

CASE REPORTS

Management of Metastatic Struma Ovarii with Papillary Thyroid Carcinoma and a Neuroendocrine Component: A Case Report

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ABSTRACT

Struma ovarii is a rare form of teratoma that is invariably benign. When neuroendocrine and thyroid tissue both coexist within the ovary, the term strumal carcinoid is often used. The incidence of malignant transformation in this lesion has been rarely described in the literature. Here, we present a case in which the thyroid component metastasized widely throughout the peritoneum.

Keywords: Peritoneal metastasis, Struma ovarii, Thyroid carcinoma.

How to cite this article: Jamal K, Jong MC de, Proctor I, Morley S, Sivabalasingham S, Abdel-Aziz TE, Kurzawinski T. Management of Metastatic Struma Ovarii with Papillary Thyroid Carcinoma and a Neuroendocrine Component : A Case Report. *World J Endoc Surg* 2018;10(3):170-172.

Source of support: Nil

Conflict of interest: None

BACKGROUND

Struma ovarii is an uncommon entity, accounting for 1% of all teratomas, and it is usually benign.¹⁻³ While small elements of thyroid tissue are found in up to 15% of all germ-cell tumors, to diagnose struma ovarii, thyroid tissue needs to comprise at least 50% of the tumor volume.⁴ Malignant transformation of these lesions occurs in less than 5% of cases, and distant metastatic disease is even rarer. Strumal carcinoid is an even more exceptional and distinctive form of ovarian teratoma, in which there is an intimate mixture of thyroid tissue and

carcinoid. Malignant transformation for this type of a tumor has been described to be very scarce.⁵

CASE DESCRIPTION

A 45-year-old woman was referred to our hospital after undergoing a hysterectomy and right salpingectomy for presumed adenomyosis. The macroscopic report demonstrated numerous soft brown sub-serosal nodules which had the histological appearance of papillary thyroid carcinoma with positive staining for TTF1, thyroglobulin, CK7, vimentin, and CD5.⁶ On reviewing her past medical history, she had undergone a right oophorectomy for an ovarian mass nine years earlier, the histology of which at the time was reported as mature cystic teratoma with no evidence of thyroid tissue. A review of these slides at our institution demonstrated evidence of malignant struma ovarii of the papillary subtype.

At her initial visit, her thyroglobulin was 174 ng/mL (reference: 0–0.9) with an anti-thyroglobulin antibody titer of 12 IU/mL, while the rest of her routine blood tests were normal. A staging Computed tomography (CT) scan and subsequent MRI-scan (Fig. 1) showed a 1.5 cm nodule anterior to the right common iliac artery, several enhancing peritoneal deposits related to the anterior abdominal wall and a 1cm nodule lateral to the caecum in the right iliac fossa. Additionally, two small subcapsular liver lesions were indeterminate. A thyroid ultrasound scan found several small hypoechoic cystic and spongiform nodules scattered throughout the parenchyma measuring up to 7 mm in diameter all classified as benign (U2).

The decision was made to perform a diagnostic laparoscopy (Fig. 2), during which several peritoneal nodules on the anterior abdominal wall were excised. The histological analysis of these confirmed metastatic papillary thyroid carcinoma. Following this, our patient underwent a total thyroidectomy, which was uncomplicated. Histology of the specimen revealed an incidental focus of papillary carcinoma (pT1aN0). Subsequently, she went on to have 3.0GBq of ¹³¹Iodine therapy with scintigraphy (Fig. 3), which demonstrated numerous peritoneal nodules and confirming the previously indeterminate

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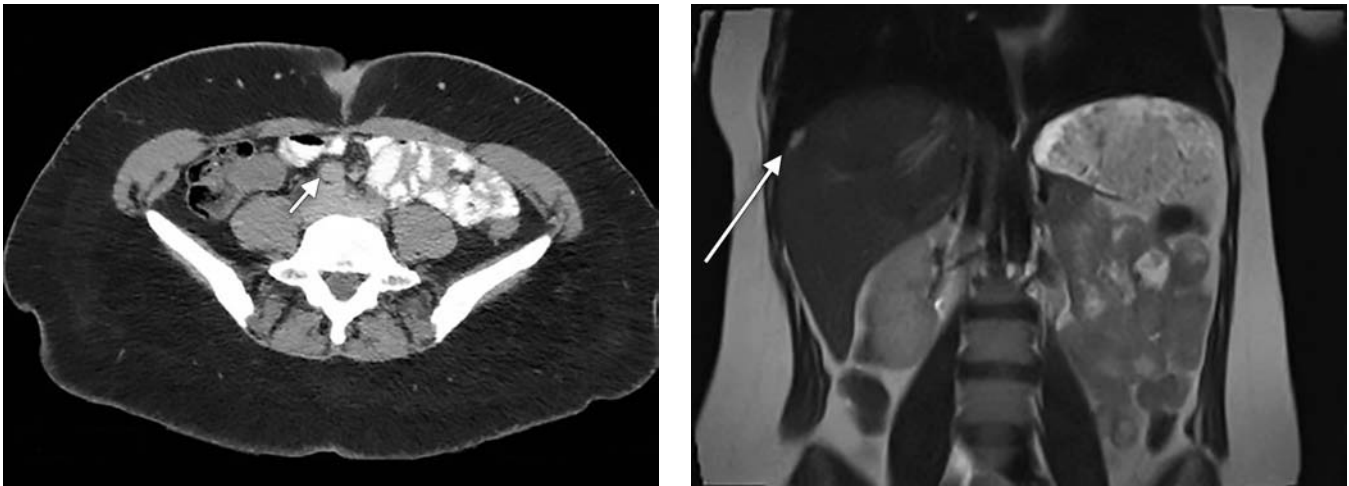


Fig. 1: Image of the staging CT-scan showing a nodule overlying aortic bifurcation MRI-scan demonstrating a subscapular liver lesion

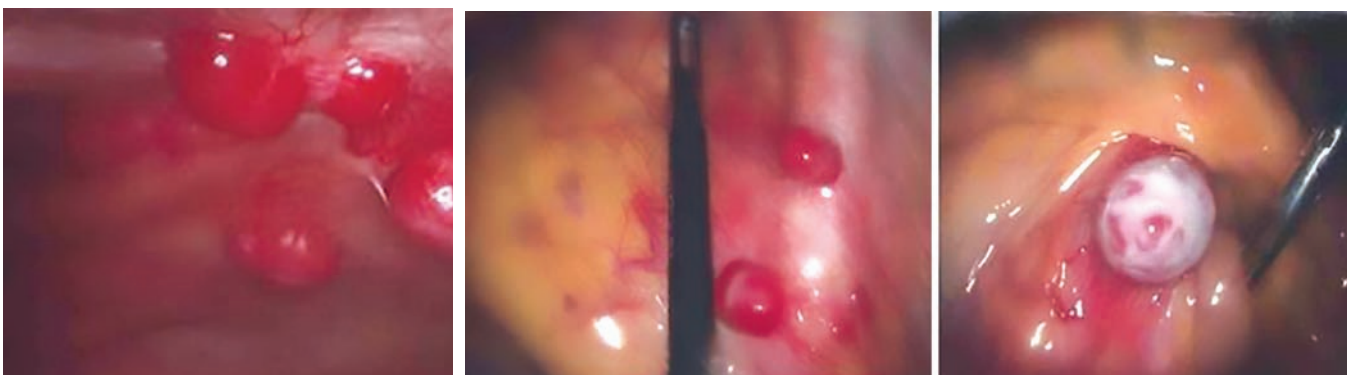
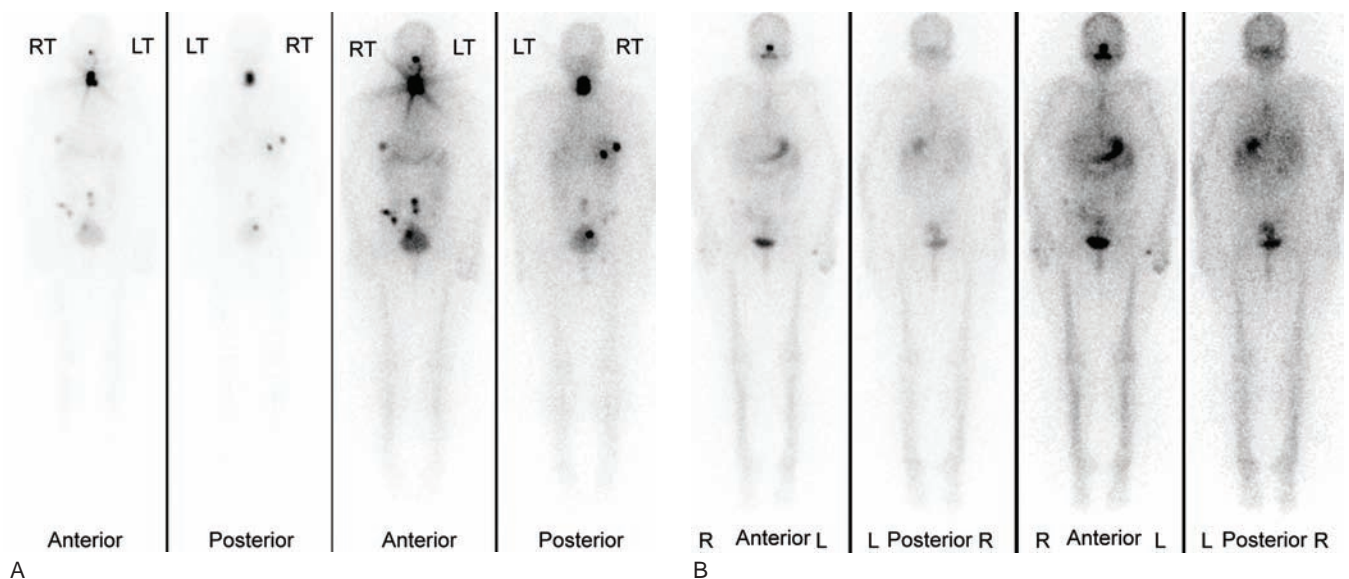


Fig. 2: Perioperative images during laparoscopy, demonstrating various locations of peritoneal nodules Nodules attached to the anterior abdominal wall Nodules located at the aortic bifurcation/overlying the right external iliac artery



Figs 3A and B: Iodine 131 scintigraphy (A) pre- and post-ablation; (B) scan demonstrating numerous peritoneal nodules and presumed metastasis to the liver

liver lesions to be iodine-avid and therefore presumed metastasis.

Due to the extent of the intra-abdominal disease, a debulking laparotomy was performed. At surgery, multiple nodules were removed varying between 3 mm and 4 cm from throughout the abdomen and pelvis, adjacent to

the colon, bladder, aortic bifurcation/right external iliac artery and vaginal vault. The right lobe of the liver and the two lesions that had previously thought to be subscapular on imaging were apparently found on the surface of the diaphragm (Fig. 4). Histology of these nodules all demonstrated evidence of metastatic papillary thyroid

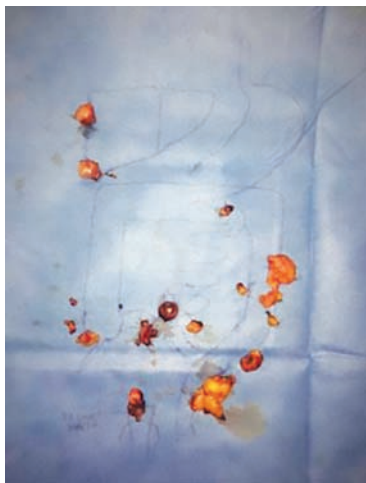


Fig. 4: A pattern of lesions removed at de-bulking

carcinoma but with admixed neuroendocrine tissue and positive staining for synaptophysin but not for calcitonin. The proliferation index, measured with MIB-1, was <1%. Following her surgery, the thyroglobulin reduced to 1.7 ng/mL and serum chromogranin A/B were both within normal limits. A post-operatively performed 131 Iodine and 68Ga-DOTATATE-PET-scan showed no signs of residual disease.

CONCLUSION

Malignant struma ovarii is a rare tumor which does not often metastasize.^{1,3} Histological differentiation of benign from malignant struma ovarii has been proven both difficult and of limited clinical value. Previous reports have shown that a proportion of histologically benign tumors will behave in a malignant fashion and that the majority of malignant tumors will not progress after local resection.⁶ For strumal carcinoid, the incidence of malignant transformation is even lower.⁵

In our case, although the peritoneal deposits demonstrated both thyroid and carcinoid tissue, histologically and biologically the thyroid element seemed to predominate. Histologically, the thyroid tissue had malignant features, and the thyroglobulin was markedly elevated with a diffuse uptake on a radioactive iodine scan. Although preoperative neuroendocrine markers were not available, post-operatively these were normal as was the 68Ga-DOTATATE-PET-scan. Therefore, we elected to treat this patient similarly to the way malignant struma ovarii is managed.

The extent of surgery necessary for malignant struma ovarii remains unclear; important factors are the age of the patient, tumor size and histology, extra-ovarian

spread and requirement to preserve fertility. In patients with a tumor confined to the ovary, options include unilateral oophorectomy, bilateral oophorectomy, and hysterectomy with bilateral oophorectomy.⁷ For patients with peritoneal disease, a more aggressive approach has been advocated encompassing additional omentectomy and lymph node dissection followed by total thyroidectomy, radioactive iodine ablation, and thyroxine suppressive therapy.⁷ Overall, survival rates seem to be generally good and not dissimilar to thyroid carcinoma, as survival rates of 89% at 10 years and 84% at 25 years have been reported.⁶

CLINICAL SIGNIFICANCE

Struma ovarii is a very uncommon entity, and stromal carcinoid is even rarer. The current report describes the management of a patient with widespread metastatic struma ovarii with papillary thyroid carcinoma as well as a neuroendocrine component. Our patient underwent treatment similar to that of malignant struma ovarii by a debulking laparotomy and total thyroidectomy. She currently has no signs of recurrent disease, after 18 months diagnosis, underlining the importance of accurate and possible aggressive management of this disease.

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