

Paediatric Cutaneous Langerhans cell histiocytosis mimicking hemangioma: A deceitful entity

Dear Editor,

Langerhans cell histiocytosis (LCH) is a rare neoplastic disorder, characterized by proliferation of Langerhans cells. In children, these diseases cause a myriad of confounding manifestations causing major diagnostic difficulties.^[1] We discuss a unique case of pediatric LCH masquerading as hemangioma of the nasion.

An 11-month-old male child presented with swelling at the root of the nose for 3 months which was insidious in onset, progressive in nature with on and off bleeding from the site with no history of trauma. There were no comorbidities or history of bleeding diathesis or any drug intake. On local examination, a firm, discolored swelling fixed to the overlying skin measuring 1 cm × 1 cm was palpated at the root of the nose. The blood counts were within the normal limits. Ultrasonography of the soft-tissue nodule over nasion revealed a 10 mm × 8 mm hypoechoic, well-defined, oval, noncompressible soft-tissue nodule. The deeper half of the nodule was showing two arterial feeders and few venous channels from underlying nasal arterial branch [Figure 1a]. The features were suggestive of hemangioma. Circular incision was made, and the mass was excised. Grossly, a small grayish white to grayish brown tissue measuring 1.2 cm × 1 cm × 0.4 cm with warty surface was seen. Cut section was grayish white, solid, and homogeneous. Microscopically, sections showed thinned out epidermis and a lesion extending up to subcutis and entrapping the appendages [Figure 1b]. Cells were spindle and arranged in storiform and fascicular pattern with focal vascular proliferations. Dense inflammatory cell infiltrations comprising predominantly of eosinophils, histiocytes, many lymphocytes, and plasma cells were also seen [Figure 1c]. The features were suggestive of LCH. Immunohistochemistry showed CD-68 and S100

show strong positive in tumor cells confirming the same [Figure 1d].

LCH is a rare disease seen in 1–4 years.^[2] Although the exact pathogenesis is not known, there is clonal proliferation of cells expressing CD1a/CD207. They constitute a sea of reactive inflammatory cells, including numerous eosinophils, lymphocytes, macrophages, natural killer cells, multinucleated giant cells, and clonal dendritic cells having a cytokine cross-talk.^[2] Histology and immunohistochemistry are essential for the diagnosis as clinically they can mimic diverse entities including vascular lesions. The latter generally present in the neonatal period, unlike this case where it has presented in infancy.^[3] Exclusive cutaneous lesions may regress spontaneously.^[3] Cutaneous mimics in this age include viral infections, pustular folliculitis, hemangiomatosis, hamartomas, juvenile xanthogranulomas, and leukemia cutis. Cutaneous manifestations constitute 30%–60% cases with pruritus, petechiae, nodules, or bullae.^[4] Histologically, the skin shows intense infiltration by histiocytic cells having folded coffee-bean or reniform-shaped nuclei and numerous eosinophils serving as clues to diagnosis. Despite clonality, these tumors rarely show mitotic figures. The mimics are readily ruled out on histomorphology along with immunohistochemical expression of CD1a and S-100.^[5]

Treatment of these patients requires customization as per their clinical presentation and severity which may be watchful observation in mild cutaneous cases or high-end systemic therapy in refractory or aggressive cases.^[2] These patients may show complete recovery or progression to multi-system disease. They need timely and accurate diagnosis where histopathology and immunohistochemistry plays a very important role as highlighted in this case.

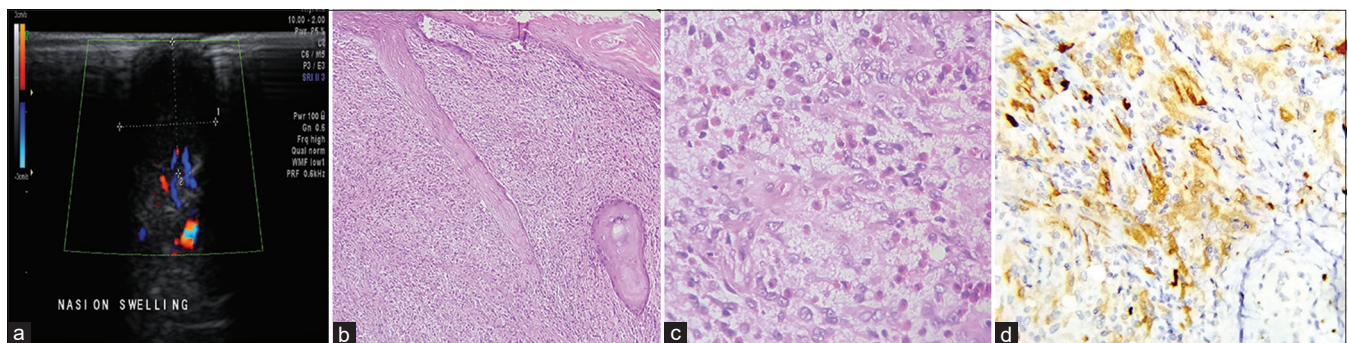


Figure 1: (a) Doppler ultrasonography showing a 10 mm × 8 mm size hypoechoic well-defined oval soft tissue with two arterial feeders and few venous channels from underlying feeder arterial branch (b and c) Section from the skin shows flattened out epidermis with sheets of inflammatory cell infiltration in dermis entrapping adnexal structures which on higher power show features histiocytic cells with coffee-bean nuclei and admixed with eosinophils and few lymphocytes (H and E [b ×40], [c ×400]) (d) Immunohistochemistry with S100 showing diffuse strong positivity (×400)

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Conflicts of interest

There are no conflicts of interest.

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