Langerhans Cell Histiocytosis over Dilated Skin Vessels

D. Bessis
A. Sotto
G. Barnéon
A.-J. Ciurana
J.-J. Guilhou

Service de Dermatologie-Phlébologie, Hôpital Saint-Charles, et Service de Médecine Interne A, Hôpital Saint-Eloi, Montpellier, et Service de Médecine Interne B, Hôpital Carémeau, Nîmes, France

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Prof. Jean-Jacques Guilhou, Service de Dermatologie-Phlébologie, Hôpital Saint-Charles, 300, rue Auguste-Broussonnet, F-34295 Montpellier Cedex 5 (France)

Cutaneous lesions of Langerhans cell histiocytosis (LCH) in the adult are characterized by clinical polymorphism and usually present as scaly or crusted papules, sometimes yellow-brown and resembling seborrhea. The eruption commonly involves the scalp, axillae and chest. Other locations, such as the inguinal and retroauricular regions, face, neck and limbs, have been observed [1]. We describe the case of

Fig. 1. Brownish papular LCH lesions (arrows) strictly overlying the portosystemic collateral vessels on the abdominal wall.

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a patient with portal hypertension suffering from LCH with unusual skin lesions, mainly located over the portosystemic collateral vessels on the abdominal wall.

A 56-year-old woman, a chronic alcoholic, was hospitalized in August 1993 for erysipelas of the left leg. Her medical history revealed that at the age of 38 years she had developed diabetes insipidus treated with vasopressin. At 43 years, she presented a chronic sub-mammary and inguinal intertrigo which was not biopsied. At 54 years, an ascites revealed alcoholic cirrhosis; the liver biopsy showed micro-nodular cirrhotic lesions without any histiocyte infiltration. At that time, clinical examination revealed left-side hemiplegia and bilateral exophthalmus. Cerebral CT localized 3 intracranial tumors; cerebral magnetic resonance imaging showed infiltration of the retroorbital cavities and of the cavernous and sphenoid sinuses; X-rays of the inferior femoral and superior tibial metaphyses showed symmetrical medullary sclerosis, associated with lytic and pagetoid cortical lesions; a CT scan showed pulmonary cysts, bilateral thickening of the pleurae as well as infiltration of the perirenal space, renal capsule and adrenal glands. Histological examination of a bone lesion and the greater omentum demonstrated dense histiocytic infiltrates surrounded by a few lymphocytes and rare eosinophils. She was treated with prednisolone 40 mg (0.5 mg/kg) daily for 2 years without progression of visceral lesions. At the time of admission in 1993, apart erysipelas on her left leg, physical examination revealed intertrigo of the submammary, inguinal, subaxillary, gluteal and navel areas with seborrhea-like lesions on the scalp; crusted brown maculopapular lesions of progressive appearance were seen on her abdomen that were strictly overlying the portosystemic collateral vessels (fig. 1).

Hepatomegaly and splenomegaly were present. Laboratory investigations gave the following results: hemoglobin, 10.7 g/dl; white blood cell count, 14,300/µl (87% neutrophils); creatininemia, 27.5 mg/l (normal, 7–15 mg/l); uremia, 1.10g/l (normal, 0.18–0.40 g/l); alanine aminotransferase, 28 U/l (normal, 5–40 U/l); aspartate aminotransferase, 17 U/l (normal, 5–55 U/l); γ-glutamyl transpeptidase, 1,414 U/l (normal, 5–80 U/l); alkaline phosphatase, 451 U/l (normal, 30–125 U/l). The prothrombin time and total bilirubin level were normal.

Histological examination of both abdominal and intertriginous skin lesions showed a dense dermal infiltration of atypical large histiocytes with numerous eosinophils and lymphocytes. Immuno-histochemical studies indicated that histiocytes were S-100 protein and CD 1α positive. Electron microscopy confirmed the presence of Langerhans cells (LC) exhibiting intracytoplasmic Birbeck granules in the superficial dermis infiltrate that extended to the epidermis. Eight days after admission, the patient died of hepatorenal failure. The family denied permission for an autopsy.

To the best of our knowledge, the unusual localization of the LCH skin lesions, overlying the abdominal wall portosystemic collateral circulation in portal hypertension, has never been described in the literature. Normal LC have been demonstrated to be of bone marrow origin and are thought to migrate via the bloodstream to the epidermis [2]. Dezutter-Dambuyant et al. [3] detected that OKT6-positive cells in blood resembled LC by being T6 positive and dendritic, although they lacked Birbeck granules, supporting the existence of LC circulating in blood. Accordingly, unlike the LC of healthy skin, histiocytes in LCH express ICAM-1 and β3- and β2-integrins, suggesting that they have an adhesive capacity and an increased ability to migrate across the vascular endothelium [4, 5].

The combination of anatomical modifications of the venous network, perturbations of portal vein hemorheology with increased blood viscosity during portal hypertension [6] and the enhanced histiocyte migratory capacity during the course of LCH could explain our patient’s unusual distribution of cutaneous lesions.
References

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