

Short Communication

Pseudo-Kaposi sarcoma and extensive skin ulceration in a patient of Klippel-Trenaunay syndrome

Sir, pseudo-Kaposi sarcoma is a rare self-limiting disorder that can be congenital or acquired. The patients develop brown macules, violaceous or purplish nodules and plaques which become verrucous or ulcerate. Pseudo-Kaposi sarcoma has been reported in patients who had chronic venous insufficiency, paralyzed extremities, amputation stump, procedures of arteriovenous vascular fistula for hemodialysis and suction-socket lower limb prosthesis. There are also reports claiming minor traumas as a cause of pseudo-Kaposi sarcoma.¹ Intractable cutaneous ulcers may complicate Klippel-Trenaunay syndrome.² We report a case of Klippel-Trenaunay syndrome with extensive ulcerations developing in the port-wine stain and a solitary pseudo-Kaposi sarcoma.

A 48-year-old male, farmer by occupation, presented to us with history of reddish patches over the trunk and limbs, and thickening and lengthening of the left upper and lower limbs since early childhood, and multiple, painful ulcers over left foot and left hand since the past one year. He also complained of an asymptomatic nodule over the left popliteal region since the past six months.

Patient complained that his left upper and left lower limbs were enlarged since early childhood with diffuse red patches over the body. As he grew older, the red patches over the other parts of the body subsided but remained over the

limbs, more on the left side than the right side, face and tongue. He complained of multiple ulceration on and off over the left upper and lower limbs, which healed on itself in 3-4 weeks. However since the past one year, he developed multiple, painful ulcers in the existing red patches over the left upper and lower limbs which did not heal.

Over the past six months, he developed an asymptomatic nodule over the left popliteal region. It was small in size to start with, gradually increased in size and the summit of the nodule ulcerated. Patient complained of difficulty in walking due to increased length of left lower limb. There was no history of bleeding from the ulcers or the nodule. There was no history of breathlessness, palpitations, seizures, mental retardation, hematuria, melena, diminished vision or hearing difficulties. Patient was born of second degree consanguineous marriage. There was no family history of similar complaints.

General physical examination and systemic examinations were normal. There were no varicosities. There was extensive port wine stain over the limbs, face and tongue. There were multiple, well-defined ulcers, measuring 0.5cmX1cm to 2cmX3cm, covered with yellowish slough, distributed in the port wine stain over the dorsum of the left foot and toes and dorsum of both hands (**Figures 1 and 2**). There was foul smelling, thick, yellowish discharge from the ulcers (**Figure 3**). Left upper and lower limbs were hypertrophic and deformed.

A well-defined nodule measuring 5cmX3cm was present over the left popliteal fossa. The surface of the nodule was ulcerated showing



Figure 1 Extensive port wine stain and ulcerations over both the upper limbs



Figure 2 Extensive port wine stain and ulcerations over the left upper limb. The ulcer shows thick, purulent discharge.



Figure 3 Extensive ulcerations over the dorsum of left foot.



Figure 4 Solitary nodule in the left popliteal fossa.

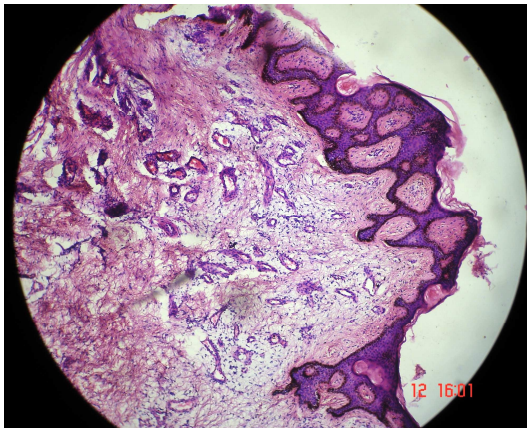


Figure 5 Histopathology of the nodule showing dilated capillaries with extravasation of erythrocytes and surrounding hyperplastic granulation tissue. Low power view. H & E stain.

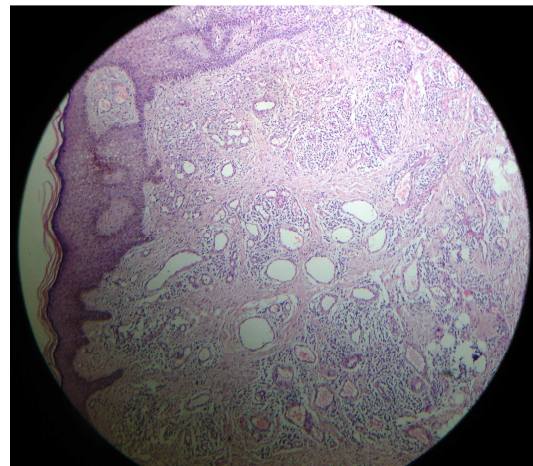


Figure 7 Histopathology of the nodule showing dilated capillaries with extravasation of erythrocytes and surrounding hyperplastic granulation tissue. High power view hematoxylin and eosin stain.

thick, yellowish, purulent discharge and slough (**Figure 4**). There was mild tenderness in the nodule. There was no palpable thrill or audible bruit. On investigation, routine hematological and biochemical parameters including renal function tests and liver function tests were normal. Pus from the ulcers was sent for culture

and sensitivity which revealed growth of *Staphylococcus aureus* sensitive to ceftriaxone. Echocardiography, ultrasound abdomen, ECG,

chest X-ray, CT-scan of the chest, abdomen and the brain were normal. Doppler studies of the limb veins revealed normal deep and superficial venous system. Arterial system was normal in all the limbs. X-ray of the left limbs revealed bony hypertrophy and deformities. The nodule over the left popliteal fossa was biopsied and subjected to histopathological examination. It revealed dilated capillaries with extravasation of erythrocytes and surrounding hyperplastic granulation tissue. There were no vascular slits (figures 5, 6 and 7). Since there was no palpable thrill or audible bruit, we did not consider the diagnosis of Stewart-Bluefarb syndrome. Based on the history, clinical data and investigation findings, we arrived at the diagnosis of Klippel-Trenaunay syndrome with extensive port wine stain and ulcerations and a solitary pseudo-Kaposi sarcoma. Patient was treated with Inj Ceftriaxone 1 gm intravenously twice daily. In about two weeks, the ulcers healed almost completely. The nodule over the left popliteal fossa was excised.

Acroangiokeratosis (pseudo-Kaposi sarcoma) is a dermatological condition characterized by purple-colored nodules, plaques or patches, mostly on the extensor surfaces of lower extremities, usually in patients with chronic venous insufficiency and arteriovenous malformations of the legs, but also in hemodialysis patients with iatrogenic arteriovenous shunts, paralyzed limbs and amputation stumps. Acroangiokeratosis in patients with chronic venous insufficiency manifests usually as bilateral skin lesions located on the dorsa of the feet, hallux and second toe, or on the medial aspect of lower legs. Acroangiokeratosis may look like Kaposi sarcoma, but in contrast to Kaposi sarcoma, acroangiokeratosis is characterized by lack of

spindle cells and silt-like vessels on histopathologic analysis.³

There are many reports of pseudo-Kaposi's sarcoma in Klippel-Trenaunay syndrome.^{4,5,6} Similarly, there is a report of intractable cutaneous ulcerations in Klippel-Trenaunay syndrome.² Association of both pseudo-Kaposi's sarcoma and extensive cutaneous ulcerations in a patient of Klippel-Trenaunay syndrome must be very rare and to the best of our knowledge, this is the first case report.

References

1. Doğan S, Boztepe G, Karaduman A. Pseudo-Kaposi sarcoma: A challenging vascular phenomenon. *Dermatol Online J*. 2007;13:22.
2. Satoshi H, Ryuhei O, Masatoshi D, Setsuya A. Skin ulcer associated with Klippel-Trenaunay syndrome. *Rinsho Derma* 2006;48:815-7.
3. Lugović L, Pusić J, Situm M *et al*. Acroangiokeratosis (pseudo-Kaposi sarcoma): three case reports. *Acta Dermatovenerol Croat*. 2007;15:152-7.
4. Irene NT, Hema OH, Rajagopalan V, Parveen B. Klippel-Trenaunay syndrome associated with pseudo-Kaposi's sarcoma. *Indian J Dermatol Venereol Leprol*. 2002;68:305-6.
5. Kofoed ML, Klemp P, Thestrup-Pedersen K. The Klippel-Trenaunay syndrome with acroangiokeratosis (pseudo-Kaposi's sarcoma). *Acta Derm Venereol*. 1985;65:75-7.
6. Del-Río E, Aguilar A, Ambrojo P *et al*. Pseudo-Kaposi sarcoma induced by minor trauma in a patient with Klippel-Trenaunay-Weber syndrome. *Clin Exp Dermatol*. 1993;18:151-3.

Pradeep Vittal Bhagwat*, **Chandramohan Kudligi***, **Shashikumar BM****, **Arun Thirunavukkarasu***, **Vijaya Veeranna Sajjan***

*Dept of Skin & STD, Karnataka Institute of Medical Sciences, Hubli, Karnataka

**Department of Skin & STD, Mandya Institute of Medical Sciences, Mandya, Karnataka