Stewart-Treves Syndrome in the Lower Limb after Arthroplasty- A Case Report

Abstract

Stewart-Treves Syndrome is a rare and fatal condition where cutaneous angiosarcoma—a high-grade malignant tumor originating in the vascular and lymphatic endothelium—classically develops in the upper limbs post-mastectomy, with radiation therapy and axillary lymph node dissection. There are very few reports of the syndrome developing in the lower limbs, without any preceding malignancy or radiation therapy. The median development time is 11 years. Angiosarcoma originates in the vascular and lymphatic vessels, and the diagnosis is based on histopathology and immunohistochemistry findings. We report an unusual presentation of the Stewart–Treves Syndrome in an elderly female involving the lower limb with preexisting chronic lymphedema, where the tumor developed 15 months after total knee arthroplasty.

Keywords: *Arthroplasty, cutaneous angiosarcoma, Stewart–Treves Syndrome, vascular tumor*

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Sir,

Stewart-Treves Syndrome (STS) is a cutaneous angiosarcoma following longstanding lymphedema in extremities classically involving the upper limb, post-mastectomy with axillary dissection. STS may develop after chronic lymphedema, (congenital or acquired), infrequently involving the lower limbs. The mean onset of the development is 11 years.[1] We report a rare presentation of STS in the leg developing soon after total knee arthroplasty (TKA) in a preexisting post-filarial chronic lymphedema.

A 77-year-old female, presented with sharply demarcated, gross swelling of the right lower limb for 6 months [Figure 1]. There was diffuse erythema, telangiectasia, blotchy bluish hyperpigmentation, and numerous variably-sized nodules clustered distally. Rugose background skin showed linear arthroplasty scar over the knee. The affected area was firm, non-tender, warm, and proximally woody-hard. Inguinal lymph nodes were not palpable. There were no systemic complaints.

The patient had filarial lymphedema in the affected limb for 15 years and underwent TKA 15 months before presentation. Seven months after surgery, an apparent bruise appeared on the lateral side of the right knee, which gradually suffused. It became aggressive in the past 1 month with rapid background edema and increased number and size of nodules.

Histopathology showed numerous oval and polygonal cells in sheets with prominent nucleoli. Neoplastic cells were in a papillaroid, sinusoid, and diffuse pattern with numerous poorly formed vascular channels, several mitotic figures (0–2/hpf) [Figure 2a]. Neoplastic cells project into numerous vascular spaces containing Red blood cells (RBCs) [Figure 2b]. On immunohistochemistry (IHC), CD31, Vimentin were positive with 90% Ki67. CD34 was negative [Figure 3a-d]. With these findings, we diagnosed epithelioid angiosarcoma. The Positron Emission Tomography - Computed Tomography (PET-CT) scan showed metastasis to deeper lymph nodes. The patient was further managed at oncosurgery.

STS is a rare, insidiously-developing angiosarcoma, classically seen in chronic lymphedema following post-mastectomy. Faulty lymphatic drainage causes defective local immunity and antigen presentation, building an immunologically privileged site as a fertile ground for

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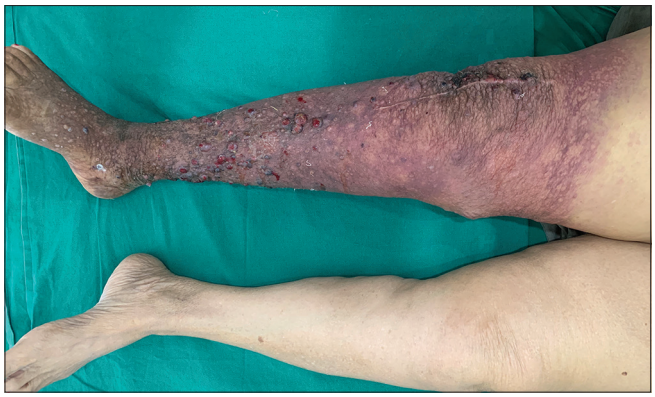

Figure 1: Patient presenting with gross edema and erythema of the right lower limb. Multiple nodules present around the lower part of the leg

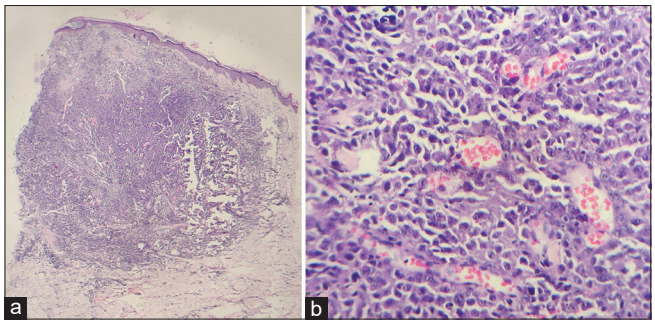

Figure 2: (a) Microphotograph shows thinned out epidermis beneath which the dermis shows neoplastic cells arranged in papillaroid, sinusoidal, and diffuse pattern (H and E stain, 10X). (b) The microphotograph shows neoplastic cells projecting into numerous vascular spaces containing RBCs. The cells are round to polygonal to spindeloid with conspicuous nucleoli, moderate pleomorphism with increased mitosis, and moderate to abundant cytoplasm (H and E stain, 40x)

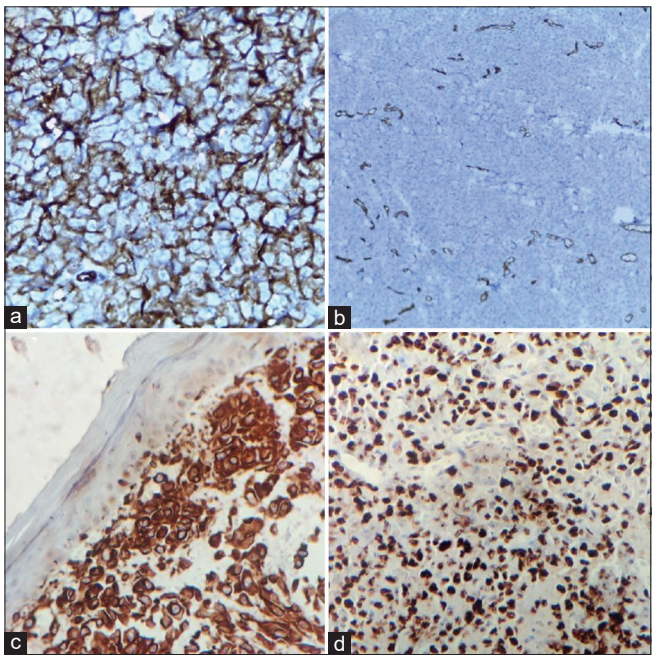

Figure 3: IHC at 40X. (a) CD31-stained positive. (b) CD34-stained negative. (c) Vimentin is strongly and diffusely positive signifying an epithelioid angiosarcoma. (d) High levels of proliferation are seen with Ki67 levels of 90%

oncogenesis.[2] In arthroplasty, the graft material may be oncogenic stimuli. Local adverse reactions to metal debris could prompt granulomatous inflammation. Commonly, the median development time post-arthroplasty is 3.5–52 years.[3] A single reported case with titanium graft was 30 years after hip arthroplasty. The short period of 15 months in our patient may be explained by the preexisting chronic lymphedema in the operated leg.

Histopathology of epithelioid angiosarcoma shows round to polygonal epithelioid cells with prominent nucleoli and infiltrative sheets of malignant cells in the dermis and subcutaneous tissue with focal anastomosing vessels, numerous mitoses with hemorrhage and necrosis.[2]

IHC markers include endothelial markers like Factor VIII, CD31, CD34, and Vascular endothelial growth factor (VEGF). The most common marker, CD31, is the gold standard. Vimentin, a marker of mesenchymal cell origin, is strongly positive in the epithelioid variant. Ki67 scores mitotic activity, and a higher percentage is seen in the epithelioid variant due to robust proliferation. CD34 positivity ranges from 40 to 100%.[4] Despite treatment, there is rapid metastasis, recurrence, and limited response.

STS in the lower limb, without any preceding malignancy or history of radiation, is rare.[5] Our case is unique because STS developed in the leg after arthroplasty in an unusually short period of 15 months.

Declaration of patient consent

The authors certify that they have obtained appropriate patient consent. The patient has given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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