CASE REPORT OLGU SUNUMU

DOI: 10.5336/dermato.2025-111195

Congenital Self-Healing Reticulohistiocytosis Presenting at Birth: Clinicopathologic Features of a Rare Neonatal Case

Konjenital Kendiliğinden İyileşen Retikülohistiyositoz: Nadir Bir Yenidoğan Olgunun Klinikopatolojik Özellikleri

Esin DİREMSİZOĞLU[a], Uğur DEMİRSOY[b], Hatice Neşe DOĞAN[c], Evren ODYAKMAZ DEMİRSOY[a]

[a]Kocaeli University Faculty of Medicine, Department of Dermatology, Kocaeli, Türkiye
[b]Kocaeli University Faculty of Medicine, Department of Pediatric Oncology, Kocaeli, Türkiye
[c]Kocaeli University Faculty of Medicine, Department of Pathology, Kocaeli, Türkiye

This case report was presented as an oral presentation at 11th National Pediatric Dermatology Days, May 23-26, 2024, Sakarya, Türkiye.

ABSTRACT Congenital self-healing reticulohistiocytosis is a rare cutaneous form of Langerhans cell histiocytosis that typically manifests in neonates and resolves spontaneously without systemic involvement. We present a case of a male infant who developed multiple papulovesicular skin lesions 2 days after birth. Histopathological examination showed CD1a and S100 positivity, confirming Langerhans cell histiocytosis. The lesions regressed without treatment within 1 month, and no recurrence or systemic involvement was observed during 1.5 years of follow-up. This case underlines the importance of considering congenital self-healing reticulohistiocytosis in neonates presenting with spontaneously regressing skin lesions and highlights the necessity of careful long-term monitoring due to the risk of late systemic involvement or relapse. Awareness of this self-limiting condition can prevent unnecessary interventions and their associated risks.

Keywords: Hashimoto-Pritzker disease; histiocytosis, Langerhans-cell; newborn; congenital; remission

ÖZET Konjenital kendiliğinden iyileşen retikülohistiyositoz, tipik olarak yenidoğanlarda ortaya çıkan ve sistemik tutulum olmaksızın spontan olarak gerileyen, Langerhans hücreli histiyositozun nadir görülen kutanöz bir formudur. Bu olgu sunumunda, doğumdan 2 gün sonra çok sayıda papülovezikül deri lezyonu gelişen erkek bir yenidoğan sunulmaktadır. Histopatolojik incelemede CD1a ve S100 pozitifliği saptanmış ve bu bulgular Langerhans hücreli histiyositoz tanısını doğrulamıştır. Lezyonlar tedavi uygulanmaksızın 1 ay içinde gerilemiş, 1,5 yıllık izlem süresince nüks ya da sistemik tutulum gözlenmemiştir. Bu olgu, spontan gerileyen deri lezyonları ile başvuran yenidoğanlarda konjenital kendiliğinden iyileşen retikülohistiyositozun akılda tutulmasının önemini vurgulamakta; ayrıca geç sistemik tutulum veya nüks riski nedeniyle dikkatli uzun dönem izlemin gerekliliğine dikkat çekmektedir. Kendi kendini sınırlayan bu tablonun farkında olunması, gereksiz girişimlerin ve bunlara bağlı risklerin önlenmesine yardımcı olabilir.

Anahtar Kelimeler: Hashimoto-Pritzker hastalığı; histiyositoz, Langerhans-hücreli; yenidoğan; doğumsal; remisyon

Langerhans cell histiocytosis (LCH) is a disease caused by clonal proliferation of Langerhans cells. According to the Histiocyte Society, it is classified into 2 main groups: single system LCH and multisystem LCH. LCH affecting only the skin is frequently seen in newborns and can appear congenital.[1]

Congenital self-healing reticulohistiocytosis (CSHR) is a rare variant of LCH first described by Hashimoto and Pritzker in 1973. Characteristics include papulovesicular lesions after birth, absence of systemic involvement, CD1a and S100 positive cells on histopathology, and spontaneous resolution within a

TO CITE THIS ARTICLE:
Diremsizoğlu E, Demirsoy U, Doğan HN, Odyakmaz Demirsoy E. Congenital self-healing reticulohistiocytosis presenting at birth: Clinicopathologic features of a rare neonatal case. Turkiye Klinikleri J Dermatol. 2026;36(1):43-6.

Correspondence: Esin DİREMSİZOĞLU
Kocaeli University Faculty of Medicine, Department of Dermatology, Kocaeli, Türkiye
E-mail: mdesinarslan@gmail.com

Peer review under responsibility of Turkiye Klinikleri Journal of Dermatology.

Received: 24 Apr 2025 ***Received in revised form:*** 09 Oct 2025 ***Accepted:*** 06 Nov 2025 ***Available online:*** 25 Nov 2025

2146-9016 / Copyright © 2026 by Türkiye Klinikleri. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

few months.[2] In this report, we present a case of CSHR lesions at birth to contribute to the understanding of this rare subtype of histiocytosis.

CASE REPORT

A 3,000 g male infant born at 38th week by C/S was admitted with the complaint of multiple lesions on his body on the 2nd postnatal day. There were multiple crusty red-brown papules and vesicles on his both palm and yellow-brown papules on his cheek, along extremities and trunk (Figure 1). There was no accompanying health problem in the neonatal period and no family history of LCH. Physical examination revealed no sign of organomegaly. Biopsy was taken from the crusted papule in the anterior aspect of the trunk with pre-diagnosis of LCH, mastocytosis, benign cephalic histiocytosis. Histopathological examination showed cells filling the papillary dermis, forming groups and layers of variable size, some of which were located intraepidermal, with large, irregular contours, eosinophilic cytoplasm, coarse chromatin, occasionally bilobulated and containing nuclei with few intranuclear clefts. Immunohistochemical examination showed diffuse strong cytoplasmic staining with CD1a and langerin, focal cytoplasmic staining with S100, negative reaction with CD68, tryptase and toluidine blue (Figure 2). Histopathological examination was compatible with LCH. Rou-



FIGURE 1: Widespread crusted papules and vesicles on the face and palmar region

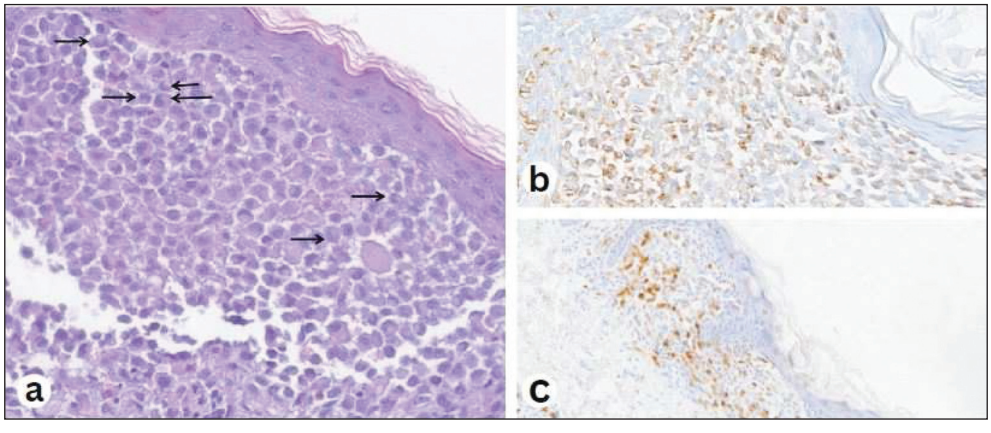


FIGURE 2: a) Tumor cells exhibiting broad eosinophilic cytoplasm, coarse chromatin, occasional bilobulation, and intranuclear clefts (black arrows) (Hematoxylin&eosin, x20); **b)** Langerin immunohistochemical staining showing cytoplasmic staining in tumor cells (Langerin, x20); **c)** Positive staining for S100 immunohistochemistry in tumor cells (S100, x20)

tine blood count, peripheral smear, bone radiographs and abdominal ultrasonography results were normal. During weekly follow-ups, the lesions gradually regressed over the course of 1 month, leaving multiple millimeter-sized hypopigmented macules on the trunk. Based on these evaluations, the case was diagnosed as CSHR. Since the lesions regressed spontaneously, no treatment was required. No recurrence or comorbidity has been observed during the follow-up period up to the age of 1.5 years, and follow-up is still ongoing. Written informed consent for publication of this case and the accompanying images was obtained from the patient's parents.

DISCUSSION

CSHR is a rare subtype of LCH and usually presents with prominent skin lesions in the neonatal period. On histopathologic examination, CD1a and S100 positivity on immunohistochemistry are compatible with the diagnosis of LCH.[2]

In our case, spontaneous resolution of the patient's complaints within one month, no recurrence until 1.5 years of age and isolated cutaneous involvement were important clinical findings. This emphasizes that CSHR usually resolves spontaneously and the prognosis is generally favorable. Since the underlying cause of histiocytosis cannot be determined and the disease regresses spontaneously, the actual incidence may be higher than the reported cases.[2] In clinical findings, diffuse red brown papulovesicular lesions may be seen throughout the body, while cases with solitary lesions and blueberry muffin syndrome like lesions have been reported.[3]

Differential diagnosis of hemorrhagic papulovesicles or purple papules are neoplastic diseases such as LCH, leukemia cutis, neuroblastoma and congenital infections as TORCH infections. Whereas differential for erythematous papulovesicles are neonatal pustular melanosis, erythema toxicum neonatorum, incontinentia pigment, juvenile xanthogranuloma and generalized eruptive histiocytoma.[4]

The absence of systemic involvement allows follow-up without adversely affecting the patient's quality of life. Although there are no definite criteria for the differential diagnosis between CSHR and disseminated LCH, careful clinical observation remains the cornerstone of management.[3]

Case reports of reactivation of early LCH after regression have been reported in the literature. Larralde et al. documented a case in which cutaneous lesions regressed by the 3rd month but recurred with systemic involvement in the 8th month, leading to death before the age of 1 year.[5] Since the most common relapse rate in LCH is reported in the first year of life, close follow-up is required in the first year of life in LCH and CSHR.[6] Complete blood count, liver function tests, evaluation of coagulation parameters, and bone radiographs should be performed to examine systemic involvement. After systemic involvement is excluded, lesions regress spontaneously in most cases; however, topical corticosteroids or tacrolimus may be administered for persistent lesions.[7]

This case highlights clinical approaches to diagnosing and managing CSHR and aims not to miss this rare disease in the differential diagnosis. Recognizing this self-healing disease is important to prevent unnecessary treatment initiation and treatment-related side effects. However, long-term follow-up is necessary to evaluate systemic symptoms.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

***Idea/Concept:** Esin Diremsizoğlu, Uğur Demirsoy, Evren Odyakmaz Demirsoy; **Design:** Esin Diremsizoğlu, Uğur Demirsoy, Hatice Neşe Doğan, Evren Odyakmaz Demirsoy; **Control/Supervision:** Uğur Demirsoy, Evren Odyakmaz Demirsoy; **Data Collection and/or Processing:** Esin Diremsizoğlu, Uğur Demirsoy, Hatice*

Neşe Doğan, Evren Odyakmaz Demirsoy; ***Analysis and/or Interpretation:*** *Esin Diremsizoğlu, Uğur Demirsoy, Hatice Neşe Doğan, Evren Odyakmaz Demirsoy;* ***Literature Review:*** *Esin Diremsizoğlu, Uğur Demirsoy, Evren Odyakmaz Demirsoy;* ***Writing the Article:*** *Esin Diremsizoğlu, Uğur Demirsoy, Hatice Neşe Doğan, Evren Odyakmaz Demirsoy;* ***Critical Review:*** *Esin Diremsizoğlu, Uğur Demirsoy, Evren Odyakmaz Demirsoy;* ***References and Fundings:*** *Esin Diremsizoğlu, Uğur Demirsoy, Hatice Neşe Doğan, Evren Odyakmaz Demirsoy;* ***Materials:*** *Esin Diremsizoğlu, Uğur Demirsoy, Hatice Neşe Doğan, Evren Odyakmaz Demirsoy.*

REFERENCES

1. Haupt R, Minkov M, Astigarraga I, Schäfer E, Nanduri V, Jubran R, et al; Euro Histio Network. Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer. 2013;60(2):175-84. [PubMed] [PMC]
2. Gökmen Z, Özkiraz S, Kıyıcı H, Ecevit A, Tarcan A. Case report: Congenital self-healing reticulohistiocytosis (Hashimoto-Pritzker disease). Turk Arch Pediatr. 2011;46:84-6. [Link]
3. Lee YH, Talekar MK, Chung CG, Bell MD, Zaenglein AL. Congenital self-healing reticulohistiocytosis. J Clin Aesthet Dermatol. 2014;7(2):49-53. [PubMed] [PMC]
4. Parimi LR, You J, Hong L, Zhang F. Congenital self-healing reticulohistiocytosis with spontaneous regression. An Bras Dermatol. 2017;92(4):553-5. [PubMed] [PMC]
5. Larralde M, Rositto A, Giardelli M, Gatti CF, Santos-Muñoz A. Congenital self-healing Langerhans cell histiocytosis: the need for a long term follow up. Int J Dermatol. 2003;42(3):245-6. [Crossref] [PubMed]
6. Larsen L, Merin MR, Konia T, Armstrong AW. Congenital self-healing reticulohistiocytosis: concern for a poor prognosis. Dermatol Online J. 2012;18(10):2. [PubMed]
7. Satter EK, High WA. Langerhans cell histiocytosis: a review of the current recommendations of the Histiocyte Society. Pediatr Dermatol. 2008;25(3):291-5. [PubMed]