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Metastatic Malignant Struma Ovarii in a Nulliparous Patient: A Case Report

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Abstract **Case Report**

Introduction*:** Malignant struma ovarii is a rare tumor, Due its rarity treatments and follow-up approaches are not clearly standardized. ***Case Presentation*:** we present a case of a 29 -year-old, nulliparous woman with metastatic malignant struma ovarii revealed by abnormal vaginal bleeding and abdominal-pelvic pain. After surgical resection, the patient had complementary radioiodine therapy (131I) after total thyroidectomy. She received a treatment dose of 100 mCi of 131I followed by post-therapy whole-body scintigraphy. ***Discussion*:** Histological diagnosis of malignant struma ovarii is similar to that of well differentiated thyroid carcinoma. there are no guidelines regarding the management of this type of cancer. After surgical resection of malignant struma ovarii, adjuvant therapy with thyroidectomy followed by radioactive 131 I therapy and suppressive thyroxine therapy is suggested. The same follow-up as thyroid cancer is commonly adopted, with abdomino-pelvic evaluation. ***Conclusion: Large clinical studies are needed to standardize the care of patients with malignant struma ovarii.

Keywords: Malignant struma ovarii - metastasis - management - surgery - iodine 131 therapy - follow up.

INTRODUCTION

Struma ovarii is a rare variant of ovarian teratoma representing 1% of all ovarian tumors, diagnosed in females at all ages, most commonly between the fifth and sixth decade of life. They contain at least 50% of thyroid tissue [1, 2]. Though typically benign, malignancy occurs in 5–10% of reported cases [2], and it is even less likely to lead to metastatic disease 5–6% [3]. The percentage of papillary thyroid carcinoma within malignant struma ovarii (MSO) is 70% [4].

When the tumor consists entirely of thyroid cancer, it is named pure MSO, and if the tumor consists of thyroid cancer and teratoma components, it is named "impure" MSO [5].

Due to the rarity of this tumor, consensual approaches for an optimal management have not yet been defined. Surgical management of the primary tumor and the thyroid gland, as well as the potential indications for adjuvant treatment, have not been standardized [6].

In this article, we present a case of young nulliparous woman with metastatic malignant struma ovarii revealed by abnormal vaginal bleeding and abdominal-pelvic pain and we discuss the management.

CASE REPORT

A 29 -year-old woman nullipara, virgin, with no medical history of either benign or malignant thyroid disease. presented to her gynecologist with menometrorrhagia and left-sided abdominal-pelvic pain evolving for 6 months. Pelvic transabdominal ultrasound revealed a solid and cystic mass in the left adnexal area. a pelvic CT scan showed a suspicious left supra- and latero-uterine solid cystic mass of ovarian origin measuring 105*68*101 mm associated with bilateral internal iliac nodes and a small peritoneal fluid effusion (Figure 1). Basal tumor markers were as follows: CA-125: 55 U/mL (range 0.0–35); beta HCG < 1.2 mlU/mL (range 0.0–5.0); CEA: 1.23 ng/mL (range 0.0–6.0); AFP: 1.83 ng/ml (range 0.0–10.0).

The patient underwent laparotomy, during the procedure, macroscopic exploration noted a bilateral ovarian tumour (larger left ovarian tumor: 11cm), torsion on the left ovary, a multicystic appearance and irregularity on the right ovary. left adnexectomy, partial

right oophorectomy and partial omentectomy were performed.

The results of histopathological examination revealed a papillary thyroid carcinoma, infiltrating both ovaries (left: 11 x 8 x 7.5 cm / Right: 5.5 x 4.5 cm) without extension to the left fallopian tube or the removed part of the omentum, no vascular emboli were found. Immunohistochemical staining revealed thyroid transcription factor-1 (TTF-1), CK7, and PAX8 positivity, Tg was not performed. The tumor was found to be BRAF mutation negative.

In view of these results, the case was discussed at the multidisciplinary team meeting, a revision surgery was indicated, thyroid function tests, and cervical ultrasonography were requested.

The TSH concentration was 1.53 mIU/L (range 0.5- 5), FT4 level was 17 pmol/l (range 12 - 22), FT3 level was 6.5 pmol/l (range 3.8 - 7.5), Thyroglobulin level was 12.55 ug/l (range 3.5 - 77) with positive Thyroglobulin anibodies to 17ui/ml (range 0 - 7). The ultrasound examination revealed a nodule in the right lobe of 7.7 x 5.5 x 3.2 mm (Tirads 4)

Two months later the surgical procedure has been completed with a right adnexectomy and multiple peritoneal biopsies. histopathological analysis showed a focus of papillary thyroid carcinoma measuring 2mm, in the remaining part of the right ovary and 2 other tumoral foci of the same nature in the right paracolic gutter and right ovarian fossa measuring respectively 2mm and 4mm. Histology of the left paracolic gutter and left ovarian fossa biopsies was normal.

Total thyroidectomy was requested, to differentiate metastatic primary thyroid carcinoma from isolated malignant struma ovarii, and to allow adjuvant treatment with radioactive iodine (I-131). Two months after, the patient underwent total thyroidectomy, Postoperative histopathological examination showed no malignant changes in the thyroid. The diagnosis of metastatic malignant struma ovarii has been made.

The patient received a treatment dose of iodine 131 of 100mCI (3.7 GBq) after thyroxine withdrawal for four weeks. The thyroglobulin level was 2.3 ng/mL (TSH =83.362 µIU/ml), and serum thyroglobulin antibodies were <12 UI/ml. Two days after treatment, the whole-body 131I scintigraphy showed an iodo-fixing focus, of residual appearance. Our decision was to keep our patient on LT4 suppressive dose and continue regular medical follow-up.

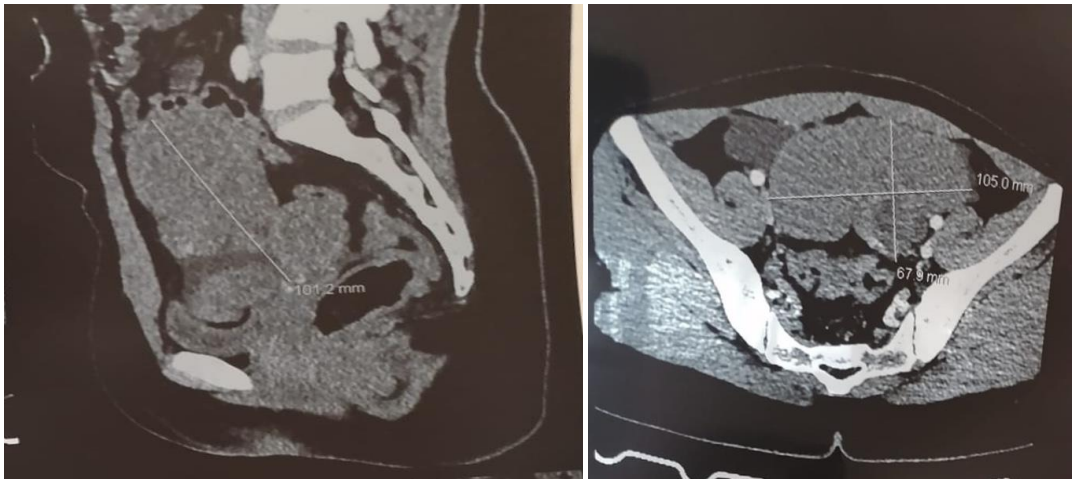

Figure 1: Pelvic CT scan: left supra- and latero-uterine solid cystic mass of ovarian origin

DISCUSSION

MSO is a rare malignant ovarian germ cell tumor. Most of the struma ovarii in which thyroid-type carcinomas arise are unilateral and more often affect the left ovary [7]. De Simone *et al*., [8], reviewed 24 cases of malignant SO, lesion sizes ranged from 5 to 20 cm, more commonly on the left side (63%), and most frequently with follicular variants of papillary thyroid carcinoma (54%) and papillary thyroid carcinoma (21%). Our presented clinical case share the same primary characteristics.

The majority of patients are diagnosed postoperatively, as was the case of our patient.

The morphological criteria of these tumors is based on classical criteria for primary thyroid carcinomas [9]. Julie *et al*., [10], proposed the following criteria for the diagnosis of malignant struma ovarii: the tumor must show clear signs of invasion, metastasis, or both, the morphologic features of the tumor must resemble thyroid tissue, and a primary carcinoma of the thyroid must be excluded.

The diagnosis can be difficult, and immunohistochemical analyses (TTF-1, Tg, and CK19...) can help in making a diagnosis [7]. In the present patient, we confirmed the positive expression of TTF-1, CK7, and PAX8.

Metastases are indicated only in 5–6% of patients with malignant struma ovarii [11]. The dissemination of the disease follows the pattern of ovarian cancer, the peritoneum, omentum, and adjacent pelvic structures, including the contralateral ovary and abdominopelvic lymph nodes, are the most common sites of metastatic involvement. [10, 11], hematologic dissemination to the bone, lung, liver, and brain is possible as well [11, 12]. Our patient had controlateral ovary and peritoneal metastasis.

Ovarian metastasis of thyroid origin and thyroid carcinoma arising from a struma ovarii are the two major differential diagnoses that have to be considered when a woman with history of thyroid carcinomas presents with an ovarian mass containing thyroid tissue and foci of thyroid carcinoma [9]. Ovarian metastases are exceedingly rare and generally associated with widespread disease. However, they must be considered in the presence of previous history of malignant thyroid carcinoma. Logani *et al*., [13], described a case of an ovarian metastasis from papillary thyroid carcinoma recurred 11 years after the initial diagnosis.

Brogioni *et al*., [14], reported to a case of bilateral ovarian metastases from a papillary thyroid carcinoma in a 38-year-old, 7 years after thyroidectomy and cervical lymph-node dissection. Our patient had no medical history of thyroid carcinoma, and the histopathological examination of the thyroid gland after thyroidectomy did not reveal any sign of malignancy.

There is no standardized management for malignant struma ovarii throughout the published literature. Surgical resection is the first phase of treatment for MSO. A conservative surgery, unilateral salpingo-oophorectomy may be acceptable in fertility-desiring patients if they have a localized tumor [3, 8].

A total hysterectomy with bilateral salpingooophorectomy is a reasonable option in postmenopausal women or in the case of extra ovarian extent [3].

Because our patient was young and nulliparous, gynecological surgery limited to unilateral left oophorectomy was initially indicated to preserve fertility and hormonal function in the second ovary. The examination of the controlateral ovary during the procedure detected a tumor, partial right oophorectomy was also done, identifing a malignant struma ovarii with metastasis to the controlateral ovary. The surgical procedure has been completed with a right adnexectomy and multiple peritoneal biopsies showing extension to the peritoneum too.

Currently, no consensus exists on the postoperative treatment and follow up of patients with MSO. It is commonly managed based on criteria for the treatment and follow up of thyroid carcinoma [3, 8, 15].

Applying these criteria to MSO, would result in thyroidectomy followed by postoperative 131I ablation therapy for most patients [15].

Yassa *et al*., [16], emphasized the role of patient stratification according to the risk of recurrence in the management of malignant struma ovarii. In patients with a low risk of recurrence (primary tumors smaller than 2 cm limited to the ovary and without aggressive histopathological features), they propose unilateral salpingo-oophorectomy and levothyroxine treatment to maintain serum TSH levels at 0.1–0.5 mIU/L. However, high-risk patients with tumors larger than 2 cm or aggressive histopathological features require additional treatment (total thyroidectomy and adjuvant I-131 therapy) to detect and potentially treat recurrences**.**

Total thyroidectomy is necessary before 131 I therapy, It confirms normal thyroid pathology and excludes a primary thyroid carcinoma with subsequent metastasis to the ovary or a second thyroid cancer, it allows radioiodine therapy and enhances its effect [3, 12, 15]**.** For our patient the histopathological analysis of the thyroid gland was normal.

131I therapy can treat residual or metastatic disease and facilitates subsequent biological monitoring by serum thyroglobulin. Indeed, serum thyroglobulin can be used as a tumor marker for follow-up of MSO in a manner analogous to patients with thyroid carcinoma [3, 10]. After 131I ablation any detectable serum thyroglobulin points to persistent or recurrent disease [4, 15].

Oudoux *et al*., [3], they recommended for monitoring after 131I therapy:

- Six months after the end of treatment: clinical examination, serum thyroglobulin measurements and diagnostic 131I scintigraphy (after rhTSH stimulation or withdrawal).
- Every 6 months for 18 months and every year thereafter for at least 20 years: the mainstay follow-up of clinical examination and thyroglobulin measurements with TSH suppression (thyroglobulin must be < 1 ng/ml). In the case of abnormal results, additional imaging must be performed (131I scintigraphy, 18FDG PET, ultrasonography, CT and/or MRI). Pelvic locations can justify complementary imaging, adapted to the initial stage of the disease.

Long-term survival rates are unknown because of the rarity of MSO and the lack of long-term follow-up data [10].

CONCLUSION

Malignant struma ovarii is a rare tumor, usually diagnosed after surgery for a suspicious ovarian mass by histological analysis. Due to the rarity of this tumor, consensual approaches for an optimal management have not yet been defined. Surgical management of the primary tumor and the thyroid gland, as well as the potential indications for adjuvant treatment, have not been standardized. Large clinical studies are needed to standardize the care of patients with malignant struma ovarii, and develop practice guidelines.

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