A 5-year-old Caucasian girl presented with a 6-month history of asymptomatic macules on her cheeks (Fig. 1). She had been treated with terbinafine hydrochloride 1% cream, hydrocortisone butyrate 0.1% cream and a course of amoxicillin-clavulanic acid with no improvement. Clinical examination showed multiple, scattered, rounded 2- to 4-mm in diameter macules localized on the cheeks. The lesions had regular orange-brown pigmentation. No other mucosal or cutaneous lesions were noted, and the child was otherwise well with no relevant past medical history. In particular, she had no constitutional symptoms and no evidence of lymphadenopathy or hepatosplenomegaly.

A complete blood count, erythrocyte sedimentation rate, a comprehensive metabolic panel, abdominal ultrasound scan and systemic skeletal radiographic survey showed no abnormalities. A skin biopsy was performed for histological examination with haematoxylin and eosin staining (Fig. 2A), integrated with immunohistochemical staining with CD68 (Fig. 2B), S-100 (Fig. 2C) and CD207 (Lang-erin, Fig. 2D).

What is your diagnosis? See page 558 for answer.
DISCUSSION

Indeterminate cell histiocytosis (ICH) is a rare disorder characterized by the proliferation of histiocytic cells that express S-100 antigens, but, unlike Langerhans’ cell histiocytosis (LCH), lack Birbeck granules. The patient reported here presented with yellow-orange macules confined to the face without visceral involvement. It should be noted that the clinical picture, with the exception of the age of patients and of the lack of regression of cutaneous lesions, shares striking similarities to that of benign cephalic histiocytosis, which represents the sole clinical differential diagnosis. Histopathological differential diagnosis includes LCH, juvenile xanthogranuloma, benign cephalic histiocytosis and the cutaneous variant of Rosai-Dorfman disease (CRDD). Besides clinical considerations, LCH can be excluded because of the lack of epidermotropism, the scarcity of eosinophils infiltrate and the absence of Birbeck granules, as documented by negativity of the CD-207 (1). Histiocytes in juvenile xanthogranuloma are vacuolated, Touton cells can be observed in the infiltrate and, finally, histiocytes are, by definition, S-100 negative; the latter feature also characterizes BCH, which can therefore be readily excluded; unlike in our case, the infiltrate in CRDD has typically a nodular appearance with emperiplois and karyophagocytosis and presents a dense collection of plasma cells.

ICH usually presents with solitary (2, 3) or multiple asymptomatic, flesh-coloured to yellow-brown maculopapules with no site of predilection. According to the literature the clinical appearance of this case resembles one of the variants of NLCH (4). Although ICH has been described in childhood (2, 5) the mean age reported in a recent large case series was 46 years (4). In general, ICH has a good prognosis, being mostly exclusively a cutaneous disorder, but a fatal paediatric case has been described (6).

Various anecdotal treatment has been described as effective for this condition, including pure coal tar and 5% 5-fluorouracil cream (5), narrow-band ultraviolet B (7) and psoralen plus ultraviolet A phototherapy (8). Moreover, acute myeloblastic leukaemia was reported following chemotherapy for ICH (9). Taking into consideration the young age of our patient, and the presence of few cutaneous lesions limited to the face with no visceral involvement, we have chosen to not treat the child, but to monitor her clinical course closely, deferring therapy for potential progression of the disease.

REFERENCES