

Papillary Thyroid Carcinoma Synchronous with Malignant Struma Ovarii

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ABSTRACT

Struma ovarii (SO) is a rare type of ovarian tumor characterized by containing more than 50% thyroid tissue. Papillary thyroid carcinoma (PTC) associated with malignant Struma ovarii (MSO) is rarely reported. Papillary carcinoma in SO was diagnosed synchronously with PTC. After the diagnosis of MSO, the thyroid was evaluated and a total thyroidectomy was performed, confirming the presence of a PTC. The literature does not present a standardised approach to these cases, emphasising the need for personalised care and the importance of submitting patients diagnosed with MSO to thyroid imaging to identify possible coexisting diseases.

Keywords: Struma Ovarii, Papillary Carcinoma, Thyroid, Ovary, Case Report.

INTRODUCTION

Struma ovarii (SO) is a rare ovarian tumor that contains thyroid tissue. *Ovarian goiter* is a term first described in 1899 that represents just 1% of ovarian tumors. SO can be differentiated from mature teratoma by the amount of thyroid tissue; the former contains more than 50% benign thyroid tissue.[1, 2]. Most SO variants are benign, corresponding to approximately 95% of cases, and reports of malignant tumors are rare, occurring in a small percentage (5-10%) of patients, known as Malignant Struma ovarii (MSO). In these cases, the most common malignant component is papillary carcinoma (PC)[3-5]. MSO associated primary papillary thyroid carcinoma (PTC) are rarely reported, making them challenging to diagnose and treat. Diagnostic and therapeutic approaches must be personalized based on the clinical and pathological characteristics of each patient.

CASE REPORT

A 45-year-old female patient was referred from the interior of Rondônia to the Hospital de Amor de Porto Velho (HA). In February 2023, she underwent a total hysterectomy and bilateral salpingo-oophorectomy. An anatomical examination revealed a MSO. The patient was referred to the Oncogynecology Department for assessment and medical management[6]. CT and MRI of the chest and upper abdomen showed no metastasis. Cervical ultrasonography revealed a suspicious thyroid nodule on the right side (ACR TI-RADS 4). A total thyroidectomy was performed. PTC was found in the right lobe, measuring 1.4 mm, without any complications or extension and free surgical margins, (FIGURE 1) [7].

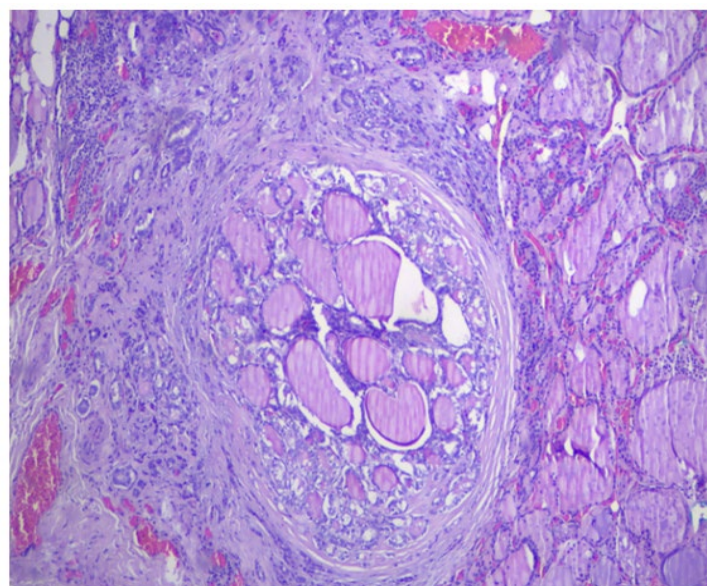


FIGURE 1: Micropapillary PTC in a Thyroidectomy Specimen at Low Power Magnification (4x)

The tumor was classified as an early-stage pT1a. Gene mutation research found the V600 variant in the BRAF gene in struma ovarii [8].

A molecular test (Hereditary Breast and Ovarian Panel) showed changes without relevant clinical significance. For this patient,

treatment with 100 mCi of iodine 131 was administered, followed by periodic clinical check-ups and a post-dose ^{131}I whole body scan (pWBS), with no recurrence in two years of follow-up.

Discussion

Diagnosing and treating SO remains challenging because of its historic significance. Many aspects of its diagnosis and management

remain unclear, and most cases are identified incidentally, with clinical data obtained mainly from case reports and retrospective studies. Pathological examination showed thyroid tissue resembling the cervical thyroid gland (FIGURE 2). The criteria used to diagnose thyroid gland carcinoma are widely applied to the diagnosis of MSO, which can present as papillary carcinoma, a follicular variant of papillary carcinoma, or with a follicular pattern, (FIGURE 3) [5].

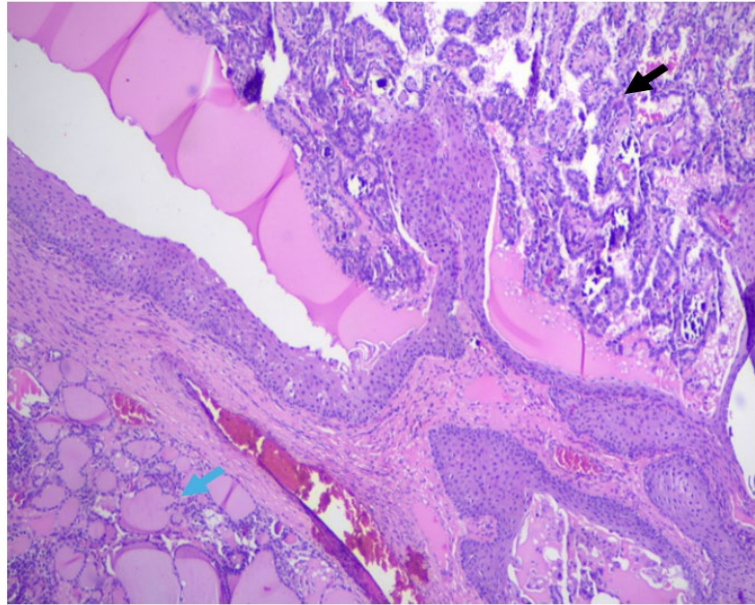


Figure 2: Hematoxylin and Eosin Staining of Ovarian. Low-power Magnification (4x) Showing Struma Ovarii with PTC. Struma Ovarii is shown in the Lower Left (Blue Arrow), Thyroid Follicular Tissue, which has the usual Appearance, and Papillary Carcinoma in the Upper Right Field (Black Arrow).

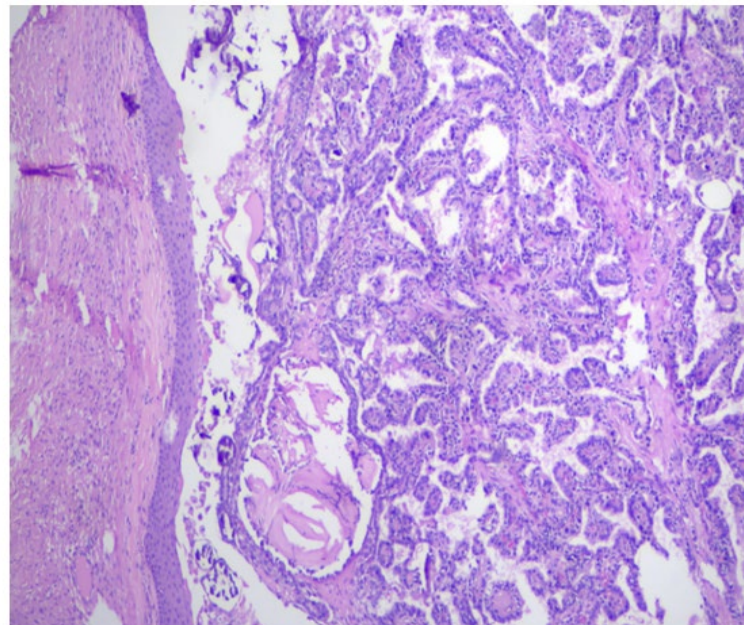


Figure 3: Hematoxylin and Eosin Staining of Ovarian PTC in Struma Ovarii (10x)

Surgery of the affected ovary is sufficient for benign unilateral SO. However, the management of malignant cases remains unclear. Total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy, and lymph node sampling is considered in MSO [6].

Some reports have documented fertility-preserving surgeries with rigorous follow-ups for selected cases. [4]. Since MSO are rare, no

treatment guidelines exist. Treatment followed thyroid cancer protocols.

In most cases, patients initially undergo debulking surgery with total abdominal hysterectomy and bilateral salpingo-oophorectomy at the time of initial diagnosis. [3]. TABLE 1 summarizes the reviewed case reports.

Table 1: Literature Review and Clinical Presentation of Malignant Struma Ovarii with Papillary Thyroid Carcinoma.

Case Report	Diagnosis	Treatment	Follow-up	Recurrence
Krishnamurthy et al. [1]	Synchronous malignant metastatic papillary thyroid carcinoma of struma ovarii and cervical cancer	Total abdominal hysterectomy with salpingo-oophorectomy and total thyroidectomy plus removal of the abdominal wall mass	For 6 years.	Not informed
Kabootari et al. [2]	Follicular variant of papillary carcinoma and from struma ovarii	Total abdominal hysterectomy with total salpingo-oophorectomy-thyroidectomy	10 years	No recurrence
Proença et al. [3]	Follicular variant of papillary carcinoma and from struma ovarii	Ablation of residual thyroid tissue was performed with a dose of 70 millicuries (mCi)	--	Not informed
Chung et al. [4]	Papillary thyroid carcinoma (50%) and follicular thyroid carcinoma (26.47%)	Unilateral oophorectomy (35.71%), bilateral oophorectomy (58.57%), radioactive iodine with an average dose of 158.22 mCi each time (38.57%).	--	Metastasis outside the ovary (51.94%). Regardless of the treatment received, the survival rate was high
Cui et al. [5]	Papillary thyroid carcinoma (54.5%)	Unilateral salpingo-oophorectomy (27.2%) and bilateral salpingo-oophorectomy (36.4%).	--	No patient in this PTC subcohort had metastatic disease.

It is unclear if cytotoxic agents are effective for malignant struma ovarii due to rare disease and lack of standardized trials, and the use of these drugs is limited owing to disease progression or side effects [1]. Radiotherapy is recommended for lesions unresponsive to iodine radioablation. NGS advances precision medicine for unresponsive tumors. The approval of pembrolizumab as an agnostic therapy for MSI-H tumors has brought hope to many patients with advanced cancer who have no available treatment options. This is not yet a reality in some countries [2]. High-risk factors for patients include extensive tumors, large lesions, elevated thyroglobulin levels, BRAF mutations, thyroid cancer, or distant metastases. Thyroidectomy and radioiodine for prevention are recommended. Adjuvant chemotherapy

includes cisplatin/etoposide/cyclophosphamide, tegafur-uracil, paclitaxel, and carboplatin, followed by oral etoposide and cyclophosphamide. MSO has a 96.7% survival rate, with a four-year recurrence time. Customized surveillance is crucial, and further studies are needed for diagnosis and management [9, 10].

Ethical approval and Consent to participate

This study was approved by the Ethics Committee of Centro Universitário São Lucas/Plataforma Brasil (opinion number 6.828.827). It was conducted in accordance with the ethical standards established in the 1964 Declaration of Helsinki and its subsequent amendments or comparable ethical standards.

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