Leonine Facies in the Cutaneous Form of Multicentric Reticulohistiocytosis

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An 85-year-old Chinese woman presented with a long history of multiple asymptomatic firm, flesh-colored papules covering her entire face, including the forehead, nose, cheeks and chin, causing thickening, furrowing and coarsening of the skin on the face, leonine facies (Picture 1A, B). Skin histology depicted diffuse infiltrates of histiocytes and...
large multinucleated giant cells with eosinophilic fine-granule cytoplasm in a ground-glass pattern in dermis (Picture 2A, B). Immunostains revealed positivity for CD68, periodic acid-Schiff staining and negative for S-100 protein. Neither joint deformities, visceral involvement nor underlying neoplasms was identified. Purely cutaneous form of multicentric reticulohistiocytosis (MRH) was diagnosed.

MRH is a rare, idiopathic, systemic non-Langerhans histiocytic disorder (1). The differential diagnosis of leonine facies includes lepromatous leprosy, chronic actinic dermatitis, scleromyxedema, amyloidosis, sarcoidosis, pseudolymphoma, lymphoma, pachydermoperiostosis, and hypereosinophilic syndrome in addition to MRH (2). The diagnosis can be confirmed by histopathology. The purely cutaneous form of MRH may involute spontaneously with a favorable prognosis.

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References