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CLINICAL RECORD

Struma ovarii and the thyroid surgeon

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Abstract

Introduction: Struma ovarii is a rare, monodermal, ovarian teratoma. The common presentation is abdominal, with pelvic mass and pain, traditionally managed by gynaecologists. The malignant form is extremely rare and consists of differentiated thyroid cancer. It is rare for struma ovarii to present with features of hyperthyroidism. We present two unusual cases of struma ovarii and discuss the role of the thyroid surgeon in their management.

Methods and results: The first case involved a 40-year-old woman with a two-month history of swelling in the lower abdomen. Investigations revealed a mass arising from the left ovary. Surgery revealed a follicular carcinoma arising in a struma ovarii. She underwent a total thyroidectomy prior to radio-iodine therapy. The second case involved a 60-year-old woman who underwent thyroidectomy for thyrotoxicosis. Three months post-operatively, she remained thyrotoxic despite stopping thyroxine. A whole body radio-iodine scan revealed high uptake in the left ovary. Histological analysis of the resected ovary showed benign struma ovarii.

Conclusion: These two cases highlight the diagnostic and therapeutic role of thyroid surgeons in the management of benign and malignant forms of struma ovarii.

Key words: Struma Ovarii; Thyrotoxicosis; Thyroidectomy

Introduction

Struma ovarii is a rare ovarian teratoma. Most are benign and managed by gynaecologists. Although they constitute predominantly thyroid tissue, they seldom present with thyrotoxic features.

Here, we present two rare but interesting cases of struma ovarii: one a malignant form and the other a benign form with thyrotoxicosis. We also discuss the thyroid surgeon's role in the diagnosis and management of these rare tumours.

Materials and methods

Case one

A 40-year-old, multiparous woman presented to the gynaecologists with a two-month history of lower abdominal swelling and altered bowel habit. There was no other significant gynaecological history, and the general medical history was uneventful. There was no family history of ovarian or any other cancer.

Examination revealed a pelvic mass extending up to the level of the umbilicus, with ascites. An ultrasound examination showed a large, solid mass in the pelvis with free fluid in the abdomen. Serum tumour markers showed an elevated CA125 level of 1099 kU/l (normal range, 0–35 kU/l).

Magnetic resonance imaging showed an irregular mass 11 cm in diameter (Figure 1) with appearances suggestive of a mucinous tumour, arising within the left ovary. The omentum showed fatty signal intensity throughout, with a thickening suggestive of spread of the disease.

The patient subsequently underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy and appendicectomy.

Histological analysis of the surgical specimens showed a well differentiated left ovarian follicular carcinoma arising in a struma ovarii. The tumour was infiltrating and breaching the capsule. The omentum and appendix were not involved. A peritoneal fluid sample did not contain any malignant cells.

In view of the tumour's size, histology and breach of the capsule, the gynaecological oncology multidisciplinary team sought help from the thyroid multidisciplinary team regarding additional radio-iodine therapy. At this juncture, the patient was advised to undergo a total thyroidectomy, prior to radio-iodine treatment. No malignancy was noted in the resected thyroid gland.

Case two

A 60-year-old woman with a 35-year history of thyrotoxicosis was referred to the thyroid clinic for surgical treatment. She had previously been treated with carbimazole followed by radio-iodine. Sixteen years later, her thyrotoxicosis had recurred and she had received a further course of radio-iodine therapy which failed to induce remission. She was taking propanolol 40 mgs thrice daily for symptomatic relief. She had a family history of thyrotoxicosis.

Having considered all three main treatment options (i.e. anti-thyroid drugs, radio-iodine therapy and surgery), the patient opted for surgery, and underwent an uneventful total thyroidectomy.

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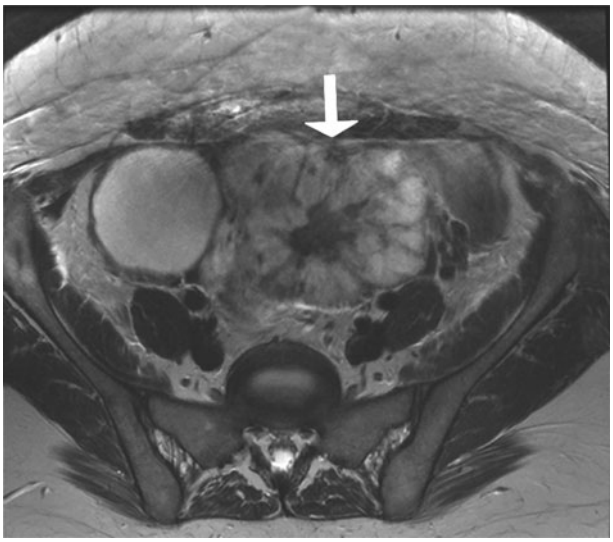

FIG. 1
Axial, T2-weighted magnetic resonance image showing tumour (arrow) in the left ovary.

Post-operatively, she was commenced on 125 μg/day thyroxine, based on her weight and age.[1] Six weeks post-operatively, her thyroid biochemical profile showed that she was still grossly hyperthyroid. This was thought to be thyroxine dose related, and therefore the dose was reduced to 75 μg/day. However, the patient remained thyrotoxic, so thyroxine replacement was stopped completely. Repeated thyroid biochemical analysis showed persistent thyrotoxicosis. An isotope scan of the neck showed no residual thyroid uptake.

At this juncture, two possibilities were considered: thyroxine overdose or an ectopic thyroxine source. A whole body radio-iodine scan (Figure 2) showed increased uptake in the left ovary, indicating a probable struma ovarii. Magnetic resonance imaging showed a partly solid, partly cystic mass arising from the left ovary.

The patient underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy and omental biopsy.

Histological analysis showed that the tumour was confined to the left ovary and consisted predominantly of thyroid tissue, i.e. it was a struma ovarii.

Discussion

Although Sir John Bland-Sutton first recognised the presence of thyroid tissue in a large ovarian tumour he removed in 1893, Gottschalk reported the first case of struma ovarii in 1901. 'Thyro-dermoid' was an alternative name proposed by Stevens.[2]

Struma ovarii is a rare, mature, monodermal, ovarian teratoma. These tumours comprise 5 per cent of all germ cell tumours and 2–4 per cent of teratomas. Struma ovarii comprises only 0.2–1.3 per cent of total ovarian tumours. A histological criterion for struma ovarii diagnosis requires more than 50 per cent of the mass to be composed of thyroid tissue.[3,4]

A malignant form is seen in only 0.3 to 5 per cent of all struma ovarii tumours. Clinical presentation is similar to that of other ovarian tumours, with symptoms such as a lower abdominal mass with pain, abnormal vaginal bleeding

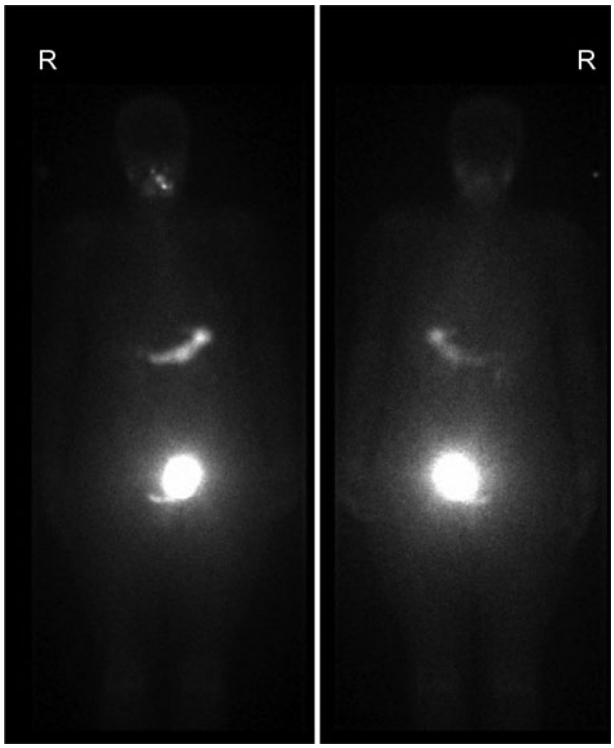

FIG. 2
Whole body radio-iodine scan showing increased uptake in the left ovary. R = right

and ascites. Hyperthyroidism has been reported in 5–8 per cent of all struma ovarii cases. Diagnostic criteria for malignant struma ovarii were suggested by Devaney *et al.*,[5] and require the presence of cytological atypia, 'ground glass' nuclei, increased mitotic activity and/or vascular invasion. These diagnostic criteria are similar to those for differentiated thyroid malignancy.

Among malignant struma ovarii, papillary carcinoma is the commonest histological subtype (44 per cent), followed by follicular carcinoma (30 per cent) and follicular variant of papillary carcinoma (26 per cent).[6] Metastases can occur to the peritoneum and liver, but lymph node spread is rare.

- **Management of malignant struma ovarii is multidisciplinary**
- **Thyroidectomy may be needed (to exclude a primary source or before radio-iodine therapy)**
- **Persistent post-thyroidectomy hyperthyroidism may be due to struma ovarii**
- **Total body radio-iodine scanning is indicated in such cases**

There has been no consensus regarding optimal management of malignant struma ovarii, due to its rarity. In general, most malignant struma ovarii tumours not invading or extending beyond the capsule can be treated with salpingo-oophorectomy with or without hysterectomy. Those that spread beyond the capsule and ovary may need combined treatment modalities. We recommend that these cases should be discussed in the thyroid cancer multidisciplinary meeting in view of the potential need for ^{131}I ablation, which will

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necessitate a total thyroidectomy to maximise efficacy. Total thyroidectomy not only enables radio-iodine treatment to be effective, but also facilitates the monitoring of thyroglobulin levels during follow up.

Due to the rarity of malignant struma ovarii, it is difficult to predict the prognosis. However, a literature review of 88 cases by Robboy *et al.*[7] showed a good long-term survival of 89 and 84 per cent at 10 and 25 years, respectively.

The incidence of secondary thyrotoxicosis due to struma ovarii alone is unknown. Struma ovarii should be suspected in patients with toxic features and an abdomino-pelvic mass. Thyrotoxicosis with or without Graves disease makes the diagnosis of struma ovarii challenging, particularly if there are no abdominal or pelvic symptoms. Persistent thyrotoxicosis after a total thyroidectomy should raise suspicion of an ectopic source of thyroid hormone. A diagnosis of struma ovarii should be strongly suspected if a high uptake of radio-iodine (^{123}I) is noted in the abdomino-pelvic region.

Conclusion

Thyroidectomy is a necessary part of the multi-modality treatment of advanced malignant struma ovarii, particularly when radio-iodine treatment is required. Although struma ovarii is not commonly seen in thyroid endocrine surgical practice, the thyroid surgeon should be aware of this entity, and should suspect it in cases of persistent thyrotoxicosis after total thyroidectomy.

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