Clinical Case Report

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Primary leiomyosarcoma of the fallopian tube

A case report and literature review

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Abstract

Rationale: Primary leiomyosarcoma (LMS) of the fallopian tube is extremely uncommon. To the best of our knowledge, so far only 21 cases of primary fallopian tube LMS have been reported in English-language literature. No new case has been reported in the past 7 years.

Patient concerns: A 44-year-old premenopausal patient presented with a 5-day history of lower abdominal pain.

Diagnoses: Pelvic ultrasonography detected an $8.8 \times 7.8 \times 6.5$ cm solid and cystic mass in the left side of the pelvic cavity. The tumor was diagnosed as a primary fallopian tube LMS on paraffin section.

Interventions: The patient treated surgically followed by 4 cycles of postoperative chemotherapy with dacarbazine and DDP.

Outcomes: The patient succumbed to the disease 27 months after the initial therapy.

Lessons: Tube LMS is a rare malignant tumor with unknown etiology, difficult early diagnosis, highly invasiveness, high local recurrence and distant metastasis rate, rapid progress, and poor prognosis. It is extremely rare so we can only summarize limited experience from limited data. Every case of tubal LMS is worth being reported.

Abbreviations: DTIC = dacarbazine, FIGO = International Federation of Gynecology and Obstetrics, LMS = primary leiomyosarcoma, OS = overall survival, PFS = progression-free survival, SMA = smooth muscle actin, STS = soft tissue sarcoma.

Keywords: case report, chemotherepy, fallopian tube, leiomyosarcoma, literature review, surgery

1. Introduction

Primary fallopian tube carcinoma is a very rare tumor accounting for about 0.1% to 1.8% of all gynecological tumors. Since diagnosis is difficult, there may be additional cases that have been misdiagnosed as ovarian malignancies.[1] Considering that the fallopian tube sarcoma is rarely seen, primary leiomyosarcoma (LMS) of the fallopian tube, a kind of tube sarcoma, is even more uncommon. To the best of our knowledge, no new case has been reported in last 7 years. Here, we describe the 22th case of primary fallopian tube LMS treated with surgery and 4 cycles of postoperative chemotherapy.

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2. Case report

A 44-year-old premenopausal patient (gravida 3, para 2) presented with a 5-day history of lower abdominal pain. It began with a sudden sharp pain lasting about 5 hours, then turned to continuously dull pain. A pelvic mass was felt during the gynecological examination. Pelvic ultrasonography detected an $8.8 \times 7.8 \times 6.5$ cm solid and cystic mass in the left side of the pelvic cavity, a uterus of normal appearance, and 2 cm-deep free fluid in the pelvic cavity. Cervical cytology showed no abnormal findings. All tumor markers including serum CA125, CA19–9, alpha fetoprotein (AFP) and carcinoembryonicantigen (CEA) were within normal limits. A chest radiograph was also normal.

A laparotomy was performed. During the operation, a $10 \times 9 \times 8$ cm hard, gray-white colored irregular tumor located in the left fallopian tube was found. A small area of rupture covered in a blood clot was observed on the surface of the tumor. Inspection of the abdominal cavity provided no evidence of metastasis. The left tube and the tumor were removed and it was identified as poorly differentiated adenocarcinoma which was highly suspected as metastasis of signet ring cell carcinoma from the frozen tissue sections. In the intraoperative exploration of intestinal canal, liver, spleen, and stomach, no obvious metastasis was found. The patient was recommended to undergo gastrointestinal endoscopy to evaluate the primary lesions for follow-up treatment. However, the tumor was diagnosed as a primary fallopian tube LMS on paraffin section. In immunohistochemistry, vimentin, desmine, and smooth muscle actin (SMA) were positive, while

cytokeratin7, cytokeratin20, and phosphoenolpyruvate carboxykinase were negative. The irrigating cytology demonstrated no evidence of malignant cells. Accordingly, a 2nd operation including abdominal hysterectomy, left oophoroectomy, right salpingo-oophorectomy, partial omentectomy, and pelvic lymphadenectomy was performed after 17 days. The histology indicated no evidence of direct invasion or metastasis into the uterus, bilateral ovaries, omentum, and pelvic lymph nodes. According to the current surgical staging for tubal cancer, this tumor was staged as International Federation of Gynecology and Obstetrics (FIGO) Ic. The patient later received 4 courses of chemotherapy with dacarbazine (DTIC, 300 mg/day, day 1–2; 200 mg/day, day 3–5) and DDP (20 mg/day, day 1–5) every 21 days without any severe complications. The patient did not follow-up regularly and succumbed to the disease 27 months after the initial therapy. Our Hospital Ethics Committee approved this case report. Patient's husband gave his consent and authorized this case report to be published.

3. Discussion

LMS is an extremely rare kind of smooth muscle tumor which affects the uterus, stomach, intestine, and posterior pelvic peritoneum. Primary fallopian tube LMS is even rarer. In 1886, Senger[2] reported the first case of primary salpingosarcoma. In the same year, Gottschalk[3] reported the first case of primary fallopian LMS, which was described as spindle-cell sarcoma at that time. Since there are only 22 cases reported in English-language literature including this case we have reported, we cannot make any conclusions with the limited data at present. However, we can still draw on past experiences in defining the characteristics of available reported cases of primary LMS of the fallopian tube summarized in Table 1.

Tubal LMS can occur at any age in adult life, with a median age of 50.47 years ranging from 21 to 70 years old. Although carcinosarcoma always occurs in postmenopausal women, the median age of incidence is 60 years old.[4]

LMS has no typical symptoms, most patients (87.5%, 7–8) have a history of lower abdominal quadrant pain. Vaginal discharge, although considered as a significant symptom, has been reported only in 2 early cases.[3,5] Pelvic mass is often found during gynecologic examination, and ultrasound or computed tomography is often considered as the origin of the ovary or uterus. In addition, it is necessary to make complete preoperative evaluation to discover all suspected lesions and evaluate if the tumor has invaded or metastasized with computed tomography or magnetic resonance imaging scans. In this case, the patient was evaluated only by an ultrasonic examination before operation. However, it seems now that the ultrasonic results are not sufficient. Serum CA125 levels remained at normal or slightly increased levels.[6]

The diagnosis is always confirmed histopathologically during or after the operation. Based on the published case reports, tube LMS is microscopically composed of spindle-shaped cells arranged in fascicular clusters with bizarre nucleus and mitoses. Hemorrhage and necrosis are also noted. Cells in our case conformed to tube LMS were observed in hematoxylin eosin specimens (Fig. 1A, B). In immunohistochemistry, desmin, h-Caldesmon, SMA, and other smooth muscle markers are always positive. In this case, both desmin and SMA (Fig. 2A, B) as well as vimentin (Fig. 2C) are positive. The frozen sections of the specimen were highly suspected as metastatic signet-ring cell carcinoma. Inspection of the abdominal cavity showed no dubious primary lesions, so it was considered appropriate to remove the tumor alone, without extending the range of resection based on that result. Further treatment was performed according to the final histology report.

Presently, there is no standard operation procedure for tube LMS. Many different surgical treatments have been reported in the published cases (Table 1). In this case, our patient underwent abdominal hysterectomy, bilateral salpingo-oophorectomy, partial omentectomy, and pelvic lymphadenectomy. Similar to the surgical treatment of ovarian neoplasm, for the nonmetastasized tube LMS (FIGO I,II), the mainstay of operation is represented by complete resection (R0) consisting of peritoneal washing, inspection of peritoneum and the surface of abdominal organs, excision of all the abdominal masses, random biopsies, total abdominal hysterectomy, bilateral salpingo-oophorectomy, partial omentectomy, and pelvic and periaortic selective lymphadenectomy. For the metastasized tube LMS (FIGO III, IV), a debulking surgery should be considered in order to excise all the primary lesion and reduce the metastatic risks as much as possible many different regimens have been reported in published works, including acombination of ifosfamide with pirarubicin,[4] gemcitabin and docetaxel,[7] and DTIC-adriablastine-vincristine-cyclophosphamide schedule (CyVADIC).[8] Of note is the report by Kobayashi et al[6] involving a case of a stage IIIc patient who survived more than 6 years without any evidence of recurrence with intraperitoneal cisplatin followed by prolonged oral etoposide for 1 year. However, due to the rarity of the tube LMS, the efficacy of the treatment has only been demonstrated via case reports making it impossible to have strong evidence supporting a best regimen. So far, the medicine use has referenced chemotherapy drugs used in uterus LMS.

Doxorubicin, ifosfamide, and DTIC are the top 3 active agents in soft tissue sarcoma. DTIC achieves an overall response of 17%. In LMS, DTIC showed a median progression-free survival and overall survival (OS) of 2 and 8.2 months respectively as monotherapy, while the figure rose to 4.2 and 16.8 months when combined with gemcitabine.[9] According to our experience in treating uterus LMS, APDTIC (epirubicin, DDP, and DTIC) shows a satisfactory results. So, we chose DTIC combined with DDP in this case. Our patient achieved a 27-months OS with 4 cycles of PDTIC regimen.

Generally, LMS has poor sensitivity to radiotherapy. Postoperative radiotherapy was used in some early LMS cases, and no recurrence was found after 3 months to 1 years of follow-up.[3,5] There is a case in German reporting telecobalt irradiation of the pelvis which showed 1-year tumor free survival time in the treatment of primary tube LMS.[10] A number of studies suggest that postoperative radiation therapy can effectively reduce pelvic recurrence, but there is still no clear evidence that radiotherapy can improve the survival rates in patients with LMS. In recent years, some researchers have discovered through clinical trials that pelvic radiation therapy does not improve the PSF and OS of stage I, II uterine sarcoma patients.[11] Therefore, currently it is considered that early stage LMS patients do not require routine radiotherapy.

4. Conclusion

Tube LMS is a rare malignant tumor with unknown etiology, difficult early diagnosis, highly invasiveness, high local recurrence and distant metastasis rate, rapid progress, and poor prognosis. It is extremely rare so we can only summarize limited experience from limited data. Every case of tubal LMS is worth

Table 1

Available reported cases of primary leiomyosarcoma of the fallopian tube.

No.	Author	Year	Country	Age	Para	Chief complaint	ϕ	CA125	Operation	Stage	IHC	Chemotherapy/Radiotherapy	Prognosis
1	Gottschalk[3]	1886	–	37	3	–	–	–	①?	–	–	–	DOD 4 d
2	Jones[3]	1893	USA	–	–	–	–	–	②	–	–	–	–
3	Jacobs[3]	1897	–	41	1	–	–	–	③④	–	–	–	DOD 1 y
4	Jacobs[3]	1904	–	27	0	–	–	–	③④	–	–		NED 2 mo
5	Scheffzeck[3]	1911	–	61	–	–	–	–	① (left)?⑤	–	–	–	No follow-up
6	Dodd[3]	1924	–	55	0	–	–	–	①	–	–		NED 1-y follow-up
7	Dodd[3]	1924	–	60	2	–	–	–	④⑥	–	–		DOD 2 y with METS
8	Reiber[3]	1939	–	44	–	–	–	–	①	–	–	Postoperative external irradiation	NED 1-y follow-up
9	Rickford[5]	1945	–	69	2	Vaginal discharge	"Grapefruit"	–	⑧	–	–	Postoperative external irradiation	NED 3-mo follow-up
10	Scheffey[3]	1946	USA	70	1	Low pelvic pain, bloody vaginal discharge			④⑦ [⑥ in 1932; ⑤ (negative) 7 mo before surgery]		–	–	NED 2-y follow-up
11	Bornstein[12]	1953	–	49	–	–	–	–	③④	–	–	–	DOD in 1y
12	Abrams[12]	1958	USA	21	1	–	3.5 (Diameter)	–	④⑦⑪	–	–	–	NED 5-y follow-up
13–15 (N/A)													
16	Jacoby[13]	1993	USA	39	0	Right lower quadrant pain	11.0 × 6.5 × 5	Normal	⑧⑨⑩ (right)⑪	I	–	4 Cycles cisplatin (100 mg/m^2) doxorubicin (50 mg/m^2)	NED 2-y follow-up
17	Ebert[8]	1995	Germany	39	1	Lower left abdominal pain	8 × 9 × 7.5	–	④⑥⑦⑪	I	–	3 Cycles CyVADIC	DOD 5 mo with METS (lung, abdominal organs, and bone)
18	Kohler[10]	1997	Germany	57	–	–	–	–	④⑦⑨⑩-⑪	–	–	Telecobalt irradiation of the pelvis (Hd 52 Gy)	NED 1-y follow-up
19	Mariani[14]	2005	Italy	48	–	–	7 × 7 × 5	Normal	④⑦ (extra-fascial) ⑪⑫⑥ at 12	Ia	Desmin, Actin+; Kerain and neuroendocrin makers–	None	NED 5-y follow-up
20	Ueda[4]	2010	Japan	69	2	Left lower quadrant pain	6 (Diameter)	Normal	④⑦⑨⑩-⑪⑬	IIc	αSMA, muscle actin+; desmin, CD34–	6 Courses pirarubicin (40 mg/m^2, d1) ifosfamide (1.5 g/m^2, d1–4)	DOD 39 mo with METS (liver, lung, and supraclavicular lymph node)
21	Kobayashi[6]	2010	Japan	70	0	Lower abdominal pain	"Child head"	500	④⑦	IIIc	Vimentin, CD10, SMA+; cytokeratin, h-Caldesmon, HMB45, Melan A, epithelial common antigen–	100 mg of CDDP intraperitoneally at the end of the operation, oral etoposide (50 mg daily for 21 d every 28 d) for 1 y	NED >6-y follow-up
22	Jacobs[7]	2010	Germany	59	0	Intermittent constipation, right lower quadrant pain	6 × 4 × 3	Normal	④ (⑦in 1991 for benign disease)	IV	Vimentin, SMA, desmin, S-100 protein, MIB1+; Ki67 80%	2 Cycles gemcitabine 900 mg/m^2 (d1 + 8) and docetaxel 100 mg/m^2 (d8), both q21, radiation + bisphosphonates (q28) for bone metastases	DOD 8 mo with METS
23	You et al (current study)	2017	China	44	2	Lower abdominal pain	10 × 9 × 8	Normal	④ (⑦⑨⑪	Ic	Desmin, SMA, vimetin +; CK7, CK20, PCK–	4 Cycles dacarbazine (300 mg/d, day 1–2; 200 mg/d, day 3–5) DDP (20 mg/d, day 1–5) q21	DOD 27 mo with METS

ϕ, Tumor size; ①, salpingectomy; ②, autopsy; ③, supravaginal hysterectomy; ④, bilateral salpingo-oophorectomy; ⑤, curettage; ⑥, appendectomy; ⑦, total abdominal hysterectomy; ⑧, unilateral salpingo-oophorectomy; ⑨, pelvic lymph node dissection; ⑩, para-aortic lymph node dissection/biopsy; ⑪, partial omentectomy; ⑫, random biopsies; ⑬, low anterior resection of the rectum. DOD = dead of disease, IHC = immunohistochemistry, METS = metastasis, N/A = not available, NED = no evidence of disease.

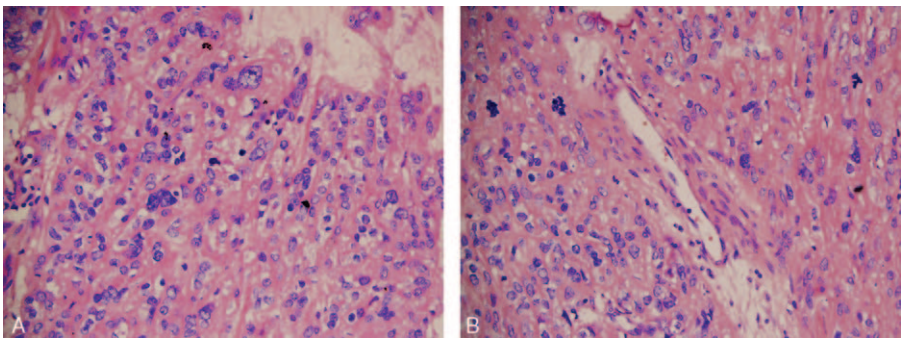

Figure 1. Tumor cells composed of fascicles of spindle cells stained with hematoxylin-eosin, (A) Bizarre nucleus and (B) mitotic cells (original magnification ×400).

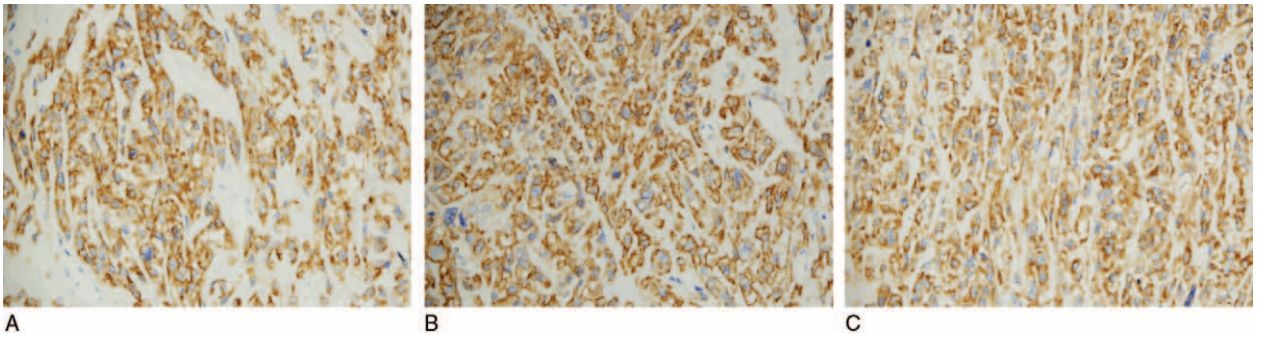

Figure 2. Immunohistochemistry: antibody reaction against (A) Desmine, (B) smooth muscle actin (SMA), (C) vimentin (×400).

being reported. In addition, except surgery, chemotherapy, and radiotherapy, other kind of therapies, such as hormonal therapy and targeted therapy, have showed potential effectiveness in soft tissue sarcoma. If they can help in the treatment of tube, LMS can only be answered from the results of future investigations.

Author contributions

Data curation: Di You, Qilin Wang, Wei Jiang, Lingjun Zhao.
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