Lymphomatoid granulomatosis presenting as angioedema

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Summary: We describe a patient with severe eyelid and lip angioedema lesions in whom biopsy specimens from angioedematous labial mucosa disclosed features of lymphomatoid granulomatosis. To our knowledge, angioedema lesions with characteristic histological findings of lymphomatoid granulomatosis have not been previously described as a presenting sign of this disease.

Introduction

Lymphomatoid granulomatosis (LG) was initially described by Liebow et al. as an angiocentric and angiodestructive lymphoreticular proliferation involving predominantly the lungs, but also other extranodal sites. Its pathogenesis remains an enigma, though it is currently accepted to be a peripheral T-cell lymphoma. We report a case of LG with an unusual mode of clinical presentation.

Case report

A 40 year old white man was admitted to our hospital because of non-pitting, waxing and waning oedema of the lips and eyelids, together with occasional hoarseness and dyspnoea. He required corticosteroid therapy for life-threatening episodes, and treatment with antihistamines, danazol, colchicine, elimination diet and antibiotics could not prevent the development of these attacks. Personal and family history were unremarkable and repeated analytical studies including ESR, blood cells count, serum biochemistry, urinalysis, serum electrophoresis and immunoglobulins, complement levels, antinuclear antibodies (ANA), antistreptolysin O (ASO) titres and chest X-rays were normal or negative. For the next 5 years, the patient continued with similar episodes, but in the last months, labial and eyelid oedema became persistent, showing only a moderate response to high dose oral corticosteroid therapy (Figure 1). Attempts to taper the dosage resulted in relapse. At that time, he complained of recurrent oral ulcers, intermittent fever and weight loss. Three mild episodes of urticaria developed. On physical examination, white patches on the tongue, soft palate and oropharynx were detected. Laboratory studies showed a mild leukopenia (3,400 leukocytes/μl) with normal differential count, ESR 19 mm/h, serum albumin 30 g/l (35–55), serum aspartate aminotransferase (AST) 72 U/l (4–40), serum alanine aminotransferase (ALT) 90 U/l (4–40) and elevated serum immune complexes (Raji cell assay). Urinalysis, complement studies including C1 esterase inhibitor, serum immunoglobulins, serum cryoglobulins, ANA and anti-extractable nuclear antigen (ENA) antibodies, rheumatoid factor, α-1 antitrypsin levels, hepatitis B surface antigen, VDRL and ASO titres, fresh stool examination, stool cultures and tuberculin test were normal or negative. Chest X-rays failed to show any abnormality. Abdominal ultrasonography and intravenous pyelogram were normal too. A sinus X-ray revealed opacification of the right maxillary antrum. Candida albicans was recovered from oral patches. The examination of a bone marrow specimen was unremarkable. Biopsies from pharynx, soft palate and upper lip mucosa showed an

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Figure 1 Persistent facial non-pitting oedema.
angiocentric proliferation of various mononuclear elements including lymphocytes, plasma cells and macrophages, with some foci of bizarre lymphoreticular cells which presented much pleomorphism (Figures 2 and 3). Tuberculoid granulomas were not present. Immunphenotyping of the infiltrate revealed a predominance of CD4-positive cells. A diagnosis of LG was made and the patient was treated with cyclophosphamide (100 mg/day orally) and methyl-prednisolone (80 mg/day orally).

Symptoms persisted over the next 7 months. His general condition deteriorated, and a right radial nerve palsy occurred. One month later recurrent epistaxis developed, and a chest X-ray revealed bilateral lung infiltrates. The patient required embolization of the internal maxillary arteries due to massive epistaxis. His condition worsened and he finally died 2 months later because of disseminated intravascular coagulation. Postmortem examination was not allowed.

Discussion

The skin is the most common organ of extrapulmonary involvement in LG. Skin lesions may be the presenting sign in as many as 20% of patients with LG, and in 10% of cases skin lesions may be seen in the absence of chest X-ray alterations. Cutaneous lesions usually take the form of erythematous macules and papules, nodules and ulcerations. However atypical manifestations may appear, such as vesicles, infiltrated annular lesions, generalized ichthyosis, patchy alopecia, loss of sweating and necrobiosis lipoidica-like lesions.

Urticarial and angioedema-like lesions are rarely described in LG. One of the patients described by MacDonald and Sarkany suffered an episode of angioneurotic oedema one year before the diagnosis of LG. Shank et al. reported a patient with LG who developed a permanent right supraorbital swelling, which was interpreted as a manifestation of her generalized disease. Guarch et al. reported bilateral palpebral oedema which subsided after chemotherapy in a patient with LG. Finally, a patient with LG developed evanescent macular and urticarial skin lesions unresponsive to therapy. In none of these cases is reference made to biopsy of the lesions. To our knowledge, ours is the first case in which typical histopathological findings of LG have been demonstrated in biopsy specimens from angioedematous labial mucosa. We cannot say if these histopathological changes were present when there was no facial oedema.
References

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