Malignant struma ovarii

Patient Information Series – What you should know, what you should ask.

What is it?

Malignant Struma Ovarii is a rare germ cell tumour that originates from a malignant transformation of a monodermal teratoma entirely or predominantly (over 50%) composed of mature thyroid tissue, resembling primary thyroid gland carcinomas. It's a very rare condition that affects women between 40 and 60 years, accounting for about 5% of all cases of Struma Ovarii and less than 0.5% of all ovarian cysts.

Macroscopically it presents as a round, smooth, multilocular-cystic or solid-cystic mass, more often unilateral, however, in 5 to 23% cases can present abdominal or metastatic diffusion, most frequently to peritoneum, omentum and mesentery. In about 10% of cases a synchronous primary thyroid gland cancer can be diagnosed.

Which are the symptoms?

This tumor can be asymptomatic in most cases. Depending on the volume of the mass, the most commonly presenting symptoms are abdominal pain or swelling, abnormal vaginal bleeding, ascites or deep vein thrombosis. If the ectopic thyroid tissue is functioning, it might cause hyperthyroidism or even thyreotoxicosis, therefore resulting in palpitations, hypertension, tremor, anxiety, weight loss and insomnia.

How can it be diagnosed?

Unfortunately, due to the aspecific morphological and imaging features of malignant struma ovarii, the rate of pre-operative misdiagnosis is extremely high, reaching up to 98%.

There is no specific biochemical or tumoral marker associated with the disease, therefore the definitive diagnosis is histopathological.

In case of an unexpected diagnosis of malignant struma ovarii, appropriate post-operative radiological staging is indicated to identify cases of advanced disease. CT scan, US or MRI are the most used techniques.

How can the condition be treated?

Due to the rarity of the disease, management is not standardized. Treatment is primarily surgical, the gold standard being total hysterectomy, bilateral salpingooophrectomy and omentectomy. Conservative surgery can be considered in younger patients with a desire for fertility preservation, however the completion of gynecologic surgery should be discussed with the patient after childbearing is completed. A full surgical staging should be performed in the presence of apparent gross metastasis.

Postoperative treatment options include total thyroidectomy, radioactive iodine therapy, thyroid suppression, external radiotherapy, and chemotherapy.

Which follow up I need?

Malignant Struma Ovarii is a rare ovarian malignancy associated with a good prognosis, with very favourable survival rates of 92–96% at 5 years and 85% at 20 years. However, recurrence has been reported in about 20% of cases.

A close surveillance is recommended, despite the lack of clinical guidelines, including clinical examination, pelvic ultrasound and dosage of thyroid-stimulating hormone (TSH) level by 6 months post-treatment. Furthermore, imaging using iodine isotopes can be used to screen for functional thyroid tissue within the pelvis.

What other questions should I ask?

- For how many years will I have to do follow-up visits?
- If a disease relapse is suspected, which imaging exams should be done?

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