Stewart-Treves syndrome is a kind of angiosarcoma associated with chronic lymphedema. It was first reported by Stewart and Treves [1] in 1948. In their study, they reported a series of six patients who had developed angiosarcoma in their lymphedematous extremities after radical mastectomy. The exact mechanism of Stewart-Treves syndrome is not clear. It may be due, however, to the accumulation of protein-rich interstitial fluid in a chronically swollen limb [2]. This fluid alters the local immune environment of the chronically edematous limb and promotes lymphangiogenesis. It also aggravates the locally immunocompromised state to easily presented malignancy. The prognosis of Stewart-Treves syndrome has been regarded as poor due to its aggressiveness. The mean survival period from this disease has been quoted to be seven months, with an approximately 35% overall five-year survival rate [3]. Because of its aggressiveness and poor prognosis, early detection and surgical management are considered the key factors in the survival of patients.

In this article, a rare case of Stewart-Treves syndrome in the lower extremities that was misdiagnosed as a pressure ulcer is reported. A 72-year-old female was referred to the authors' clinic for the management of a protruding purple to black skin lesion with central ulceration, which developed about six months before the patient’s presentation, in her right lower leg (Fig. 1). Her physical examination showed that the size of the lesion was about 9 cm × 8 cm, and there was a mild pitting edema in her right lower leg. Since the initial development of the lesion, the patient had been treated in the local clinic under the impression of

![Fig. 1. Preoperative findings of a 9 cm × 8 cm purple to black protruding skin lesion with central ulceration on the upper lateral side of the right lower leg with diffuse edema.](http://dx.doi.org/10.5999/aps.2013.40.3.275)
lateral aspect of the right lower leg with possible
extension to the adjacent soft tissue and a unilateral
subcutaneous honeycomb-appearance that suggested
long-standing lymphedema (Fig. 3). There was no
evidence of metastasis.

A wide excision was performed with a 3 cm safety
margin that included part of the fibular head. The
frozen section was margin-free of the tumor, and the
defect was covered with a split-thickness skin graft.
The histologic examination showed diffuse extravas-
ation of red blood cells with rich vascularity, and the
vascular endothelial cells showed severe pleomor-
phism (Fig. 4A). The immunohistochemistry of
CD31, CD34, and vimentin (Fig. 4B) were positive,
so the pathologist confirmed the diagnosis of angio-
sarcoma.

The oncology department recommended adjuvant
radiotherapy, but the patient refused it because of her
poor economic status. She underwent right lower leg
amputation above her knee one month after her first
operation, and she expired three months later due to
pleural metastasis with malignant pleural effusion.

Angiosarcoma is a rare soft tissue malignancy of
vascular endothelial cell origin. Angiosarcomas can
be classified into several subtypes [3]. Primary
cutaneous angiosarcoma occurs on the face and scalp
of older patients. Lymphedema-associated angiosar-
coma, also known as Stewart-Treves syndrome, is
associated with chronic lymphedema. Other
angiosarcoma subtypes include radiation-induced
angiosarcoma, primary breast angiosarcoma, deep
soft-tissue angiosarcoma, and visceral angiosarcoma.

The histopathological appearance of Stewart-
Treves syndrome can vary from a well-differentiated
lesion to a poorly differentiated one [3]. Low-grade,
well-differentiated lesions show irregular vascular
channels and cutaneous angiosarcoma with an

---

Fig. 2. Magnetic resonance imaging of a 6.7 cm × 3.0 cm × 8.3 cm enhanced irregular mass with an intermediate signal intensity on the T2 weighted image and good development of the vascular structure accompanied by diffuse subcutaneous edema.

Fig. 3. The positron emission tomography-computed tomography of the fludeoxyglucose-avid malignant tumor in the upper lateral aspect of the right lower leg with possible extension to the adjacent soft tissue and a unilateral subcutaneous honeycomb-appearance (white arrows) suggests long-standing lymphedema. There was no evidence of metastasis.
irregular or sinusoidal vessel pattern. High-grade, poorly differentiated lesions can comprise undifferentiated cells, so they are difficult to differentiate from other malignancies. These tumors can be confirmed via immunohistochemistry and are usually positive for factor-VIII-related antigen, vimentin, CD34, and CD31.

Wide excision is the only known curative treatment option for the localized disease, and there is no known effective therapy for the metastatic disease [4]. Thus, only early diagnosis and prompt radical excision with or without adjuvant radiotherapy can promote survival.

The diagnosis can be delayed, however, because it may show only erythema, swelling, and a papule-like skin lesion initially, and ulceration, hemorrhage, and mass fungation later [3]. Thus, if there is pain, sudden enlargement, or purple-red nodules on the skin of the involved extremity, recognition of their significance and their prompt evaluation by a surgeon are very important.

In conclusion, the authors experienced a case of Stewart-Treves syndrome in the lower leg, which was misdiagnosed initially and therefore, the treatment of which was delayed. It was reported herein because an atypical skin lesion in a chronically edematous limb can be malignant even if it looks benign, and a high level of suspicion and early management of the lesion are very important in improving the potential for patient survival.

References
1. Stewart FW, Treves N. Lymphangiosarcoma in postmastectomy lymphedema; a report of six cases in elephantiasis chirurgica. Cancer 1948;1:64-81.
2. McHaffie DR, Kozak KR, Warner TF, et al. Stewart-Treves syndrome of the lower extremity. J Clin Oncol 2010;28:e351-2.
3. Young RJ, Brown NJ, Reed MW, et al. Angiosarcoma. Lancet Oncol 2010;11:983-91.
4. Abraham JA, Hornick EF, Kaufman AM, et al. Treatment and outcome of 82 patients with angiosarcoma. Ann Surg Oncol 2007;14:1953-67.

Images

Fig. 4. Histologic findings. (A) H&E stain (×40) demonstrated abnormal, pleomorphic, and malignant endothelial cells. (B) Positive immunohistochemistry of vimentin (×100).

Treatment of Refractory Lower Extremity Ulcer Associated with Sneddon’s Syndrome

Hyo Hyun Seok¹, Yongjoon Noh¹, Eui Cheol Jeong¹,², Ji Ung Park¹,², Yoon Ho Hong³
¹Department of Plastic and Reconstructive Surgery, Seoul National University College of Medicine, Seoul; Departments of ²Plastic Surgery and ³Neurology, SMG-SNU Boramae Medical Center, Seoul, Korea

Correspondence: Eui Cheol Jeong
Department of Plastic Surgery, SMG-SNU Boramae Medical Center, 20 Boramae-ro 5-gil, Dongjak-gu, Seoul 156-707, Korea
Tel: +82-2-870-2331, Fax: +82-2-831-2826
E-mail: ejjeong.md@gmail.com

No potential conflict of interest relevant to this article was reported.

A patient with livedo reticularis and ischemic cerebrovascular disease was first reported by Sneddon in 1965, after which the disease was named Sneddon’s syndrome [1], a very rare disease with the incidence of four new cases per million per year [2].

A 49-year-old male patient was referred from the dermatologic clinic to our department for painful