Systemic Anaplastic Large Cell Lymphoma with cutaneous involvement

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ABSTRACT

Systemic anaplastic large cell lymphoma (sALCL) is a rare, malignant T-cell lymphoma characterized by CD30 expression. There are two subtypes: ALK-positive sALCL and ALK-negative sALCL. In either condition, patients commonly present with lymphadenopathy and B symptoms, such as fever, night sweats, and weight loss. Extranodal involvement is uncommon, but does occur to the skin, liver, and lungs. We present a unique case of ALK-negative sALCL with diffuse secondary cutaneous involvement.

CASE PRESENTATION

A 75-year-old woman presented to the dermatology clinic with complaints of bumps on her back and arms for the past 3-4 weeks. The bumps were associated with occasional pruritus. Physical exam revealed multiple crops of papules in different stages of healing, some with central areas of necrosis and ulcerations (Figure 1B,1C). Bilateral forearms showed erythematous papules (Figure 1A). Severe cervical lymphadenopathy was also noted.

The patient was followed by an oncologist due to the severe lymphadenopathy. Findings from the oncologist included a CT of the head and neck, showing high volume disease with bilateral axillary and mediastinal adenopathy. A bone marrow biopsy demonstrated cells staining positive for T-cell origin and negative for anaplastic lymphoma kinase (ALK).

A punch biopsy from the right upper back was taken, which showed large lymphocytes scattered throughout the dermis accompanied by a few scattered eosinophils (Figure 2C). The lymphocytes stained positive for CD3 and CD30 (Figure 2A,2B). A diagnosis of ALK-negative sALCL was made based on the findings from the oncologist and skin biopsy. The patient was started on a chemotherapy regimen, however, was lost to follow-up.

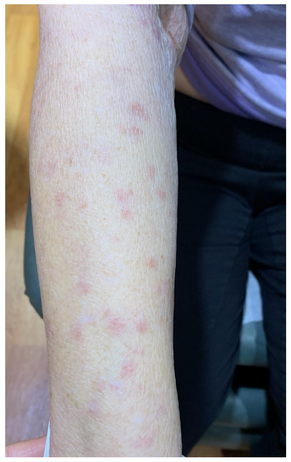

Figure 1A. Erythematous papules of right forearm

CLINICAL PRESENTATION

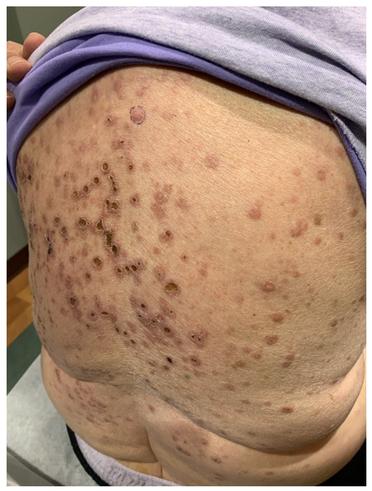

Figure 1B. Violaceous to erythematous papules, with central areas of necrosis and ulcerations

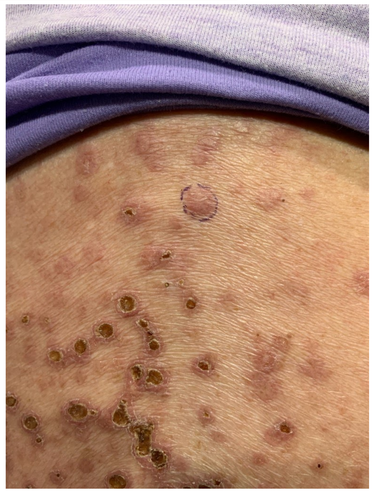

Figure 1C. Up-close image showing biopsy site on right upper back

HISTOPATHOLOGY

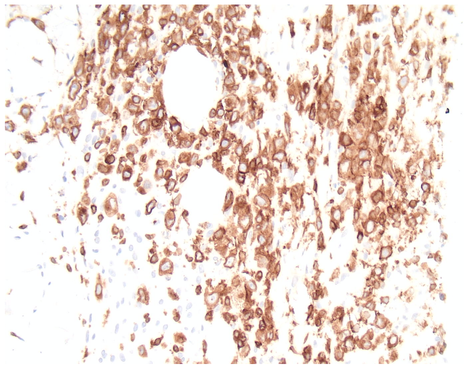

Figure 2A. Cells staining positive for CD3

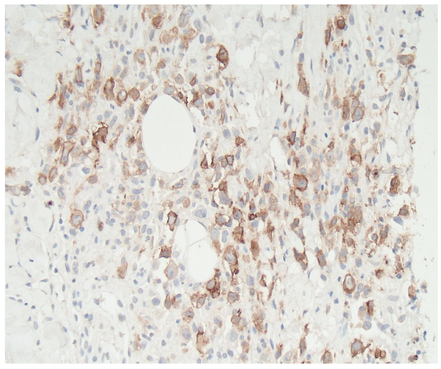

Figure 2B. Cells staining positive for CD30

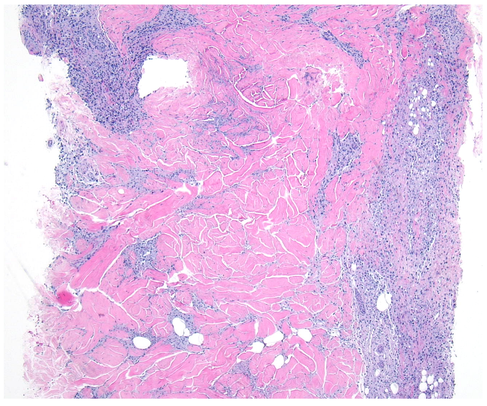

Figure 2C. Punch biopsy showing large lymphocytes throughout the dermis

DISCUSSION

Systemic ALCL (sALCL) is a rare T-cell lymphoma that is part of the larger group of anaplastic large cell lymphomas. The disease represents 2-3% of all non-Hodgkin lymphomas (NHL) in adults and 10-20% of NHL in children. By definition, CD30 is expressed in at least 75% of the T-lymphocytes. One characteristic feature of sALCL is the appearance of neoplastic cells with eccentric horseshoe or kidney-shaped nuclei called “hallmark cells” under microscopic examination. Systemic ALCL can be classified based upon the presence or absence of anaplastic lymphoma kinase (ALK) protein into ALK-positive sALCL and ALK-negative sALCL. ALK-positive sALCL is more common in children and young adults and is associated with a better prognosis. In contrast, ALK-negative sALCL presents more often in the 6th decade of life and is typically more severe.

Dissemination to the skin in sALCL occurs in approximately 20% of cases and is typically observed in advanced stages of the disease. Involvement of multiple regions of the body, as in our case, occurs in only 10% of patients and is rarely reported. The infrequency of such cutaneous manifestations can lead to misdiagnosis with other diseases, such as lymphomatoid papulosis and primary cutaneous ALCL. These conditions are also CD30+ cutaneous T-cell lymphoproliferative disorders and present with similar findings on histology. However, these diseases differ from sALCL in terms of prognosis and management. Current treatment for sALCL is predominantly chemotherapy. Monoclonal antibodies against CD30+ cells, such as brentuximab vedotin, and ALK inhibitors are used to treat refractory disease.

CONCLUSIONS

Cutaneous involvement is an uncommon presentation of systemic anaplastic large cell lymphoma. This case demonstrates the importance of correlating pathologic findings with the clinical course of symptoms to arrive at the correct diagnosis and maximize the chance of remission for the patient.

REFERENCES

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