Case Report

A rare and highly malignant Stwart-Treves syndrome case after breast cancer surgery: a case report

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Background: Stewart-Treves syndrome (STS) is a lymphatic sarcoma secondary to chronic lymphedema of the extremities. Most STS patients have a history of breast cancer and have undergone radical mastectomy and postoperative radiation and chemotherapy. It usually occurs 11 to 12 years after surgery, and about 0.45% of patients are estimated to have the disease. The characteristics of STS include that it is clinically relatively rare, has a high degree of malignancy, can spread easily in the absence of timely treatment, and has low survival rate. Herein, we report a case of STS which developed 13 years after breast cancer-related lymphoedema (BCRL). It allows doctors to recognize and detect the disease earlier.

Case Description: A 74-year-old woman had undergone modified radical mastectomy 13 years ago for invasive ductal breast cancer in her left breast. After multiple rounds of postoperative chemoradiotherapy, multiple purple lesions were found in the left upper limb during physical examination in April 2021. The lesions spread rapidly and were varied in size. An immediate skin biopsy reported the lesions as STS. The patient was diagnosed with lymphangiosarcoma with metastasis (STS). The surgical method was shoulder joint amputation, chest wall resection, and local flap transfer. After surgery, the patient underwent 6 rounds of paclitaxel 300 mg + carboplatin 300 mg chemotherapy. After chemotherapy, the patient's wound healed and the suspected metastasis disappeared. At the time of writing, she has survived for more than 13 months, and her quality of life has improved significantly, to the satisfaction of the patient and her family. The patient is able to eat normally and lead a normal life with some assistance, without significant weight loss.

Conclusions: Although rare, STS is a serious invasive complication of breast cancer surgery. To increase their relative survival time, patients with BCRL need to identify and thoroughly investigate rapidly progressing skin lesions, and undergo timely surgery.

Keywords: Case report; Stewart-Treves syndrome (STS); lymphedema; lymphangiosarcoma

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Introduction

Lymphangiosarcoma is a rare type of tumor secondary to chronic lymphedema of the limbs. The term was first used by Stewart and Treves in 1948 to describe a very rare tumor, also known as Stewart-Treves syndrome (STS), which almost always occurs on the basis of chronic lymphedema. It is often associated with long-term chronic lymphedema of the upper limb secondary to radical mastectomy (1,2). It may occur as a sequela of primary or secondary chronic lymphedema. It usually occurs 11 to 12 years after breast cancer surgery, and about 0.45% of patients are estimated to have the disease. The vast majority of STS cases occur in the upper limbs, with less than 10% reported in the lower extremity lymphedematous sites (3), and even

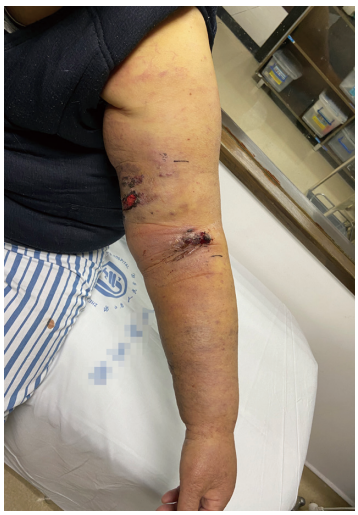

Figure 1 Before surgery, there were multiple purple patches on the upper limbs.

less frequently due to factors related to knee replacement surgery (4). Most of STS patients have a history of breast cancer and have undergone radical mastectomy and repeated chemoradiotherapy. As STS develops, the general limb edema aggravates, local tenderness occurs, and erythema appears on the local skin, which further develops into purplish-red demarcated papules or blisters. The early manifestations are red, blue, and bluish-purple porphyritic or papular nodules, which are often multiple and scattered subcutaneous nodules that are not easily detected, gradually transforming into stasis spots and cellulitis, and later fusing into large ulcerated and hemorrhagic masses. Tumors grow rapidly, often spread along subcutaneous tissue or deep fascia, invade deep muscles, or spread through subcutaneous lymphatic vessels. We present the following case in accordance with the CARE reporting checklist (available at https://gs.amegroups.com/article/view/10.21037/gs-22-344/rc).

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). This report has been approved by the Ethics Committee of Zhejiang Provincial People's Hospital (Approval No. QT2022191) with the informed consent of the patients and their families. A copy of the written consent is available for review by the editorial office of this journal.

A female patient, born on 1 February 1948, with no relevant family history, underwent modified radical mastectomy for infiltrating ductal breast cancer in her left breast in 2008. The postoperative pathology suggested invasive ductal breast cancer with ipsilateral axillary metastasis of 10/31. After surgery, taxotere and adriamycin (TA) regimen chemotherapy was performed for 6 rounds and radiotherapy was performed for 24 times. Swelling of the left upper arm, forearm, back of hand, and finger occurred immediately after surgery, which was diagnosed as upper limb lymphedema. In April 2021, due to the sudden appearance of multiple purple erythema on the left upper limb, which rose higher than the skin surface and was accompanied with pain, the patient presented to the Zhejiang Provincial People's Hospital for diagnostic resection of local lesions on the left upper limb. The pathological findings of the left upper limb were consistent with lymphedem-associated lymphangiosarcoma. The immunohistochemical staining results were as follows: tumor cells ERG(+), CD34(+), c-myc(+), Ki67(+, 90%), P53(mutant), EMA(–), CK(Pan)(–), Fli-1(–), general photos of the patient are shown in *Figure 1*.

Physical examination yielded the following findings: the patient's left upper limb had obvious edema, with multiple irregular purplish-red plaques elevated above the skin surface, some of which were ulcerated and bleeding; the skin was hard and some of the plaques showed leather-like changes. The patient had severe pain and poor upper limb mobility.

On 16 April 2021, the preoperative PET-CT results were as follows: left chest wall soft tissue thickening with increased ^{18}F-fluorodeoxyglucose (FDG) metabolism, which led us to consider the possibility of tumor invasion. Right axillary lymph node metastasis was possible. Multiple thickening of the skin in the subcutaneous fat space of the left upper limb was flocculent, nodular, and cord-like. Patchy, low-density shadow of the muscle bundles were present around the left humerus, ulna, and radius, and increased metabolism of FDG was observed above this area, which was considered consistent with the influencing manifestations of sarcoma. The preoperative pathological results of multiple tissues were consistent, indicating a high degree of malignancy, and multiple metastases were found in the axillary lymph nodes and chest wall skin. Combined with preoperative biopsy, MR and PET-CT, it was determined that the patient had metastases in the left chest wall, neck, and right axilla.

The decision to perform amputation and resection of

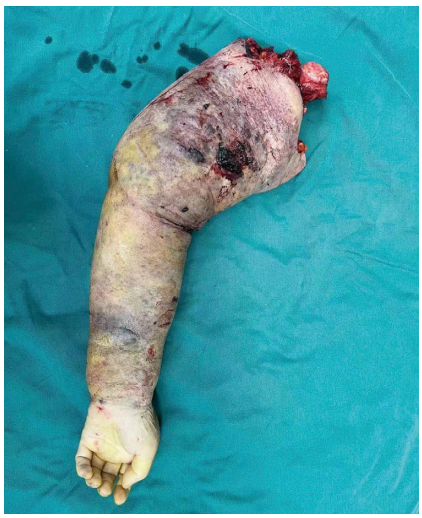

Figure 2 Amputated left upper limb.

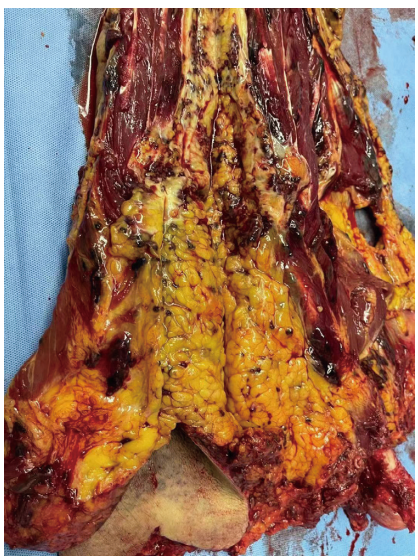

Figure 3 Internal lesions of the left upper limb.

the chest wall lesions was made after multidisciplinary preoperative discussion. The malignancy of the tumor was very high, and the 1-year survival rate was reported to be very low. After our multidisciplinary discussion, the patient and her family were made aware of the characteristics of high malignancy and low survival rate of the tumor. The patient accepted our treatment plan of amputation + chemotherapy and signed the informed consent. The patient underwent shoulder amputation, chest wall metastasis resection, and local flap transfer under general

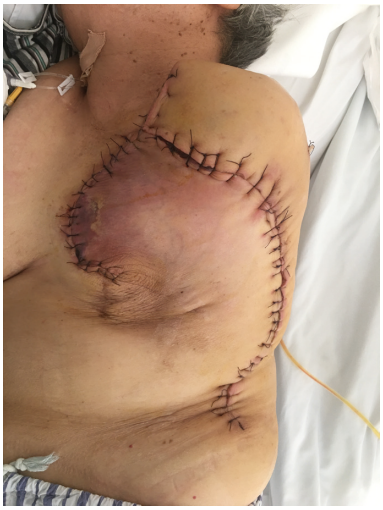

Figure 4 Postoperative resection margin was partially necrotic.

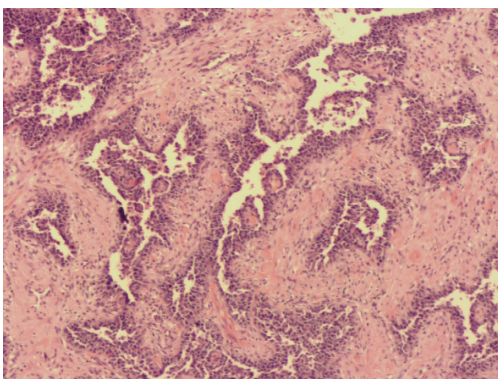

Figure 5 Pathological image of lymphangiosarcoma: ERG(+), CD34(+), c-myc (+), Ki67 (+90%), P53 (mutant), EMA (–), Fli-1 (+), CK(Pan) (–), Fli-1 (–). HE dyed, 100×.

anesthesia on 23 April 2021. The operation went smoothly, as shown in *Figure 2* and *Figure 3*. Postoperative skin necrosis still occurred in the chest of the patient (*Figure 4*).

Tumor markers were in the normal range during treatment. Postoperative pathological photographs are shown in *Figure 5*. After surgery, the patient received chemotherapy in the oncology department of our hospital, using paclitaxel 300 mg + carboplatin 300 mg chemotherapy for 6 rounds (24 June 2021 to 7 October 2021).

The right axillary lymph nodes were repeatedly inspected during chemotherapy and found to shrink and disappear. A year after surgery, the patient's wound had healed, the previous metastasis disappeared, and her quality of life had improved (*Figures 6*,*7*). To date, the patient has survived

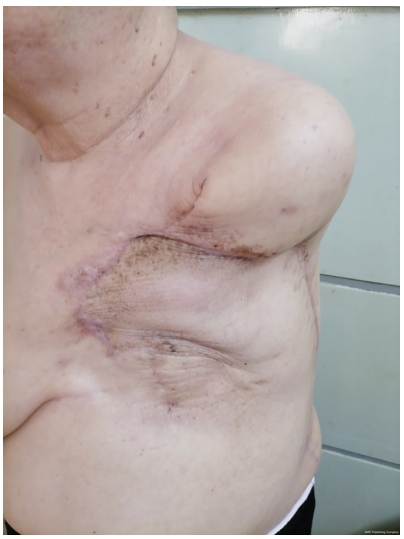

Figure 6 Thirteen months after surgery, the wound has healed.

Figure 7 Thirteen months after surgery, the wound has healed.

for 13 months after surgery and more than 6 months after chemotherapy (*Figure 8*). At present, the patient is eating normally and living almost normally with the help of her family members without significant weight loss. The patient and her family members are very satisfied with our treatment. We will continue to conduct follow-up on to the patient's condition, and the relevant medical information which requires further attention.

Discussion

The overall prognosis of STS is poor; it is characterized by rapid progression, and is prone to distant metastasis. Its incidence is about 0.07–0.45% of patients who survive more than 5 years after breast cancer surgery (5-7). Therefore, the clinical understanding of STS in the lower limbs is generally insufficient, and it is easily misdiagnosed and missed. Lymphangiosarcoma of the upper extremity is often found in patients with long-term lymphedema after breast cancer surgery. The clinical manifestations are not specific at the early stage, the lesions progress quickly, the prognosis is poor, and it is difficult to distinguish from other skin diseases at the early stage. Doctors have different cognitive levels of hemangiosarcoma, which influences whether early detection and treatment are initiated. The level of medical treatment also affect the prognosis of this disease. Early pathological biopsy is helpful for diagnosis and treatment. Due to the rarity of STS, even relatively experienced physicians may overlook its potential occurrence, and it is recommended that similar signs be detected and eliminated by biopsy as soon as possible. Lymphangitic sarcomas, once found, are more likely to metastasize to the chest wall and the contralateral axilla, and even to the whole body. Recurrence of left axillary metastasis and eventual death have been reported months after amputation of right lymphosarcoma (8). In the case reported here, the potential of STS was recognized in a timely manner, the treatment process of pathologic diagnosis was performed in time, and our diagnosis was timely and accurate. On preoperative PET-CT, we found that the tumor had spread rapidly to the entire upper limb, left chest wall, and contralateral axilla, and we chose shoulder amputation and side breast tissue resection. The advantage of surgery is that the tumor lesions can be removed as much as possible, but the disadvantage is that the trauma is relatively large, and the side chest wall does not heal easily in one stage. After dressing change, the site can be healed. In this case, the suspected metastasis in the right axilla could not be completely removed after evaluation, so the surgery was not complete and the second stage chemotherapy was needed. However, the choice of surgery was correct, and the patient and her family agreed with our treatment plan. Fortunately, the patient was very sensitive to our chemotherapy regimen, the chemotherapy effect was good, the metastasis decreased and disappeared,

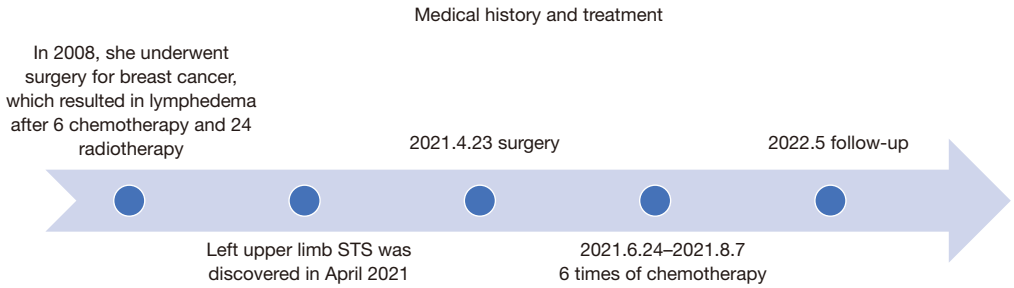

Figure 8 Medical history and treatment.

and the chest wall wound healed. She has survived for more than 13 months with improved quality of life through active management after chemotherapy. The patient is able to eat normally and lead a normal life with some assistance, without significant weight loss. The patient and her family are very satisfied with our treatment.

The confirmation of STS is mainly via physical examination and pathology, and can be assisted by Doppler ultrasound, MR and PET-CT. Laboratory indicators including tumor indicators are not remarkable for this case. Surgical treatment as soon as possible after diagnosis combined with postoperative chemotherapy, immunotherapy, and targeted therapy can improve patient survival, among which targeted therapy may be the development direction of future treatment for cutaneous angiosarcoma. Aggressive treatment of BCRL may be a good way to prevent STS.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://gs.amegroups.com/article/view/10.21037/gs-22-344/rc

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://gs.amegroups.com/article/view/10.21037/gs-22-344/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). This report has been approved by the Ethics Committee of Zhejiang Provincial People's Hospital (approval No. QT2022191) with the informed consent of the patients and their families. A copy of the written consent is available for review by the editorial office of this journal.

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