Chapter 30
Yellowish Erythematous Desquamative Lesion in a Middle Aged Man

Torello M. Lotti and Olga Olisova

A 51 years-old Caucasian man has been referred to our Institute for the first time 3 years ago for progressive itching diffuse and plane erythematous yellowish discoloration of the skin of the trunk and neck (Figs. 30.1 and 30.2). A tiny desquamation was evident at the center of the lesions. Blood tests were all normal or negative according to our laboratory while in the past serum protein electrophoresis had shown the presence of an immunoglobulin M lambda Monoclonal component with negative Bence-Jones urine protein. The serum protein electrophoresis repeatedly showed a normal profile after the initial detection of the IgM lambda monoclonal peak.

Repeated computed tomography never showed any skeletal alterations. The bone marrow contained a normal cellular population.

Based on the Case Description and The Photographs, What Is Your Diagnosis?

1. Diffuse plane normolipaemic xanthomatosis
2. Generalized eruptive histiocytosis
3. Non-X histiocytosis
4. Histiocytosis of mononuclear phagocytes other than Langerhans cells
5. Multicentric reticulohistiocytosis
6. Letterer-Siwe disease

T. M. Lotti (✉)
Science of Communication, Marconi University, Rome, Roma, Italy
e-mail: professor@torellolotti.it

O. Olisova
Department of Dermatology & Venereology, Sechenov University, Moscow, Russia

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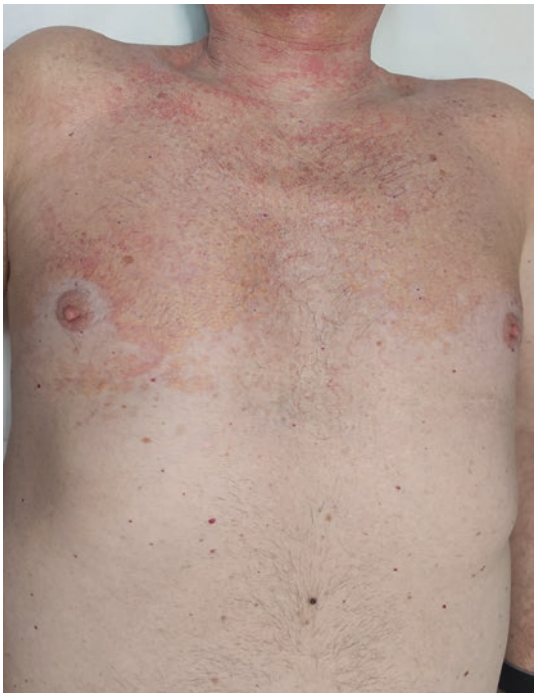

Fig. 30.1 Plane erythematous yellowish discoloration of the skin of the trunk and neck

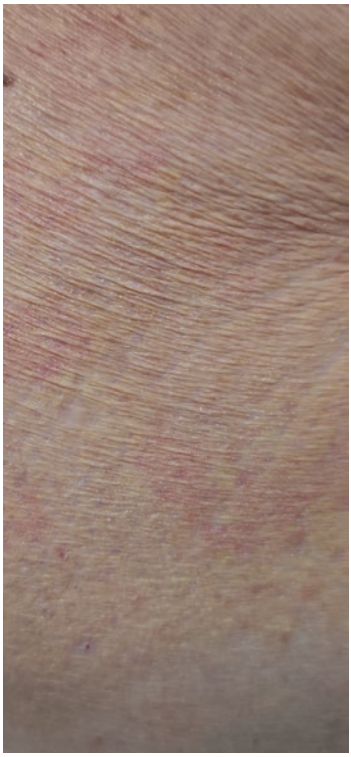

Fig. 30.2 Higher magnification of Plane erythematous yellowish discoloration of the skin of the trunk and neck

Diagnosis

Diffuse plane normolipaemic xanthomatosis.

Skin biopsy showed aggregates of foamy macrophages in the upper and medium dermis.

Plasma cells and limited zones of necrobiosis with cholesterol clefts were evident. Touton giant cells were not visible.

Discussion [1–5]

Diffuse plane normolipaemic xanthomatosis (DPNX) is a rare acquired dermatosis characterized by yellow-orange-erythematous plaques usually symmetrically distributed. DPNX is in fact a type of non-Langerhans histiocytosis due to the deposition of cholesterol and lipids in the skin in subjects who may have normal levels of blood cholesterol and no underlying disorders. Oral manifestations have been described including verruciform and plane xanthomas. DPNX is often associated with lymphoproliferative malignancies, multiple myeloma and hepatitis C and monoclonal gammopathies. Aggregation of foamy histiocytes in the superficial and medium dermis is always evident. Patients should be kept under surveillance for many years for the appearance of other associated diseases.

Key Points

- Diffuse plane normolipaemic xanthomatosis is a rare form of acquired non-Langerhans histiocytosis showing as yellowish plaques of the skin and of the oral mucosa. Multicentric reticulohistiocytosis should be excluded always.
- Deposition of cholesterol between the collagen fibers is evident in the upper and medium dermis with aggregates of foamy histiocytes.
- Underlying monoclonal gammopathies, multiple myeloma, hepatitis C and other lymphoproliferative malignancies should be always suspected, keeping the affected subjects under surveillance.

References

1. Cota C, Pellacani G, Lora V, Brancorsini D. Diffuse (generalized) plane xanthoma misdiagnosed as carotenoderma: usefulness of reflectance confocal microscopy. Am J Dermatopathol. 2020;42(12):e167–9.
2. Souza MCC, Martins PHT, Vivian A, Luzzatto L. Diffuse normolipemic plane xanthoma and hepatitis C: chance? An Bras Dermatol. 2020;95(2):261–2.

3. Gupta M, Sharma R, Gulati A, Gupta A. Diffuse normolipemic plane xanthoma with underlying photosensitive dermatoses. BMJ Case Rep. 2019;12(11):e230720.
4. Morsink LM, Nijhof IS. Diffuse plane normolipaemic xanthomatosis as a manifestation of monoclonal gammopathy. Br J Haematol. 2019;187(4):411.
5. Lotti T, Santucci M, Casigliani R, Fabbri P, Bondi R, Panconesi E. Multicentric reticulohistiocytosis. Report of three cases with the evaluation of tissue proteinase activity. Am J Dermatopathol. 1988;10(6):497–504.