DERMATOLOGY PRACTICAL & CONCEPTUAL
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Waldenström macroglobulinaemia presenting as tingling ulcers on the feet

Isabelle Fischer,[1] Delwyn Dyall-Smith,[1] Birgit Peters,[1] Wilhelm Stolz,[1] Brigitte Coras-Stepanek[1]

[1] Department of Dermatology, Klinikum Schwabing, Munich, Germany

Key words: peripheral neuropathy, storage papules, macroglobulinaemia cutis

Citation: Fischer I, Dyall-Smith D, Peters B, Stolz W, Coras-Stepanek B. Waldenström macroglobulinaemia presenting as tingling ulcers on the feet. Dermatol Pract Conc. 2013;3(3):3 . http://dx.doi.org/10.5826/dpc.0303a03.

Received: May 1, 2013; **Accepted:** June 1, 2013; **Published:** July 31, 2013

Funding: None.

Competing interests: The authors have no conflicts of interest to disclose.

All authors have contributed significantly to this publication.

Corresponding author: B. Coras-Stepanek, M.D., Department of Dermatology, Klinikum Schwabing, Koelner Platz 1, 80804 Munich, Germany. Tel. 0049 89 3068; Fax . 0049 89 3068 3918. Email: brigitte.coras@klinikum-muenchen.de.

ABSTRACT A 56-year-old man presented with tingling ulcers on the feet. On further skin examination, papules on the knees were observed. Biopsies revealed extravascular eosinophilic deposits of IgM, and Waldenström macroglobulinaemia was diagnosed. The skin manifestations have resolved with chemotherapy. Peripheral neuropathy and storage papules are rare manifestations of Waldenström's macroglobulinaemia.

Case presentation

A 56-year-old man presented with a 12-month history of tingling ulcers on both feet (Figure1A). He was otherwise well. On examination there were ulcers on both feet, 1-1.5 cm in diameter with a macerated edge. Skin-colored to red-brown papules were noted on both knees (Figure 1B) as well as irregular red circles on both calves. Skin biopsies taken from knee papules and red circles on the lower leg showed extravascular eosinophilic deposits in the dermis (Figure 1C), which stained positive with PAS, IgM (Figure 1D) and kappa stains, but negative with Congo red. Investigations are summarised here: Serum kappa free chains, 85.9 mg/l (normal 3.3-19.4 mg/l); serum lambda free chains, 17 mg/l (normal 7.7-26.3 mg/l); serum free kappa chains /free-lambda chains ratio, 5.05 (normal 0.26-1.65); serum electrophoresis, elevated Ig M gradient; serum immunofixation, monoclonal Ig M/kappa-type immunoglobulin; urine immunofixation, monoclonal kappa-free chain; bone marrow microscopy, infiltrate of lymphoplasmocytic cells (15%) (most CD20 and CD138 positive) with preferential expression for IgM.

Discussion

The cutaneous manifestations of Waldenström macroglobulinaemia have been classified as either infiltration by neoplastic cells or those related to the paraproteinaemia, such as immunoglobulin deposition, cryoglobulinaemia or blood hyperviscosity. A number of other nonspecific cutaneous presentations have been reported. Approximately 5% of patients with Waldenström macroglobulinaemia develop cutaneous manifestations, with neoplastic infiltration being the least common. Skin signs may either precede or follow the diagnosis. Prognosis does not appear to be influenced by the presence or absence of cutaneous disease [1].

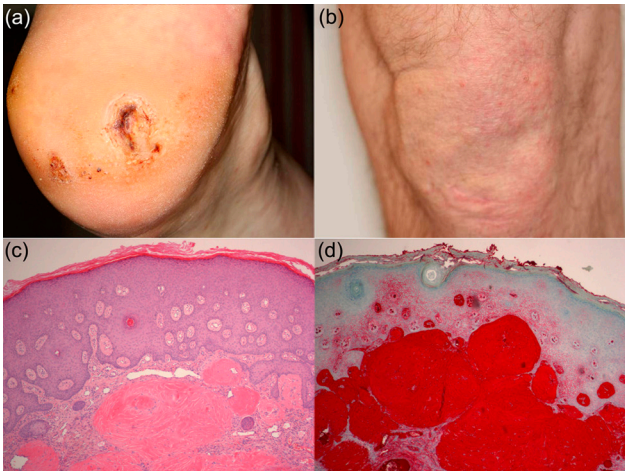

Figure 1A. The patient presented with tingling ulcers on the feet. [Copyright: ©2013 Fischer et al.]

Figure 1B. On further examination, papules were noted on the knees. [Copyright: ©2013 Fischer et al.]

Figure1C. Biopsy from a knee papule showed homogeneous extravascular eosinophilic hyaline deposits throughout the papillary and reticular dermis (H&E 40x). [Copyright: ©2013 Fischer et al.]

Figure 1D. IgM stain demonstrates the extravascular IgM deposits in the dermis (40x). [Copyright: ©2013 Fischer et al.]

Neoplastic cell infiltrates present as red or purple patches and plaques. In one case the patient presented with a burning red face which was initially diagnosed as rosacea but with time the more typical purple infiltration developed [2].

Peripheral neuropathy is a well recognised presentation of dysproteinaemias, particularly IgM [3]. In Waldenström macroglobulinaemia the neuropathy is typically sensorimotor, initially with paraesthesias and numbness in the feet and hands. Motor symptoms, such as weakness or foot drop, develop later. The possible mechanisms in Waldenström macroglobulinaemia include microangiopathy, direct infiltration of nerves, endoneural deposition of IgM protein or amyloidosis, hyperviscosity or immunoglobulin deposit in blood vessels. The tingling ulcers in our patient are probably an example of this and, although biopsies were not taken from the ulcers, may have been due to IgM deposition given he also had macroglobulinaemia cutis.

There has been only one other case reported, to our knowledge, of ulcers on the sides of the feet [4]. These developed in areas of hyperkeratosis and were associated with dysaesthesia and a burning sensation. However, the biopsy showed a leukocytoclastic vasculitis with a neutrophilic infiltrate. Storage papules were present on the knees and perianal skin. Another report described IgM dermal deposits that presented as painful hyperkeratotic papules on the soles of the feet that were not ulcerated [5].

On further examination, our patient had the papules of macroglobulinosis. Storage papules, also known as macroglobulinaemia cutis, are rare but well described and are usually found on the knees or other extensor surfaces. They are usually skin-coloured, red-brown or translucent, and may be umbilicated, crusted or haemorrhagic. Biopsy shows dermal eosinophilic hyaline material, which is IgM.

Our patient was treated with six cycles of bendamustine infusions. After five cycles the lower limb ulceration and neurological symptoms had resolved, although the blood IgM level did not change.

Conclusion

Waldenström macroglobulinaemia is a low-grade lymphoplasmacytoid lymphoma that usually presents with nonspecific systemic symptoms, but cutaneous changes may be the initial manifestation and hence may present first to a dermatologist. Although our patient presented with tingling ulcers, the diagnosis was reached after skin biopsy from knee papules demonstrated dermal immunoglobulin deposition.

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