Mottled hyperpigmentation on the face and neck of a woman 35 years of age

Case report

A Taiwanese woman 35 years of age presented with asymptomatic, mottled hyper- and hypopigmented macules on her neck and face, which had presented in the last 2 years. Spotted macules were first noted on her neck and had slowly progressed to involve her face as time went by. Her medical history was unremarkable. She did not have photosensitivity or any other systemic or cutaneous diseases before the onset of the lesions. There were no family members with similar cutaneous presentations. Examination revealed diffuse hyperpigmentation with numerous and varying sizes of hypopigmented spots over her neck and face (Figure 1). There were no apparent erythemas, blisters, telangiectasia, atrophy, or papular lesions. Her mental and developmental milestones were normal.

A skin biopsy specimen was obtained from her neck. The histopathologic evaluation revealed that mildly hyperkeratotic epidermis and amorphous eosinophilic masses were deposited in the papillary dermis (Figure 2). There were no alternations in the reticular dermis. Congo red staining was positive for eosinophilic materials (Figure 3).

Figure 1 Diffuse hyperpigmentation with numerous and varying sizes of hypopigmented spots on the neck and face.

Figure 2 Mildly hyperkeratotic epidermis and amorphous eosinophilic masses deposited in the papillary dermis (hematoxylin & eosin, 200×).

Figure 3 Congo red staining was positive for eosinophilic materials (Congo red, under a fluorescent microscope, 400×).
Diagnosis

Amyloidosis cutis dyschromica-like macular amyloidosis on the face and neck.

Discussion

Primary cutaneous amyloidosis (PCA), a pruritic skin disorder, is the deposition of amyloid in the papillary dermis without any systemic involvement. It is more commonly seen in South America and Southeast Asia compared with North America and Europe. It is also more common in Chinese compared with Malays or Indians.1

The clinical presentations of PCA could be divided into lichen, macular, nodular, or tumefactive amyloidoses. Lichen amyloidosus is the most common type and clinically characterized by an intensely pruritic eruption. Multiple discrete hyperkeratotic papules may coalesce into plaques and are often located on the extensor surfaces of the legs. Macular amyloidosis presents as brownish macules and are often distributed in a rippled pattern on the upper aspect of the back. Nodular amyloidosis is a rare form of PCA and presents as a solitary or multiple erythematous to brownish waxy nodules on the legs or the trunk.

The differential diagnoses include dyschromatosis universalis hereditaria, xeroderma pigmentosum, amyloidosis cutis dyschromica (ACD), and poikilodermatous amyloidosis. The deposition of amyloid in the papillary dermis is not observed in the first two diseases. ACD, which was described first by Morishima in 1970,2 is characterized by the following features: (1) stippled, reticular hyperpigmentation with hypopigmented spots without papulation almost over the whole body, (2) no or little itch, (3) onset before puberty, and (4) small foci of amyloids close beneath the epidermis. Familial ACD has been reported in Taiwan.3 Our case presented with mottled skin lesions, like ACD, but they were limited to the face and neck, and onset occurred after puberty. Poikilodermatous amyloidosis is another rare type of PCA and is associated with short stature, blisters, photosensitivity, and palmoplantar keratoderma.4 The present case was of normal stature, and we did not observe any dermatologic abnormality except for the hyperpigmentation on her neck and face.

The main treatment strategy is to reduce the irritation and friction of the skin. Sedative antihistamine and topical steroids are beneficial for pruritus relief. Administration of retinoid may also be effective for both the pruritus and hyperkeratotic papules.5 In addition, 532-nm and 1064-nm Q-switched Nd:YAG laser therapy may reduce pigmentation in macular amyloidosis patches.6 The patient received oral isotretinoin for 5 months, but neither obvious clinical improvement nor progression of the pigmentation was observed.

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References


Received: Dec 5, 2011
Revised: Jan 12, 2012
Accepted: Jan 18, 2012