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REVIEW

The diagnosis and management of struma ovarii

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Abstract

Struma ovarii are a rare type of monodermal teratoma. Preoperative diagnosis of struma ovarii is difficult as the symptoms, clinical presentation and image on ultrasound are often similar to that of ovarian carcinoma. These patients, with mostly benign disease, often have more extensive surgery than necessary. We present the cases of four women who were diagnosed with struma ovarii postoperatively. They illustrate a range of symptoms and varied surgical approaches. The objective of this mini review is to raise awareness of the diagnosis of struma ovarii. The extensiveness of surgery, mostly in premenopausal women, can be reduced if there is a high suspicion of struma ovarii prior to surgery. An ovarian cyst with a large solid component may be suggestive of this diagnosis and magnetic resonance imaging if possible, and serum thyrogglobulin may aid in diagnosis. It is also prudent to test for hyperthyroidism if the diagnosis is suspected, to avoid thyroid crisis during surgery.

Keywords: diagnosis, struma ovarii, surgical management, thyroid follicles

Introduction

Germ cell tumours are a heterogeneous group of neoplasms arising from primordial germ cells. Teratomas are the most common type of germ cell tumours. Struma ovarii are a rare type of monodermal teratoma containing more than 50% thyroid tissue, and account for approximately 2–3% of teratomas.[1]

Struma ovarii typically occur between the ages of 30 and 50 years.[1] As with germ cell tumours, women usually present with abdominal distension due to the mass and ascites, abdominal pain due to torsion, and occasionally with symptoms due to hormonal production.[2] Ascites occurs in approximately 10% of women, and in some cases this can be combined with a pleural effusion (modified Meigs syndrome).[3] Peritoneal implants can also occur from benign lesions, called peritoneal strumosis.[4]

Roughly 5–8% of patients with struma ovarii have clinical signs of hyperthyroidism.[5] As benign struma ovarii contain normal thyroid tissue, they have a high uptake of iodine and can secrete thyroid hormones.[6] A small percentage of benign struma undergo malignant transformation. Often malignant struma do not produce thyroid hormone, however, when hyperthyroidism exists in malignant struma 83% of patients have distant metastases.[6] Hyperthyroidism can be life threatening if not diagnosed preoperatively as thyrotoxic crisis can occur.[7] These patients should be treated with a beta blocker and attain a euthyroid state with the use of a thionamide for 4–6 weeks prior to surgery.[4]

There is no typical ultrasound feature of struma, although it could be suspected if there is a large solid component on ultrasound.[8] Tumour markers are of little clinical value. The cancer antigen (CA)-125 may be raised in some patients, especially those with ascites, but is not reliable in women of reproductive age.[1] If the patient has functioning thyroid tissue in the tumour and a normal thyroid gland, then thyroid-stimulating hormone, thyroxine, triiodothyronine and thyroglobulin levels can be used to assist in diagnosis. Thyroglobulin can be used for follow-up as a marker of response to treatment, especially in cases of malignancy and metastases.[9] The diagnosis of struma ovarii is most often made postoperatively on histology.

Method

The Tygerberg National Health Laboratory Service database was searched for diagnoses of struma ovarii from 2012–2014 by International Classification of Diseases, Revision 10 code. Four women were found who had had surgery for ovarian masses which were diagnosed as struma ovarii postoperatively. Their case notes were retrospectively reviewed.

Results

The four patients had a wide variation in age (20–52 years) and different presenting complaints, and there was a differing approach to surgery for all of them based on the findings of the preoperative investigations.

Case 1

A 20-year-old nulliparous woman presented with a one week history of nausea, epigastric tenderness and abdominal distension. She had regular menses and was not on any hormonal contraception. On examination, her thyroid was not enlarged and her pulse rate was 78 beats per minute (bpm). She had a central mobile mass arising from the pelvis of approximately 25 cm, with no ascites clinically. The ultrasound revealed a complex mass with solid and cystic components (102 x 112 x 64 mm) posterior to the uterus and moderate ascites. Evidence of metastases was not seen on further imaging. Investigations showed a lactate dehydrogenase of 595, alpha-fetoprotein of 2.2, beta human chorionic gonadotropin of < 1 and CA-125 of 486 kU/l.

These results raised the suspicion of primitive germ cell malignancy. Three weeks after presentation, the patient underwent a laparotomy and left adnexectomy. At surgery, a smooth-walled, 25 x 15 cm left ovarian mass was seen. There was no capsular invasion or sign of peritoneal spread. The uterus and right ovary looked normal.

Histology

Macroscopically, the ovarian tumour comprised multiloculated cysts containing gelatinous brown, mucinous material. On microscopy, the ovarian mass showed predominantly thyroid follicles, some filled with colloid. There were no malignant features. The immunohistochemical stains were positive for thyroglobulin and thyroid transcription factor-1 (TTF-1). Thus, a diagnosis of struma ovarii was made.

Case 2

A 52-year-old postmenopausal woman presented with a four-month history of increasing abdominal distension, shortness of breath and loss of appetite. She had no other known medical illnesses. She had undergone five vaginal deliveries, and her only previous surgical procedure was a sterilisation. On examination, her thyroid was not enlarged and her pulse rate was 60 bpm. Her abdomen was distended, a mass of approximately 10 cm could be felt, and ascites was demonstrated with a fluid thrill. An ultrasound and computed tomography scan confirmed a complex solid and cystic mass arising from the right ovary measuring 11 x 12 cm. Massive ascites were also present with left-sided pleural effusion; both cytologically negative for malignancy. Tumour markers showed a CA-125 of 1 115 kU/l. The patient was suspected of having ovarian carcinoma. Thus, she underwent a staging laparotomy with a total abdominal hysterectomy, bilateral salpingo-oophorectomy and omental biopsy. At surgery, the mass was removed with the capsule intact. There was no sign of peritoneal spread.

Histology

Sections from the ovarian mass showed histological features of pure struma ovarii without any associated dermoid components. The struma was composed of thyroid tissue with a predominantly microfollicular pattern, and large cysts filled with colloid. Malignant features of papillary thyroid carcinoma were absent. Thyroglobulin and TTF-1 confirmed a thyroid origin.

Case 3

A 50-year-old woman presented with post-coital bleeding. An ultrasound was performed, and an incidental finding of a 9 x 8 cm left adnexal cyst with a large solid component was seen. There were no ascites on ultrasound. The patient had no known medical conditions. She had been amenorhoiec for five months and was thought to be perimenopausal. She had had two previous Caesarean sections, and there was no other surgical history. There was no family history of breast or ovarian cancer. On examination, her thyroid and pulse rate was normal. Her abdomen was not distended and there were no palpable masses. The investigation showed a CA-125 of 16 kU/l. The patient had a low Risk of Malignancy Index score. However, the ultrasound features were concerning. Thus, she underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy and omental biopsy.

Histology

The right ovary showed histological features in keeping with struma ovarii. The cyst wall contained benign thyroid tissue, with no evidence of mature or immature tissue elements. The uterus contained multiple small fibroids, with no evidence of any other pathology.

Case 4

A 47-year-old premenopausal woman presented to the surgeon with symptoms of cholelithiasis. On ultrasound scan, there was an incidental finding of a dermoid cyst measuring 7 x 5 cm, comprising solid and cystic components. The patient had had two previous Caesarean sections and was a type 2 diabetic on insulin and oral treatment. The investigation showed a CA-125 of 13 kU/l. There were no concerns about malignancy. Therefore, the surgery was planned as a laparoscopic procedure. The patient underwent a laparoscopic cholecystectomy and right adnexectomy, and had an uncomplicated postoperative course.

Histology

A multiloculated cyst, primarly made up of thyroid follicles containing colloid, was seen on microscopy. There were also foci of mature adipose tissue, squamous epithelium and respiratory epithelium. A diagnosis of struma ovarii with associated dermoid tissue was made.

Discussion

As the diagnosis of struma ovarii is usually made postoperatively, most women undergo a staging laparotomy, total abdominal hysterectomy, bilateral salpingo-oophorectomy and omentectomy, with the removal of macroscopic disease, where possible, as for any other suspected ovarian malignancy. Our patients had surgery which varied in extent from a laparoscopic adnexectomy to a full-staging laparotomy, depending on the pre-surgical suspicion of malignancy and their menopausal status. A unilateral salpingo-oophorectomy is adequate in women who

wish to retain fertility, and this would be the definitive treatment in benign disease. All our patients had benign disease.

The definition of struma ovarii is a mature teratoma comprising thyroid tissue exclusively or predominantly. The histopathological appearance is characteristic, resembling mature thyroid tissue composed of variably sized thyroid follicles filled with colloid. Some areas are cellular, containing microfollicles, and others show a more trabecular to solid growth pattern. Immunhistochemically, these cells stain for thyroglobin and TTF-1, which confirms the thyroidal nature of the tissue.[7]

Malignant transformation occurs in less than 5% of struma, and the histological criteria are the same as those for thyroid malignancies.[10] Thyroid-type carcinoma originating in struma ovarii needs to be differentiated from thyroid carcinoma with metastases to the ovaries.[7] If a primary ovarian malignancy is present, then adjuvant treatment with a total thyroidectomy and radioactive iodine (I-131) therapy has been shown to decrease recurrence and mortality rates.[8] Serum thyroglobulin can be used as a marker for the recurrence of disease.[10] Women should be advised not to fall pregnant for six months following I-131 treatment as there is an increased risk of miscarriage. The side-effects of 131-I therapy also include transient amenorrhoea and early menopause.[6]

It is difficult to distinguish between struma ovarii and ovarian carcinoma preoperatively as the symptoms, clinical presentation and image on ultrasound are often similar. These patients with benign disease often have more extensive surgery than necessary, resulting in a large abdominal scar, a longer hospital stay and a more lengthy recovery. The removal of both ovaries may induce or worsen menopausal symptoms, and leads to an increased risk of all-cause mortality and coronary heart disease.

An ovarian cyst with a large solid component on ultrasound may raise suspicions of struma ovarii. Nurliza Binti Md *et al.* recommend using magnetic resonance imaging and serum thyroglobulin to preoperatively diagnose struma, and then proceed to a laparoscopic procedure.[11] If struma ovarii is suspected, thyroid functions should be tested. This test does not only aid in diagnosis, but also avoids thyroid crisis during surgery. I-131 scintigraphy can be used to confirm the presence of ovarian thyroid tissue and locate metastatic disease.[3]

Although struma ovarii are rare, a high index of suspicion and relevant preoperative testing may facilitate in diagnosis, allow appropriate surgery to be performed, and spare women from unnecessary morbidity.

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