Case report

A CASE OF MORBIHAN'S DISEASE
Chronic Upper Facial Erythematous Oedema

G. Leigheb, P. Boggio, M. Gattoni and G. Bornacina

ABSTRACT

A 61 year-old male patient with recurrent episodes of reddish-violet erythema and oedema of the forehead, nose, periocular regions, eyelids and cheeks, prevalently on the left side of the face, was followed for a period of 9 years. Pachydermatous sequelae developed in the affected areas. The chronic course of the manifestation, the nonspecific histological picture and the lack of response to treatment suggested a diagnosis of Morbihan's disease. The peculiar features of the condition and its resemblance to rosaceous lymphoedema are discussed.

KEY WORDS

Morbihan's disease, rosaceous lymphoedema, chronic upper facial erythematous oedema.

INTRODUCTION

Morbihan's disease, a rare condition of which 10 cases have been reported to this date, mostly by French authors (1, 2, 3), is characterized by chronic recurrent erythemato-oedematous manifestations of the upper half of the face (nose, forehead, periocular and palpebral regions) with a typical deep reddish-violet hue and marked oedema. The course is marked by recurrent flare-ups with intervening periods of partial regression of the symptoms and pachydermatous scleromyxoedematous sequelae. The dermatosis has a chronic course, usually lasting many years.

The differential diagnosis usually involves various forms of dermatoses due to exposure to sunlight, chronic eczema, rosacea, chronic lupus erythematosus, dermatomyositis and actinoreticulosis (2). The chronic course, nonspecific laboratory and histological findings and refractoriness to treatment are the findings criteria upon which the diagnosis is usually based.

CASE REPORT

A male patient aged 61 years. Family history uninformative. Normal development. The patient had a history of mild hypothyroidism due to a hypofunctioning struma with persistent enlargement of the right lobe and low normal iodine uptake values. Stabilized bilateral fibronodular tuberculosis of the lung apex for which he had been treated with isoniazide, etambutol, rifampicin and betametasone during three months prior to observation. Treatment had

been discontinued shortly before he was admitted to the hospital.

The patient stated that the dermatosis had started three or four years previously. The first area affected was the left eyelid, from which the manifestations spread with bursts lasting 15-30 days. During this time severe oedema was present, followed by variable periods of relative remission. When the patient was first examined the manifestations had been present for 10-15 days.

Physical examination of the integuments (Fig. 1) revealed a redish-violet coloured erythemato-oedematous plaque irregularly covered with grouped translucent oedematous pseudo-bullae associated with teleangiectasias. There was a ptosis of the left upper eyelid with a moderate oedema of the lower eyelid. Patches of diffuse lilac-reddish oedema of firm elastic consistency, more extensive on the left side, were also present in the zygomatic regions. The skin of the nose, especially towards the tip, was also oedematous, with a reddish-violet hue, and with dilated pores, giving an "orange peel" like appearance. The only subjective symptom the patient complained of was a "taut" sensation in the left orbital region. The bulbar and palpebral conjunctivae were unaffected. Moderate bright red oedema with slight pityriasic scaling was present on the forehead, especially in the supra-orbital regions, where the skin had a pachydermatous appearance, with accentuated folds.

Laboratory tests were normal. The TINE test was intensely positive. The porphyrins were normal. FAN and Ab anti-DNA were negative. Sedimentation rate 3. Thyroid hormone assays were normal.

While the patient was in hospital the oedema of the face cleared off in 10 days, but this was followed by a relapse. A histological diagnosis of lymphangioma, based on the presence of marked lymphangiectasias in the dermis, with infiltration of lympho-monocytic elements and plasma cells, was made after examination of a surgical biopsy taken from the left upper eyelid. Reductive blepharoplasty was done to correct the ptosis caused by the excess of palpebral tissue. A biopsy sample, excised simultaneously from the forehead, revealed a normal epidermis with partly dilated follicles, and oedema of the dermis with diffuse dilatation of the lymph vessels and mostly perivascular infiltration of lympho-monocytic cells and plasma cells (Fig. 2). The patient improved and was discharged with a prescription of rifampicin and betametasone (8 mg daily). After three months the patient had a relapse, with a virtually identical picture, also affecting the left upper eyelid despite the blepharoplasty. Another biopsy of the lesions of the forehead revealed actinic elastosis and superficial infiltration of the dermis associated with ectasia of the blood vessels with minimal dermotropism. The cellular infiltrate was rather composite, with presence of small lymphocytes and isolated cells with an indented or cerebroid nucleus, but without involvement of the epidermis. During the following

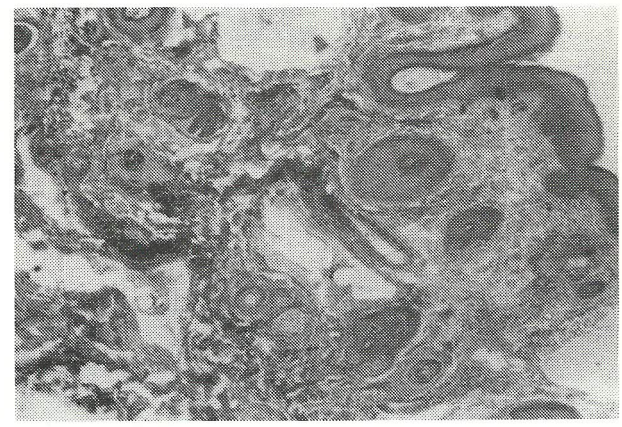

Fig. 2: Biopsy: normal epidermis and dilated follicles. Diffuse oedema of the dermis

month there was only a slight remission of the manifestations in spite of systemic corticosteroid treatment. Laboratory tests, thyroid function tests, T-lymphocyte subpopulations and FANs were persistently normal. At a follow-up examination 8 months later, persistent oedema of the eyelids and a pachydermatous appearance of the forehead and nose were observed, but the erythematous component was no longer present (Fig. 3).

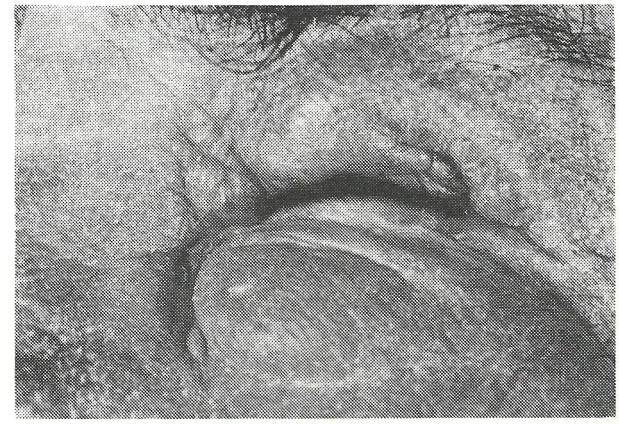

Fig. 4: Relapse after two years

Two years later the patient again referred to us for another relapse, with a perfectly identical picture (Fig. 4). In particular, there were oedema and ptosis of the left eyelids and the circumference of the palpebral border was reduced by one third. The eyelids were covered with pseudo-vesicles and grouped oedematous pseudo-bullae. The eyelids of the right eye were normal. The patient experienced no discomfort. During the period of observation (about one month) the

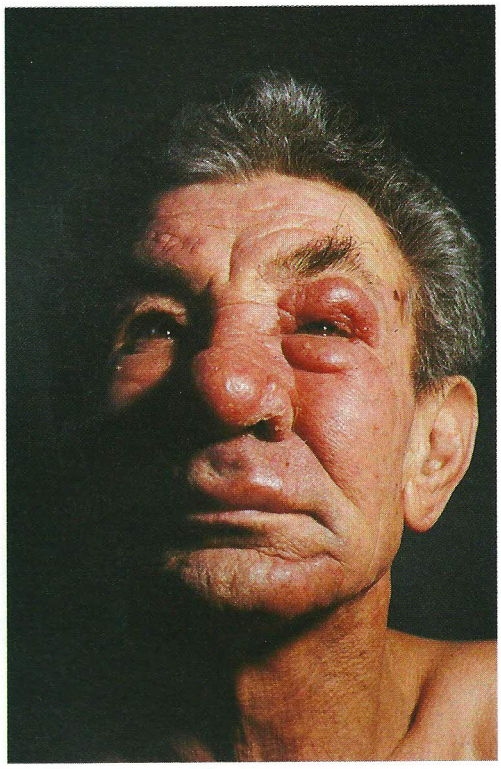

Fig. 1: Aspect of the patient at the first examination

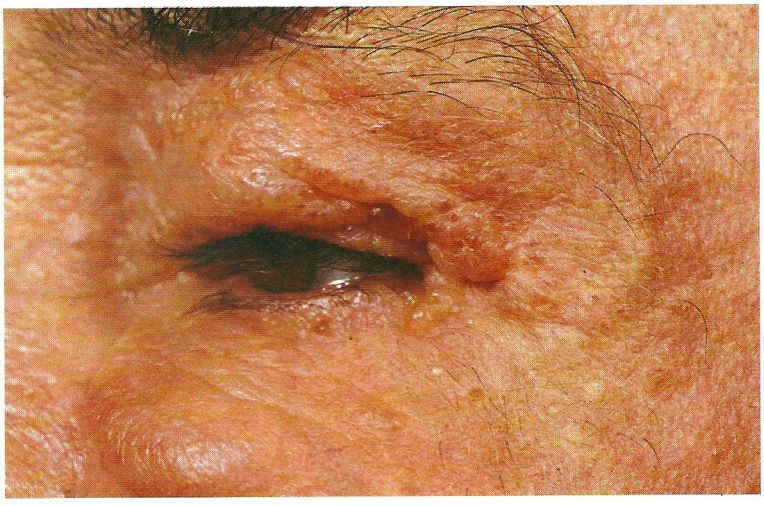

Fig. 3: Clinical aspect after 8 months

oedema and erythema were at first unaffected by corticosteroid treatment (deflazacort, 30 mg) but then improved rather rapidly although not completely. MED was normal. A further biopsy from the right supraciliary region revealed orthokeratosis and focal parakeratosis with lympho-histiocytic perivascular and periadnexial infiltration in the upper and mid dermis, associated with a few eosinophilic cells and moderate actinic elastosis.

We then lost sight of the patient for 6 years but recently we had the opportunity to examine him again (9 years after he had been first referred to us). Pachydermatous oedematous manifestations without an appreciable erythematous component were present in the same regions (forehead, nose, zygomatic regions). During the intervening period he was given no particular treatment and had other periodical relapses similar to those described above. His general condition was good.

DISCUSSION

This case is similar to the ten other cases described, first by French, and more recently by Italian authors (2, 3, 4) as Morbihan's disease. The clinical diagnosis was established after other similar conditions had been ruled out on the grounds of the clinical, laboratory and histopathological findings. The nonspecific histological picture enables us to rule out sarcoidosis, lymphoma and lupus erythematosus, whereas the histopathological picture, clinical course, refractoriness to treatment, irrelevance of exposure to sunlight and absence of pustulation allow the exclusion of dermatomyositis, actinoreticulosis, chronic eczema, dermatitis due to sunlight, and acne rosacea. Only the picture described by English workers as "rosaceous lymphoedema" (5) seems rather similar to Morbihan's disease in some respects, but it is not possible to form an opinion as to the possible identity of the two forms because detailed information on the incidence, course and outcome of the former is lacking.

Peculiar features of the present case are the intense oedematous component of the relapses producing a histological picture very similar to lymphangioma, and the resulting marked pachydermic sequelae. The co-existence of latent hypothyroidism (reported in at least one other case (2)), which may have enhanced the scleromyxoedematous features of this case, is also noteworthy. No evidence of an associated neoplastic condition, observed by other workers (3) was found.

The etiopathogenesis of Morbihan's disease is still obscure, although the hypothesis of a mechanism based on recurrent episodes of congestion of the local microcirculation, somewhat similar to the mechanism responsible for rosacea, can reasonably be considered.

Various forms of treatment, including antibiotics, have proved ineffective, only partly useful, as in the case of thalidomide during the early stages of the disease (1).

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