Dermatology Case Reports

Case Report Open Access

Primary Cutaneous Extranodal NK/T-Cell Lymphoma Mimicking a Large B-Cell Lymphoma (Leg Type)

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Abstract

Introduction: Natural killer/T-cell lymphoma, nasal type is a distinct entity, rarely reported in sub-Saharan Africans. Although the skin is the second most common involved site after the nasal cavity, very few cases of primary cutaneous extranodal natural killer/T-cell lymphoma (PC-ENK/T-NT) have been reported so far. Their clinical features reported in the literature are variable.

Case report: We report a case of a 48-year-old man with a PC-ENK/T-NT presenting as a multinodular and ulcerated tumor of 9 cm in diameter on his left leg, mimicking a large B-cell lymphoma, leg type at the onset and only developed nasal and lymph node involvement at a later stage.

Conclusion: PC-ENK/T-NT in our case is remarkable by its presentation simulating a B-cell lymphoma of leg type.

Keywords: Cutaneous lymphomas; Natural killer/T-cell lymphoma; Leg type B-cell lymphoma

Introduction

Natural killer/T-cell lymphoma, nasal type is a rare and distinct entity, which represents less than 0.5% of non-Hodgkin lymphomas [1]. It is common in Latin America as well as in Asia while it is infrequently seen in Western and African countries [2]. It most commonly arises from the nasal cavity and its vicinity. The skin is the second most common site of involvement. However, the incidence of primary cutaneous extranodal natural killer/T-cell lymphoma (PC-ENKTCL) is known to be extremely rare. Very few cases affecting the skin as the primary site have been reported to date [3]. In most cases of PC-ENKTCL previously reported in the literature, the morphology of the skin lesions commonly appears as subcutaneous nodules, cellulitis-like swelling, erythematous papules, nodules or plaques, sometimes ulcers, usually present on the trunk and limbs [1].

The presentation of ENTCL as a multinodular tumor of the leg is extremely rare and may be easily misdiagnosed. In this report, we describe an unusual case with a diagnostically challenging clinical presentation, as a tumor of the leg, mimicking large B-cell lymphoma, leg type.

Case Report

A 48-year-old Senegalese man, without any past medical history, initially presented with a 4-month duration of a gradually enlarging nodular tumor located on his anterior right lower leg developed over four months. The tumor had started as multiple, painless, occasionally pruritic nodules over the lower third of his right leg, progressively increasing in size, becoming confluent in a large and ulcerated mass.

The patient did not experience any episodes of fever or night sweats and there was no history of weight loss. Physical examination revealed a healthy appearing man with a swelling of the entire right leg, covered of a multinodular tumor of 9 cm in diameter which was ulcerated on the surface (Figure 1). A firm, solitary, subcutaneous, painless nodule of 1.5 cm was also noted on his left thigh. Further physical examination including palpation of lymph nodes as well as inspection of the mucous membranes showed no other abnormalities. The remainder physical examination was unremarkable.

Radiological examination showed a soft tissue swelling in the leg region without bone abnormalizes. Laboratory investigations revealed normal total white blood cell, lymphocyte, platelet and erythrocyte count and normal haemoglobin level. There were no atypical cells in the peripheral blood. Erythrocyte sedimentation rate was 33 mm/h and serum C-reactive protein, lactate dehydrogenase level, liver and renal function tests were within normal range. ELISA for HIV and HBSAg were negative.

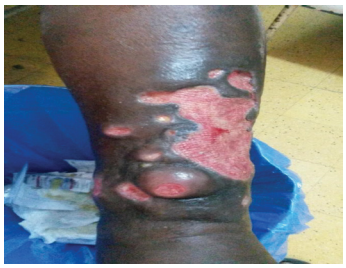

Figure 1: Primary cutaneous extranodal natural killer/T-cell lymphoma presenting as ulcerated multi nodular tumor of the leg.

Histopathological examination of the skin biopsy revealed a diffuse infiltrate of small and medium- sized atypical lymphocytes extending from the dermis into the subcutaneous fat (Figures 2A-2D). The tumour cells had a pale cytoplasm and irregular hyperchromatic nuclei, and mitoses were frequent. There were a few plasma cells and neutrophils among the lymphocytes. There was no perivascular accentuation of the infiltrate. No epidermotropism was noted. The features were suggestive of cutaneous lymphoma possibly B-cell lymphoma. The tissue paraffin block was sent to a higher centre for immunohistochemical analysis.

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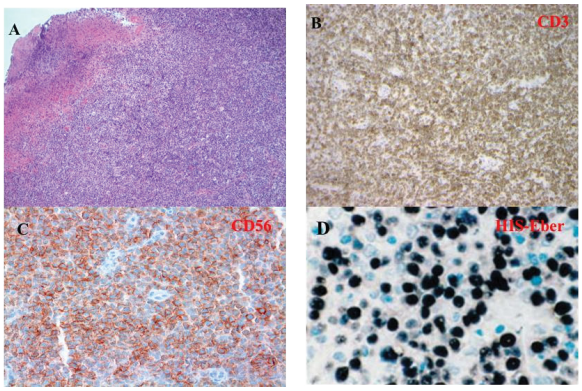

Figure 2: Diffuse lymphomatous infiltrate in the dermis with epidermal ulceration: (A) HEX40, (B) Immunohistochemically, the tumor cells express CD3ε intracytoplasmic, (C) CD56 and (D) EBV by *in situ* hybridization.

Further investigations, including chest X-ray, abdominal ultrasound, CT scan of the chest/abdomen/pelvis did not indicate extracutaneous involvement. Bone marrow examination was not performed. In the interim, the patient was treated with CHOP chemotherapy regimen (cyclophosphamide, doxorubicin, vincristine, prednisolone) which was well tolerated. After temporary improvement, he presented with rapid progression of the tumor with concomitant occurrence of new tumefaction of the left side of the nose and enlargement of submandibular lymph nodes. Immunohistochemical results from skin biopsy were obtained which revealed positivity to CD56, surface CD3ε intracytoplasmic+, CD2, granzyme-B, perforin and TIA-1 and negativity to CD20, CD4, CD8 and CD30. In addition, positive staining for Epstein-Barr virus-encoded RNA (EBER) was found by *in situ* hybridisation. Thus, based on these findings, the final diagnosis of primary cutaneous extranodal natural killer/T-cell lymphoma (ENKTL), nasal type, was made.

ENT examination and nasoendoscopy did not reveal any further tumor infiltration in the area of pharynx. A biopsy from the nasal cavity showed similar findings to that of the skin with diffuse infiltration by lymphoid cells with hyperchromatic nuclei in the subepithelial region, admixed with plasma cells and neutrophils. A few blood vessels showed fibrinoid degeneration and angiolysis. Nuclear dust and necrosis were seen.

The patient was referred to the haematology-oncology service, where he was treated with EPOCH chemotherapy (etoposide, prednisone, vincristine, cyclophosphamide and doxorubicin). In the course of hospitalization, the patient died from sepsis related to complications of the chemotherapy.

Discussion

In our case, the characteristic cell morphology, the positive expression of CD2, CD56 and cytotoxic markers and the detection of EBER by *in situ* hybridisation in atypical lymphocytes confirmed the diagnosis of ENTCL. This lymphoma has been very rarely described in sub-Saharan Africans despite the endemicity of chronic Epstein-Bar virus infections in this region. It is more prevalent in East Asia and South-and Central America [1].

In over 80% to 90% of cases, it mainly affects the nasal cavity and the nasopharynx [4]. It may also primarily or secondarily involve extra nasal sites, like the skin. Skin involvement as the primary site is known as primary cutaneous extranodal NK/T-cell lymphoma, nasal type (PC-ENK/T-NT). Although the skin is the second most common involved site, very few cases of PC-ENK/T-NT have been reported so far [3].

In PC-ENK/T-NT, clinical features reported in the literature are variable. The most frequently observed cutaneous lesions are usually subcutaneous nodules, erythematous papules, infiltrative plaques. They are often ulcerated, commonly located on the trunk or extremities [5-8]. They also can mimic cellulitis, panniculitis, fasciitis and facial swelling [1,9]. Our case is unusual as it presented with large multinodular and ulcerated tumefaction on leg at the onset and only developed nasal and lymph nodes involment at a later stage. This unusual clinically presentation may lead to misdiagnosis, including large B cell lymphoma of leg type and other cutaneous malignancies (like squamous cell carcinoma). This presentation of ENTCL resembling a primary cutaneous large B-cell lymphoma of leg type is extremely rare [1,4-6,10].

In case of a such presentation, the careful recognition of marked angiocentric disposition and presence of infarctive-type necrotic areas should raise the suspicion of PC-ENK/T-NT [2,11]. But, such typical histopathological findings of ENTCL are known to be less frequent in skin lesions, as it was interestingly in our case [4]. Immunohistochemical analysis is also essential for diagnosis. The most common immunophenotype is CD2+, CD56+, surface CD3, and cytoplasmic CD3ε intracytoplasmic+ [6].

Furthermore, lymphoma cells are infected by Epstein-Barr virus (EBV). Then, the *in-situ* hybridization for EBV-encoded RNA is critical for the diagnosis. The prognosis of this entity is usually poor in spite of treatment and most patients die within few months. As in our case, distant dissemination on the nasal cavity in the course of an extra nasal ENK has been well documented [11].

Several studies have indicated that the efficacy of CHOP against ENTCL is inadequate [12]. In the present case, the CHOP regimen which was the only available in our countries was used.

Conclusion

We reported a case of a PC-ENK/T-NT remarkable by its presentation simulating a B-cell lymphoma of leg type.

The increasingly reported number in the literature of PC-ENK/T-NT presented as ulcerated tumor of the leg should logically lead to isolate a new sub entity of ENKTCL of leg type.

Conflict of Interests

None

References

1. Lee WJ, Jung JM, Won CH, Chang SE, Choi JH et al. (2014) Cutaneous extranodal natural killer/T-cell lymphoma: A comparative clinico histopathologic and survival outcome analysis of 45 cases according to the primary tumor site. J Am Acad Dermatol 70: 1002-1009.
2. Peck T, Wick MR (2015) Primary cutaneous natural killer/T-cell lymphoma of the nasal type: A report of 4 cases in North American patients. Ann Diagn Pathol 19: 211-215.
3. Felcht M, Varelis G, Marx A, Klemke CD (2014) Progressive nodules on the right shin of a 74- year-old woman. J Dtsch Dermatol Ges 12: 358-361.
4. Santucci M, Pimpinelli N, Massi D, Kadin ME, Meijer CJ, et al. (2003) Cytotoxic/natural killer cell cutaneous lymphomas. Cancer 97: 610-627.
5. Vasconcelos PD, Ferreira C, Soares-Almeida L, Filipe P (2016) Multifocal primary cutaneous extranodal NK/T lymphoma nasal type. An Bras Dermatol 91: 219-221.
6. Little AJ, Subtil A, Choi J (2014) Rapidly enlarging nodular plaque on the leg. JAMA Dermatol 150: 1107-1108.
7. Liaw TY, Hu SC (2016) Nasal-type extranodal natural killer/T-cell lymphoma

presenting as a solitary non-healing lower leg ulcer. Australas J Dermatol 57: e17-19.

8. Chia HY, Tey HL, Tan KB, Chong WS (2009) Nasal-type extranodal natural killer/T-cell lymphoma presenting with extensive leg ulcers. Clin Exp Dermatol. 34: e693-295.

9. Park GH, Won CH, Chang SE, Lee MW, Choi JH (2016) Extranodal natural killer/T-cell lymphoma preceded by persistent hemifacial swelling. Int J Dermatol 55: 450-452.

10. Stokkermans-Dubois J, Jouary T, Vergier B, Delaunay MM, Taieb A (2006) A case of primary cutaneous nasal type NK/T-cell lymphoma and review of the literature. Dermatology 213: 345-349.

11. Moon IJ, Kang HJ, Lee MW, Lee WJ (2017) Primary cutaneous extranodal natural killer/T-cell lymphoma presenting as bilateral erythematous patches on the arms. Indian J Dermatol Venereol Leprol 83: 453-456.

12. Tian C, Yu Y, Zhang Y (2016) A case report of primary cutaneous natural killer/T-cell lymphoma. Mol Clin Oncol 5: 777-778.