Case report

Unusual Coexistence of Castleman's Disease and Pityriasis Lichenoides in a Child

ABSTRACT

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| Castleman disease is a rare lymphoproliferative disorder, particularly in children. Its association with pityriasis lichenoides has not been previously reported. This report presents a 5-year-old girl born to consanguineous parents, who developed recurrent necrotic and bullous skin lesions diagnosed as chronic pityriasis lichenoides (CD8+). One year later, she developed bilateral cervical lymphadenopathy, and imaging revealed multiple necrotic nodes. A lymph node biopsy confirmed hyaline-vascular Castleman disease. This case suggests a potential immunological link between chronic pityriasis lichenoides and Castleman disease. The patient is scheduled for surgical excision of the lymph node mass and will undergo continued dermatologic follow-up. This is the first pediatric case reporting this rare coexistence, highlighting the need for systemic evaluation in chronic dermatoses with lymphadenopathy. |

*Keywords:* Castleman disease, Pityriasis lichenoides, CD8+, Hyaline-vascular, Child, Lymphadenopathy*.*

1. INTRODUCTION

Castleman disease (CD) is an uncommon lymphoproliferative disorder characterised by benign but often massive lymph-node hyperplasia (Christian Hoffmann, et al. 2024). The hyaline-vascular (HV) histological variant predominates in the unicentric form and represents roughly 80 % of cases, yet paediatric presentations are exceptionally rare, accounting for fewer than 10 % of reported series (Jiang P, et al., 2024), (Yavasoglu I, et al., 2009). Unicentric HV-CD usually manifests as a slow-growing, painless cervical or mediastinal mass and is frequently curable by complete surgical excision (Hu S, et al., 2023), (Imen BI, et al., 2020).

Pityriasis lichenoides (PL) is an equally rare spectrum of inflammatory dermatoses—including pityriasis lichenoides chronica (PLC) and pityriasis lichenoides et varioliformis acuta (PLEVA)—that classically affects children and young adults with recurrent crops of papulo-vesicular or ulceronecrotic lesions that heal with hypopigmented scars (Teklehaimanot F, et al., 2025). Histologically, PL demonstrates an interface dermatitis rich in cytotoxic CD8-positive lymphocytes, and clonal T-cell receptor rearrangements have led some authors to regard it as a cutaneous lymphoproliferative disorder.

Although PL has occasionally been described as a paraneoplastic manifestation in Hodgkin and non-Hodgkin lymphomas, its coexistence with CD has not, to our knowledge, been reported. We present what appears to be the first documented case of HV-CD associated with chronic PL in a child, discuss potential pathogenic links, and highlight the diagnostic obstacles raised by this unusual constellation.

2. PRESENTATION OF CASE

We report the case of a 5-year-old girl born to first-degree consanguineous parents, who had developed—since the age of one—recurrent pustular and bullous skin lesions that progressed to necrosis and ulceration. These eruptions appeared in flare-ups alternating with remission and eventually left hypopigmented residual scars (Figure 1).

A punch biopsy of a lesion demonstrated chronic pityriasis lichenoides with immunohistochemically dense CD8-positive lymphocytic infiltrates (Figure 2).

Over the past year the child also noted painless, bilateral cervical swellings. Examination revealed multiple firm, mobile lymph-node masses bilaterally, the largest measuring 2 cm. Cervical ultrasound and CT imaging showed numerous well-circumscribed, heterogeneous hypoechoic latero-cervical nodes with central necrosis, ranging from 6 to 12 mm.

Excisional biopsy of one node revealed an atrophic nodal architecture containing only a few lymphoid follicles; several atretic follicles displayed hyalinised capillaries traversing the germinal centres. Extensive interfollicular hyaline deposits and numerous small vessels with hyalinised walls were present. This pattern is diagnostic of the hyaline-vascular variant of unicentric Castleman disease.

3. discussion

The temporal sequence in our patient—cutaneous lesions beginning at one year of age and persistent lymph-node enlargement appearing later—raises the possibility that PL acted as an early paraneoplastic or immune-dysregulatory clue to underlying CD. PL has long been thought to reflect either an aberrant immune response to infection or a low-grade clonal T-cell proliferation; CD, conversely, is driven by dysregulated cytokine signalling, notably interleukin-6, within germinal-centre B-cells and follicular dendritic cells (Sikora M, et al., 2025). A shared milieu of chronic T-cell activation and cytokine overproduction may therefore provide a mechanistic bridge between the two entities. Elevated interleukin-6 levels, typically seen in Castleman disease, and CD8+ cytotoxic T-cell infiltrates in PL suggest a possible shared cytokine-driven immunopathology This implies that the inflammation and immune cell behavior observed in both conditions might stem from similar dysregulation of cytokine signaling pathways, specifically involving IL-6 and cytotoxic T-cells (Sumiyoshi R, et al., 2022), (Farias MG, et al., 2022), (Li Yu, et al. 2017).

The hyaline-vascular variant is the most common form of Castleman disease, characterized by specific microscopic features including increased lymphoid follicles with regressed germinal centers and hyalinized vessels (Naghashpour M, et al., 2010). This subtype of CD is typically localised and haemodynamically silent. In children it preferentially involves cervical nodes, and complete surgical resection yields long-term remission rates exceeding 95 % (Hu S, et al., 2023), (Korbi AE, et al., 2020), (Carbone et al., 2021). Our patient’s imaging findings of well-circumscribed, heterogeneous, partly necrotic nodes measuring up to 2 cm were consistent with a localized lymphoproliferative process. Histological analysis revealed an atrophic nodal parenchyma with sparse lymphoid follicles, hyalinised blood vessels traversing atretic germinal centers, and extensive interfollicular hyaline deposits—features characteristic of the hyaline-vascular variant of Castleman disease.

Management of chronic PL remains empirical; macrolide antibiotics, narrow-band UVB phototherapy and low-dose methotrexate are most frequently employed. Interestingly, isolated reports describe improvement of PL after successful treatment of an associated haematological malignancy, supporting the paraneoplastic hypothesis (Kempf W, et al., 2005), (C. Damak, et al., 2015). Whether excision of the unicentric CD focus in our patient will attenuate her cutaneous relapses warrants close dermatological follow-up.

Consanguinity in our case may point to an inherited tendency toward immune dysregulation, as suggested for familial forms of both CD and PL, although the precise genetic drivers remain elusive. This highlights the need for genetic and cytokine-profiling studies in similar presentations.

4. Conclusion

This case underscores an unrecognised association between chronic pityriasis lichenoides and unicentric hyaline-vascular Castleman disease in a paediatric patient born of consanguineous parents. The coexistence of persistent, relapsing ulceronecrotic skin lesions with latero-cervical lymphadenopathy should prompt clinicians to consider an underlying lymphoproliferative process. Early lymph-node biopsy remains pivotal for diagnosis, and complete surgical excision of the unicentric CD lesion offers an excellent prognosis. Further reports are needed to clarify whether PL may serve as a cutaneous marker of Castleman disease and to unravel the immunopathogenic links between these two rare disorders.

Consent

All authors declare that ‘written informed consent was obtained from the patient’s legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

Ethical approval

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

Disclaimer (Artificial intelligence)

Authors hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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Figure 1 (a,b) : Crusted papular lesions and hypopigmented scarring in chronic pityriasis lichenoides.



Figure 2 : Immunohistochemical staining showing strong CD8-positive lymphocytic infiltrate in chronic pityriasis lichenoides.