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Malignant Struma Ovarii Detected Incidentally Due to Contralateral Ovarian Torsion

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Abstract

Struma ovarii is a rare and highly specialized form of ovarian teratoma, in which thyroid tissue is the exclusive element (>50%). It comprises about 2% of all teratomas, and the majority of struma ovarii is benign tumor. But malignant struma ovarii, which showed malignant cell change arising in the benign struma ovarii, rarely reported. Here, we report an incidentally detected case of papillary thyroid carcinoma arising in the struma ovarii due to contralateral ovarian torsion in a 80-year old female.

Key Words : Malignant struma ovarii, Papillary carcinoma, Teratoma, Torsion

Introduction

Struma ovarii is a monodermal variant of ovarian teratoma, initially described by Bottlin in 1888. Later, by Pick in 1902 and 1903, who recognized that struma ovarii was composed of thyroid tissue [1]. Although 5-15% of all ovarian teratomas may contain some thyroid tissue; only 2% of these are made exclusively of struma ovarii. The vast majority of struma ovarii is benign tumors (95%) [2]. But malignant struma ovarii, which showed malignant cell change arising in the benign struma ovarii, rarely reported. The most common histologic type of malignancy arising in the struma ovarii is papillary thyroid carcinoma [3].

Case Report

A 80-years-old woman was admitted, due to lower abdominal pain. There were no specific findings in past history and family history. On physical examination, a huge palpable abdominal mass was found. Ultrasound examination in the emergency department of Dong San Medical Center showed a large round multiseptated mass in the cul-

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de-sac (Fig. 1A). On pelvic examination, vagina showed no specific lesion but the cervix showed motion tenderness. On dynamic abdomen and pelvic CT, there was a multiseptated cystic mass, measuring 13×10.5×10 cm in maximum dimensions (Fig. 1B). Pelvic MRI examination also revealed a large multiloculated cystic mass suspicious of mucinous cystic neoplasm (Fig. 1C).

In laboratory finding, tumor markers such as CA-19-9 and CA-125 were within normal range. Total abdominal hysterectomy with bilateral salpingoopherectomy was done along with Culdoplasty.

Received specimen for frozen section was the right ovary attached to the fallopian tube. Grossly, the uterus was normal sized. The right ovary was 16.0×10.0×9.5 cm in dimensions and 707.0 gm in weight. Cut surface of the right ovary showed a large, markedly congested, multilocular cystic mass containing mucoid material (Fig. 2A). In contrast to right ovary, the left ovary was small, measuring 3.1×2.0×2.0 cm . Cut surface of the left ovary shows a pale tan to pink glistening solid and cystic lesion with an area of ill-defined whitish solid and firm tissue (0.6×0.5×0.5 cm) at the upper portion of the left ovary (Fig. 2B).

Microscopically, the right ovary and fallopian tube showed massive hemorrhage, congestion, and coagulative necrosis without viable lining epithelial cells. Most of the left ovary showed benign thyroid follicles, and diagnosed as struma ovarii. The upper portion of the left ovary showed typical papillary fronds composed of optically clear nuclei, irregular nuclear membrane, nuclear groove and pseudoinclusion, representing a papillary thyroid carcinoma (Fig. 3). There was no evidence of lymphovascular invasion or perineural invasion of the tumor cells. Additional immunohistochemical stains showed positivity for thyroglobulin, galectin-3 and HBME-1 in the tumor cells (Fig. 4) Malignant struma ovarii was incidentally detected by operation of a wrenched large contralateral mucinous neoplasm.

Ultrasound examination of neck after operation showed an about 10.3×14.5×14.9 mm sized heterogeneous slightly hypoechoic mass in the lower pole of the right thyroid lobe. Some color signals are noted within the mass. Possibility of nodular hyperplasia was considered. An about 4.8×8.8×8.5 mm sized heterogeneous isoechoic nodule in the left thyroid lobe. Possibility of nodular hyperplasia was considered. Several small colloid

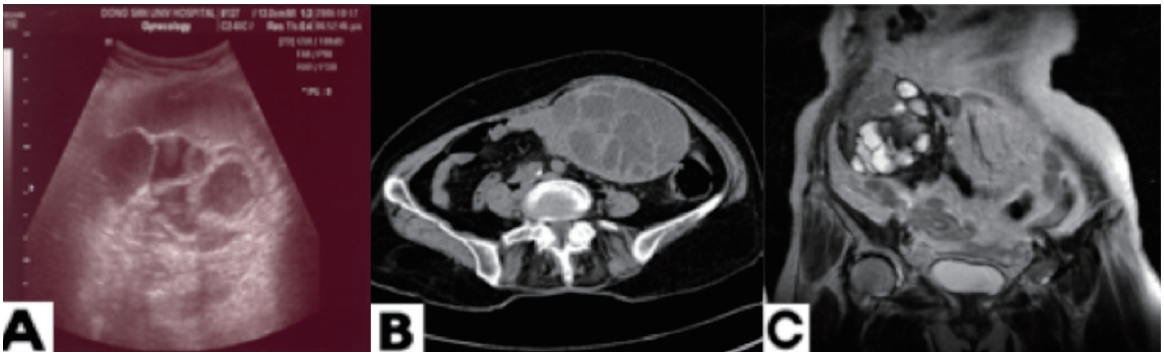

Fig. 1. A: Transvaginal ultrasound examination showed multiseptated huge mass in right adnexa. B: Abdominopelvic computed tomography showed a large ovoid multiseptated cystic mass of right ovary, suspicious of mucinous cystadenoma. C: Magnetic resonance imaging of pelvis showed a large ovoid multiloculated cystic mass in right adnexa.

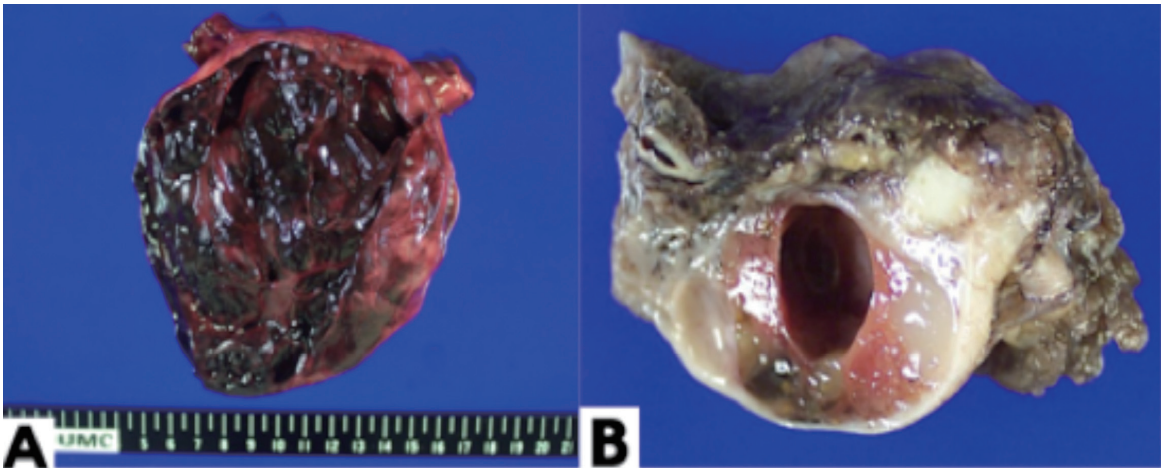

Fig. 2. A: Right ovary shows a large multiloculated cystic mass containing hemorrhagic material. B: Left ovary shows pale tan to pink glistening solid and cystic lesions with an area of ill-defined whitish firm tissue.

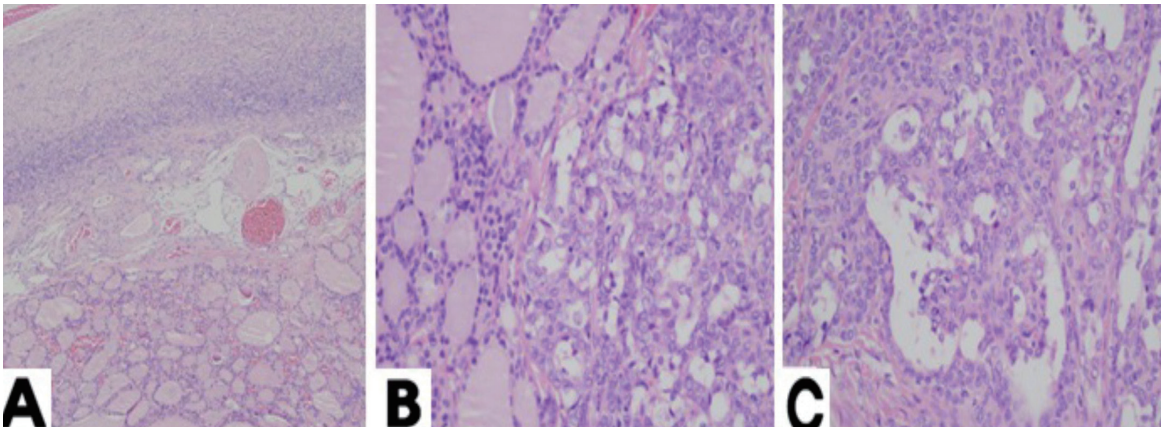

Fig. 3. A: Numerous benign thyroid follicles within the left ovarian stroma are seen (H&E, ×100). B: Benign thyroid follicles (left) are compared with malignant papillary thyroid carcinoma (right) within the left ovary (H&E, ×400). C: Papillary structure composed of optically clear nuclei, irregular nuclear membrane, nuclear groove and pseudoinclusion represents papillary carcinoma in left ovary (H&E, ×400).

cysts were noted in both thyroid lobes. An about 13.1 mm sized enlarged lymph node in the left supraclavicular region probably a reactive lymph node was found. Thyroid function test after operation was in normal range. The patient is still alive for 6 years after the diagnosis of malignant struma ovarii, papillary thyroid carcinoma arising in struma ovarii.

Discussion

Ovarian teratoma is a common benign ovarian tumor consist of ectodermal, mesodermal, and

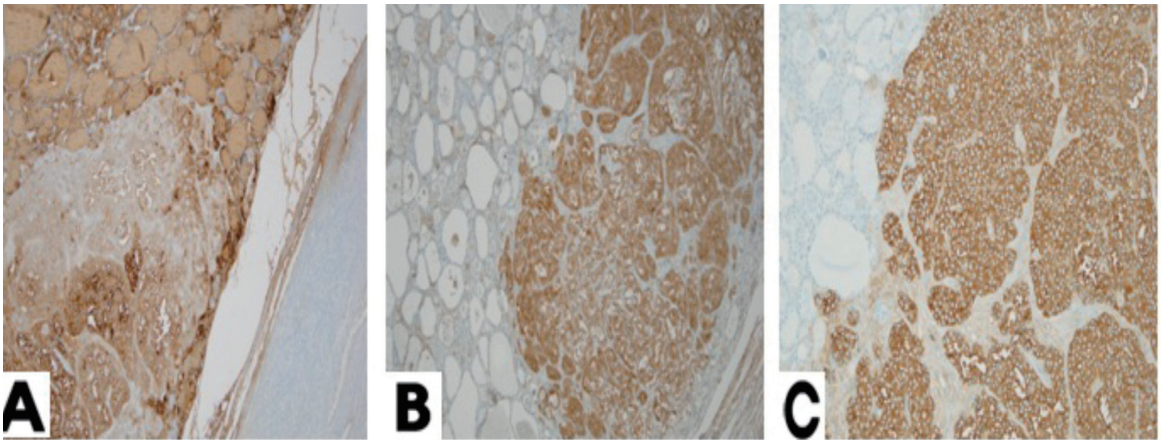

Fig. 4. A: Both papillary thyroid carcinoma components and benign thyroid follicles are all stained for thyroglobulin (×100). B&C: The tumor cells of papillary thyroid carcinoma (right) only shows positive cytoplasmic reaction for galectin-3 (B, ×100) and HBME-1 (C, ×100).

endodermal components, and thyroid tissue component may be found in about 20% of the ovarian teratoma. But struma ovarii containing 50% or more thyroid tissue, is rare, accounting for only 2% of all ovarian teratomas [4]. The majority of struma ovarii is benign tumor, and malignant struma ovarii is rare. Generally malignant struma ovarii occurs in the fifth decade and preferentially involves the left ovary rather than the right ovary. In all reported cases, a total of 39 cases up until 2004, the average age at clinical presentation was 44. Patients predominately presented 45% with a pelvic mass, 40% with abdominal pain, 9% with menstrual irregularities and 5% with hyper-thyroidism [5]. Commonly the patient undergoes surgery, because a malignant ovarian tumor is suspected on radiologic examination [6]. Metastasis from malignant struma ovarii is uncommon, seen in 5-23% of patients with malignant struma ovarii. However, the recent studies suggest a higher potency of recurrence [3].

Diagnosis of struma ovarii is usually made after operation, because frozen section often only reveals a teratoma [6]. Histopathologically, struma ovarii is composed of various-sized thyroid follicles filled with pink-staining, homogenous, gelatinous colloid, lined with cuboidal or columnar epithelium [7]. Malignant struma ovarii showing histopathologic features of thyroid carcinoma, are found in 5-37% of all struma ovarii [6]. Papillary carcinoma is the most common histologic type [8-10]. Common criteria for diagnosis of malignant struma ovarii is the presence of nuclear features of papillary thyroid carcinoma. Nuclei of the tumor cells are enlarged, clear, ground glass, empty and overlapping. The papilla consists of a central fibrovascular stalk with loose connective tissue and variably sized, thin-walled vessels. Mitoses are rare. Intranuclear inclusions of cytoplasm and nuclear grooves are often found. These neoplasms exhibit nuclear features similar to those of their counterparts in the thyroid gland of neck. The identification of psammoma bodies in the context of a thyroid-type papillary neoplasm is highly supportive in the diagnosis of carcinoma [7]. Immunohistochemical staining with thyroglobulin is very useful in supporting a diagnosis of struma ovarii [11]. Antibodies such as Hector Battifora mesothelial [cell]-1 (HBME-1) and galectin-3, can also help to diagnosis papillary thyroid carcinoma [12,13]. Molecular analysis revealed that v-raf murine sarcoma viral oncogene homolog B1 (BRAF)

mutations, common in papillary thyroid carcinoma, were also present in 66% of malignant struma ovarii with papillary features [14]. RET/PTC (*RET* proto-oncogene rearrangements) were seen in 70% of malignant struma ovarii with follicular variant papillary thyroid carcinoma [15].

Rare cases of thyroid carcinoma metastasizing to the ovary, have been reported, and it should be differentiated from malignant struma ovarii. In those cases, the primary thyroid tumor should be detected by clinical thyroid examination and ultrasonography, and the ovarian tumor should show no component of teratoma [16]. In our patient, ultrasound examination of thyroid did not reveal any pathologic lesions. Malignant struma ovarii should be differentiated from other primary ovarian tumors, such as granulosa cell tumor, Brenner tumor, papillary serous cystadenoma or cystadenocarcinoma. Differential diagnosis can be made by the cytologic features representing typical thyroid follicles, and by immunohistochemical staining of organ specific antibodies such as thyroglobulin, TTF-1, inhibin, WT-1 (Wilms tumor-1), and CA-125 [10].

There is no consensus on management of malignant struma ovarii. Surgical removal of the ovarian tumor is main treatment. Surgical treatment varies from total abdominal hysterectomy, plus bilateral salpingo-oophorectomy with omentectomy, to conservative surgery involving unilateral oophorectomy or strumectomy (cystectomy), for preservation of fertility. Some authors have suggested that total abdominal hysterectomy with bilateral salpingo-oophorectomy is a reasonable option in postmenopausal women or in premenopausal women who have completed childbearing. In cases where preservation of fertility is desired, conservative surgery, such as unilateral oophorectomy or strumectomy (cystectomy), should be attempted only if there is an unilateral lesion without evidence of capsular invasion or metastasis [10]. However postoperative management is still controversial [17]. Adjuvant treatment options are including thyroxine, to reduce TSH secretion, near-total thyroidectomy with radioactive iodine ablation.

Because of its rarity and the lack of consensus in treatment, the prognosis of thyroid type carcinoma arising in struma ovarii is difficult to estimate. If only histological criteria are considered, 14% with typical follicular carcinoma, 7% with papillary carcinoma, 100% with undifferentiated (anaplastic) carcinoma, and 0% with highly differentiated follicular carcinoma of ovarian origin (HDFCO) died of neoplasm [18]. Even when clinically malignant, the neoplasm shows long survival, as evidenced by an 84% 25-year survival. Unless obviously poorly differentiated, no single histologic or clinical feature reliably predicts which neoplasms will be biologically malignant. However dense fibrous adhesions and larger strumal size, especially over 12 cm, are suggestive of neoplasms that will have spread at the time of operation or are likely to recur [19].

Summary

Ovarian teratoma is a common benign ovarian tumor, but struma ovarii containing 50% or more thyroid tissue, is rare. Among them, malignant struma ovarii arising in old women is rarer. So we report the malignant struma ovarii, detected incidentally due to the torsion of contralateral ovarian mass in 80-year-old women.

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