with which 98.2% of diagnoses were wrong, wrongly classifying the OS as a benign dermoid cyst. The main finding on ultrasound is the presence of "struma pearls". When studying OS through magnetic resonance imaging (MRI) and computed tomography, it appears as a complex multilobulated mass with thickened septa, multiple cysts, enhanced solid components, and a smooth exterior.²⁻⁹ In addition, on MR images, the cysts contain hyperintense and hypointense areas on T1W1 and T2W2, the areas with colloidal material and hormone

content are hypointense on T1W1 and T2W2, and when contrast is applied, the colloid does not enhance and in most cases, tumors exhibit moderate or no wall enhancement. Another important characteristic is that this type of tumor does not present fat.² Tumor markers such as CA-125 and inhibin A have been studied, however, they are not clinically useful due to their poor sensitivity and specificity. The definitive diagnosis is confirmed by histopathological analysis.¹⁰

Treatment

Due to the great heterogeneity and low prevalence of this neoplasm, therapeutic options are very limited, for which the surgical approach is the only therapeutic option and must be individualized for each patient. Therapeutic options range from abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy, and lymphadenectomy, to unilateral oophorectomy with fertility preservation, conservative treatment, and thyroidectomy with radioactive iodine ablation.¹³

Prognosis

If there is no metastasis, the prognosis is good, with an overall survival rate of 89% and 84% at 1 and 25 years, respectively.¹⁴ The most common sites of metastasis are the lung (50%) and bone (25%) and their presence indicates an unfavorable prognosis with a 10-year survival rate of 40%.¹⁵ It has been reported that after surgical resection of a tumor without data suggestive of altered thyroid function, patients presented symptoms of weakness, myalgia, drowsiness, and hypotension, the cause being laboratory-confirmed primary hypothyroidism (Hashimoto's thyroiditis). It is believed that the previous euthyroid state was probably maintained by the autonomous production of thyroid hormones by the struma ovarii.¹⁶

CASE REPORT

A 34-year-old female with a BMI of 58.3, attends the gynecology service due to a history of chronic hypogastric pain, uterine myomatosis, and polycystic ovary syndrome, with 12 and 20 years of evolution, respectively.

She reports 1 year with a progressive increase in symptoms, accompanied by intermittent fatigue, anxiety, panic attacks, tachycardia, photophobia, and intense headache.

On physical examination, a mass was palpated in the right pelvic region with pain on compression. Laboratory studies reported hypertriglyceridemia, hypercholesterolemia, and hypochromic anemia. Ultrasonography described a 10.2×12.5 cm fibroid pedicled in the uterine fundus (Figure 1).

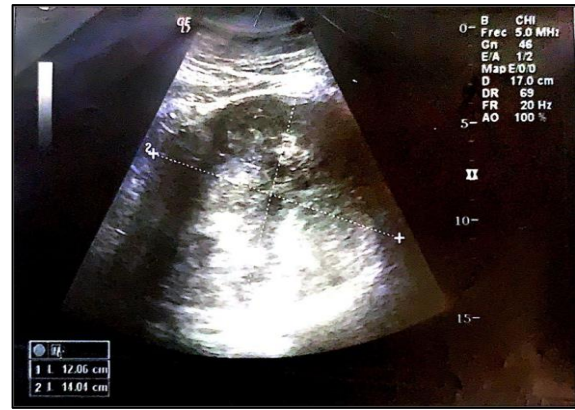


Figure 1: Ultrasound of adnexal mass with measurements of 14×12 cm, heterogeneous echogenicity and partially defined borders. Multiple calcifications in its interior, in addition to a hypoechoic solid component in its periphery, suggestive of teratoma, initially classified as a pedunculated myoma.



Figure 2: Right ovary with ovoid morphology, grayish-brown surface, nodular and with a visible vascular network. Weight 800 grams and dimensions of 15×10×9 cm.

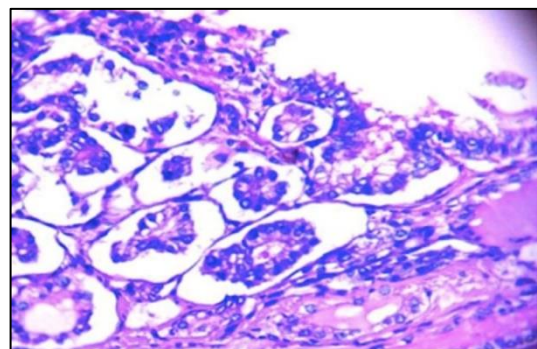


Figure 3: Optical micrograph stained with hematoxylin and eosin, magnification 10X. Mature thyroid tissue is seen with follicles of various sizes, surrounded by hyperplastic cuboidal follicular cells.

It was decided to perform a total abdominal hysterectomy via laparoscopy. During the transoperative period, a mass

was observed in the right ovary with macroscopic data suggestive of malignancy. It was decided to perform a total abdominal hysterectomy with bilateral salpingo-oophorectomy (TAH-BSO).

The histopathological report reports a tumor of 15×10×9 cm, weighing 800 gms, consistent with struma ovarii, with foci of papillary carcinoma and extensive areas of necrosis, an intact capsule, and pathological stage pT1a (Figure 2). Immunohistochemical markers (AE1/AE3, thyroglobulin, TTF-1) were suggestive of struma ovarii; Papillary carcinoma foci were identified by the presence of nuclei with scattered chromatin and an optically empty appearance, enlargement, and clefts (Figure 3, 4).

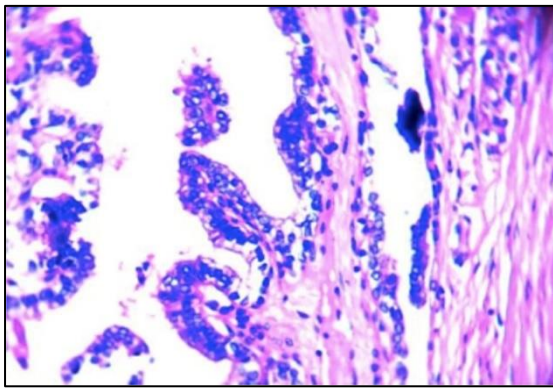


Figure 4: Optical micrograph stained with hematoxylin and eosin, magnification 40X. Focal hyperplasia of the lining epithelium is observed with cylindrical cells with a central, round and basophilic nucleus, forming papillary projections.

At 24 postoperative hours, he was discharged without complications. At 3 months of outpatient follow-up, the thyroid profile is normal, with no signs of hyperthyroidism.

DISCUSSION

The preoperative clinical diagnosis of struma ovarii is a challenge for the surgeon. Sometimes the diagnosis can be facilitated by the presence of symptoms of hyperthyroidism, although this is only found in less than 10% of patients.¹⁰ In most cases, the diagnosis is obtained by histopathological study. The laparoscopic approach can be performed in most cases, although it confers the risk of intra-abdominal tumor rupture with the possibility of dissemination, which would worsen the prognosis.

In cases in which diagnostic laparoscopy is performed and the diagnosis is confirmed postoperatively, it is necessary to perform a second procedure for staging, either by open or laparoscopic technique.¹⁷

Despite the low frequency of this pathology, its risk of malignancy makes it important to keep this possibility in

mind as a differential diagnosis in any pelvic mass under study.

CONCLUSION

OS represents a diagnostic challenge for surgeons. Your suspicion may never present itself and your symptoms may be attributed to other conditions. Misdiagnosis has a direct impact on the patient's quality of life. For this reason, it is recommended that in patients between the second and third decades of life and older women, with single pelvic masses and menorrhagia, struma ovarii be considered within the diagnostic suspicions.

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