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Case Report

Overlap of histological features of pigmented purpuric dermatosis and kaposi sarcoma – pseudo kaposi sarcoma

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ABSTRACT

Pseudokaposi sarcoma can be congenital or acquired. Due to prolonged venous stasis, leads to peripheral tissue hypoxemia, which results in neovascularisation and fibroblast proliferation. Treatment includes rectification of the underlying pathology such as graded compression stockings or compression pumps for venous stasis and ulcer management with regular wound care. Medical management includes oral antibiotics, along with compression therapy with good results. Our patient presented mild lower limb pitting edema unilaterally with erythematous barely elevated plaque over the anterior aspect of the lower limb.

1. Introduction

Pseudo-Kaposi sarcoma is an extremely rare usually self-limiting disease. Can be congenital or acquired. Although etiology remains unknown, due to prolong venous stasis along with insufficiency of the calf muscle to pump the capillaries leads to chronic edema resulting in chronic tissue hypoxemia and induces new vessels and fibroblast proliferation.

2. Case

A 14-year-old boy presented with right lower limb multiple erythematous to slightly vascular plaque since 6 months. He complains of mild to moderate swelling in the right lower limb on prolong standing subsides after taking rest. On cutaneous examination, lateral aspect of the lower half of right leg shows discrete erythematous to slightly purpuric ill-defined barely elevated plaques [Figure 1] measuring from smallest of 1x1 cm and largest being 3x6 cm, of which a plaque shows white to yellow crusting slight verrucous surface [Figure 2]. No local rise of temperature, nor tenderness, thrill, compressibility. No ulceration, no palpable pulsation. No hypertrophy of limbs, or spinal deformity. While examining we found soft compressible skin-colored swelling suggestive of incompetent perforators on prolonged standing. Brodie- Trendelenburg test for SFJ was normal, for perforators were positive. A differential diagnosis of pseudo-Kaposi sarcoma, verrucous hemangioma, angiokeratoma circumscriptum. Advised for Venous doppler of right lower limb which shows competent saphanofenoral, saphenopopliteal perforators, prominent GSV, multiple focal incompetent perforators, and varicosities with hard nodular subcutaneous fat suggestive of venous malformation secondary to incompetent perforators. Dermoscopy shows radial vascular streaks in pink to erythematous background which blanches on pressure [Figure 3]. Biopsy from two sites taken which shows hyperkeratosis with focal acanthosis [Figure 4], superficial and mid dermis shows multiple small dilated capillaries lined with plump endothelial cells [Figure 5],

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perivascular lymphocytic cells, and extravasated RBCs suggestive of Acroangiodermatitis of mali (PseudoKaposi sarcoma).

We prescribed topical timolol, calcium dobesilate, limb elevation, and compression therapy. The lesion showed a slight decrease in size and vascularity after 2-3 weeks of follow-up [Figure 6].

Table 1: show differentiation features of pseudo kaposi and kaposi sarcoma

	Pseudo Kaposi sarcoma	**Kaposi sarcoma**
Promontory sign	Absent	Present
Vessels	Tortuous venules	Prominent jagged vessels
Arrangement of vessels	Angiomatous capillaries separated by edematous matrix	Back to back appearance of angiomatous capillaries
Atypia	No to minimal	Atypia of endothelial cells
Hyaline globules	No	Small hyaline globules seen
Perivascular infiltrate	No plasma cells	Plasma cells
Vascular proliferation	Small dilated vessels lined by plump endothelium with hyperplasia of existing vessels	Slit like spaces and spindle cell proliferation, vascular proliferation independent of pre existing vessels
PAS staining of vessels	Present	Absent
Factor VIII associated antigen	Present	Absent
CD34 Positivity	Present in endothelial cells	Present in endothelial cells and spindle cells
Dermal hemosiderin and RBC extravasation	Present	Present
HHV 8	Negative	Positive

3. Discussion

Pseudo-Kaposi sarcoma is an extremely rare usually self-limiting disease. Earhart et al. used the term *pseudo-Kaposi sarcoma* in 1974 for the first time,[1] later the same clinical picture was introduced by Mali in 1965 as kaposiform skin lesion, in chronic vascular insufficiency patients. Causes can be congenital or acquired (as in our case). Presents as brown to hyperpigmented macules, purplish plaque, violaceous nodules which progresses to become verrucous later ulcerates. Pseudo-Kaposi sarcoma seen in chronic venous incompetency,[2] paresis of extremities,[3] amputation stump,[4]

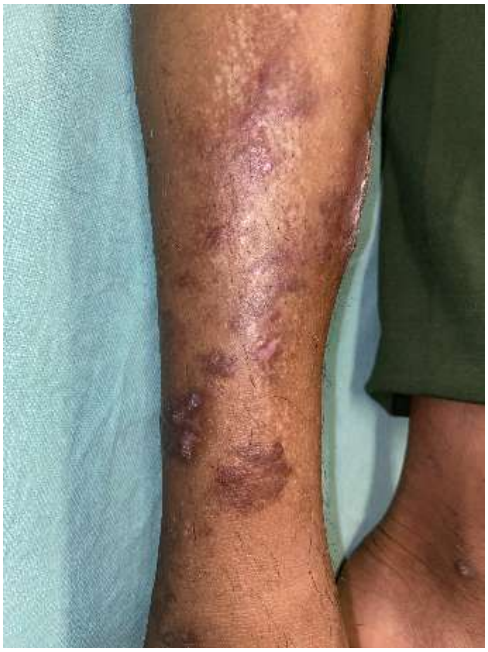
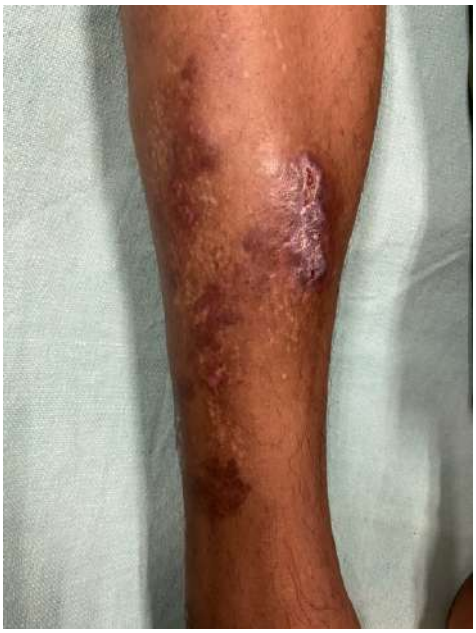


Figure 1:



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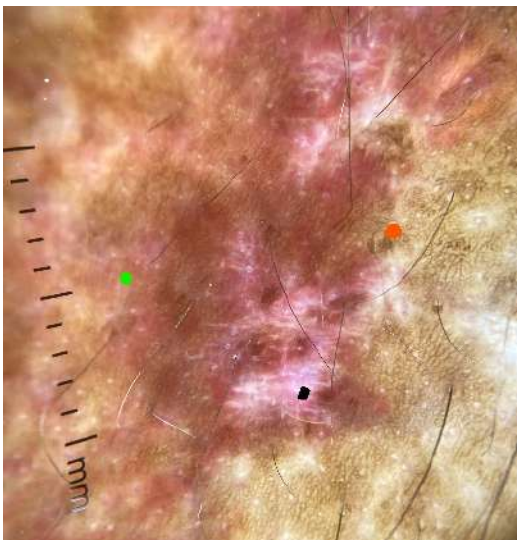

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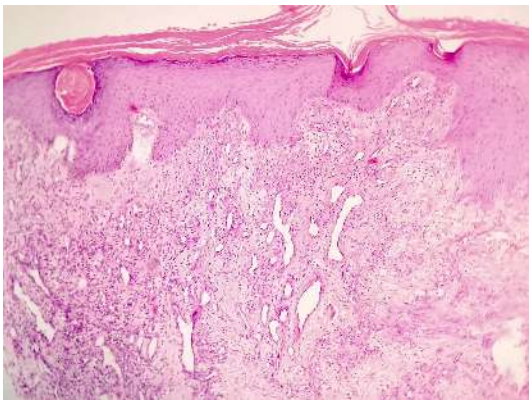

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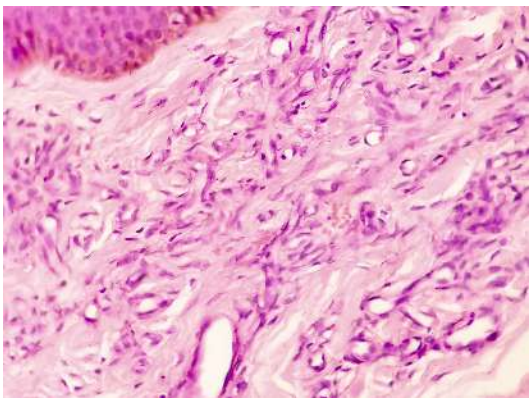

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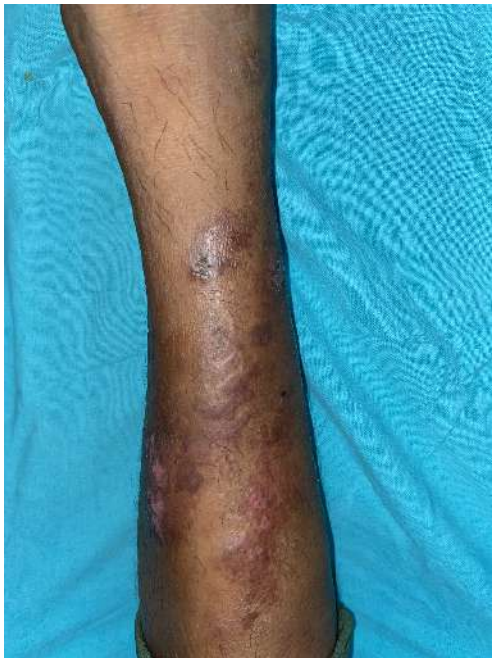

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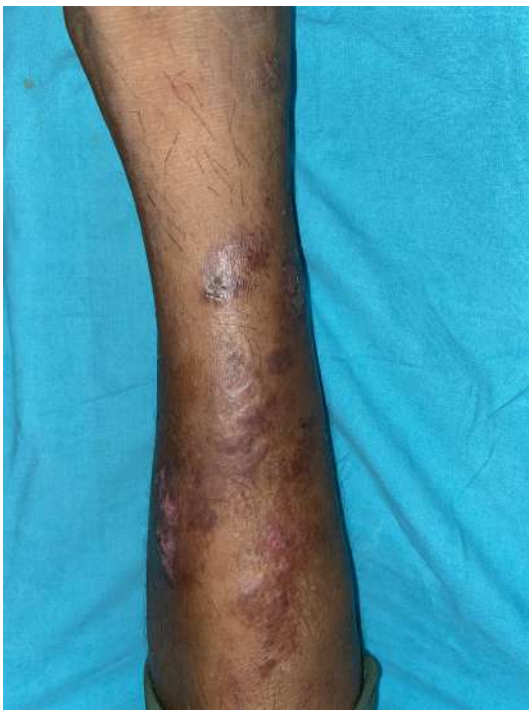

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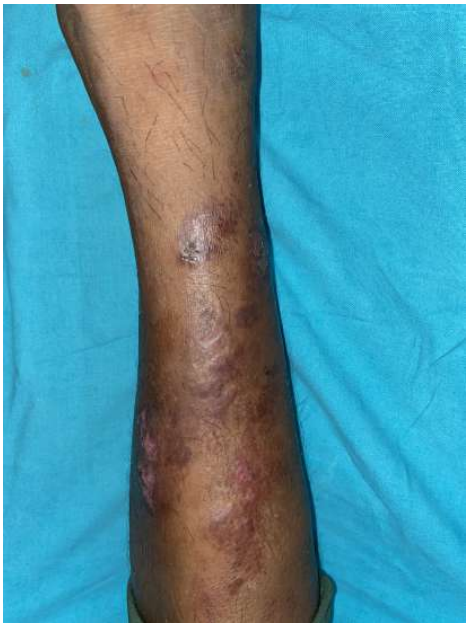

Figure 8:

procedures such as arteriovenous fistula for hemodialysis,[5] and suction-socket lower limb prosthesis,[6] following minor trauma.[7] Differentiating features of pseudo Kaposi sarcoma and Kaposi sarcoma is summarized in Table 1.

Various variants of acroangiodermatitis are:

1. Stewart-Bluefarb syndrome: Due to congenital arteriovenous malformation along with multiple arteriovenous fistula in lower limb. Starts in younger age as unilateral painful violaceous macules and papules which ulcerates in lower limb. On palpation increased warmth and thrill, with varicose veins.[8]
2. Mali type is exaggerated stasis or gravitational dermatitis, seen in old patients, usually bilateral with chronic venous incompetency over dorsum of feet and first two toes and over medial aspect of lower legs. Begins as purplish macules and patches that gradually evolves into soft, nontender, red to violaceous papules and indurated plaques and nodules.[8]
3. Acroangiodermatitis in first pregnancy starts as gravity purpura (Dermite ocre of Favre) over lower limbs at sites of varicosities from dorsa of feet and toes.[9]
4. Angiodermatitis begins in patients with chronic renal failure after arteriovenous shunt for hemodialysis[9]

Although etiology remains unknown, due to prolong venous stasis along with insufficiency of the calf muscle to pump the capillaries leading to chronic edema resulting in chronic tissue hypoxemia induces new vessel and fibroblast proliferation.[10]

Management of acroangiodermatitis is correction of underlying pathology, such as chronic venous insufficiency which includes, graded compression stockings, compression pump for venous stasis and local antibiotic treatment and regular wound care in case of venous ulcers. For gravitational eczema topical corticosteroids is included.[9] Oral erythromycin 500mg QID and oral Dapsone 50mg BD for 3months showed best results.[11] Pulsed dye laser can be considered for therapy. In arteriovenous malformation embolization of small fistulae, surgical elimination of shunts can be considered.[12]

4. Conclusion

Our patient has Mali type of variant with atypical presentation with younger age of onset, presenting unilateral limb on both medial and lateral aspect as lichenified keratotic palque which usually get under diagnosed.

5. Source of Funding

None.

6. Conflict of Interest

None.

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