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CASES FOR DIAGNOSIS

Asymptomatic Facial Papules

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Clinical History

The patient was a 15-month-old girl with no personal or family history of interest. She was seen in the dermatology outpatient clinic for a 2-month history of asymptomatic lesions on the frontal region of the head; there had been no improvement after topical corticosteroid treatment, and the lesions had increased in number.

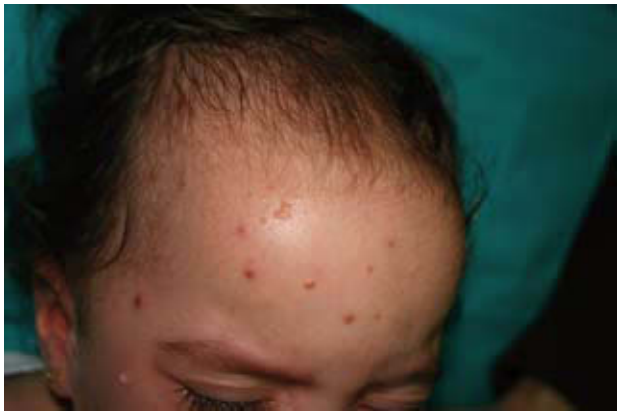

Figure 1.

Physical Examination

Physical examination revealed a number of flat, brownish-yellow papules on right lateral part of the frontal region of the head (Figure 1). There were no associated general symptoms, and the Darier sign was negative. There was no family history of interest.

Complementary Tests

Biopsy was performed of one of the lesions: in the superficial dermis, beneath a normal epidermis, there was a diffuse proliferation of histiocytic cells with clear, foamy cytoplasm; in some areas Touton giant cells could be identified (Figure 2). There was also a moderate degree of vascular proliferation. Immunohistochemistry was positive for S100, CD68, and for CD34 (in the vessels) and was negative for CD1a. No further laboratory tests were requested.

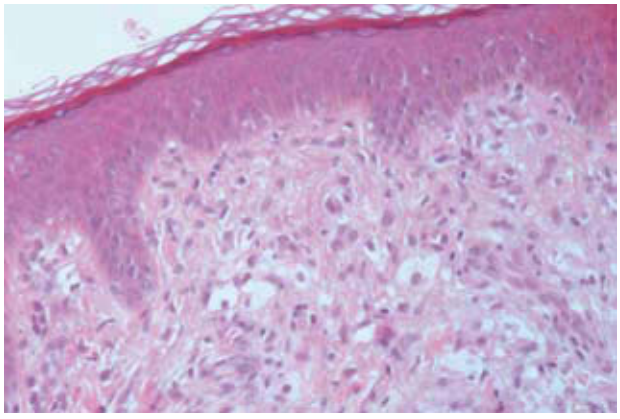

Figure 2. Hematoxylin-eosin, ×100.

What Was the Diagnosis?

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Diagnosis

Benign cephalic histiocytosis.

Clinical Course and Treatment

The patient is being followed up periodically and there has been no change in the size or number of the lesions.

Discussion

Benign cephalic histiocytosis is a rare, benign non-X histiocytosis, that presents clinically as asymptomatic papules on the scalp of children and resolves spontaneously.[1] Systemic involvement is uncommon, although there has been a report of a case in which it was associated with diabetes insipidus,[2] another associated with osseous Langerhans cell histiocytosis,[3] and a third with insulin-dependent diabetes mellitus.[4]

Since it was first described by Gianotti, Caputo, and Ermacora in 1971,[2] only 40 cases have been published in the literature.[3]

Clinically it presents with flat, brownish-yellow or reddish-brown maculopapules of 1 to 8 mm in diameter.[1] Lesions can also appear on the neck, trunk, limbs, and even in the pubic region; the muscosas and acral regions are always respected.[1-6]

According to Jih et al,[1] the mean age at appearance of the lesions is 15 months, and the incidence is identical in boys and girls.

Of the 40 cases published, 18 have shown initial signs of resolution and a further 10 presented complete resolution after a mean of 50 months.

Histologically there are 3 possible patterns of infiltration (papillary dermal, lichenoid, and diffuse), and most commonly the infitrate is in the papillary or superficial dermis.[1] Immunohistochemistry is characteristic but not diagnostic; it is positive for factor XIIIa, OKM1, and Leu-3, and is negative for the Langerhans cell marker, CD1a. Protein S100 is usually negative, but can occasionally be weakly positive.[1,6]

Electron microscopy is specific but not pathognomic, with structures similar to desmosomes, cytoplasmic comma-shaped bodies, and vermiform particles, and no Birbeck granules.

The differential diagnosis includes flat warts, multiple Spitz nevus, juvenile xanthogranuloma, Langerhans cell histiocytosis, urticaria pigmentosa, generalized eruptive histiocytosis, and lichenoid sarcoidosis.[1]

Flat warts, Spitz nevus, and lichenoid sarcoidosis are differentiated through histological study.

The micronodular form of juvenile xanthogranuloma presents with papules that grow to form larger nodules before undergoing spontaneous involution; the nodules have a wider distribution.

Langerhans cell histiocytosis preferentially affects the flexor surfaces and presents with a crusted, desquamating, papular rash. It may also affect internal organs and cause fever or general malaise. Immunohistochemistry is positive for S100 and CD1a.

Urticaria pigmentosa characteristically presents a positive Darier sign and a mast cell infiltrate on histological study.

Generalized eruptive histiocytosis is more common in adults and the lesions have a wider distribution.

Although the etiology of the benign cephalic histiocytosis is unknown, it is believed that generalized eruptive histiocytosis and the juvenile xanthogranuloma are different clinical manifestations of the same types of disease.[3]

We add a new case of benign cephalic histiocytosis to those already published in the literature.

Conflicts of Interest

The authors declare no conflicts of interest.

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