Acknowledgments

We thank Dr Miguel Ángel Marín Cárdenas of the Radiodiagnostic Unit at Hospital Universitario Miguel Servet in Zaragoza, Spain, for providing us with the magnetic resonance imaging report and images.

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Poliosis and Status Epilepticus as the Presentation of Tuberous Sclerosis in an Infant

Poliosis y estatus epiléptico en un lactante como presentación de esclerosis tuberosa

To the Editor:

Tuberous sclerosis (TS) is a neurocutaneous syndrome with an autosomal dominant pattern of inheritance and a high penetrance. It is characterized by the appearance of hamartomatous lesions in several organs,[1] and the most usual clinical signs of this disease are skin lesions, epilepsy, learning difficulties,[2] and behavioral disorders.

We report the case of a 51-day-old infant, born at term of nonconsanguineous healthy parents after an uncomplicated pregnancy and delivery, who was seen in the pediatric emergency department due to persistent convulsive seizures that had lasted 3 days. Physical examination revealed the presence of lanceolate hypopigmented macules on the limbs and trunk that were 3 to 11 mm in diameter (Figure 1). Of particular interest was the presence of poliosis of the medial half of the left upper eyelid and a hypopigmented macule at the medial angle of the eye (Figure 2), which were present at birth. The clinical suspicion of tuberous sclerosis led us to perform cerebral magnetic resonance imaging, which revealed multiple cortical tubers (Figure 3), and an

Figure 1 Lanceolate hypopigmented macule on the thigh.

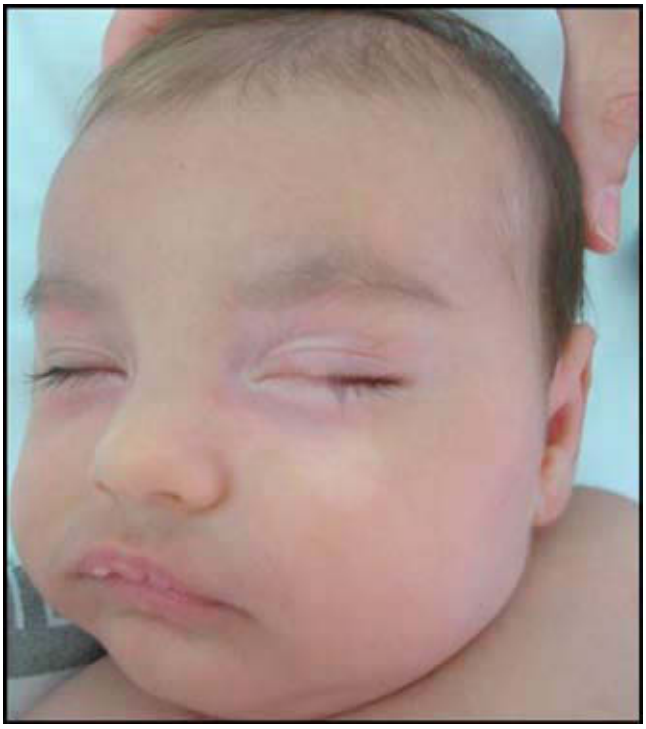

Figure 2 Palpebral poliosis and oval hypopigmented patch.

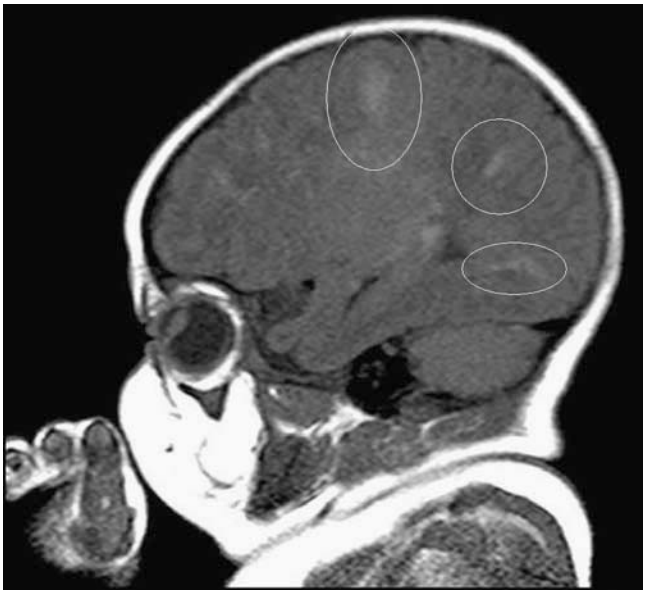

Figure 3 Sagittal magnetic resonance image of the brain showing multiple cortical tubers.

electroencephalogram, which revealed irritative foci. The other diagnostic tests, including fundus examination, were normal. The initiation of treatment with valproic acid and vigabatrin controlled the symptoms and was tolerated well by the patient. The study of the parents was negative.

The diagnosis of tuberous sclerosis is based on major and minor criteria, many of which are skin manifestations.

The earliest skin manifestation of tuberous sclerosis is the presence of lanceolate, hypopigmented patches, which are found in 90% of affected infants under 2 years of age.[3] The pigment is decreased due to a reduced size and number of melanosomes.[1,3] The patches are usually multiple and lanceolate with irregular edges, measuring 1 to 3 cm and distributed on the trunk and limbs.[1] Areas of terminal hair may be involved, leading to poliosis, as in the case presented. Confetti lesions, measuring 1 to 3 mm and distributed symmetrically on the limbs, are another type of hypopigmented macule that is associated with tuberous sclerosis, although they usually appear in the second decade of life.[3] The remaining skin manifestations of the disease appear later. Facial angiofibromas and shagreen plaques in the sacral region appear in early childhood (2-5 years),[3] periungual fibromas in adolescence,[1,3] and fibrous plaques on the forehead usually in early childhood, although the latter may appear at any age.[3]

The noncutaneous clinical manifestations of tuberous sclerosis, which determine morbidity and mortality, are secondary to the development of hamartomas in various organs.[1,2] Of these, the most important are neurological (85% of patients have their first episode of epilepsy in the first 2 years of life,[3] as occurred in our patient), cardiac, and renal manifestations. Though cardiac and renal tumors (rhabdomyoma and angiomyolipoma, respectively) may lead to premature death, recent studies show that they regress in childhood or early adolescence.[2] These hamartomatous lesions also appear as polyps in the colon and as retinal phakomas.[2,3]

A multidisciplinary approach is therefore of great importance in the assessment of patients with this genodermatosis.[1,3] Early detection of tuberous sclerosis is essential in order to initiate appropriate treatment and offer genetic counseling.[4] Ash-leaf macules are a very useful diagnostic sign as they are present in almost all patients. This sign is present at birth in two-thirds of cases, although it is not always easy to detect at this age, especially in fair-skinned children. We report a case in which the typical hypopigmented ash-leaf macules were associated with poliosis, a sign that has been reported little in the literature to date. Other diseases characterized by depigmentation of the hair and skin should be considered in the differential diagnosis. These include piebaldism, an autosomal dominant disease that presents at birth and whose main clinical manifestation are areas of leukoderma in a characteristic distribution associated with poliosis; Waardenburg syndrome, an inherited disease that, in addition to achromic spots, presents with characteristic ocular and phenotypic alterations; and other diseases such as albinism and those included in white forelock syndrome.[5]

In conclusion, we believe that poliosis in tuberous sclerosis, reported in only 1 of the articles we reviewed, is a more common sign than is indicated in the literature. It should also be taken into account that it can be the first sign of this disease.[4]

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Erythema Elevatum Diutinum or Extrafacial Granuloma Faciale?

¿Eritema elevatum diutinum o granuloma facial extrafacial?

To the Editor:

Erythema elevatum diutinum (EED) and granuloma faciale (GF) are rare diseases. EED presents with multiple plaques or nodules typically located on extensor areas,[1,2] whereas GF is usually a single lesion in the facial region.[3] However, differential diagnosis is sometimes difficult due to clinical and pathological similarities.[4]

We report the case of a 57-year-old man who came to our department because of the appearance at least 15 years earlier of an asymptomatic brownish plaque of soft consistency on the anterior aspect of the left thigh; the plaque had grown slowly but progressively, becoming harder and darker (Figure 1). A further 3 brown, plaque-like lesions with an atrophic appearance had developed years later on the back of the right leg, on the left thigh, and over the left iliac crest; the patient said they were similar to the early appearance of the initial lesion. There were no systemic symptoms. The biopsy of the nodular lesion (Figures 2 and 3) showed fibrosis and perivascular interstitial infiltrates formed of lymphocytes, plasma cells, and neutrophils with karyorrhexis, and vascular proliferation with intraluminal fibrin thrombi. Laboratory tests revealed an immunoglobulin (Ig) A concentration of 384 mg/dL (normal range, 100-300 mg/dL) and IgM of 78.9 mg/dL (normal range, 80-250 mg/dL), as well as a ferritin of 402 ng/mL (normal range, 30-400 ng/mL), the high level of which led us to perform a study to exclude hemochromatosis. Protein electrophoresis, other laboratory values, electrocardiogram, and chest radiograph were normal. Serology for hepatotropic viruses and human immunodeficiency virus was negative. In agreement with the patient, no treatment was applied.

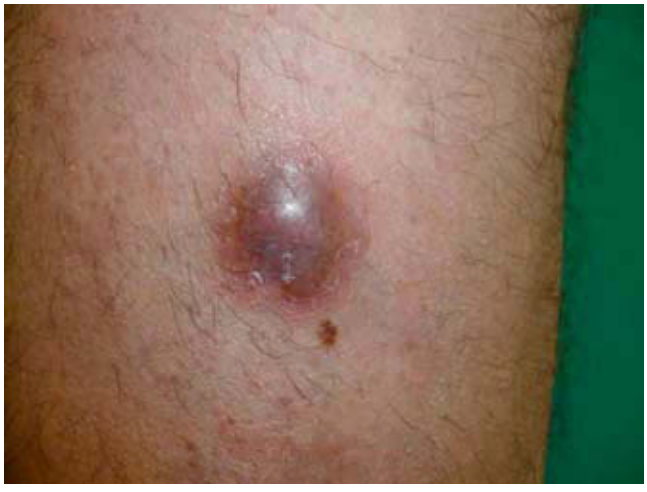

Figure 1 Well-defined brown nodule of 2 cm diameter on the anterior aspect of the left thigh, adherent to the superficial tissues.

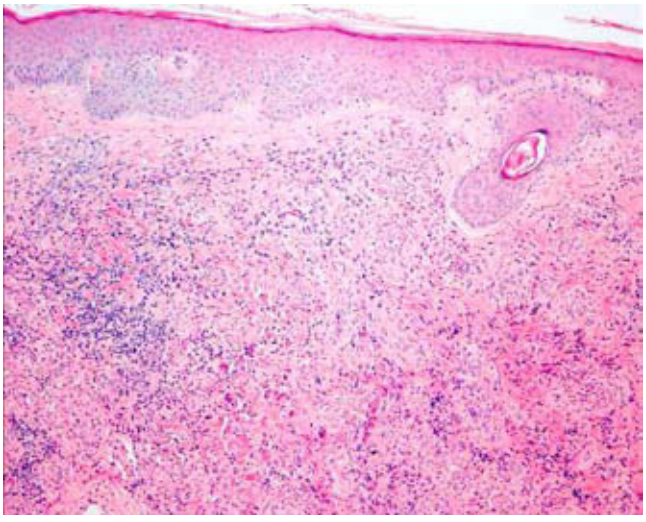

Figure 2 Mixed infiltrate with abundant neutrophils in the dermis, sparing the most superficial part of the papillary dermis (Grenz zone) (hematoxylin-eosin, original magnification ×100).