Large Wolffian adnexal tumor of the ovary: A case report and literature review

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Received May 7, 2016; Accepted April 13, 2017

DOI: 10.3892/ol.2017.6859

Abstract. Female Wolffian adnexal tumor (WAT) is a rare neoplasm arising from the remnants of the mesonephric duct and <100 cases have been reported globally. The present case report describes a 73-year-old female patient with WAT in the left ovary which, to the best of our knowledge, is the largest benign WAT tumor to be reported. In addition, the present case report reviewed previous studies on the clinical characteristics and therapy for WAT and the surgery methods for female WAT of ovary were summarized. WATs are typically benign; however, a number factors may increase the risk of malignancy.

Introduction

Female Wolffian adnexal tumor was first reported in detail in 1973 by Kariminejad and Scully (1) in a study of 9 cases and named 'female adnexal tumor of probable Wolffian origin (FATWO)'. The World Health Organization subsequently renamed FATWO as Wolffian adnexal tumor (WAT) in 2003. WAT is a rare neoplasm arising from the remnants of the mesonephric duct and predominantly occurs in the broad ligament; however, WAT may occur in the ovaries, fallopian tubes and peritoneum (2). WAT is typically known to behave in a benign manner; however, under certain circumstances more aggressive behaviors have been observed (3). WAT is misdiagnosed due to its rarity and non-specific clinical features and pathological histology forms (4). Therefore, owing to the limited cases and literature, there are no recommended therapeutic approaches (5). In the present study, a case of WAT in the ovary was examined, literature associated with WAT was reviewed and the surgery methods for patients with WAT were summarized.

Case report

On 28 October 2015, 20 years following a hysteromyomectomy, a 73-year-old female visited the outpatient department of the Department of General Surgery, Ninth People's Hospital (Shanghai, China), presenting with abdominal pain and bloating for the previous 2 weeks. An abdominal examination revealed that the patient exhibited abdominal distention, palpation revealed a large lump with an unclear boundary, but there was no tenderness or rebound tenderness. An ultrasound scan identified a large mass and the patient required additional computed tomography (CT) or magnetic resonance imaging (MRI) examination. The patient was admitted to the Ninth People's Hospital (Shanghai, China) to conduct a more detailed examination. Chest X-ray and serum tumor markers (including a-fetoprotein, carcinoembryonic antigen, cancer antigen (CA) 199, CA153, CA724 and CA125) were unremarkable and cardiopulmonary function was acceptable. A routine blood test identified a moderate inflammatory reaction and slightly decreased levels of serum albumin. Whole abdomen CT scan and MRI imaging examinations were conducted (Fig. 1) which confirmed that there was a large abnormal lobulated soft tissue mass, the origin of which was uncertain. Since the patient's abdominal pain and bloating did not improve, and may have worsened with the increasing tumor size, an exploratory laparotomy was conducted, following consent being obtained from the patient's family.

When the abdominal cavity was opened, a large tumor occupying the patient's entire abdominopelvic cavity was observed. The tumor was ~26x24x15 cm in size, with a well-encapsulated solid, ovoid or lobulated appearance (Fig. 2) and exhibited partial adhesion to surrounding tissue. No traces of ascites were identified. The tumor was gradually separated which enabled the origin, of the left ovary, to be identified. The ovarian tumor was removed and examined as frozen sections (FS) to confirm its pathological features and decide whether to enlarge the surgery. No enlarged abdominal or pelvic lymph nodes were identified and the remaining abdomen and pelvis were identified as normal. When the FS proved negative for malignancy, simple tumor resection was achieved.

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Key words: Wolffian adnexal tumor, ovary, benign, surgery method, literature

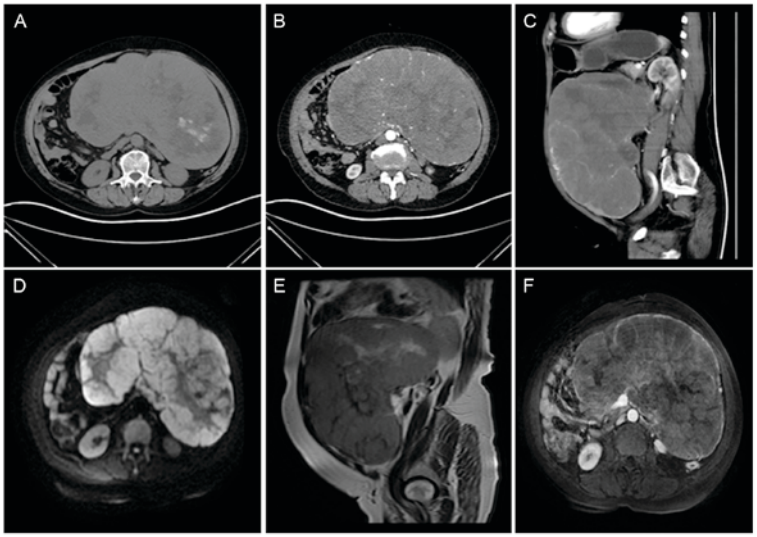

Figure 1. Imaging examination of the patient. (A) Abdominopelvic cavity CT scan, cross plane; (B) abdominopelvic cavity contrast-enhanced spiral CT scan, cross plane; (C) abdominopelvic cavity contrast-enhanced spiral CT scan, sagittal plane; (D) MRI scan using DWI sequence, cross plane; (E) enhanced MRI scan using T2WI sequence, sagittal plane; and (F) enhanced MRI scan using T2WI sequence, cross plane. CT, computed tomography; MRI, magnetic resonance imaging; T2WI, T2-weighted image.

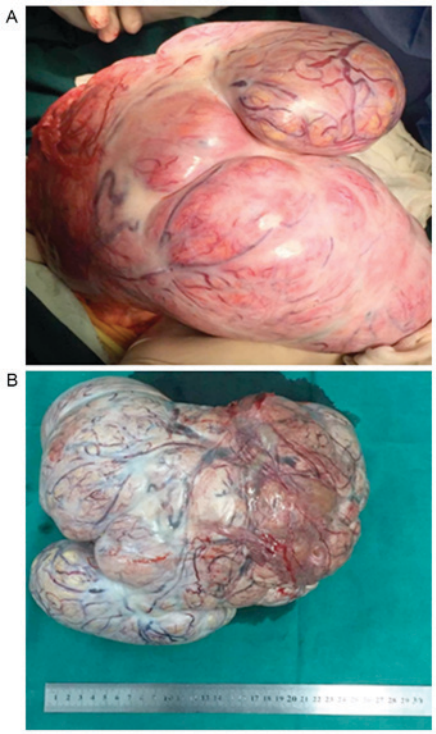

Figure 2. (A) Large mass identified following opening of the peritoneal cavity, which occupied the patient's entire abdominopelvic cavity; and (B) measurement of the excised mass using a ruler in cm.

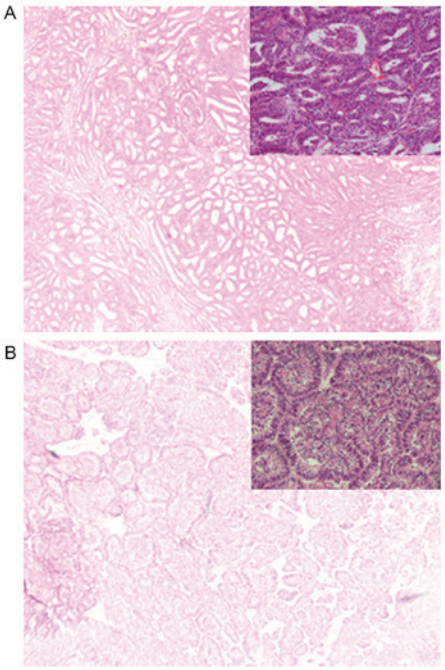

Figure 3. Characteristic histopathological patterns identified on microscopic examination, hematoxylin and eosin staining. (A) A typical sieve-like pattern with hollow tubules varying in size and shape with occasional cysts (magnification, x40 and enlarged to x200). (B) A solid or diffuse arrangement of the neoplastic epithelial cells, closely packed and winding (magnification, x40 and enlarged to x200).

The postoperative period was uneventful and the patient was discharged on day 9 following surgery with no further treatment offered. At the time of writing (~6 months following this surgery), the patient remained alive without evidence of tumor metastasis or recurrence and received regular follow-ups.

The final pathology report identified the tumor as an ovary-derived female WAT. The cross-section of mass (4 μm thick) contained solid and microcystic parts and exhibited a yellow-gray appearance with focal hemorrhage and necrosis. Characteristic histopathological patterns identified on microscopic examination included a solid or diffuse arrangement of the neoplastic epithelial cells, small or medium-sized cells arranged in a microcystic, sieve-like, trabecular and closely packed pattern (Fig. 3). Tissue sections were cut into 4 μm thick pathological section and immunohistochemical staining was performed using strept avidin-biotin complex dyeing at room temperature for 40 min. We found that the sections

Table I. Summarized literature review of ovary-derived Wolffian adnexal tumor cases.

Case no.	Age, years	Metastasis status	Size, cm/external surface	Surgery method	Follow-up
1	56	Situ	14, smooth	H, BSO	NED, 7 years
2	51	Situ	Large, smooth	H, BSO	NED, 9 years
3	52	Situ	15, smooth	H, BSO	NED, 15 years
4	28	Situ	2, smooth	H, BSO	NED, 2 years
5	64	Metastasis	8, smooth	BSO, omentectomy	LFU
6	51	Situ	11, smooth	USO	NED 4 years
7	Reproductive age	Situ	11, smooth	USO	NED, 1 years
8	56	Situ	20, smooth	H, BSO	NED, 1 years
9	58	Situ	12, smooth	H, BSO	LFU
10	41	Situ	10, smooth	USO	NED, 1 years
11	52	Situ	8, smooth	H, BSO	Lung metastasis, 8 years later
12	51	Situ	10 and 4.5, smooth	H, BSO	NED, 37 months
13	27	Situ	10, smooth	STR	NED, 3 years
14	75	Metastasis	15, smooth	USO	LFU
15	87	Metastasis	4.5, smooth	H, BSO	NED, 7 months
16	62	Metastasis	/	H, BSO	NED, 19 years
17	51	Situ	2.5, smooth	H, BSO	LFU
18	73	Situ	26, smooth	STR	NED, 5 months

Situ, the tumor lesions were in the ovary and not expanded or no metastasis; H, BSO, hysterectomy with bilateral salpingo-oophorectomy; BSO, bilateral salpingo-oophorectomy; USO, unilateral salpingo-oophorectomy; STR, simple tumor resection; NED, no evidence of disease; LFU, lost to follow-up; /, not given in the literature.

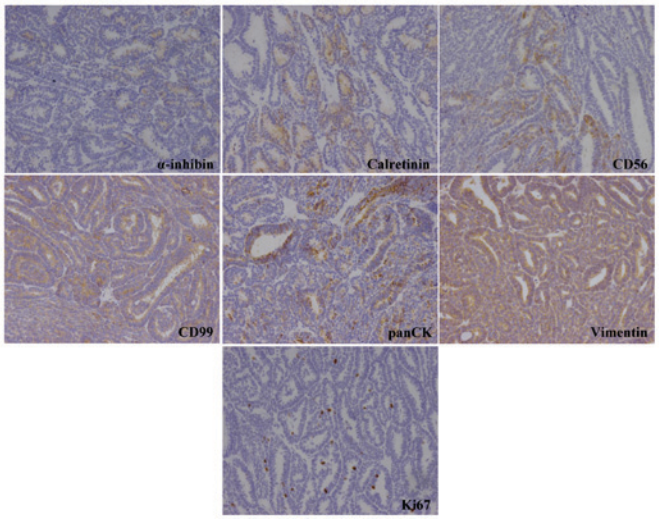

Figure 4. Positive immunohistochemical staining of Ki-67. CD, cluster of differentiation; panCK, pan-cytokeratin.

were positive for vimentin, cluster of differentiation (CD)99 and pan-cytokeratin (CK) and focally positive for calretinin, α-inhibin and CD56. The positive proportion of Ki-67 was 3%; however, staining was negative for chromogranin, epithelial membrane antigen, CD10, CD34, synaptophysin and S-100 protein (Fig. 4).

The present case report was approved by the Ethics Committee of the Ninth People's Hospital, School of Medicine, Shanghai Jiao Tong University. Preoperative informed consent was obtained from the patient in accordance with institutional guidance. The pathological samples were obtained from the surgical resection specimens which did not result in any disadvantages to the health and prognosis of patient. The present case report maintains the privacy of the patient.

Discussion

WAT is a rare neoplasm, with <100 cases worldwide, and arises from the rare persisting remnants of the mesonephric duct (6). The age at diagnosis ranged between 18 and 81 years, with a mean age of 50 years (7). The prognosis of the patient's tumor was not associated with its clinical presentation and cytology, which made it difficult to diagnose (5). Therefore, there were no clear recommendations regarding the patient's preoperative diagnosis. The diagnosis of the present case relied primarily on histopathological features, which were characterized by a tubular pattern with either closely packed tubules or solid cords, a sieve-like growth pattern produced by cysts of various sizes and a diffuse growth composed of

spindle or polygonal cells (8). The primary differential diagnosis included Sertoli-Leydig cell tumors, clear cell tumors and granulosa cell tumors since the microscopic appearance of the aforementioned tumors exhibited similarity to that of WAT (4-9). A previous study (10) revealed that CD56-positivity may be a diagnostic biomarker to differentiate between malignant FATWOs and benign lesions; however, this required further validation. Although the patient in the present case report had been identified as ovary-derived WAT, the tumor did not initially induce gynecological symptoms but induce abdominal pain and bloating. In addition, to the best of our knowledge, no previous studies had reported a WAT as large as the tumor in the present patient. Therefore, it was increasingly difficult to diagnose the present patient and it was elected to only conduct an exploratory laparotomy.

WAT typically behaves as a benign lesion; however, in a number of cases, more aggressive behavior has been encountered (11). Previous studies identified that ~1/5 of the cases were associated with an adverse outcome and the principal metastatic sites were the liver and the lung (6,9). Owing to the rarity of this type of tumor, there is no standard surgical therapy for WAT patients. Lesin *et al* (12) demonstrated that the majority cases of relapsed WAT had occurred in patients who were initially treated with tumor resection only, and hypothesized that the optimal therapy for WAT was complete surgical resection with hysterectomy and bilateral adnexectomy. In addition, adjuvant chemotherapy or radiation therapy was controversial and typically not an effective treatment (7).

The literature review of the present case report identified 34 studies associated with WAT. To the best of our knowledge, a total of 18 ovary-derived WAT cases, including the present case, have now been reported (Table I). In Table I, the first 11 cases had been reported before (13). It was revealed that the reported ovary-derived WAT cases age ranged between 28 and 87 years, and 17 cases were patients who exhibited tumors on only one side of the body. In addition, the tumors identified in the literature report typically extended to the fallopian tubes, uterus, appendix, lung, liver and other sites; however, the tumors rarely extended to the contralateral ovary. Only 1 patient (case no. 12) was diagnosed with tumors in two ovaries. A total of 16 cases (88.89%) had a hysterectomy with bilateral salpingo-oophorectomy or bilateral salpingo-oophorectomy alone, and only 2 cases had a simple tumor resection. However, there was not enough information to analyze the statistical difference of disease-free survival in these 18 cases. Therefore, unless the patient is an unmarried female or too old to tolerate surgery, it is suggested to select hysterectomy with bilateral salpingo-oophorectomy. Following the initial surgical treatment, it is recommended that patients are to be appropriately followed up for a long-term period. Additionally, the present case report identified that there was limited optional therapy to treat recurrent or postoperative metastatic WAT tumors (14). A previous study (6) reported that molecular targeted therapy, including the tyrosine kinase inhibitor Gleevac® (STI 571), may be considered. However, additional studies are required to determine the effectiveness of this option.

Acknowledgements

The authors thank Dr Hong-Xiu Han for the pathological and immunohistochemical assistance.

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