



SKIN MANIFESTATIONS OF INTERNAL DISEASE

DIFFUSE NORMOLIPEMIC PLANE XANTHOMA ASSOCIATED WITH PULMONARY SARCOIDOSIS AND MONOCLONAL GAMMOPATHY

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Background: Xanthomas represent lipid deposits in the skin. They include hyperlipemic xanthoma, normolipemic xanthoma, and related condition, necrobiotic xanthogranuloma. Diffuse normolipemic plane xanthoma (DNPX) is a rare form of xanthomatosis, now considered as an uncommon subtype of non-Langerhans histiocytosis. DNPX is characterised by presence of yellow-orange plaques on the neck, trunk, buttocks, flexural folds and usually accompanied by xanthelasma. The lesions may initially present as urticarial papules and plaques. This type of xanthoma can be associated with hematological diseases, especially with paraproteinemia, monoclonal gammopathy and multiple myeloma. However, other malignant hematological or lymphoproliferative disorders, such as acute monoblastic leukemia, chronic myeloid leukemia, chronic lymphatic leukemia, adult T-cell lymphoma/leukemia, Sézary syndrome, histiocytosis X, Waldenstrom's macroglobulinaemia, cryoglobulinemia and Castleman's disease have been reported in association with DNPX.

Observation: We report a 63-year-old female patient with a 5 years history of yellow plaques on upper eyelids and reddish-yellow plaques localized to her neck, upper back and extremities. Histopathology showed a perivascular infiltration of histiocytes with foamy cytoplasm and scarce lymphocytes in the papillary dermis. The lipid profile was normal, but laboratory data showed significantly higher chitotriosidase and ACE levels. Serum protein electrophoresis and imunofixation detected monoclonal IgG kappa protein. The patient has also been followed by pulmonologist. Pulmonary sarcoidosis was diagnosed by imaging exams, transbronchial needle aspiration biopsy of mediastinal lymph node and histopathological confirmation. The patient was treated for sarcoidosis with oral prednisone, which led to significant remission of the skin lesions after 8 months of therapy.

Key message: A few cases of hyperlipemic xanthoma associated with cutaneous sarcoidosis have been reported, but, to the best of our knowledge, this is the first case showing association of diffuse normolipemic plane xanthoma with pulmonary sarcoidosis.

