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Case Report

CD30+ anaplastic large-cell lymphoma masquerading as lupus vulgaris – A case report

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ABSTRACT

Primary cutaneous anaplastic large-cell lymphoma (PC-ALCL) constitutes 9% of cutaneous T-cell lymphoma, which usually presents as a solitary ulcerating nodule or indurated plaque but sometimes has varied presentations mimicking eczema to squamous cell carcinoma. In our case, PC-ALCL presented as a noduloulcerative lesion. Since PC-ALCL is rare, there is a high chance to misdiagnose and hence high index of suspicion is necessary.

Keywords: Anaplastic lymphoma, Cutaneous large cell lymphoma, Lupus vulgaris

INTRODUCTION

Cutaneous lymphomas form the second most common group of extranodal non-Hodgkin lymphoma. Primary cutaneous anaplastic large-cell lymphoma (PC-ALCL) accounts for 9% of cutaneous T-cell lymphoma. The diagnosis of PC-ALCL is made only if lesions are limited to skin for at least 6 months after initial identification. Hence, regular follow-up of patients is important to rule out systemic involvement.[1] An average of 50% cases is diagnosed at the age of 61 years, reaches an overall peak in the 6th decade of life.[2] The diagnosis of cutaneous lymphoma is difficult and often delayed because of varied presentations and large number of differential diagnosis involving entire spectrum of primary or secondary CD 30+ lymphoproliferative diseases. Histologically, they show a diffuse inflammatory infiltrate with large-sized T-lymphocytes with characteristic round, oval or irregular nuclei, prominent eosinophilic nucleoli, and abundant cytoplasm.

CASE REPORT

A 60-year-old male presented with painful ulcer over the right side of the face for 6-month duration. Lesions started as nodule, which broke down to form an ulcer involving right side of the face such as the right ala of the nose and right side of the upper lip, with progressive increase in size over the past 1 month. 9 years ago, he took treatment for cervical lymphadenopathy for 6 months, details of which are not available. Examination revealed well-defined crusted plaque with ulcerations over the right side of the nose and perinasal area extending to the right side of the upper lip [Figure 1a]. Keeping differential diagnosis of leishmaniasis and lupus vulgaris in mind, incisional biopsy was performed. The Mantoux test was done, the result of which was negative. Meanwhile, histopathology report showed acanthotic epidermis with horn cyst and inflammatory

infiltrates, dermis with lymphoplasmacytic infiltrate and focal neutrophilic collections, and few areas with ill-formed collections of histiocytes, which was in favor of lupus vulgaris. He was started on antituberculous treatment under Directly Observed Treatment, Short-course and was followed up. The lesion started healing in initial 2 months, after which we lost follow-up. Six months after initiation of treatment, he presented with ulceroproliferative growth involving the tip of the nose and the right side of the face, more extensive than initial lesion [Figure 1b]. With high suspicion for cutaneous malignancy, incisional biopsy was repeated from the edge of the ulcer, which showed epidermis with orthokeratosis, parakeratosis, crust formation, and focal ulceration. Acanthosis and neutrophilic microabscess were also noted. Dermis showed neoplasm composed of sheets of lymphoid cells [medium to large sized cells with prominent nucleoli], with small lymphocytes and scattered eosinophils [Figure 2a]. To know the clonality of cells, immunohistochemistry was done. Neoplastic cells showed cytoplasmic positivity for leukocyte common antigen and strong membrane positivity for CD30 [Figure 2b] and CD3. CD4 showed strong membrane positivity. MIB staining was highly positive (>20% positive). CD20, alkaline phosphatase, myeloperoxidase, and epithelial membrane antigen were all negative. The diagnosis was made as CD30+ ALCL, which could be either primary or a cutaneous involvement of systemic diseases. Anaplastic lymphoma kinase was negative, favoring PC-ALCL. PET scan [Figure 3] showed activity localized to the right side of the face, confirming it as PC-ALCL.

Figure 1: (a) Initial presentation with ulcer over the right side of the face. (b) Presentation after 6 months as ulceroproliferative growth involving the tip of the nose right side of the face.

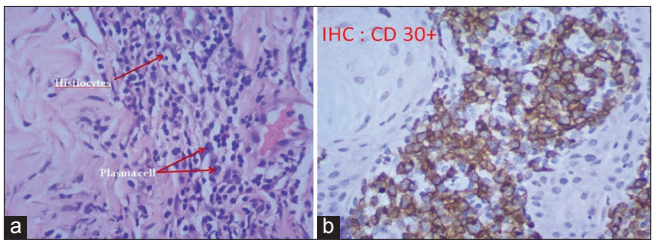

Figure 2: (a) Histopathology showing ill-formed collections of histiocytes and plasma cells in the background of inflammatory infiltrate (H and E, ×200). (b) Immunohistochemistry showing CD30+ cells (DAB chromagen, ×200).

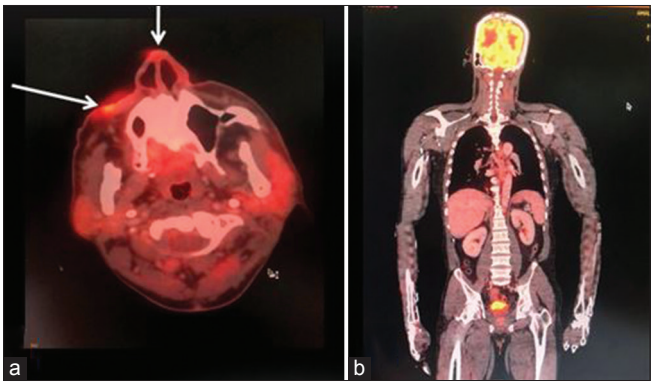

Figure 3: (a) PET scan showing activity confined to the right side of the face. (b) PET scan showing no other areas of activity/involvement.

DISCUSSION

Lymphoproliferative disorders of primary cutaneous CD30+ type with T-cell origin include lymphomatoid papulosis and CD30+ anaplastic large-cell lymphoma. PC-ALCL affects elderly with the median age of 55 years, with the male-to-female ratio as 1.5–2:1. It commonly affects trunk and extremities and presents as solitary or multiple nodules with ulceration. PC-ALCL can be called a mimicker, as it may resemble eczema, pyoderma gangrenosum, pyogenic granuloma, morphea, and squamous cell carcinoma. In our case, it presented as a lupus vulgaris-like lesion. PC-ALCL presents as rapidly growing ulcer with spontaneous regression seen in 20–42% of cases.[3] Extracutaneous spread occurs in 10% of cases and most commonly involves regional lymph nodes. The mainstay of treatment is wide excision and local radiation.[4] The overall prognosis of the disease is good with 5 years survival between 70% and 90%. Cutaneous recurrences account for around 39%. In our cases, what was perceived as response to anti-tuberculous therapy could be spontaneous regression seen in ALCL. In all cases of nodular ulcerative lesions, cutaneous malignancy should be considered as a differential diagnosis, and dermatologists and pathologists should work as a team for early and accurate diagnosis of the disease.

CONCLUSION

To conclude, in all cases of noduloulcerative lesions dermatologist should consider cutaneous malignancy as a differential diagnosis and dermatologist and pathologist should work hand in hand for early diagnosis of the disease.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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