Struma ovarii: clinic presentation with Pseudo-Meigs Syndrom Struma ovarii: apresentação clínica com Síndrome de Pseudo-Meigs

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Abstract

Struma ovarii is a form of mature teratoma consisting predominantly of thyroid tissue. In 5% of the cases, it may occur with ascites (20%) and pleural effusion, constituting the Pseudo-Meigs Syndrome. We describe the case of a 62-year-old woman monitored at a Pulmonology consultation for dyspnea on moderate exertion and parapneumonic pleural effusion with a one-year evolution of non-malignant etiology. Ascites and solid adnexal lesion with increased cancer antigen-125 (1475.8 U/ml) were diagnosed . Histological examination revealed a lesion with characteristics of struma ovarii, with no signs of malignancy. Being a benign disease case, the prognosis is excellent.

Keywords: Struma ovarii, Pseudo-Meigs Syndrome, Elevated cancer antigen-125.

Resumo

Struma ovarii é uma forma de teratoma maduro constituído predominantemente por tecido tiróideu. Pode cursar com ascite (20%) e derrame pleural, constituindo o Síndrome de Pseudo-Meigs, presente em 5% dos casos. Descreve-se o caso de uma mulher de 62 anos vigiada em consulta de Pneumologia por dispneia para médios esforços e derrame pleural parapneumónico com um ano de evolução, de etiologia não maligna. Diagnosticada ascite e lesão anexial sólida com *Cancer Antigen-125* aumentado (1475.8 U/ml). O exame histológico revelou lesão com características de *struma ovarii*, sem sinais de malignidade. Tratando-se de um caso de patologia benigna o prognóstico é excelente.

Palavras-chave: Struma ovarii; Síndrome de Pseudo-Meigs; Cancer antigen-125 elevado.

INTRODUCTION

S truma ovarii is a form of mature teratoma consisting predominantly of thyroid tissue¹. It corresponds to about 5% of ovarian teratomas, with a peak incidence between 40-60 years old¹. It represents 1% of ovarian tumors and approximately 3% of all dermoid tumors².

Depending on its histological characteristics, it can be classified as benign or malignant and most of the time it presents with a unilateral adnexal mass³.

The symptoms are variable and may present with pain, abnormal uterine bleeding or pelvic mass in the gynecological examination, with a considerable percentage of asymptomatic patients, being the diagnosis incidentally made by ultrasound or other imaging exam³. In 5% of the cases, it may occur with ascites (20%) and pleural effusion, constituting the Pseudo--Meigs Syndrome. This syndrome is characterized by the presence of pleural effusion and ascites caused by a pelvic tumor other than a fibroma^{4,5}.

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Symptoms and analytical alterations of hyperthyroidism are uncommon, appearing in 5-8% of cases without anatomical changes of the thyroid gland^{1,3}.

The serum marker cancer antigen-125 (CA-125) is used as a classic marker in the surveillance of women treated for epithelial ovarian carcinoma, however with poor specificity⁵. CA-125 elevation increases the suspicion of malignancy and it is described in some cases of malignant struma ovarii. Rarely, it is associated with Pseudo-Meigs Syndrome, being highly suggestive of malignancy^{4,5}. This elevation can be explained by the irritation and subsequent inflammation of the pleura and peritoneum produced by the presence of fluids in these spaces⁶.

From the radiological point of view, it appears as a solid ovarian mass, without specific characteristics of this entity. The diagnosis is histological, with signs of malignancy being found in 5-37% of cases, being more frequent in large tumors⁷. The histological criteria for malignancy are the same as for other types of thyroid tumors¹. The struma ovarii is composed by variably sized macro and microfollicles containing colloid⁸. Cells are flattened to cuboidal/columnar with small round to oval nuclei, even chromatin and pale to eosinophilic cytoplasm⁸. There are no histologic signs of malignancy such as papillary thyroid carcinoma (the most commonly associated malignancy in struma ovarii) cytologically characterized by crowded or overlapping elongated nuclei with irregular contours or chromatin clearing, and papillary architecture⁸. Follicular carcinoma is the second most common malignancy form in this tumor, and its histologic diagnosis implies the demonstration of tumor invasion trough capsule into surrounding ovarian tissue, vascular invasion or metastases^{1,9}.

Metastasis is rare, occurring in 5% of cases, to the lung, bone, liver, and brain.

The treatment of malignant struma ovarii is not consensual and may involve bilateral oophorectomy, total hysterectomy, and adjuvant therapy^{1,3,10}. In the benign form, the surgical tumor removal is sufficient for treatment^{3,10}.

The prognosis is generally favorable, with a 5-year survival rate of 92-96.7%¹⁰. The prognostic factors associated with malignant disease are not entirely known, however, the lesion size and the histological subtype seem to have an impact on the survival¹.



FIGURE 1 and 2. Medium/large pleural effusion on the right, multiloculated at the bases. Thin layer of pleural effusion on the right.

CLINICAL CASE

Female, 62 years old, with a personal history of arterial hypertension, type 2 diabetes mellitus, dyslipidemia and asthma, polymedicated. Multiparous, menopause at age 55, not having undergone hormone replacement therapy. Monitored in a Pulmonology consultation due to dyspnea for medium efforts with one year of evolution, with parapneumonic pleural effusion (Figures 1 and 2), requiring repeated thoracocentesis, with a negative cytological study. Pleural biopsy was performed, which was negative for malignancy. In this context, she underwent positron emission tomography (PET) which, in addition to a large pleural effusion, revealed the presence of ascitic fluid in the pelvis. The patient was referred to a Gynecology consultation to exclude a malignant gynecologic disease, not presenting changes in the gynecological examination.

A transvaginal ultrasound was performed, which revealed a solid lesion of $70 \times 54 \times 28$ mm, in the right ovarian, with peripheric increased vascularity on Doppler examination (score 3) and a slight ascitic effusion in the cul-de-sac. A pelvic magnetic resonance imaging (MRI) was performed, confirming this finding with a description of a solid adnexal lesion measuring $75 \times 60 \times 30$ mm, strongly enhancing contrast, with a low-uptake central region (Figure 3). In the analytical study performed, the patient had increased CA-125 (1475.8 U/ml – reference value of 35 U/ml) and normal thyroid function.

Patient underwent exploratory laparotomy with collection of ascitic fluid and right adnexectomy, having the extemporaneous examination revealed a lesion compatible with struma ovarii. Total hysterectomy and



FIGURE 3. Solid right adnexal lesion in MRI, in an axial plane (red arrow). Lesion measuring 75×60×30 mm, solid, strong contrast uptake, with a low uptake central region.



FIGURE 4. Definitive histological examination in paraffin sections demonstrates an ovarian lesion with characteristics of struma ovarii, without signs of malignancy. H&E stain.

left adnexectomy were performed, with no macroscopic residual disease at the end of the procedure.

The postoperative period was uneventful, with spontaneous resolution of the pulmonary condition.

Histological examination revealed an right ovary measuring 70×60×50 mm with a lesion with characteristics of struma ovarii without signs of malignancy (Figure 4).

Observed in a Pulmonology consultation one month after gynecological surgery with complete resolution of the respiratory condition, having been discharged from the Pulmonology consultation. (Figure 5)



FIGURE 5. Chest radiography, 5 weeks after the surgical procedure, without signs of pleural effusion, spontaneous resolution.

Patient oriented to a Gynecology Oncology consultation for postoperative evaluation, with no complaints and no evidence of gynecological pathology and with normalization of CA-125 values.

DISCUSSION

Struma ovarii is a rare form of mature teratoma consisting of more than 50% of thyroid tissue¹. Most tumors are unilateral and benign, rarely appearing at menopause, with a peak incidence during childbearing age. They are mostly asymptomatic, being an imaging finding³. In the present case, the patient started with respiratory symptoms, being a diagnostic challenge, constituting the Pseudo-Meigs Syndrome. The pathophysiology of pleural effusion can be explained by the drainage of ascites through the diaphragm⁶. In this case, the pleural effusion and its characteristics became the pillar for the differential diagnosis of the patient's pathology. The respiratory condition, despite multiple interventions, had spontaneous resolution after surgical removal of the teratoma. Elevation of CA-125 increases the suspicion of malignancy in the study of adnexal masses and is described in about 5 to 37% of cases of struma ovarii⁷. Rarely, it is associated with Pseudo-Meigs Syndrome, being highly suggestive of malignancy^{4,5}. This elevation can be explained by the irritation and subsequent inflammation of the pleura and peritoneum produced by the presence of fluids in these locals⁶. The patient had a considerable elevation of this marker, despite the benignity of the pathology, thus demonstrating its low specificity for malignancy.

Most of these tumors are functionally inactive, which is why symptoms and analytic changes of hyperthyroidism are uncommon. The patient under study never presented anatomical or analytical changes in thyroid function during her illness.

Imaging diagnostic suspicion is difficult, as it appears as a solid ovarian mass, without specific characteristics, and it is not easy to differentiate it from other types of ovarian tumor. In a postmenopausal woman, a solid mass with ascites and elevation of CA-125 should be assumed to be a high probability of malignancy and probably in an advanced stage. The patient had all these characteristics, with the hypothesis of ovarian cancer being the most likely. The diagnosis of struma ovarii is therefore histological, thyroid tissue is found on microscopic examination and the criteria for malignancy are the same as for other types of tumors of the gland¹. Malignant transformation is described in 5-37% of cases⁷, being more frequent in large tumors (greater than 10 cm) and with more than 80% of stromal tissue¹¹.

Treatment of malignant struma ovarii may involve bilateral oophorectomy, total hysterectomy and adjuvant therapy that may include radioactive iodine^{1,3}. If benign, surgical treatment is sufficient³.

In this clinical case, the initial presentation with pleural effusion became a clinical challenge because the study was guided by the signs and symptoms presented by the patient, who did not present any gynecological complaints. Not being a frequent diagnosis in post-menopause, the atypical presentation made the diagnosis difficult. After surgery, the patient had spontaneous resolution of symptoms. In the presence of pleural effusion, pelvic mass, ascites and elevation of CA-125, although rare, it is possible to have a diagnosis of a benign situation.

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CONFLICTS OF INTEREST

Authors declare they have no conflicts of interest.

INFORMED CONSENT

The patient gave informed consent for publication.

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