SKIN MANIFESTATIONS OF INTERNAL DISEASE

PAPULOERYTHRODERMA OF OFUJI AS A PARANEOPLASTIC MANIFESTATION OF A MARGINAL ZONE B-CELL LYMPHOMA

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Background: Papuloerythroderma of Ofuji is a rare reactive disease, clinically characterized by widespread, pruritic, red–brown papules that can progress into a confluent erythroderma that typically spares skin folds (“deck-chair” sign). Various underlying aetiologies have been implicated, including infections and malignancy.

Observation: An 80-year-old Caucasian woman presented with widespread intensely pruritic red-brown papular lesions, that latter coalesced into an erythroderma sparing the face and abdominal and thoracic folds. Additionally she had multiple tender hard cervical, supraclavicular, axillary and inguinal lymph nodes. The patient denied accompanying complaints, namely constitutional symptoms. Two months earlier, she had a similar episode, but lesions were restricted to the lower limbs, persisted a few weeks and improved spontaneously.
Laboratory investigations showed blood eosinophilia (8%, 900 G/ul) and marked elevation of serum immunoglobulin E (31.400 IU/ml) with no other significant alterations on blood, urinary and imaging exams. Skin biopsy revealed a dense superficial and deep dermal infiltrate of lymphocytes, numerous eosinophils and a significant contingent of neutrophils, with occasional large lymphomononucleated cells. Immunophenotyping of the skin infiltrate was normal. These findings fit within the spectrum described in Ofuji's papuloerythroderma.
Histopathology of a supraclavicular lymph node showed almost complete involvement by a small cell lymphoma and immunophenotyping revealed a marginal zone B-cell lymphoma.
Skin lesions resolved completely and spontaneously within a month. There was also normalization of the blood eosinophil count. Taking into consideration the indolent behaviour of the marginal zone B-cell lymphoma and patient’s age no specific therapy was performed and a watchful waiting approach, with close reappraisal was decided.

Key message: Papuloerythroderma of Ofuji is rare, with approximately 200 cases reported, predominantly in Asians and males. Due to its possible association with neoplastic diseases, oncological screening is mandatory.