

Authors' reply

Sir,

We thank the authors for their interest in the case "Familial gigantic melanocytosis" and appreciate the opportunity to elucidate the differential diagnosis in this case.¹

Griscelli syndrome type 3 is a rare autosomal recessive disorder of hypopigmentation characterized by silvery gray hair, congenital hypopigmentation and presence of large clumps of pigment in hair shafts.² The hypopigmentation seen in Griscelli syndrome type 3 is caused by defective transport of melanosomes resulting in abnormal accumulation of melanosomes in melanocytes. Histopathology reveals increased deposition of melanosomes in melanocytes. Thus, an absence of the abovementioned clinical features made the diagnosis of Griscelli syndrome type 3 unlikely. The histopathological evidence of giant melanocytes, in addition, pointed to the diagnosis of familial gigantic melanocytosis.

Antimalarial-induced pigmentation is commonly seen following treatment with chloroquine, quinine or quinidine.³ It is characterized by bluish-gray pigmentation on the face, neck, legs and arms with involvement of nail beds in the form of diffuse or transverse melanonychia. Oral mucosa and hard palate involvement may also be seen. Histopathology of drug-induced pigmentation shows increased epidermal and dermal pigmentation with melanophages in the papillary dermis. The absence of drug intake, sparing of the oral mucosa and nails and gigantic melanocytes in the histological specimens ruled out the diagnosis of drug-induced pigmentation.

Fanconi anemia is a chromosomal instability disorder characterized by developmental defects, progressive bone marrow failure and cancer susceptibility. Easy bruising between the ages of 4 and 10 years is the most common presentation. Cutaneous pigmentation is presented in the form of generalized brownish pigmentation of the whole body, with discrete hypopigmented macules. Histopathological examination shows sparse superficial perivascular lymphocytic infiltrate with melanophages within papillary dermis.⁴

Hemochromatosis is a disorder in which iron overload leads to deposition of iron in organs such as skin and liver. Cutaneous pigmentation is diffuse and bronze colored. It is associated with other stigmata of chronic hepatic failure. Biliary cirrhosis is characterized by intense pruritus, excoriation and hyperpigmentation.

Lastly, as demonstrated in the histopathologic picture already published in the case report, such large melanocytes in increased numbers are absolutely characteristic of this rare

condition.¹ In this case, histopathologic findings were pivotal in establishing the diagnosis.

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Conflicts of interest

There are no conflicts of interest.

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