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Short Communication Article

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An Atypical Presentation of Epitheloid Hemangioma Mimicking Lupus Pernio Sarcoidosis

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Dear editor,

Epithelioid hemangioma, also known as angiolymphoid hyperplasia with eosinophilia, histiocytoid hemangioma, inflammatory angiomatous nodule, atypical granuloma and pseudopyogenic granuloma [1] is a rare clinicopathological entity.It is a benign vascular tumor composed of epitheloid endothelial-cell-lined channels. We report an unusual case of epitheloid hemangioma of the nose with morphological appearance and dermoscopic features of lupus pernio.

A 53-year-old women presented with a swelling of the nose evolving for 2 years.There was no history of pain, bleeding, or ulceration. Dermatological examination revealed a solitary smooth erythematoviolaceous non-pulsatile nodule of the left ala of the nose (figure 1). There was no mucosal involvement or regional lymphadenopathy. Dermoscopy revealed translucent orange globular structures and multiple linear and branching vessels (figure 2a).Clinical and dermoscopic features were suggestive of lupus pernio diagnosis. However, histopathological examination revealed a vascular proliferation with circumscribed lobules and blood vessels lined by epitheliod endothelial cells with perivascular lymphocytes and eosinophils (figure 2b).The histopathological features were compatible with the diagnosis of epitheloid hemangioma. Hemogram did not show eosinophilia. Thoracic abdominal and pelvic computed tomography and bone imaging didn't show any extradermatological localization. Treatment with intralesional steroids was proposed.

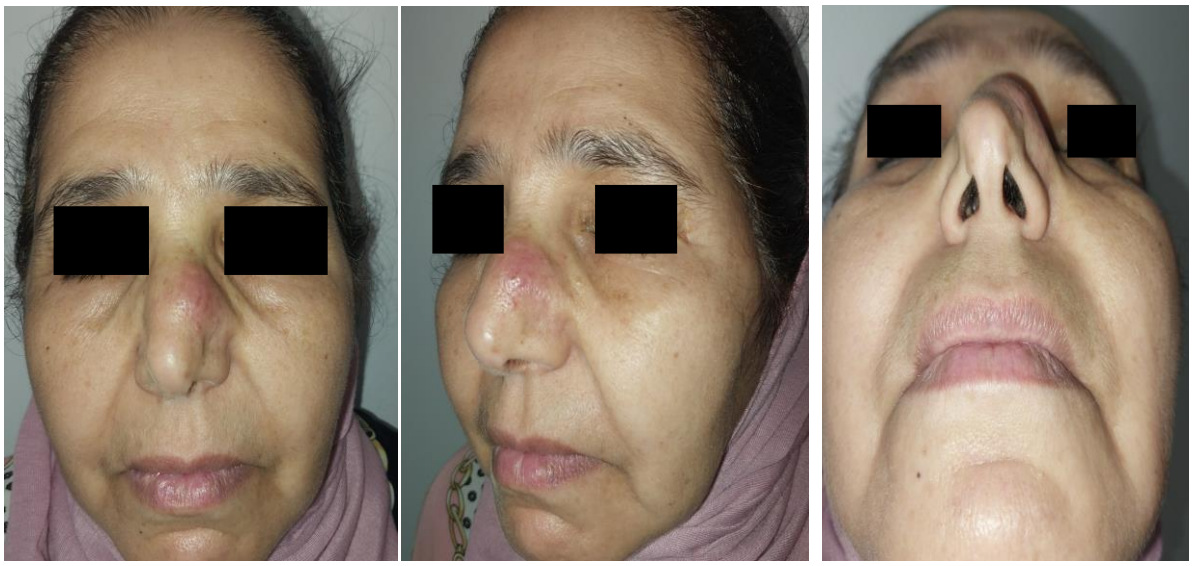

Figure 1: Solitary erythematoviolaceous nodule of the left ala of the nose

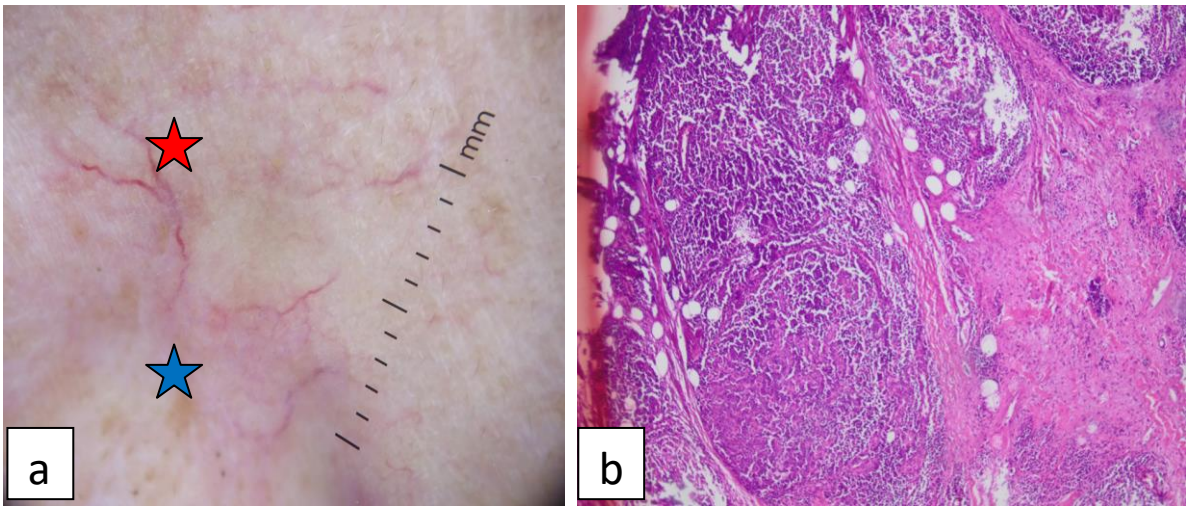

Figure 2:
a: Dermoscopic features: Linear and branching vessels
Translucent orange globular structure
b: Vascular proliferation with circumscribed lobules and vessels lined by epitheliod endothelial cells

Epitheloid hemangioma (EH) is a rare, non-malignant affection characterized by an epithelioid endothelial cell. EH belongs to the heterogeneous group of epithelioid vascular tumors. It is usually occurring in females between the second and fifth decade of life .It generally appears in the dermal and subcutaneous tissues of the head and neck [2].Nasal localization is unusual since only few cases of epithelioid hemangioma localized to the nose have been described in the literature. EH clinically appears as a solitary or multiple erythematous to violaceous papules or plaques[3].Many extradermaological localizations of EH are reported,mainly bones and lymph nodes .Other sites are much rarer: the lungs, the penis ,eyes (internal canthus),tongue, breasts, arteries, colon, heart, spleen and testicles [4]..The main differential diagnosis are Kimura disease, IgG4-related skin disease,bacillary angiomatosis, cutaneous epithelioid angiomatous nodule, pyogenic granuloma , Kaposi sarcoma and malignant epithelial vascular tumors such as epithelioid sarcoma-like hemangioendothelioma,epithelioid hemangioendothelioma and epithelioid angiosarcoma [1]. In our case, the clinical presentation was confusing mimicking a lupus pernio especially in the presence of typical dermoscopic features of sarcoidosis. Pathologic features of EH typically includes wellcircumscribed nodules with many vessels lined by characteristic plump epithelioid endothelial cells, intracytoplasmic vacuoles and perivascular inflammatory infiltrate of lymphocytes and eosinophils [3]. Surgical excision, especially micrographically controlled excision (Mohs surgery) is currently considered the standard therapy and has high recurrence rates of 33-50 %.The other therapeutic options include ablative,systemic,intralesional and topical treatments including cryosurgery,intralesional corticosteroids, lasers, photodynamic therapy,propanolol and isotretinoin due to its antiangiogenic effect [5].

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