CASE REPORT

Multiple skin nodules as a sign of relapsed systemic anaplastic large cell lymphoma in an adolescent

Sarangini Yoganandamoorthy[1], **Nadeesha Dinaratne**[1], **Fathima Rahna Sehu Mohamed**[1], **Thuwaraha Kalaivathanan**[1], **Ramani Punchihewa**[2], **Janaka Akarawita**[1]

[1]Dermatology Unit, National Hospital of Sri Lanka, Colombo, Sri Lanka.
[2]Histopathology Unit, National Hospital of Sri Lanka, Colombo, Sri Lanka.

Correspondence: Sarangini Yoganandamoorthy, sarangini.y@gmail.com

Abstract

Anaplastic large cell lymphoma (ALCL) is a rare T-cell non-Hodgkin lymphoma, accounting for 2–3% of all non-Hodgkin lymphomas. While primary cutaneous ALCL is well documented, secondary cutaneous involvement from systemic ALCL is an uncommon presentation and can signify disease progression or relapse. Early recognition is key to timely management.

A 15-year-old patient with a history of systemic ALCL presented with multiple, rapidly enlarging, firm nodules and plaques over the face, trunk, and extremities. Lesions were tender and non-ulcerated, and this was her second episode within the past year, after which she defaulted on treatment. She also complained of exertional dyspnoea. Skin biopsy revealed acanthosis of the epidermis and a heavy, nodular dermal infiltrate composed of atypical lymphoid cells, including large pleomorphic cells, with small reactive lymphoid cells observed in the background. Immunohistochemistry for CD30 showed strong diffuse membrane positivity in large cells, confirming cutaneous metastasis of systemic ALCL. CD3 and CD20 were negative, and ALK-1 testing was not performed due to unavailability.

Cutaneous metastasis in systemic ALCL is rare, especially among paediatric and adolescent populations. It can mimic infections, drug reactions, or other malignancies, thereby complicating diagnosis. Recognising skin involvement is significant, as it can indicate relapse. Histopathology with immunophenotyping is essential for establishing a definitive diagnosis. Early detection facilitates timely systemic therapy and clinical management. This case highlights the importance of vigilance for new skin lesions in patients with a history of systemic ALCL, emphasising the crucial role of dermatological assessment in ongoing follow-up care.

Keywords: anaplastic large cell lymphoma; systemic ALCL; skin nodules; relapse; cutaneous metastases

Conflicts of Interest: None declared

Funding: None

Received: 30 March 2026 Revised: 23 April 2026 Accepted: 02 May 2026

How to cite this article: Yoganandamoorthy S, Dinaratne N, Sehu Mohamed FR, Kalaivathanan T, Punchihewa R, Akarawita J. Multiple skin nodules as a sign of relapsed systemic anaplastic large cell lymphoma in an adolescent. Sri Lanka J Dermatol. 2026;25(1):95-100. doi: https://doi.org/10.4038/sljd.v25i1.394

Introduction

Anaplastic Large Cell Lymphoma (ALCL) is a subtype of non-Hodgkin T-cell lymphoma characterised by strong CD30 expression. ALCL accounts for around 13% of all non-Hodgkin lymphomas in the paediatric population.[1]

According to the World Health Organization (WHO), ALCL is classified into four main groups: ALK (Anaplastic Lymphoma Kinase) -positive ALCL, ALK-negative ALCL, breast implant–associated ALCL, and CD30-positive cutaneous lymphoproliferative disorders, which are further subdivided into cutaneous ALCL and lymphomatoid papulosis.[2] Cutaneous ALCL can be further categorised as either primary cutaneous ALCL, which originates in the skin, or systemic ALCL with secondary cutaneous metastases.3 The skin is the most common site of extranodal involvement in systemic ALCL.1 Although primary cutaneous ALCL has been extensively studied, there is limited data available regarding secondary cutaneous ALCL.[3]

This report describes a 15-year-old patient with systemic ALCL and secondary cutaneous involvement, emphasising the associated diagnostic and staging challenges.

Case Presentation

A 15-year-old female with a previously diagnosed systemic Anaplastic Large Cell Lymphoma (ALCL) presented with multiple, rapidly enlarging, firm nodules and plaques on the face, trunk, and extremities, which had developed within two weeks.

The initial diagnosis of ALCL was made three years prior, following evaluation for low-grade fever, cervical lymphadenopathy, and hepatosplenomegaly. The patient received chemotherapy, achieving complete remission 18 months earlier. Treatment was complicated by neutropenia, which was treated with granulocyte colony-stimulating factor (GCSF).

The patient reported the recent onset of multiple painful skin nodules that rapidly increased in number and size on the face, trunk, and extremities over a two-week duration. There was no preceding history of trauma, insect bites, or new medications. Constitutional symptoms, including intermittent fever, night sweats, cough, and dyspnoea, were present at the time of presentation. A non-healing ulcer on the left knee had persisted for two months. This was her second episode within the past year, and she had self-discontinued follow-up for the last seven months.

Dermatological examination revealed multiple erythematous to violaceous nodules, measuring approximately 0.5–2 cm in diameter, distributed over the face, trunk, and extremities (Figure 1A-D). The nodules were firm to soft, dome-shaped, and tender, some with evidence of ulceration. Mucosal involvement was absent. Cervical lymphadenopathy and hepatosplenomegaly were noted on systemic examination.

Baseline laboratory investigations, including complete blood count and liver function tests, were performed. The erythrocyte sedimentation rate (ESR) was elevated at 60 mm/hr. Lactate dehydrogenase (LDH) was not done due to unavailability. Ultrasound examination demonstrated hepatomegaly, massive splenomegaly, and bilaterally enlarged cervical lymph nodes with fatty hilum. Chest X-ray showed a globular, dilated cardiac silhouette, suggestive of pericardial effusion.

Diagnosis and Management: Based on the clinical findings, cutaneous relapse of ALCL was considered the primary differential. Differential diagnosis, including disseminated mycobacterial infection and deep fungal infection, was considered.

Skin biopsies from two nodules were obtained for histopathological evaluation. Microscopic examination revealed acanthosis of the epidermis and a dense, nodular dermal infiltrate composed of atypical lymphoid cells, including large pleomorphic cells, with small reactive lymphoid cells in the background (Figure 2A-D). Immunohistochemistry for CD30 revealed strong diffuse membrane positivity in large cells, confirming cutaneous metastasis of systemic ALCL. CD3 and CD20 were negative, and ALK-1 testing was not performed due to unavailability. These findings are consistent with cutaneous involvement by systemic ALCL.

Based on the clinical history and histopathological findings, a diagnosis of secondary cutaneous involvement of systemic ALCL was established. The patient was referred to the oncology team for chemotherapy and further management. Despite these interventions, the patient succumbed to the disease during oncological treatment within a few weeks.

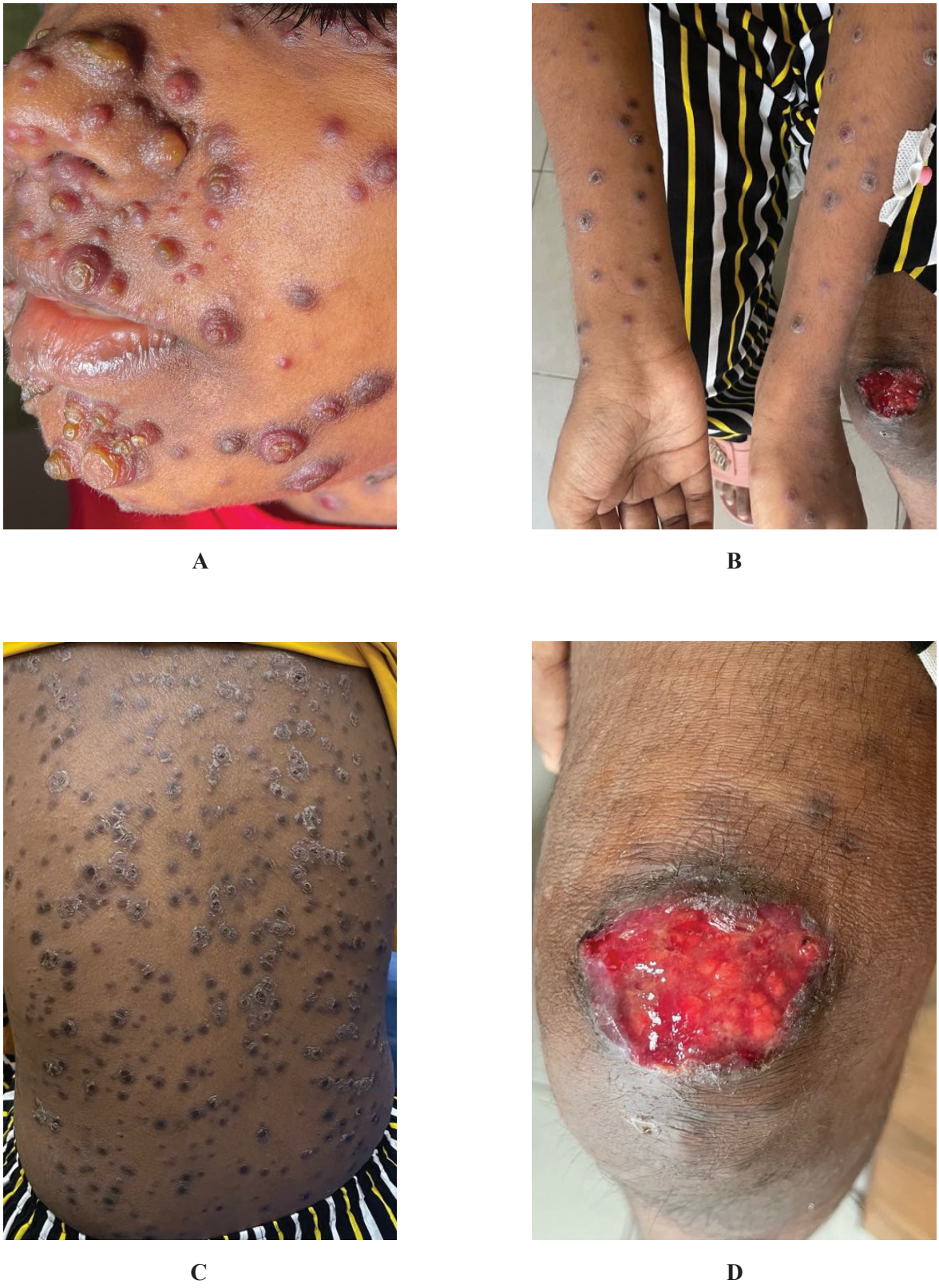

Figure 1. Erythematous to violaceous nodules distributed over the (A) face, (B) extremities, and (C) trunk; (D) non-healing ulcer on the left knee that had persisted for two months prior to presentation

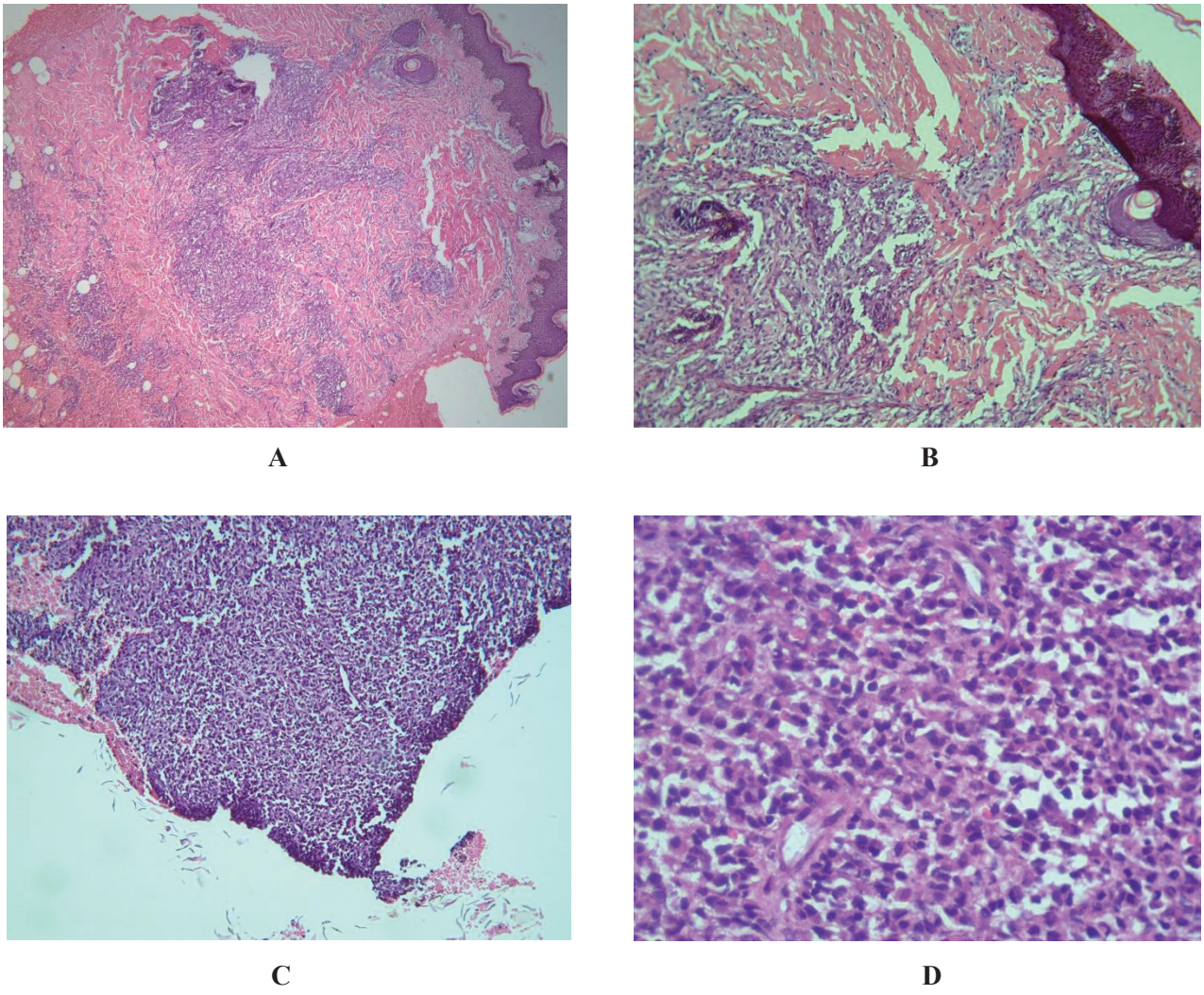

Figure 2. (A, B) Biopsy from a nodular lesion on the upper limb (H&E, × 40 and ×100); (C, D) Biopsy from a nodular lesion on the face (H&E, ×100 and × 400).

Discussion

Cutaneous manifestations of lymphoma arise either as primary cutaneous lymphomas or as secondary involvement from systemic disease.

Primary cutaneous lymphomas are classified as cutaneous T-cell lymphomas (CTCL) and cutaneous B-cell lymphomas (CBCL). Most skin-associated lymphoid infiltrates are of T-cell origin. Among CTCLs, mycosis fungoides is the most common subtype, typically presenting with patches, plaques, or tumours. Sézary syndrome is a more aggressive leukaemic variant, characterised by erythroderma, lymphadenopathy, and circulating malignant T cells. Other subtypes include CD30-positive lymphoproliferative disorders, such as lymphomatoid papulosis and primary cutaneous anaplastic large cell lymphoma. CBCLs include indolent forms such as primary cutaneous marginal zone lymphoma and follicle centre lymphoma, as well as the more aggressive diffuse large B-cell lymphoma, leg type.[4]

Secondary cutaneous involvement occurs more often with non-Hodgkin lymphoma than Hodgkin lymphoma. It may present as nodules, plaques, or ulcerative lesions and often indicates advanced disease.[4]

ALCL is a subtype of non-Hodgkin lymphoma that can be categorised into two broad groups: (1) primary cutaneous ALCL and (2) primary systemic ALCL. Although they are similar in morphology, the clinical features, genetics, and immunohistopathology differ.[5]

Primary systemic (nodal) ALCL is more common than the cutaneous form, and it comprises 2-8% of all non-Hodgkin lymphomas. Its prognosis depends on ALK-1 positivity, which is commonly expressed in the systemic type. Extranodal involvement of systemic ALCL most frequently affects the skin in about 20% of patients, followed by bone (17%), soft tissue (17%), lung (11%), and liver (8%). Gastrointestinal tract and central nervous system involvement is rare.[5,6]

Skin involvement in systemic ALCL may occur at initial presentation or during a relapse. Cutaneous lesions are variable and can present as nodules, papules, or ulcerative lesions that can mimic other dermatological conditions, such as mycobacterial or fungal infections.[5]

In contrast, in primary cutaneous ALCL, skin lesions arise de novo and are usually seen in older patients (around 60 years). These lesions typically present as a red-brown, solitary nodule or papule that may superficially ulcerate. Very rarely, clusters of multiple tumour nodules may develop, either in a localised area or at multiple sites.[6,7]

In systemic ALCL, most patients (around 60%) have advanced-stage disease with extranodal involvement at diagnosis. They may also experience constitutional symptoms such as fever, arthralgia, bone pain, migratory rash, and pruritus, which can be attributed to paraneoplastic phenomena caused by cytokines produced by the tumour cells.[8]

The presence of multiple skin nodules, as described in this case, should raise suspicion for systemic disease recurrence in patients with a prior history of ALCL. Relapses of systemic ALCL can involve lymph nodes, skin, bone marrow, and other extranodal sites. Cutaneous relapse can be isolated or occur with systemic disease progression.

The diagnosis is via histological examination of a skin biopsy. Histology will show diffuse proliferation of large, atypical, pleomorphic lymphoid cells with abundant cytoplasm, and hallmark cells with kidney-shaped nuclei and wreath cells with multiple nuclei arranged in a circular pattern can be seen. Immunohistochemistry plays a critical role. CD30 expression is a defining feature. Additional markers, including ALK, epithelial membrane antigen (EMA), and CD3, can help with diagnosis and distinguishing other lymphoproliferative disorders.[8]

Upon identification of relapse, patients require comprehensive restaging, including PET-CT and bone marrow evaluation. The gold standard for management is multi-agent chemotherapy, such as the CHOP regimen. (cyclophosphamide, doxorubicin [hydroxydaunomycin], vincristine [Oncovin], and prednisone) Autologous or allogenic haemopoietic stem cell transplantation, anti-CD30 monoclonal antibody-targeted therapy (brentuximab vedotin) in patients with refractory or recurrent disease may be considered.[5,8]

Systemic ALCL has a less favourable prognosis than primary cutaneous ALCL overall. ALK-1 expression in systemic ALCL is associated with better survival than ALK-1-negative disease.

However, cutaneous or haematogenous spread is an indicator of poor prognosis in systemic ALCL, with a 5-year survival of 23-28% after the development of cutaneous involvement.5 Although ALK-positive ALCL is associated with relatively favourable outcomes, relapse adversely impacts prognosis.

Conclusion

Cutaneous involvement in Anaplastic Large-Cell Lymphoma (ALCL) is an important clinical manifestation that can indicate a relapse and systemic metastasis.

The appearance of new skin lesions in patients with a known history of systemic lymphoma should prompt a thorough dermatological evaluation. These lesions can mimic other benign dermatologic conditions, in turn delaying diagnosis. Therefore, the early recognition and prompt histopathological examination of suspicious cutaneous lesions is essential to establish the correct diagnosis. This will facilitate timely multidisciplinary management and early detection of relapse, which is critical for optimising patient outcomes. Vigilance, consistent follow-up, and proactive monitoring are essential. Patients and their families should be educated about warning signs and symptoms to facilitate early medical intervention.

This case highlights the importance of awareness of these presentations of lymphoma relapse and maintaining a high index of suspicion when evaluating new skin lesions, particularly when lesions are rapidly progressive, in a patient with a history of lymphoma.

Ethics statement

Informed consent was obtained from the patient's guardian.

Authors' contribution

SY drafted the manuscript. All authors reviewed and approved the final version.

References

1. Kadin ME, Carpenter C. Systemic and primary cutaneous anaplastic large cell lymphomas. Semin Hematol. 2003;40(3):244–56. doi: 10.1016/s0037-1963(03)00138-0
2. Alaggio R, Amador C, Anagnostopoulos I, Attygalle AD, Araujo IB de O, Berti E, et al. The 5th edition of the World Health Organization Classification of Haematolymphoid Tumours: Lymphoid Neoplasms. Leukemia. 2022;36(7):1720–48. doi: 10.1038/s41375-022-01620-2
3. Lee WJ, Moon IJ, Lee SH, Won CH, Chang SE, Choi JH, et al. Cutaneous anaplastic large-cell lymphoma (ALCL): A comparative clinical feature and survival outcome analysis of 52 cases according to primary tumor site. J Am Acad Dermatol. 2016;74(6):1135–43. doi: 10.1016/j.jaad.2015.12.053
4. Whittaker SJ. Cutaneous lymphomas. In: Griffiths CEM, Barker J, Bleiker TO, Hussain W, Simpson RC, editors. Rook's Textbook of Dermatology. 10th ed. Oxford: Wiley-Blackwell; 2024.
5. Yang S, Khera P, Wahlgren C, Ho J, Jukic D, Geskin L, et al. Cutaneous anaplastic large-cell lymphoma should be evaluated for systemic involvement regardless of ALK-1 status: Case reports and review of literature. Am J Clin Dermatol. 2011;12(3):203–9. doi:10.2165/11537520-000000000-00000
6. Stein H, Foss HD, Durkop H, Marafioti T, Delsol G, Pulford K, et al. CD30+ anaplastic large cell lymphoma: A review of its histopathologic, genetic, and clinical features. Blood. 2000;96(12):3681–95. doi:10.1182/blood.V96.12.3681
7. Ortiz-Hidalgo C, Pina-Oviedo S. Primary Cutaneous Anaplastic Large Cell Lymphoma—A Review of Clinical, Morphological, Immunohistochemical, and Molecular Features. Cancers (Basel). 2023; 15(16):4098. doi: 10.3390/cancers15164098
8. Shustov A, Soma L. Anaplastic Large Cell Lymphoma: Contemporary Concepts and Optimal Management. In: Cancer Treatment and Research [Internet]. Cham: Springer International Publishing; 2019. p. 127–44. Available from: doi.10.1007/978-3-319-99716-2_6