

Cutis Verticis Gyrate Secondary to Acute Monoblastic Leukemia

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A 64-year-old man affected by acute monoblastic leukemia developed a cutis verticis gyrata during the terminal phase of hemopathy. The association between these two diseases is rare. The classification of cutis verticis gyrata in primary essential, primary non-essential and secondary forms is reviewed. Performing a skin biopsy is necessary in the diagnostic approach to patients with cutis verticis gyrata. Key words: Scalp; Specific skin lesions.

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Skin lesions in patients suffering from acute myelogenous leukemia (AML) are often reported. In acute monoblastic leukemia (M 5), specific cutaneous manifestations occur in 10–50% of the cases (1). Clinically, they are brownish-red patches or violaceous nodules localized on the trunk and limbs. Scalp and mucosal surfaces may also be affected. Cutaneous involvement usually develops during the medial or terminal phase of the disease, even though leukemic infiltration of the skin may occasionally precede hematological diagnosis.

We report a patient with AML who developed both specific nodular cutaneous lesions on the trunk and limbs and typical cutis verticis gyrata (CVG) on the occiput scalp.

CASE REPORT

In August 1991, a 64-year-old male suffering from an acute monoblastic leukemia (M 5 according to F.A.B.) was admitted to the Geriatric Ward of the Imola General Hospital.

The laboratory data showed a hyperleukocytosis (WBC $33.6 \times 10^9/l$; differential count: 70% neutrophils, 7% lymphocytes, 6% myelocytes, 4% metamyelocytes and 4% blasts cells) and a thrombocytopenia (platelets $18 \times 10^9/l$). The hemoglobin level was normal.

Bone marrow biopsy, Jamshidi-needle biopsy and cytochemical stainings were performed. The results obtained were characteristic of acute undifferentiated monoblastic leukemia (M 5A). Cytogenetic abnormalities were represented by trisomy 8.

Skin examination showed brownish-red patches and nodules, sometimes overlapping to form indurated plaques, on the trunk and limbs.

Microscopic examination of a skin biopsy sample showed perivascular and periadnexal infiltrates extending into the dermis and subcutaneous tissue. These infiltrates consisted of wide monomorphic cells with large kidney-shaped folded nuclei and with a fairly eosinophilic cytoplasm.

Immunohistochemical examination of the cutaneous infiltrate was performed. The myelomonoblastic cells were CD68 positive and partially CD68R positive.

On the basis of morphologic and immunohistologic findings, a diagnosis of myelomonoblastic leukemia with cutaneous involvement was made.

Cutaneous lesions cleared after chemotherapy and flared up during relapses. Chemotherapy was administered in 4 cycles during a 6-month period, but no complete remission was obtained, neither in the peripheral blood nor in the bone marrow.

Fifteen days after the end of the 4th cycle of chemotherapy with Idarubicin (20 mg/die for 3 days), Cytarabine (1.700 mg/die for 5 days) and Ethoposide (200 mg/die for 5 days), the patient presented high bone marrow suppression with a very low blood cell count (WBC $0.3 \times 10^9/l$; platelets $8.0 \times 10^9/l$; hemoglobin 9.5 g/dl).

After hair loss, the skin of the occipital scalp revealed parallel and predominantly vertical folds as well as deep furrows with thin and short hairs. The clinical aspect resembled the outer surface of the cerebrum, suggesting a diagnosis of CVG.

The performance of a cutaneous biopsy on the scalp was delayed for 2 weeks, due to the seriousness of the clinical conditions and the ease with which bleeding was induced by thrombocytopenia.

The histological examination of the biopsic specimen showed the presence of a thick infiltrate consisting of large cells with irregular nuclei and fairly eosinophilic cytoplasm in the dermis and hypoderma. The histologic and immunohistochemical pattern was that of a specific dermo-hypodermic infiltrate in AML (M 5).

The patient's condition deteriorated and cutaneous plaques and nodules quickly spread to all skin areas. The death of the patient was due to infective pneumonia.

DISCUSSION

CVG is a descriptive term for a rare condition of the scalp, characterized by skin folds and deep furrows that mimic the outer surface of the cerebrum, typically localized between the vertex and the occiput. The folds are predominantly parallel and vertical. Hair appears more dense in furrows than on folds (2).

CVG is classified in three forms: a primary essential CVG, a primary non-essential CVG and a secondary CVG (3, 4).

Primary CVG usually presents symmetrical folds and has a post-puberty onset. The histopathology shows thickened connective tissue bundles and hypertrophy or hyperplasia of the hair follicles and sebaceous glands (3). CVG may be present with no associated abnormalities (the essential primary form) or, as is more often seen, combined with neuro-psychiatric conditions such as mental retardation, cerebral palsy, epilepsy, chronic schizophrenia, ophthalmologic abnormalities (the non-essential primary or symptomatic form).

Secondary CVG presents asymmetrical folds and may have its onset at any age (5). It depends on a local underlying process such as an inflammatory disease (eczema, psoriasis, folliculitis, impetigo, pemphigus) or a neoplasia (naevi, neurofibromas and other tumours). Why an infiltrate below the epidermis produces folds and furrows resembling the cerebrum surface is not known. Some authors suggest that it may be due to the anatomy of the scalp and its attachments (5). CVG can also occur as part of a systemic disease such as pachydermoperiostosis, acromegaly, myxedema, diabetes, amyloidosis and tuberous sclerosis.

In secondary CVG, the histopathological findings are typical of the underlying disease.

In the literature CVG is reported in association with malignant tumours (6–8). Some authors have suggested that CVG may be a paraneoplastic syndrome in these cases (7).

Up to now, only one other patient with CVG associated with acute leukemia has been reported (3).

In our case the CVG developed in the terminal phase of an AML (M 5). The skin biopsy showed leukemic infiltration in the skin of the occipital scalp, suggesting that it was a CVG that came secondary to a local underlying process.

In accordance with the case described we recommend that a skin biopsy should always be performed to identify the etiology of CVG.

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