Clinical Case Report

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Pancreatic metastasis of angiosarcoma (Stewart–Treves syndrome) diagnosed using endoscopic ultrasound-guided fine needle aspiration: A case report

Junpei Sasajima, MD, PhD[a], Jiro Uehara, MD[b], Takuma Goto, MD[a], Shugo Fujibayashi, MD[a], Kazuya Koizumi, MD, PhD[a,c], Yusuke Mizukami, MD, PhD[a,d], Akemi Ishida-Yamamoto, MD, PhD[b], Mikihiro Fujiya, MD, PhD[a,*], Toshikatsu Okumura, MD, PhD[a]

Abstract

Background: Pancreatic involvement of angiosarcoma is extremely rare.

Methods: We herein report a rare case of angiosarcoma associated with chronic lymphedema (Stewart–Treves syndrome) with pancreatic metastasis that was diagnosed using endoscopic ultrasound (EUS)/fine needle aspiration (FNA).

Results: A 43-year-old woman with a history of radical hysterectomy with bilateral inguinal lymphadenectomy and chemoradiotherapy for cervical cancer 15 years prior noticed the presence of erythematous indurative plaques on her right femoral region, where chronic lymphedema had developed. Contrast-enhanced computed tomography (CT) revealed not only multiple nodules in the subcutaneous tissue of the right femoral region but also a 25 mm × 20 mm solid mass in the region of the pancreatic tail. A histological analysis of the specimens obtained using EUS/FNA revealed angiosarcoma that was immunohistochemically positive for platelet/endothelial cell adhesion molecule-1 but negative for cytokeratin. The patient was diagnosed as Stewart–Treves syndrome that had metastasized to the pancreas. Chemotherapy was performed, but the patient died 14 months after her diagnosis.

Conclusion: Unfortunately, this patient was not followed up, even though she had chronic lymphedema of the right femoral region due to the repeated occurrence of phlegmon. To improve the survival rate of this fatal secondary malignant complication of radical lymphadenectomy, an early diagnosis with consecutive and long-term clinical follow-up and close monitoring for Stewart–Treves syndrome is therefore important.

Abbreviations: EUS = endoscopic ultrasound, FNA = fine needle aspiration, MAID = mesna doxorubicin dacarbazine and ifosfamide.

Keywords: Angiosarcoma, Endoscopic ultrasound/fine needle aspiration, Pancreatic metastasis, Stewart–Treves syndrome

1. Introduction

The development of angiosarcoma associated with chronic lymphedema, which sometimes occurs after radical lymphadenectomy, is known as Stewart–Treves syndrome.[1] Patients with Stewart–Treves syndrome have a very poor prognosis.[2] We herein report a rare case of Stewart–Treves syndrome with pancreatic metastasis that was diagnosed using endoscopic ultrasound (EUS)/fine needle aspiration (FNA) without any complications. Metastasis to the pancreas is an uncommon condition, representing only about 2% to 4% of malignant lesions in the pancreas in a series of surgical resection.[3] Renal cell carcinoma, colon cancer, and lung cancer are the most common tumors that metastasize to the pancreas, and pancreatic involvement of angiosarcoma is extremely rare.[4–6]

2. Case report

A 43-year-old woman noticed the occurrence of erythematous indurative plaques on her right femoral region. Fifteen years prior to this presentation, she had undergone radical hysterectomy with bilateral inguinal lymphadenectomy for cervical cancer, and adjuvant chemoradiotherapy had been subsequently performed at another hospital. Despite developing chronic lymphedema of the right femoral region and the repeated occurrence of phlegmon, she was not followed up after the termination of adjuvant chemoradiotherapy.

A physical examination revealed light-red and partially purpuric irregular-shaped erythematous indurative plaques with 2-cm-wide dark-red ulcerated nodules on the right thigh and in

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[a] Division of Gastroenterology and Hematology/Oncology, Department of Medicine, [b] Department of Dermatology, Asahikawa Medical University, Hokkaido, [c] Gastroenterology Medicine Center, Shonan Kamakura General Hospital, Kanagawa, [d] Center for Clinical and Biomedical Research, Sapporo Higashi Tokushukai Hospital, Sapporo, Japan.

[*] Correspondence: Mikihiro Fujiya, Division of Gastroenterology and Hematology/Oncology, Department of Medicine, Asahikawa Medical University, 2–1–1–1 Midorigaoka-Higashi, Asahikawa 078–8510, Hokkaido, Japan (e-mail: fjym@asahikawa-med.ac.jp).

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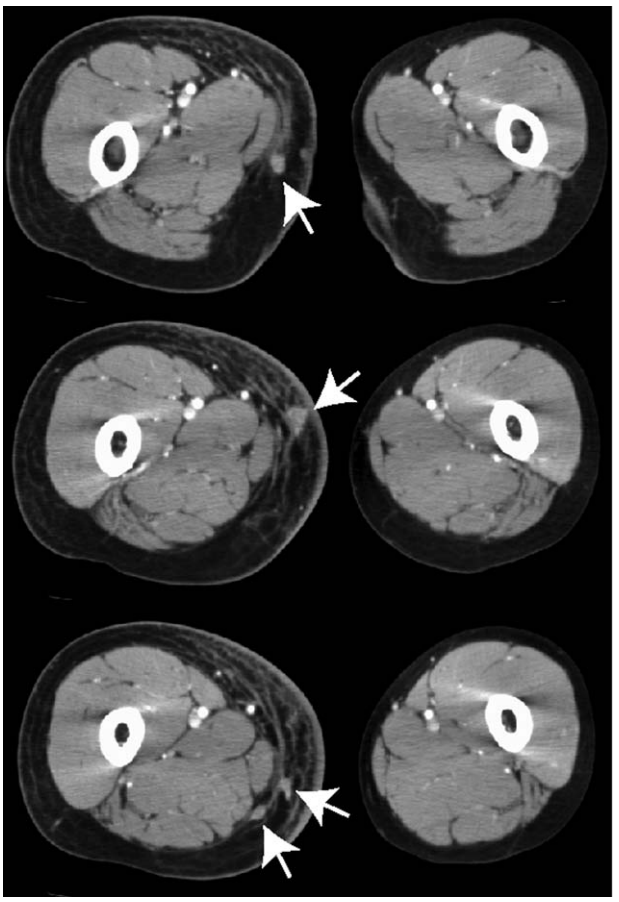

Figure 1. Contrast-enhanced computed tomography of the femur demonstrated right femoral enlargement due to chronic lymphedema. Multiple nodules (see the arrows) with contrast enhancement were detected in the subcutaneous tissue of the right femoral region.

the genital area. A blood test showed normal findings except for elevated levels of vascular endothelial growth factor (67 pg/mL; normal range 0.0–38.3 pg/mL). Contrast-enhanced CT revealed multiple nodules in the subcutaneous tissue of the enlarged right femoral region (Fig. 1). The pathological findings of a biopsy of the right femoral region revealed angiosarcoma, and therefore Stewart–Treves syndrome was suspected. Multiphase contrast-enhanced CT revealed a 25 mm × 20 mm solid mass in the region of the pancreatic tail with poor enhancement (Fig. 2). Endoscopic ultrasonography (GF-UC240P; Olympus Medical Systems Corp., Tokyo, Japan) showed a 30-mm-wide hypoechoic mass in the pancreatic tail (Fig. 3). Subsequently, EUS/FNA was performed by 2 passes of a 22-gauge needle (EchoTip Ultra Endoscopic Ultrasound Needle; Cook Japan Inc., Tokyo, Japan) without any complications related to the procedure. A histological analysis of the obtained specimens revealed angiosarcoma that was immunohistochemically positive for platelet/endothelial cell adhesion molecule-1 but negative for cytokeratin (Fig. 4).

Chemotherapy using docetaxel was administered after the patient was diagnosed as Stewart–Treves syndrome. After 3 courses, chemotherapy was changed to paclitaxel due to leucopenia. After 11 courses of paclitaxel chemotherapy, multiple liver metastases developed, and mesna, doxorubicin, dacarbazine, and ifosfamide (MAID) chemotherapy was administered as third-line chemotherapy. However, MAID chemotherapy was performed only for 2 courses, as the patient developed febrile neutropenia. Of the 4 drugs included in the MAID regimen, only dacarbazine was continued and combined with teceleukin for 2 courses. However, the patient died 14 months

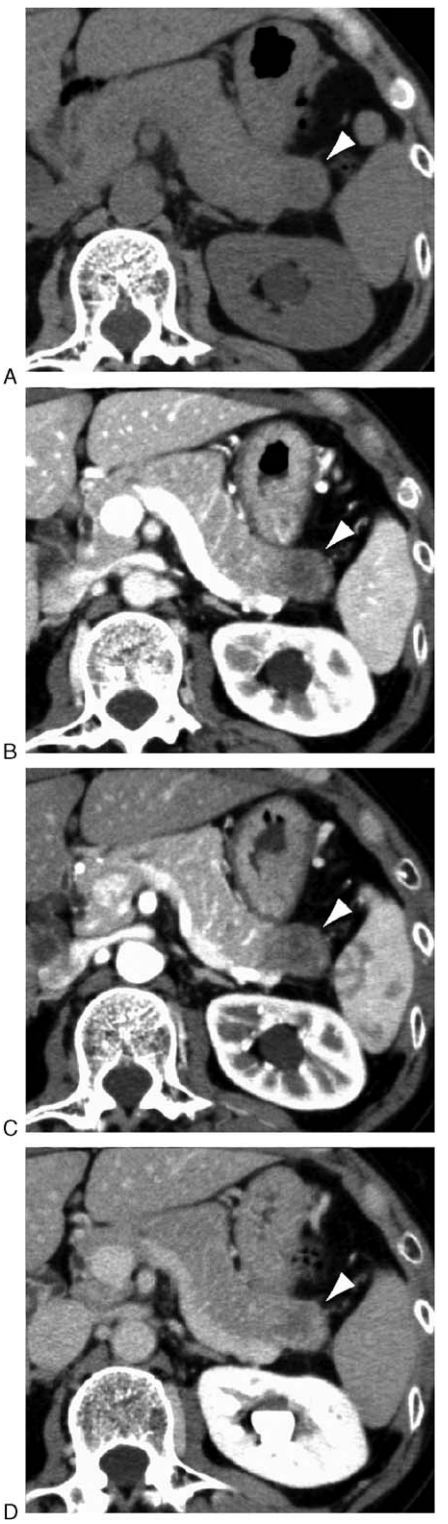

Figure 2. Multiphase contrast-enhanced computed tomography revealed a 25 mm × 20 mm low-attenuation solid mass (see the arrow heads) in the pancreatic tail with poor contrast enhancement. A) Plain, B) arterial phase, C) pancreatic phase, D) portal venous phase.

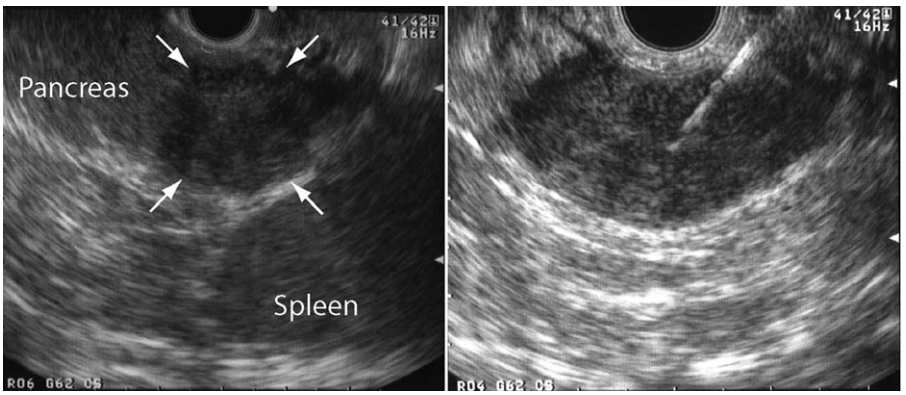

Figure 3. Endoscopic ultrasonography revealed a 30-mm hypoechoic mass (see the arrows) of the pancreatic tail. EUS/FNA was performed by 2 passes of a 22-gauge needle with no complications related to the procedure. EUS/FNA = endoscopic ultrasound/fine needle aspiration.

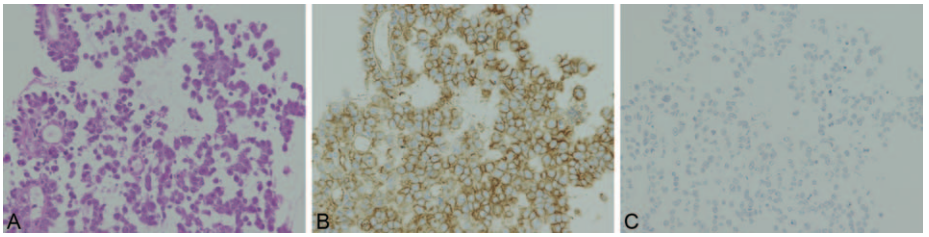

Figure 4. A histological analysis of the specimens obtained through EUS/FNA revealed angiosarcoma that was immunohistochemically positive for platelet/endothelial cell adhesion molecule-1 but negative for cytokeratin. A) Hematoxylin and eosin staining, B) platelet/endothelial cell adhesion molecule-1 staining, C) cytokeratin (magnification: approximately 400×). EUS/FNA=endoscopic ultrasound/fine needle aspiration.

after her diagnosis due to the progression of metastatic angiosarcoma.

Ethical approval was not required for this case report as it did not relate to the patient's privacy or treatment. Informed consent for the publication of this case report could not be obtained because the patient had died.

3. Discussion

To our knowledge, this is the first case report in which pancreatic metastasis of angiosarcoma was diagnosed using EUS-FNA. Pancreatic involvement of angiosarcoma is extremely rare,[4,5] and only 4 cases have been reported in the English literature.[4,5,7,8] In addition, only 2 reports have been published concerning the performance of EUS-FNA in patients with angiosarcoma.[7,9] The spleen was punctured in the first case,[7] while the retroperitoneum was punctured in the second.[9] No complications associated with EUS-FNA have been reported in any of the three cases, including the present patient, suggesting that EUS-FNA is a safe procedure for use in diagnosing such organ angiosarcoma.

In the present case, angiosarcoma developed on the right femoral region with chronic lymphedema 15 years after radical hysterectomy with bilateral inguinal lymphadenectomy followed by adjuvant chemoradiotherapy for cervical cancer. Therefore, the patient was diagnosed with Stewart–Treves syndrome, which is characterized by the development of angiosarcoma associated with chronic lymphedema. Lymphedema is known to play a role in the pathogenesis of angiosarcoma,[1,10] indicating that any kind of radical lymphadenectomy has the potential to induce the development of angiosarcoma. In addition, radiation therapy has also been reported to be a risk factor for Stewart–Treves syndrome.[1,10] Because angiosarcoma is believed to develop from 5 to 15 years after surgery,[1] patients who undergo radical lymphadenectomy, especially with adjuvant radiotherapy, should be followed up in the long term.

Patients with Stewart–Treves syndrome have a very poor prognosis.[2] Unfortunately, this relatively young patient was not followed up, even though she had chronic lymphedema of the right femoral region due to the repeated occurrence of phlegmon. To improve the survival rate with this fatal secondary malignant complication of radical lymphadenectomy, early diagnosis with consecutive and long-term clinical follow-up and close monitoring for Stewart–Treves syndrome is important.

4. Conclusion

We encountered a rare case of Stewart–Treves syndrome with pancreatic metastasis that was diagnosed using EUS/FNA without any complications. To improve the survival rate of Stewart–Treves syndrome, early diagnosis with consecutive and long-term clinical follow-up and close monitoring is important.

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