Stewart-Treves Syndrome – Endangered Not Extinct
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INTRODUCTION

Cutaneous angiosarcoma is a high-grade malignant neoplasm of endothelial cells, which develops in one of three settings: on the head and neck of the elderly, at sites of prior radiotherapy, and in the setting of chronic lymphedema. More than 90% of all angiosarcomas associated with lymphedema arise after mastectomy and lymph node dissection. This is referred to as Stewart–Treves Syndrome (STS). The condition was first described in 1948 by Stewart and Treves who presented six cases of angiosarcoma in postmastectomy lymphedema. STS is relatively rare, with an incidence between 0.07% and 0.45% in patients who survive at least five years after radical mastectomy.1,2 The peak age for developing STS is 65-70 years, corresponding to trends in breast cancer occurrence, with additional years for the subsequent development of chronic lymphedema.3

The pathophysiology of STS is controversial. Although it is acknowledged that lymphedema can induce angiosarcoma, the mechanism is unknown.4 Within an area of chronic lymphedema, the initial lesion typically presents as a palpable subcutaneous mass and develops reddish-blue macules or nodules with satellite lesions. Ultimately, the lesion ulcerates and in advanced disease, can become necrotic.7

CASE PRESENTATION

A 79-year-old Caucasian female presented for evaluation of a lesion on her left forearm. The lesion began approximately one month prior and had been rapidly enlarging, becoming painful and bleeding on occasion. Her past medical history was significant for hypertension, diabetes and breast cancer of the left breast 14 years prior, for which she had been treated with mastectomy, lymph node dissection, radiation, and chemotherapy. The patient states she developed lymphedema of her left arm shortly after the surgery and radiation therapy. Family history was significant only for colon cancer in her mother. On exam, the patient had a firm, friable, erythematous to violaceous tumor with a blue cast, which was one of several similar nodules. The patient returned for follow-up three months later and unfortunately had developed new lesions around the graft site. She decided against any further intervention.

REFERENCES