Diffuse Skin Erosions as an Atypical Manifestation of Congenital Self-Healing Reticulohistiocytosis

Dear Editor,

Congenital self-healing reticulohistiocytosis (CSHRH), also known as Hashimoto–Pritzker disease, is a rare benign variant of Langerhans cell histiocytosis (LCH). It is characterized by skin lesions in healthy infants, which then spontaneously regress, with a Langerhans cell infiltrate upon histopathological analysis.[1] A full-term, low birth weight (2 kg) male child with generalised erosions over the body, was born to a 27-year-old primipara mother. On cutaneous examination, multiple erosions ranging from 0.5 cm to 2 cm in size were distributed over the scalp, face, trunk, and limbs along with multiple purpuric lesions and blisters on the palm and soles [Figure 1a and b]. The mucosae, nails and hair were uninvolved. The child was afebrile with stable vital signs, and no organomegaly or lymphadenopathy. The routine blood investigations including haemogram, serum biochemistry, blood culture, urine analysis, and serology for hepatitis B virus (HBV), hepatitis C virus (HCV) and human immunodeficiency virus (HIV) did not reveal any abnormality. Serology for veneral disease research laboratory (VDRL) and toxoplasma, rubella, cytomegalovirus and herpes infection (TORCH) were negative for the mother and child. Tzanck smear from the erosions did not show any multinucleated giant cells or acantholytic cells. Systemic evaluation, including chest radiography, ultrasound of the abdomen, and radiological skeletal survey did not reveal any abnormality.

After 3 weeks, multiple greasy yellow crusted papules and vesicles were noticed over the scalp and face with scaly erythematous papules over the trunk and limbs, with a few lesions resolving with hypopigmented scars [Figure 2]. A biopsy was performed on a characteristic lesion. On histopathology, a dense lichenoid infiltrate of histiocytes with abundant amphophilic cytoplasm and a large bilobed nucleus was seen, suggestive of LCH. To confirm the diagnosis, immunohistochemistry (IHC) staining with CD1a was done and it was found to be positive [Figure 3a and b].

Based on the clinical characteristics, histopathology, and IHC, a diagnosis of CSHRH or Hashimoto–Pritzker disease, a variant of LCH was made. The child was followed up every 2 weeks, and at 3 months, most of the lesions had resolved, leaving behind atrophic hypopigmented scars.

LCH is a rare clonal disease of the monocyte-macrophage system, characterized by uncontrolled proliferation and accumulation of CD1a/CD207 dendritic cells. CSHRH was first described by Hashimoto and Pritzker in 1973, who reported a case of congenital LCH with red-brown nodules with spontaneous resolution within 5 weeks leaving residual scars.[1]

As per the newer classification developed by the histiocyte society in 2016, LCH can be divided into LCH single system, LCH lung, LCH MS-RO$^+$, LCH MS-RO$^-$ and LCH associated with another myeloproliferative/myelodysplastic disorder.[2] CSHRH, a congenital form of LCH with only skin involvement and no systemic involvement, is a single-system LCH.

CSHRH typically presents as multiple, painless, erythematous to brown papulonodular lesions on the skin that may also develop vesicles and crusting. In our case, the neonate presented with erosions, which are very rarely reported in the literature.[3,4]

Erosions in neonates can be a primary manifestation of several diseases, including impetigo, staphylococcal scalded skin syndrome, autoimmune blistering disorders, epidermolysis bullosa, neonatal herpes, and congenital syphilis. Early recognition and evaluation are critical to differentiate benign transient causes from serious systemic diseases.

CSHRH is regarded as a benign, self-limiting disorder with a good prognosis. The characteristics of CSHRH appear to be (1) congenital lesions, (2) an otherwise well infant with no or mild systemic symptoms, (3) histopathologically demonstrating LCH, and (4) spontaneous involution of skin lesions.[5]

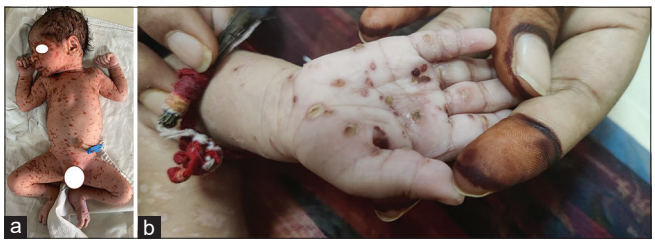

Figure 1: (a) Multiple erosions with peripheral scaling, diffusely distributed involving the scalp, face, trunk, and limbs. (b) Multiple purpuric lesions and blister on the palm

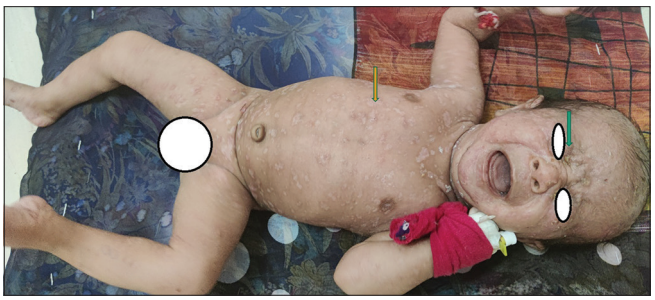

Figure 2: Multiple hypopigmented atrophic and scaly lesions on the trunk (yellow arrow) along with yellow greasy papules on the face and neck (green arrow)

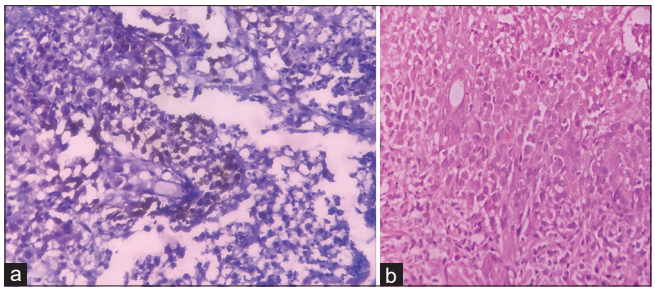

Figure 3: (a) CD1a IHC marker showing membranous staining of Langerhans cells. (b) Langerhans cell with a moderate amount of amphophilic cytoplasm and bilobed nuclei having prominent nuclear grove and fine chromatin. (H&E; 40x). CD1a IHC: Cluster of differentiation 1a Immunohistochemistry

In a review of 125 published cases of CSHRH, a 10% relapse rate was noted. There have been reports of patients developing permanent disabling diseases such as diabetes insipidus or progression to involve the lungs, eyes, or bones.[5]

There is no explanation why a few lesions self-heal while others disseminate, and there is no means to differentiate either by histopathology or morphology between the CSGRH and multisystem LCH. A recently proposed hypothesis of "misguided myeloid differentiation" states that the differentiation of precursor cells in whom mitogen-activated protein kinase (MAPK) mutation occurs defines the clinical extent and severity of the disease. Clinical data support a fetal liver-origin progenitor cell for self-healing reticulohistiocytosis, while stem cell from bone marrow leads to high-risk multisystem LCH.[6] Thus, analysis of MAPK mutation may provide additional information about clinical types and prognosis of the disease. In resource-poor countries like India, mutational analysis is not readily available, and thus a long-term follow-up is required to rule out the possibility of relapse, progression, or systemic involvement.[7]

There is no recommended treatment for CSHRH, but if lesions persist, topical corticosteroids, tacrolimus, or nitrogen mustard can be used.[5] In conclusion, this case illustrates the need to include LCH in the spectrum of congenital erosive lesions.

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Consent statement

The authors certify that they have obtained appropriate consent from the parents to publish photographs of the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Chinmai Yadav, Ranjana Beniwal, Yamini Verma[1]

Departments of Dermatology and [1]Pathology, Dr. Sampurnanand Medical College, Jodhpur, Rajasthan, India

Address for correspondence:
Dr. Ranjana Beniwal,
Sector-C, Shastri Nagar, Jodhpur - 342 005, Rajasthan, India.
E-mail: ranjanabeniwal@gmail.com

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