

A rapidly growing crateriform nodule on the nose

Case

A 34-year-old otherwise healthy woman presented for a rapidly growing skin lesion over the nose of 5 weeks' duration. She attempted treatment with topical antibiotics with no improvement. Physical examination revealed a 1.5 cm crateriform erythematous nodule with a central crusted keratinous core over the left nasal sidewall [Figure 1]. The clinical differential diagnosis mainly included epithelial neoplastic tumors such as keratoacanthoma or infections such as cutaneous leishmaniasis, atypical mycobacterial infection or deep fungal infection.

A biopsy specimen [Figures 2 and 3] was taken and revealed irregular epidermal hyperplasia, and a dense dermal mixed cell infiltrate consisting of numerous neutrophils, small

lymphocytes, and numerous large, atypical and mitotically active lymphoid cells. These atypical cells exhibited large, pleomorphic, vesicular and hyperchromatic nuclei with prominent nucleoli and abundant cytoplasm [Figure 2]. Immunohistochemical staining [Figure 3] of these cells showed positivity for CD3, CD4, lymphocyte-associated antigen and CD30 (more than 75% of tumor cells) with negative staining for CD15, CD20 and anaplastic lymphoma kinase. Ki-67 revealed high proliferative index. Special stains and skin tissue cultures were negative for infectious etiology.

What Is Your Diagnosis?

Diagnosis

Primary cutaneous anaplastic large cell lymphoma



Figure 1: A crateriform erythematous nodule with a central crusted keratinous core over the left nasal groove

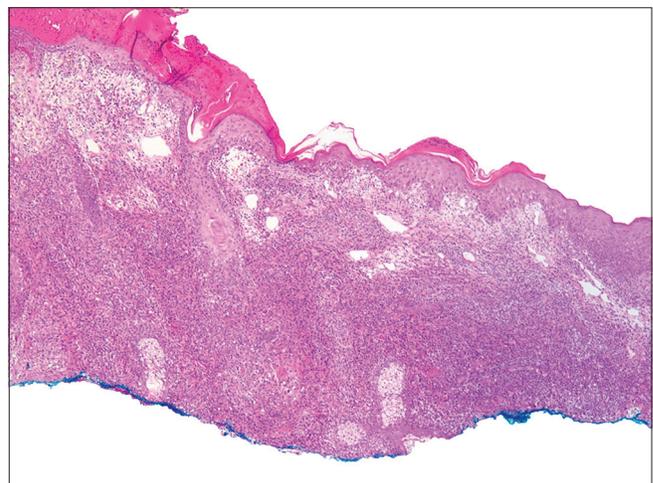


Figure 2a: Histopathologic examination exhibited dense dermal mixed cell infiltrate consisting of small lymphocytes, numerous neutrophils and numerous large, atypical CD30-positive lymphoid cells (H and E, ×400)

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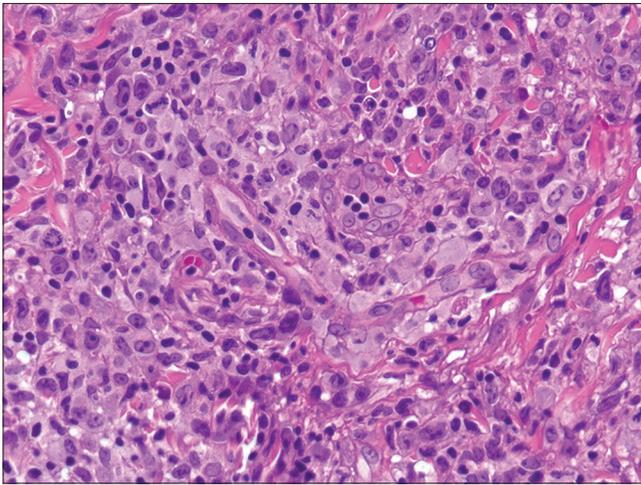


Figure 2b: Close-up view illustrating the atypical large lymphocytes ($\times 400$)

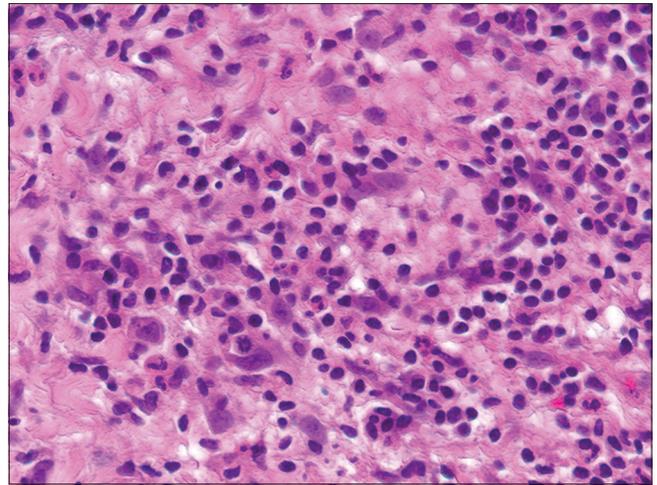


Figure 2c: Close-up view illustrating the neutrophils that can obscure the neoplastic lymphoid cells ($\times 400$)

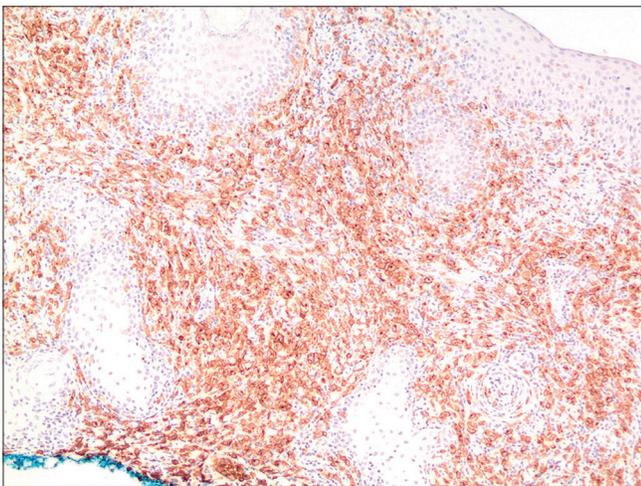


Figure 3a: Positive CD30 ($\times 100$)

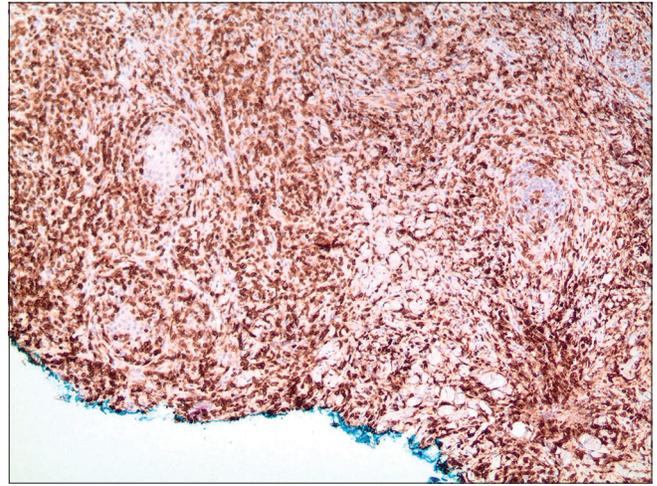


Figure 3b: Positive CD3 ($\times 100$)

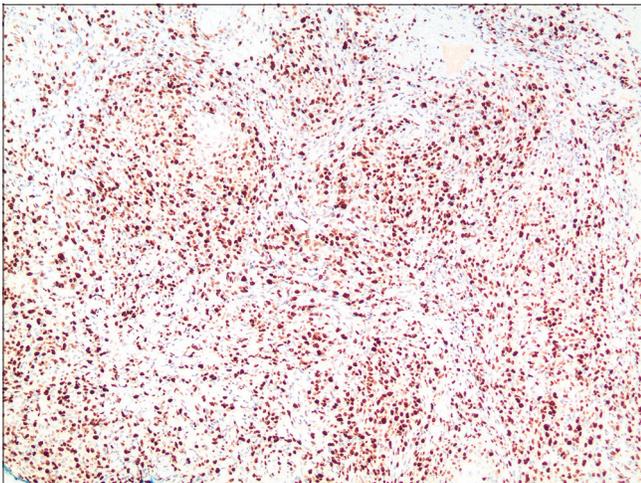


Figure 3c: Positive Ki67 ($\times 100$) immunostaining

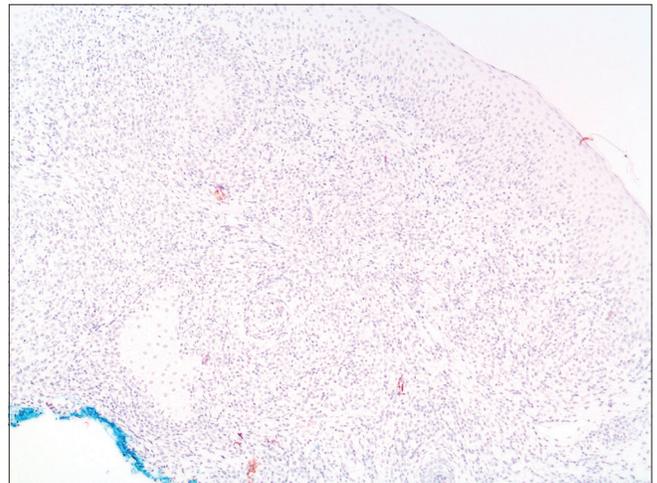


Figure 3d: Anaplastic lymphoma kinase ($\times 100$) was negative

Discussion

Primary cutaneous anaplastic large cell lymphoma is grouped into the spectrum of the primary cutaneous CD30+ lymphoproliferative disorders, constituting the second most common form of primary cutaneous T-cell lymphoma following mycosis fungoides and accounting for 25% of cases. It is an indolent neoplasm with a 5-year survival of up to 95%. It predominantly affects adults (age range 50-70 years) and has a male predominance (male to female ratio 2:1).^{1,2}

Clinically, it typically presents with a solitary nodule or tumor which often ulcerates. Multifocal involvement has also been described. Lesions may spontaneously regress, indicating a favorable prognosis; however, relapses are common.² Histologically, the tumor cells are usually atypical, pleomorphic, irregularly shaped with prominent nucleoli, abundant basophilic cytoplasm and high mitotic index. These cells comprise at least 30% of the inflammatory cell infiltrate. Epidermotropism, adnexal and lymphatic involvement may also be seen. The majority of the neoplastic cells express CD2, CD3 and CD30 (at least 75% of the tumor cells). Positive immunohistochemical staining with anaplastic lymphoma kinase was previously considered in favor of systemic anaplastic large cell lymphoma and against the diagnosis of primary cutaneous anaplastic large cell lymphoma; however, anaplastic lymphoma kinase positivity has recently been well documented in rare cases of primary cutaneous anaplastic large cell lymphoma.^{2,3}

Primary cutaneous anaplastic large cell lymphoma presenting as a clinical mimicker of keratoacanthoma has been reported in the literature. In the cases described, the lesions presented as a rapidly enlarging tumor with a central keratinous plug, similar to our patient's presentation.^{4,5} Furthermore, primary cutaneous anaplastic large cell lymphoma may also mimic squamous cell carcinoma or keratoacanthoma histologically by displaying pseudocarcinomatous hyperplasia. In such cases, the keratinocytes may also exhibit atypia, and the hyperplasia may easily obscure the tumor cells, leading to an erroneous diagnosis.^{4,5} Growth factors secreted by the tumor cells, such as epidermal growth factors and transforming growth factors, are believed to cause this hyperplasia.⁵

Moreover, a neutrophil/eosinophil-rich primary cutaneous anaplastic large cell lymphoma, albeit rare, has also been reported. The largest and most recent case series conducted by Kong *et al.* included a total of 9 patients.⁶ They described a neutrophil/eosinophil-rich inflammatory cell infiltrate which was not confined to ulcerated or purulent areas with the absence of infection or necrosis. This constitutes a diagnostic challenge since the dense inflammatory infiltrate may mask the atypical lymphoid cells and lead to an erroneous diagnosis of keratoacanthoma or squamous cell carcinoma.⁴ Studies suggest that tumor cells produce interleukins 5 and 8 which lead to the dense neutrophilic and eosinophilic infiltrate.⁴

Though the morphologic and immunophenotypic characteristics of primary cutaneous anaplastic large cell lymphoma and lymphomatoid papulosis overlap significantly especially lymphomatoid papulosis type C whose microscopic features may be indistinguishable from primary cutaneous anaplastic large cell lymphoma, differentiation can be achieved by correlating the pathologic features with the clinical presentation. Lymphomatoid papulosis is usually characterized by recurrent waxing and waning crops of papules and nodules that tend to be smaller than those of primary cutaneous anaplastic large cell lymphoma. Its other histologic types have histologic features that allow their differentiation from primary cutaneous anaplastic large cell lymphoma. These variants may exhibit wedge-shaped infiltrate of scattered/small clusters of large atypical CD30+ lymphocytes admixed with inflammatory cells in lymphomatoid papulosis type A, epidermotropic infiltrate of small-medium-sized lymphocytes variably expressing CD30 in lymphomatoid papulosis type B, marked epidermotropism of atypical CD8+ and CD30+ lymphocytes in lymphomatoid papulosis type D and angioinvasive atypical CD30+ lymphocytic infiltrates with hemorrhage and necrosis in lymphomatoid papulosis type E.

Treatment options include surgical excision, local radiation and topical bexarotene, mainly for solitary or localized primary cutaneous anaplastic large cell lymphoma. Multifocal or disseminated disease is treated with systemic chemotherapy, photochemotherapy, targeted therapies such as brentuximab (monoclonal antibody to CD30) and even allogeneic and autologous stem cell transplantation with variable outcomes.² Systemic work-up in our patient was negative, and the lesion was surgically excised with no recurrence after 3 months of follow-up.

This case illustrates a diagnostic challenge whereby a primary cutaneous anaplastic large cell lymphoma can mimic a keratoacanthoma clinically and display a dense neutrophil-rich inflammatory cell infiltrate histologically. Given the clinical overlap with keratoacanthoma, it is important to consider it in patients with no known risk factors for keratoacanthoma or squamous cell carcinoma presenting with rapidly growing keratoacanthoma-like lesions.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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