Cutis verticis gyrata in a 24 year-old young man revealing a T-cell lymphoblastic lymphoma

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Abstract

T-cell lymphoblastic lymphoma (T-LBL) is frequently revealed by a mediastinal mass or peripheral lymphadenopathy. Skin lesions in T-LBL usually present as multiple nodules associated with multiple peripheral lymphadenopathy and bone marrow invasion. Our patient is particular by the revealing presentation of the lesions as Cutis verticis gyrate.

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Case Report

Key clinical message : T-cell lymphoblastic lymphoma (T-LBL) is frequently revealed by a mediastinal mass or peripheral lymphadenopathy. Skin lesions in T-LBL usually present as multiple nodules associated with multiple peripheral lymphadenopathy and bone marrow invasion. Our patient is particular by the revealing presentation of the lesions as Cutis verticis gyrate.

Key Words : Cutis verticis gyrate, Malignant hemopathy, T-cell lymphoblastic lymphoma

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INTRODUCTION

Cutis verticis gyrate (CVG) or "washboard" pachydermia is a specific semiological aspect of the scalp and forehead characterized by hypertrophy and redundancy of the integument forming "waves" separated by deep folds. Secondary CVG have a wide variety of causes: genodermatoses, endocrinopathies, overload disease, paraneoplastic or tumor diseases. Lymphoblastic lymphoma (LBL) is a rare neoplasm with a poor prognosis.[1] We describe a case of CVG revealing a T-cell lymphoblastic lymphoma (T-LBL) in a young man.

CASE REPORT

A 24 year-old man presented to our dermatology department for an asymptomatic deep rippling lesions on the frontal level of the scalp evolving for 2 months. He had no past medical history. Dermatology examination revealed multiple large nodules on deeply infiltrated erythematous-violaceous skin limited to the frontal area of the scalp resulting in folds that mimick the surface of the cerebral cortex (Figure1A). Dermoscopy revealed dotted vessels, fine short linear vessels and scales over a salmon-pink background (Figure 1B). Physical examination revealed no locoregional adenopathies. Histopathologic examination revealed a dense infiltration in the dermis, made by medium sized immature lymphoid cells, with a high mitotic index (Figure 2A). There was no epidermotropism. Immunohistochemical stains were positive for terminal deoxynucleotidyl transferase (TdT), CD3, CD4 and CD8 and negative for CD20 (Figure 2B, 2C). The diagnosis of secondary CVG associated with T-LBL was made. The patient's general condition worsened within a few days and he started complaining of debilitating dyspnoea and productive cough. The patient underwent computed tomography of the chest, abdomen and pelvis, which revealed a large mediastinal mass measuring 15.5cm*10cm*22cm associated with pleural and pericardial effusions as well as multiple bilateral renal masses. A transthoracic needle biopsy was performed, confirming the diagnosis of pulmonary involvement with T-cell lymphoblastic lymphoma. The patient received chemotherapy.

DISCUSSION

T-cell lymphoblastic lymphoma is a very rare form of non-Hodgkin lymphoma, accounting for approximately 20% of LL.[1,2]It commonly occurs in young male adults, more than children or elderly.[2] T-LBL is frequently revealed by a mediastinal mass or peripheral lymphadenopathy. Skin localizations of all LBL are noted in fewer than 20% of affected patients.[3] Unlike B-cell LBL (B-LBL), skin involvement concerns only a minority of patients with T-LBL, as well as lymph node involvement. To our knowledge, only 15 cases of T-LBL with cutaneous involvement have been reported in the literature.[1,4,5]Skin lesions mostly present as nodular lesions, located on the scalp, the trunc and the limbs. Skin lesions in T-LBL usually present as multiple nodules associated with multiple peripheral lymphadenopathy and bone marrow invasion. Our patient is

particular by the revealing presentation of the lesions as CVG without other distant lesions. Only four cases of T-LBL diagnosed by cutaneous involvement have been reported so far.[1]

To our knowledge, secondary CVG revealing T-LBL has not been reported previously.

Dermoscopic features of skin lésions are not specific for LBL.[6] However, they are comparable to that primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorders presenting as short fine irregular or serpiginous vessels over salmon-pink background. The authors correlate the presence of pinkish background with atypical lymphocytic infiltrates related angiogenesis.[6]

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Legends:

Figure 1: Clinical and dermoscopic presentation:

A: Multiple large nodules on deeply infiltrated erythematous-violaceous skin limited to the frontal area of the scalp resulting in folds.

B: Dermoscopy: dotted vessels (white circle), fine short linear vessels (green circle) and scales (blue circle) over a salmon-pink background.

Figure 2: Histology and immunohistochemistry:

A: Dense infiltration in the dermis, made by medium sized immature lymphoid cells, with a high mitotic index, (H&E 400).

B,C: Immunohistochemistry: positive for CD3 (x200) and TdT (x100).

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