PAEDIATRIC DERMATOLOGY

PAPULAR XANTHOMAS WITH DESTRUCTIVE ARTHRITIS-A RARE ASSOCIATION

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Background: Papular xanthoma (PX) is a special type of xanthoma and belongs to the juvenile xanthogranuloma family, most of which are manifested as disseminated, asymptomatic papules and nodules. Mucosa is occasionally involved, however, system involvement is rare. Multicentric reticulohistiocytosis (MRH) is a rare type of systemic non-Langerhans cell histiocytosis and commonly occurs in adult populations.

Observation: We report the case of a 7-year-old girl who presented with xanthomatous lesions in the periungual regions of both hands and on the back of her tongue, associated with bilateral knee and shoulder joint swelling and pain for 6 months, which was clinically compatible with multicentric reticulohistiocytosis. Nevertheless, the histopathology of xanthomatous lesions in the skin was more suggestive of papular xanthoma. The patient was treated with low-dose methotrexate. Two years later, the skin lesions subsided, but no improvement was observed in the lingual mucous membrane lesions and arthritis symptoms.

Key message: This was an interesting case due to the mucosal involvement and the presence of destructive arthritis which were the clinical features of MRH. However, the histological characteristics supported the non-Langerhans cell histiocytosis favoring PX over MRH.