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CASE REPORT

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

Sarra Saad[1] | **Nadia Ghariani Fetoui**[1] | **Jacem Rouatbi**[2] | **Sana Mokni**[1] | **Najet Ghariani**[1] | **Baderedine Sriha**[2] | **Mohamed Denguezli**[1]

[1]Department of Dermatology, Farhat Hached University Hospital, Sousse, Tunisia

[2]Department of Anatomopathology, Farhat Hached University Hospital, Sousse, Tunisia

Correspondence
Sarra Saad, Department of Dermatology, Fathat Hached University Hospital, Sousse, Tunisia.
Email: drsaadsarra@gmail.com

Abstract

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

KEYWORDS

cutis verticis gyrata, Dermoscopy, scalp, skin nodules, T-cell lymphoblastic lymphoma, TdT

1 | INTRODUCTION

Cutis verticis gyrata (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. CVG may be classified into primary (essential and non-essential) and secondary types.[1] Primary CVG is rare, characterized by the absence of neurological and ophthalmological changes and by exclusion of secondary causes of the disease. Secondary CVG has a wide variety of causes: genodermatoses, endocrinopathies, overload disease, neoplastic or paraneoplastic disease.[1] The pathogenesis in this case is related to the underlying pathology. Lymphoblastic lymphoma (LBL) is a rare neoplasm with a poor prognosis.[2] We describe a case of secondary CVG revealing a T-cell lymphoblastic lymphoma (T-LBL) in a young man.

2 | CASE REPORT

A 24-year-old man presented to our dermatology department with 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 (Figure 1A). Dermoscopy revealed dotted vessels, fine short linear vessels, and scales over a salmon-pink background (Figure 1B). Physical examination revealed no locoregional adenopathies and the general condition of the patient was good. Histopathologic examination of scalp nodule biopsy 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 (IHC) stains were

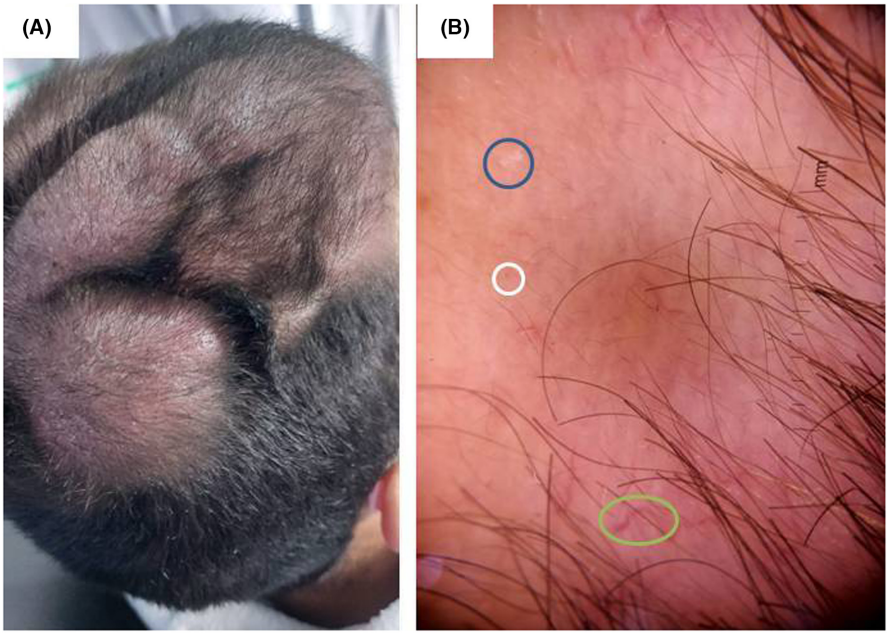

FIGURE 1 Clinical and dermoscopic presentation: (A) Multiple large nodules on deeply infiltrated erythematous-violaceous skin limited to the frontal area of the scalp resulting in folds. (B) Dermoscopy: dotted vessels (white circle), fine short linear vessels (green circle), and scales (blue circle) over a salmon-pink background.

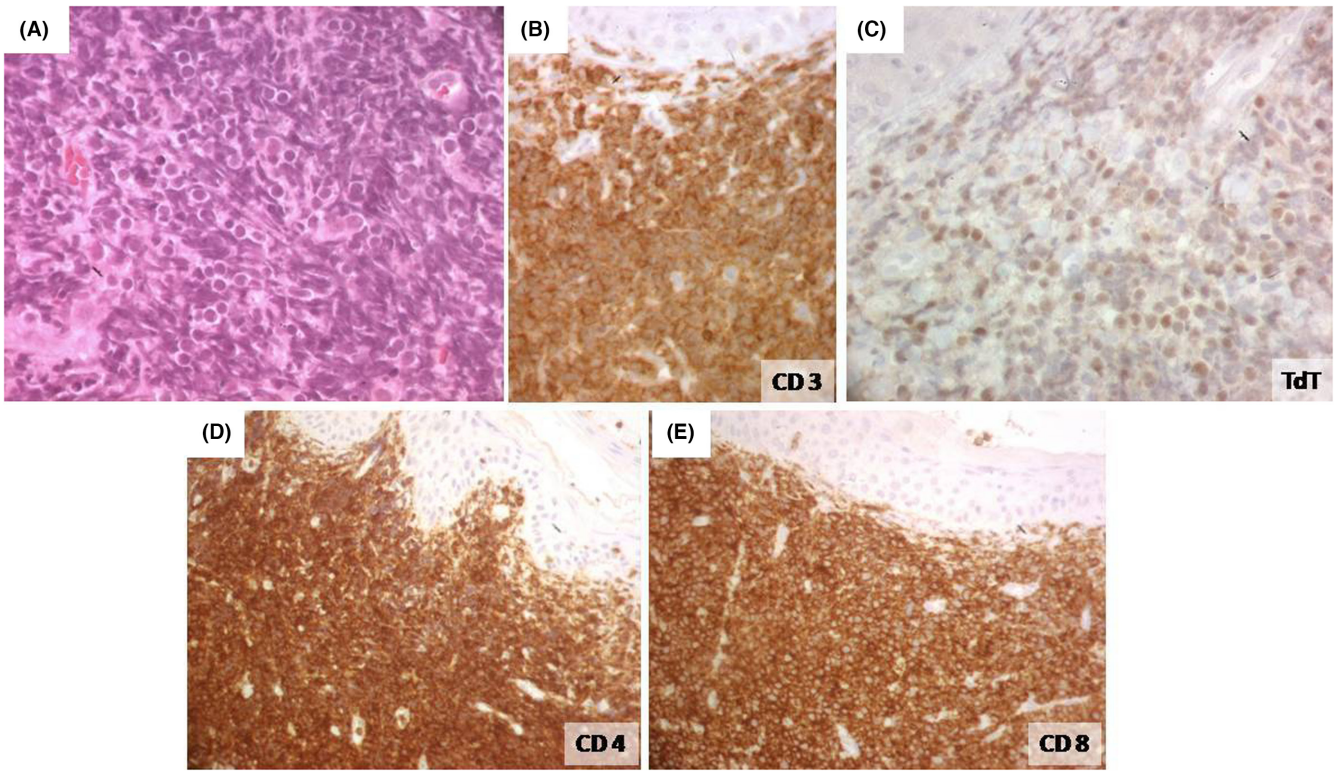

FIGURE 2 Histology and immunohistochemistry: (A) Dense infiltration in the dermis, made by medium sized immature lymphoid cells, with a high mitotic index, (H&E 400). (B–E) Immunohistochemistry: positive for CD3 (x400), TdT (x200), CD4 (x200) and CD8 (x200).

positive for terminal deoxynucleotidyl transferase (TdT), CD3, CD4, and CD8 and negative for CD20 (Figure 2B–E). The biological investigations showed normochromic normocytic anemia at 9 g/dl and a normal red blood cell count, white blood cell count and platelets counts. 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 dyspnea and productive cough. The patient underwent computed tomography of the chest, abdomen, and pelvis, which revealed a large mediastinal mass measuring 15.5*10*22 cm 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. The protocol used including cyclophosphamide, vincristine, doxorubicine, dexamethasone (Decadron, Dexasone), methotrexate, and cytarabine. He is still undergoing treatment.

3 | DISCUSSION

T-cell lymphoblastic lymphoma is a very rare form of non-Hodgkin lymphoma, accounting for approximately 20% of LBL.[2,3] It commonly occurs in young male adults, more than children or elderly.[3] T-LBL is frequently revealed by a mediastinal mass or peripheral lymphadenopathy. The diagnosis of LBL can be confused with an acute lymphoblastic leukemia. In order to find the appropriate diagnosis, the analysis of the bone marrow must be made, showing a difference in the percentage of blasts. In fact, the marrow contains less than 25% of blast in the LBL.[4] Skin localizations of all LBL are noted in fewer than 20% of affected patients.[5] 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.[2,6,7] Skin lesions mostly present as nodular lesions, located on the scalp, the trunk, 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.[2,7,8] They all had erythematous and purplish nodule, that was painful or asymptomatic. The location was on the arm, on the occipital level of the scalp, on the sternal region and breasts, and on the forehead.

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

We emphasize the importance of dermoscopy in our patient who was in favor of a secondary origin of CVG. Dermoscopic features of skin lesions are not specific for LBL.[9] 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.[9]

T-cell lymphoblastic lymphoma is a very rare disease. The clinical presentation of our patient is original by a revelation of the disease by an unusual cutaneous localization. The diagnosis of CVG is usually clinical, although multiple investigations, such as dermoscopy, histopathological examination, and IHC stainging of the skin biopsy as well as blood tests can be performed to distinguish primary from secondary forms of CVG.

AUTHOR CONTRIBUTIONS

Dr Saad and Dr Ghariani Fetoui examined the patient and contributed to the first draft of the manuscript. Dr Mokni and Dr Ghariani made the diagnosis and helped in writing the manuscript and literature search. Dr Rouatbi and Dr Sriha contributed to the histological data. Dr Denguezli revised and approved the final version of the manuscript. All the authors contributed to and have approved the final manuscript.

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None.

DATA AVAILABILITY STATEMENT

Data sharing not applicable - no new data generated

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

ORCID

Sarra Saad https://orcid.org/0000-0002-7637-635X
Nadia Ghariani Fetoui https://orcid.org/0000-0001-8556-7919
Jacem Rouatbi https://orcid.org/0000-0003-3795-0846

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