

Stewart-Treves syndrome

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Stewart-Treves syndrome is a rare cutaneous angiosarcoma with a poor prognosis. Physicians must be aware of this lethal syndrome, especially in patients who have been treated for breast cancer with radiation and lymph-node dissection (such as the case reported here). Patients who develop unexplained enlarging plaques of coalescing purple papules should have immediate biopsy for early diagnosis of Stewart-Treves syndrome. MRI, along with PET/CT, can help evaluate the extent of disease and help with treatment strategies.

Case report

In September of 2012, a 70-year-old female presented with erythema and induration in the left arm that she presumed was caused by a recent mosquito bite. The patient's past medical history revealed left-sided breast cancer that was treated with mastectomy and lymph-node dissection, chemotherapy, and radiation therapy six years earlier. The new presenting lesion appeared on the ipsilateral side as her left mastectomy. The patient's initial assessment was thought to be cellulitis of the left arm, and she was treated with multiple courses of antibiotic therapy for two months with no significant improvement. An incision and drainage at that point did not demonstrate any purulent drainage. Approximately one month later, multiple blue satellite lesions developed within the area of erythema and induration. The mass also advanced in size at a rapid rate, to 20 centimeters in circumferential length extending from the proximal arm down to the elbow. The enlarging plaque of coalescing purple papules appeared necrotic, with multiple small satellite lesions that had erupted within the plaque (Fig. 1).

After biopsy of the lesion, histology results confirmed an angiosarcoma. The patient was consequently treated with



Figure 1. 70-year-old female with Stewart-Treves syndrome. Photograph of the left arm demonstrates an area of violaceous skin with surrounding satellite lesions.

multiple doses of chemotherapy. Since the diagnosis and treatment of the left-arm angiosarcoma, both the size of the mass and metabolic activity on CT/PET imaging showed significant improvement without evidence of metastatic disease.

Magnetic resonance imaging (MRI) performed after the initial incision and drainage revealed marked edema-like signal with decreased signal on the T1-weighted sequence and increased signal on the T2-weighted images involving the subcutaneous soft tissues of the arm, without muscle or osseous involvement (Fig. 2).

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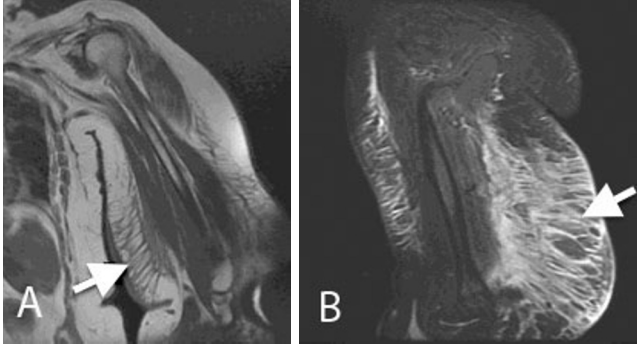


Figure 2. 70-year-old female with Stewart-Treves syndrome. A: T1-weighted image. B: STIR image demonstrating edema-like signal in the subcutaneous tissues of the arm (arrows).

A subsequent PET/CT scan after the biopsy confirmed an angiosarcoma for staging purposes that demonstrated marked hypermetabolism. It was associated with severe subcutaneous edema confined to the left arm without evidence of adenopathy or distant metastasis (Fig. 3).

Discussion

Stewart-Treves syndrome is a rare and deadly cutaneous angiosarcoma, with around 400 reported cases in the world literature (1). It develops in cases of long-standing chronic lymphedema (2). The pathogenic mechanism causing the angiosarcoma is not known. However, it has been postulated that since lymphangiomas (proliferation of lymph vessels) is often seen in uninvolved areas of the affected edematous arm, lymphatic blockage stimulates the growth of lymphatic vessels, possibly via cytokines such as vascular endothelial growth factor (VEGF). It is unclear whether radiation contributes to the risk of Stewart-Treves syndrome. It could indirectly contribute to an increased risk by causing axillary-node sclerosis, resulting in lymphatic blockage and lymphedema (1).

The majority of reported cases have occurred in association with lymphedema in postmastectomy patients, with over 90% involving the upper extremity (2).

The prevalence of angiosarcoma is estimated to be 0.45% in patients living five years after a modified radical mastectomy (3).

Although this tumor is not directly related to breast cancer or radiotherapy, chronic lymphedema is the common factor that leads to Stewart-Treves syndrome. Edema secondary to cardiac or renal disease has not been associated with this malignancy, however, suggesting that factors other than chronic edema alone must be present to result in this syndrome. The median time from mastectomy to the development of angiosarcoma is 5 to 10 years (4, 5). The mean interval between the original breast-cancer diagnosis and the subsequent angiosarcoma was 9.7 years for patients with upper-extremity tumors and 4.4 years for those with chest or breast tumors, respectively (6).

Angiosarcomas are extremely aggressive tumors with a high local recurrence rate and a tendency to metastasize early to many areas. Metastatic angiosarcoma to the lungs and chest wall is a common cause of death in patients with Stewart-Treves syndrome and often occurs even after aggressive surgical treatment (2). The survival of patients with Stewart-Treves syndrome is poor, ranging from 19 months to 31 months (5). Patients treated with amputation had a slightly better prognosis than those treated by radiation therapy (3).

Physicians must be aware of this rare but lethal syndrome, especially in patients who have been treated for breast cancer with radiation and lymph-node dissection. Patients who develop unexplained enlarging plaques of

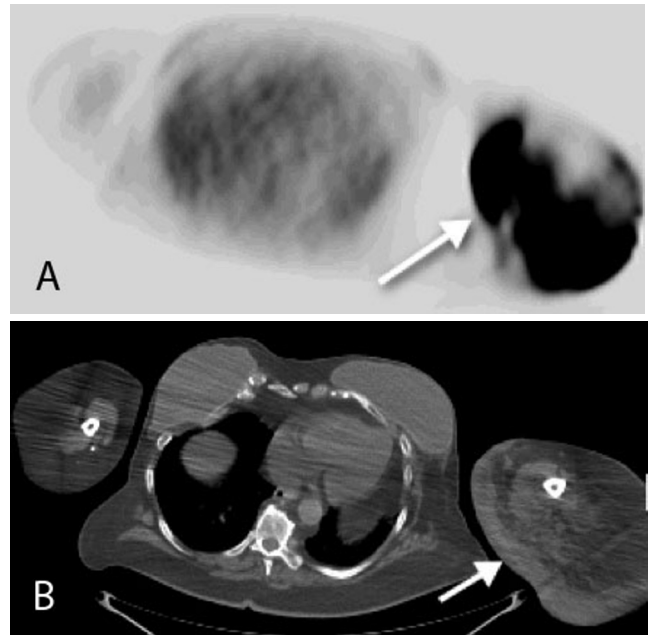


Figure 3. 70-year-old female with Stewart-Treves syndrome. Axial PET (A) and CT images (B) demonstrate a hypermetabolic region along the posterior left arm with marked subcutaneous edema and skin thickening on CT (white arrows).

coalescing purple papules should have immediate biopsy for early diagnosis of Stewart-Treves syndrome. MRI, along with PET/CT, can play a role in evaluating the extent of disease and for helping with treatment strategies (7)

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