A case of primary cutaneous anaplastic large cell lymphoma on eyelid

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Abstract

Primary cutaneous anaplastic large cell lymphoma is a kind of cluster of differentiation 30+ primary cutaneous lymphoproliferative disorders with a relatively good prognosis in the absence of high-stage disease. Primary cutaneous anaplastic large cell lymphoma shows a higher frequency in males and commonly affects the head and neck. Palpebral involvement is very rare. We present a 42-year-old lady patient with primary cutaneous anaplastic large cell lymphoma involving the eyelid which was initially misdiagnosed as stye. The patient underwent a total excision of the lesion and showed complete regression of the lesion after surgery without any other treatment. There was no evidence of local or systemic disease during follow-up after nine months.

Key words: Cluster of differentiation (CD 30), eyelid, primary cutaneous anaplastic large cell lymphoma

Introduction

Primary cutaneous anaplastic large cell lymphoma (PCALCL) is a cluster of differentiation (CD) 30+ lymphoproliferative disorder of the skin with a relatively good prognosis.[1] Primary cutaneous lymphomas form approximately 25–30% of CD 30+ lymphoproliferative disorder.[2] Anaplastic large cell lymphoma was initially described in 1985 which includes three types: Primary cutaneous anaplastic large cell lymphoma, primary systemic anaplastic lymphoma kinase-positive anaplastic large cell lymphoma and primary systemic anaplastic lymphoma kinase-negative anaplastic large cell lymphoma.[3] Diagnosis of primary cutaneous anaplastic large cell lymphoma relies strongly on clinicopathologic correlation and is made if the lesions are limited to the skin for at least half a year. Therefore, we must follow-up the patient to rule out any systemic disease.

Case Presentation

A 42-year-old lady presented with a painful erythematous hard nodule on the left lower eyelid of 11 days duration [Figure 1a]. She gave a history of progressive growth of the nodule after a surgical treatment of this lesion which was diagnosed as stye from the department of ophthalmology four days earlier. Physical examination revealed a firm, tender and fixed mass of 2.5 cm × 1.5 cm on the left lower eyelid [Figure 1a]. Laboratory tests showed increased white blood cell count (13.1×10^9/L) and neutrophilic granulocytes (69%). There was no palpable lymphadenopathy. After seven days of antibiotics treatment, the mass enlarged remarkably and developed edema and ulceration [Figure 1b]. Direct microscopic examination, fungal and bacterial cultures, tuberculosis antibody, extractable nuclear antigen and screening for human immunodeficiency virus were negative.

Histopathologically, the tumour revealed a diffuse dermal infiltrate composed of numerous large atypical lymphoid cells with abundant mitotic figures and apoptotic cells [Figures 1c and 1d]. The tumour cells were positive for CD 2, CD 3, CD 4, CD 30 [Figure 1e] and Ki-67 (70%) and were negative for CD 8, CD 19, CD 20, anaplastic lymphoma kinase 1, paired box 5, CD 10, Melan-A, CD 56 [Figure 1f], T cell intracytoplasmic antigen-1, programmed cell death 1,

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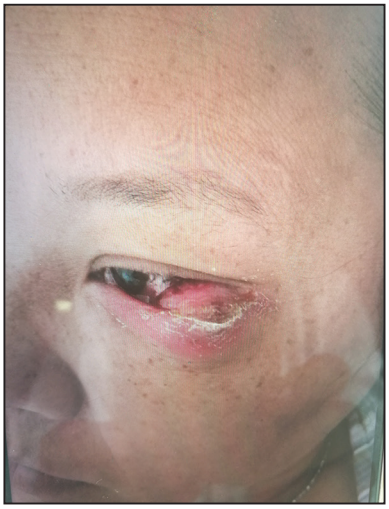

Figure 1a: The nodule on the left lower eyelid

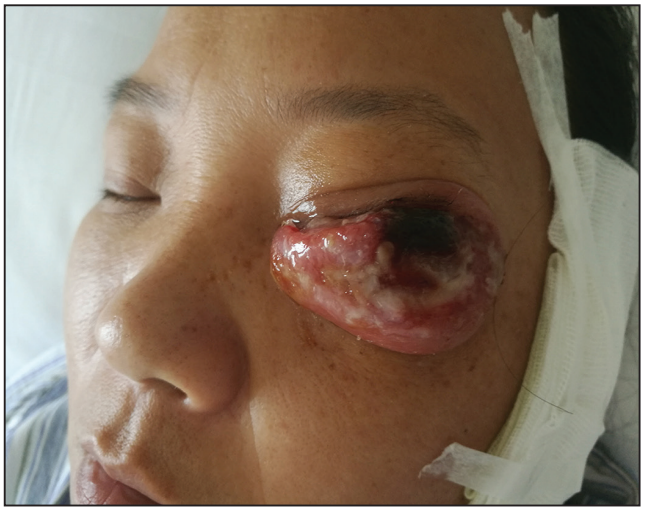

Figure 1b: Progressive growth of the lesion after seven days

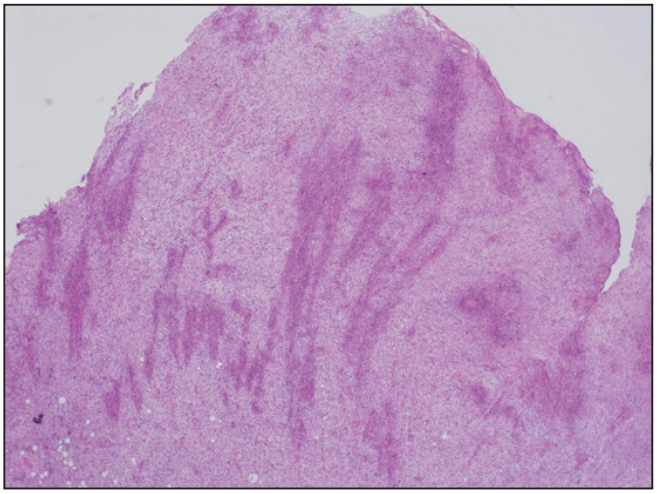

Figure 1c: Diffuse atypical lymphoid cells infiltration in the dermis (Haematoxylin and eosin, ×20)

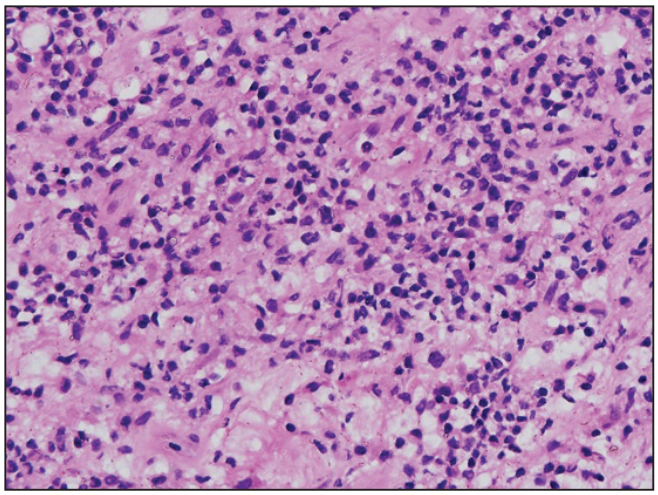

Figure 1d: Diffuse atypical lymphoid cells infiltration in the dermis (Haematoxylin and eosin, ×400)

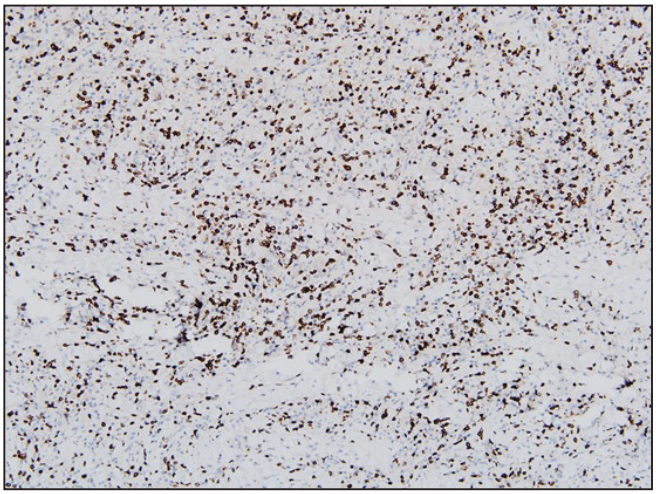

Figure 1e: Immunohistochemistry positive for CD30

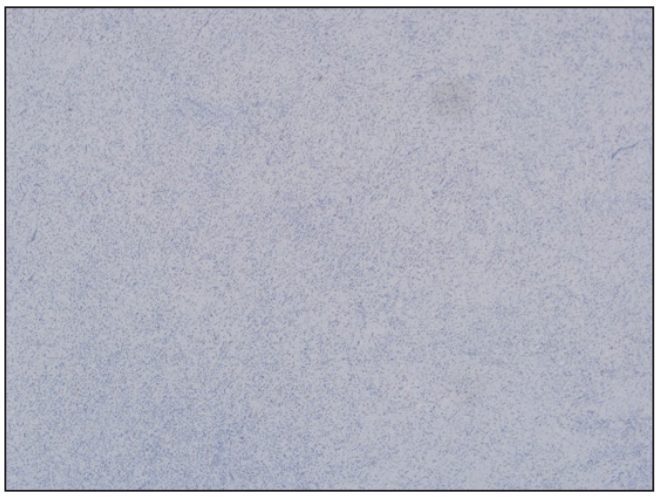

Figure 1f: Immunohistochemistry negative for CD56

Figure 2: In clinical remission after one month of a total excision

CD 15, epithelial membrane antigen and Epstein-Barr encoding region. Full body computed tomography scan and bone marrow biopsy indicated no positive findings. The final diagnosis was primary cutaneous anaplastic large cell lymphoma. The patient received a complete excision of the lesion without radiotherapy or chemotherapy [Figure 2]. Two months later, the patient received a positron emission tomography and computed tomography scan which showed no abnormal findings. There were no findings of local or systemic disease during follow-up after nine months.

Discussion

Primary cutaneous anaplastic large cell lymphoma is a type of CD 30+ primary cutaneous lymphoproliferative disorders with a high survival rate which represents the second most common cutaneous lymphomas after mycosis fungoides.[2,4] It mainly affects adults 50–70 years of age (male:female ratio: 3–2:1).[4] It presents in the skin without systemic involvement at the time of diagnosis which affects the head and neck area commonly and rarely happens on the eyelid alone. Moreover, it affects more frequently the upper eyelid compared to the lower eyelid.[1,5] Patients present with solitary or localised nodules, sometimes ulcerated and 20% may present with multifocal lesions.[6] Histologically, it is composed of large cells with an anaplastic, pleomorphic or immunoblastic cytomorphology that expresses CD 30 antigen by the majority of the tumour cells.

It has a 90% survival rate at five years.[4] Extracutaneous disease, age above 60 years, leg involvement, multiple skin lesions and regional nodal involvement may confer a worse prognosis. It can be treated with excision or local radiation, but relapse is frequent.[2,4]

Differential diagnosis includes infection disease and other malignant tumour of eyelid such as systemic anaplastic large cell lymphoma, secondary cutaneous involvement, basal cell carcinoma, squamous cell carcinoma, lymphomatoid papulosis and extranodal NK/T-cell lymphoma. The most common malignant tumour of eyelid is basal cell carcinoma (80–90%), followed by squamous cell carcinoma (40%), sebaceous carcinoma (5%) and melanoma (1%).[7] Basal cell carcinoma commonly affects the elderly with the highest incidence among patients of 70–75 years.[8] Squamous cell carcinoma presents with a longer course of disease. However, in our case, a 42-year-old patient presented with a duration of 11 days. Moreover, microscopic features of our case are also different from basal cell carcinoma and squamous cell carcinoma. Primary cutaneous anaplastic large cell lymphoma involving the eyelid exclusively has been reported rarely in the literature,[9] orbital anaplastic large cell lymphoma is likewise rare and a case of anaplastic large cell lymphoma involving both the skin and orbit is also reported.[10] But in our case, there was no orbital involvement. In addition, the nodule on the eyelid was misdiagnosed as stye initially from the department of ophthalmology and evolved with progressive growth after a surgery of the lesion. The negative direct microscopic examination as well as fungal and bacterial cultures could rule out infection. Primary cutaneous anaplastic large cell lymphoma with extracutaneous involvement can sometimes be confused with systemic anaplastic large cell lymphoma with secondary cutaneous involvement but full body computed tomography scan and bone marrow biopsy indicated no positive findings in our case. Considering the histopathological and immunohistochemistry, the extranodal NK/T-cell lymphoma was also be ruled out. Lymphomatoid papulosis is characterised by a papulonodular skin eruption which may be solitary, grouped or generalised. Type C lymphomatoid papulosis must be distinguished from primary cutaneous anaplastic large cell lymphoma,[11] but lymphomatoid papulosis can regress spontaneously.

A number of factors must be considered when it comes to the treatment for primary cutaneous anaplastic large cell lymphoma. Radiotherapy or surgical excision can be used to treat the cases of solitary or localised disease.[12] It has a very high five years overall survival rates. Although the prognosis is generally favourable, appropriate diagnosis and management as well as a long time follow-up are important. In our case, the patient underwent surgical excision and showed good response to treatment and was in clinical remission at her last follow-up. Primary cutaneous anaplastic large cell lymphoma presents as a single, ulcerated lesion that grows rapidly. We should pay more attention to any ulcerated lesion which grows rapidly and shows no response to antibiotic.

Conclusion

Primary cutaneous anaplastic large cell lymphoma has a relatively good prognosis. The diagnosis relies strongly on clinicopathologic correlation and is made if the lesions are limited to the skin for at least half a year. Therefore, as in our case, there were no findings of local or systemic disease during follow-up after nine months of surgical excision. But It is also important for a longer follow-up time to rule out any systemic disease.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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