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CASE REPORT

Highly Differentiated Follicular Carcinoma Arising from Struma Ovarii (Strumosis) Presenting as Pseudo-Meigs Syndrome with Elevated CA125 Level: A Case Report

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Abstract

A 75-year-old woman presented with the left ovarian mass, ascites, unilateral pleural effusion, and elevated CA125 levels. Clinical and radiological findings were highly suspicious for a malignancy. The patient underwent surgical excision of the uterus and infracolic omentum, and an intraoperative consultation was performed. Ovarian mass was diagnosed as "struma ovarii" and no other surgical procedure was performed. Histopatologic examination showed struma ovarii on the left ovary and bland-looking thyroid tissue implants on omentum and hernia sac. The patient diagnosed as "highly differentiated follicular carcinoma arising from struma ovarii" (HDFCO). No additional therapy was recommended to the patient and after a 53 months of follow-up, no recurrence was identified. HDFCO previously named strumosis is characterized by the presence of thyroid tissue in the peritoneal cavity with a bland morphology. Pseudo-Meigs syndrome with elevated serum CA125 levels in a 75-year-old lady is highly suspicious for malignancy, but HDFCO is an entity with a benign clinical course that shows the importance of intraoperative consultation. We describe the first case of HDFCO (strumosis) presenting as Pseudo-Meigs syndrome with elevated serum CA125 levels.

Keywords: CA125; pseudo-meigs' syndrome; struma ovarii; strumosis.

Struma ovarii is a monodermal teratoma exclusively or predominantly consisting of mature thyroid follicular cells[1]. However, some struma ovarii cases have been associated with extra-ovarian spreading of benign thyroid follicles. The term "peritoneal strumosis" is applied for the presence of thyroid tissue in the peritoneal cavity, which has a bland morphology indistinguishable from normal benign thyroid tissue, in the absence of a distinct thyroid carcinoma. A nomenclature "highly differentiated follicular carcinoma arising from struma ovarii (HDFCO)" has been proposed for this entity, as the extraovarian spread is considered as an evidence of malignancy[2]. Meigs' syndrome refers to a benign and solid tumor accompanied by ascites and pleural effusion; and Pseudo-Meigs syndrome is characterized by same clinical features associated with other ovarian or gynecological tumours[3].

A postmenopausal woman presenting with Pseudo-Meigs syndrome (a pelvic mass, ascites, and pleural effusion) and elevated serum CA125 levels, is highly suspicious for an ovarian malignancy; however, it should be kept in mind that it can have a benign origin.

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In the literature, a few struma ovarii cases and one malignant struma ovarii case are reported as associated with Pseudo-Meigs' syndrome and elevated CA125 serum levels, but there are no reports about HDFCO (strumosis) with these clinical features. A case report of 75-year-old woman with struma ovarii and strumosis associated with pseudo-Meigs' syndrome is presented.

Case Report

On March 2016, a 75-year-old postmenopausal woman was admitted to our hospital with abdominal pain. Her medical history included hypertension, coronary artery disease, and cholecystectomy. Menopause occurred 25 years ago, at the age of 50. Computed tomography (CT) revealed a left ovarian mass (11×7 cm in diameter) with cystic and solid components, widespread ascites, and a right pleural effusion. Omental cake consistent with disseminated diease was also detected. Thoracentesis yielded 1200 mL of serous fluid, and cytological examination of pleural fluid revealed reactive mesothelial cells, without any malignant cell. Tumor marker levels were normal, except CA125 levels of 1127 U/mL (normal value below 35 U/mL). Thyroid hormone levels were within normal limits as follows: Free thyroxine (FT4)=14.9 pmol/L (7.86–14.41) and thyroid-stimulating hormone (TSH)=0.52 microIU/mL (0.34–5.6).

A total abdominal hysterectomy, bilateral salpingo-oophorectomy, infracolic omentectomy, and video-assisted thoracoscopic surgery (VATS) were performed. Before the abdominal surgery, the patient was referred to the cardiothoracic surgery unit for VATS. The pleural effusion was aspirated; however, no mass was observed. During abdominal surgery, the intraoperative findings showed a 2000 mL ascites, an adnexal mass originating from the left ovary, and a normal right ovary, fallopian tubes, and uterus. There were not any macroscopically observed tumors on omentum and peritoneal sites. The left ovary was sent for intraoperative pathology consultation, and reported as "mature cystic teratoma." A pelvic-paraaortic lymph node dissection was avoided.

Macroscopic examination of the left ovary showed an encapsulated mass that measured 14×9×5 cm, and there was no extension of the tumor to the surface of the ovary. The mass was composed of both solid and cystic areas. The multilocular cystic area contained hair, teeth, and sebum, and the solid component's cut surface had a gelatinous glossy yellow appearance suggestive of a struma ovarii.

On histopathologic examination, the ovarian mass was predominantly composed of thyroid tissue and, to a lesser extent, skin, and cartilage (Fig. 1a). The glossy yellow parts appeared to be a characteristic nodular goiter that showed various-sized thyroid follicles with colloid and lined with bland cuboidal epithelium.

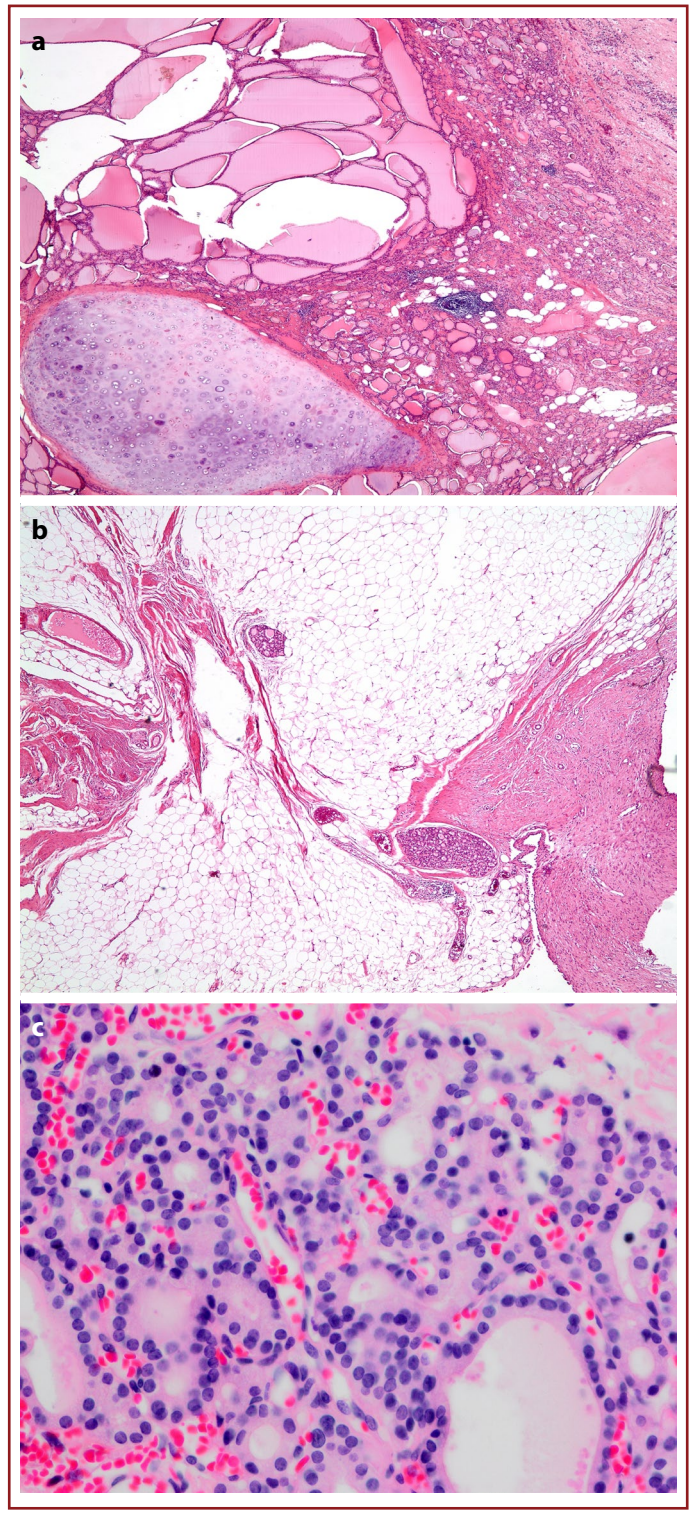

Figure 1. (a) Microscopic appearance of the left ovarian mass showed predominantly thyroid follicules of varying sizes, and to a lesser extent cartilage (H and E, ×40). **(b)** Thyroid tissue implants were identified in the omentum and hernia sac (H and E, ×40). **(c)** These implants showed variably-sized thyroid follicles with bland thyrocytes (H and E, ×400).

Microscopically, thyroid tissue implants were seen in the omentum and hernia sac (Fig. 1b), although there were not any macroscopic findings. These nodules also showed variably-sized thyroid follicles with bland thyrocytes that lack papillary thyroid carcinoma nuclear features (Fig. 1c). An immunohistochemical study was performed on the omental nodules, and strong expression was observed with thyroglobulin and thyroid transcription factor 1 (TTF-1), which confirmed the thyroid epithelial nature. Ki-67 proliferation index was 1–2%.

Pleural and peritoneal effusion regressed just after the surgery. The case was discussed by the gynecology-oncology tumor board, a 2–3 months follow-up without any adjuvant therapy and to perform a thyroid function test after the surgery was decided.

The patient's thyroid function was at normal levels at the time of surgery, and at the 9th month after the surgery. An umbilical hernia developed in the patient 8 months after the surgery that was surgically removed and sent for a pathological review; no tumoral lesion consistent with a struma ovarii was recognized. Now, the patient has had no recurrence after 53 months of follow up.

Discussion

Struma ovarii is a monodermal teratoma that is composed either exclusively or predominantly of thyroid tissue. It is the most common type of monodermal teratoma and it presents almost 3% of all ovarian teratomas. The term "malignant struma ovarii" is recommended for describing the entity "thyroid-type carcinoma arising in struma ovarii"[4]. Malignant transformation occurs in 5% of cases, and the most common thyroid type carcinomas arising in struma ovarii are papillary and follicular types respectively[5]. The term "peritoneal strumosis" or "strumosis" is used for the presence of innocuous thyroid tissue that histologically resemble non-neoplastic thyroid tissue in the peritoneal cavity. However, Roth and Karseladze proposed that such cases should be designated as highly differentiated follicular carcinoma arising from struma ovarii (HDFCO), as extraovarian spread is considered as the evidence of malignancy. It is recommended not to use the term of "peritoneal strumosis"[2].

In a clinical perspective, Roth et al.[4] reported 18 HDFCO cases with review of the literature. None of the cases died of neoplasm, although one patient was living with persistent disease. They reported that HDFCO is the least aggressive form among malignant struma and metastasis from thyroid gland. Robboy et al.[6] reported seven cases in their series, consistent with HDFCO. Only one patient have had multiple recurrences and eventually died from her disease. Moreover, they proposed that it is not true to call this entity as "benign" and should be avoided. Although this entity is usually has an indolent clinical course, occasional cases have had recurrence and a worse behaviour.

Meigs syndrome was defined first in 1954 as serous ascites and pleural effusion associated with a fibroma or thecoma of the ovary and the serosal effusions must resolve after removal of the tumour. Pseudo-Meigs syndrome is characterized by same clinical features associated with other gynecological tumours[3].

In the literature, a few cases of struma ovarii associated with Pseudo-Meigs syndrome and elevated CA 125 level have been published[1]. Moreover, one malignat struma case was reported[7]. We describe the first case of highly differentiated follicular carcinoma arising from struma ovarii (HDFCO) or as previously named "strumosis with Pseudo-Meigs' syndrome and elevated CA 125 serum levels."

Our case, a postmenopausal patient with a pelvic mass, ascites, pleural effusion, and an elevated CA125 level, who also had an appearance of omental cake on radiological examination was regarded as highly suspicious for a malignant ovarian mass. However the intraoperative consultation and final histopathology report revealed a struma ovarii and strumosis.

Histopathologic differential diagnosis of strumosis includes malignant struma ovarii and metastasis from thyroid gland. The most common throid-type carcinomas arising in struma ovarii is papillary and follicular type thyroid carcinomas. Papillary nuclear features are required for papillary carcinoma. Moreover, invasion into the surrounding ovarian tissue, vascular invasion, or metastases is required for follicular carcinoma[8]. None of them was detected on histopathologic examination. A detailed clinical history or thyroid examination should be assessed to evaluate a metastasis to ovary from the thyroid gland. Furthermore, other teratomatous elements are not expected to be seen in the metastatic tumor foci. Our patient's ultrasonography imaging of thyroid revealed no abnormal findings.

As it is a comparatively rare lesion, it is challenging to diagnose as well as to choose the adequate treatment. The treatment of struma ovarii is surgical resection of the ovarian tumor, which is typically a unilateral oophorectomy. In malignant struma ovarii and HDFCO, so called "strumosis" surgery is also the first step of treatment. Conservative surgery (unilateral salpingo-oophorectomy) may be performed in fertility-desiring patients. Total abdominal histerectomy with bilateral salpingoopherectomy is a rational option in postmenopausal women. After surgery patients may need a total thyroidectomy and I131 ablation to prevent further growth

of the peritoneal lesions. An adequate period of follow-up is required, that is, 10–20 years for a safe judgment[8].

Our case was discussed in the gynecology-oncology tumor board; any additional therapy was not recommended to our patient after surgery; and it was decided to follow-up patient with thyroid function tests. She has been followed-up for 53 months and still had no recurrences.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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