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SHORT COMMUNICATION

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Multiple ocular impairment in a patient affected by Waldenström's macroglobulinaemia

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Abstract ● Background: We studied a bilateral tumefaction of the lacrimal gland in a female patient. ● Method: Ocular and general clinical examinations were carried out. ● Result: Computerized tomography (CT) of the cranial orbit showed a tumefaction of solid density in the lacrimal gland. Histological examination of material removed by needle aspiration revealed the presence of elements of a lymphoplasmacytoid nature. Fluorescein angiography showed dilatation of the veins, intraretinal flame haemorrhages and small ischaemic areas. Chest CT showed an increase in the size of the middle and upper mediastinal lymph nodes, and examination of a specimen of bone marrow from the chest revealed the presence of small lymphocytes with a plasmacytoid tendency.
● Conclusion: On the basis of the findings, we diagnosed Waldenström's disease with rare multiple ocular impairment (lacrimal gland and retina) in an early stage.

Introduction

Waldenström's macroglobulinaemia is caused by a monoclonal proliferation of lymphatic B cells producing IgM and can be considered one of the clinical presentations of lymphoplasmacytoid lymphomas.

Lymphomas of the orbit and its adnexa, both primary and secondary, are rare: they comprise 0.24% of all lymphomas for primary and 1–4% for secondary localizations [2, 11, 14, 19].

Shields et al. [20] found 71 cases of lymphoid hyperplasia, lymphoma and plasmacytoma by biopsy examination of 645 lesions in the orbital space. Twelve of these were in the cavity of the lacrimal gland, while 59 were in other parts of the orbit.

The rarity of this affection, the difficulty that histological diagnosis may present, and the scarcity of published cases [9, 10, 17, 21] prompted us to present a newly diagnosed case of Waldenström's macroglobulinaemia with bilateral localizations in the lacrimal gland together with retinal involvement in both eyes.

Case report

A 74-year-old woman was examined. Informed consent was obtained for all examinations performed.

Clinical observation

The pathological anamnesis was of special interest. The patient made reference to surgical removal of a tumefaction in the right mandibular area in 1977. This tumefaction did not involve the mandible bone. It recurred three times in the next 4 years. The histological examination revealed a cytomorphology consistent with plasmacytoma.

X-rays of chest, cranium, spinal column and pelvis showed no significant pathological alteration. Radiotherapy was then performed (22 times at 5.5 Gy) on the right mandibular area. This was preceded by melphalan therapy (two tablets per day for 6 days).

Only in 1993 did laboratory analysis not show elevated erythrocyte sedimentation rate (ESR) and hypergamma-globulinaemia (IgM 4730 mg/dl, K-chain 2440, L-chain 261). Immunoelectrophoresis showed IgM/K gammopathy.

In July 1994 the patient reported a palpebral tumefaction in the right eye, of soft consistency, not painful, and with a normal color of overlying cutis. A similar tumefaction also appeared in the left

eye. The patient was admitted to the ophthalmic ward of the Fatebenefratelli Hospital, Rome.

Objective examination

The patient presented best corrected visual acuity of 8/10 in the right eye (RE) and 10/10 in the left eye (LE).

The objective examination showed bilateral hypertrophy of the lacrimal gland in both eyes (RE>LE) and swollen eyelids (Fig. 1). The conjunctive was chemotic in RE and normohaemic in LE. Transparent, avascular and specular cornea, anterior chamber of normal profundity and optically empty, normotrophic iris, centred, round normal-reacting pupil, transparent lens and normal vitreous were observed in both eyes.

The intraocular pressure was 14 mmHg in RE and 16 mmHg in LE.

The examination of the ocular fundus in both eyes showed flame haemorrhages along the greater vascular arcades, tortuous and dilated veins and diffused microaneurysms.

Fluorescein angiography, performed in both eyes, showed a normal filling time but increased retinal circulation time. Furthermore, dilatation of the venous vessels, intraretinal flame-shaped haemorrhages, microaneurysms and small areas of capillary nonperfusion were present.

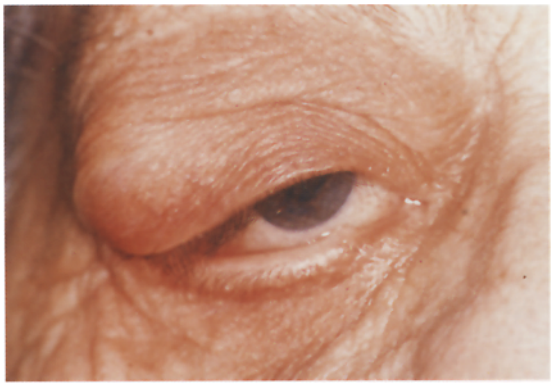

Fig. 1 The tumefaction in the upper temporal eyelid area at the time of first observation

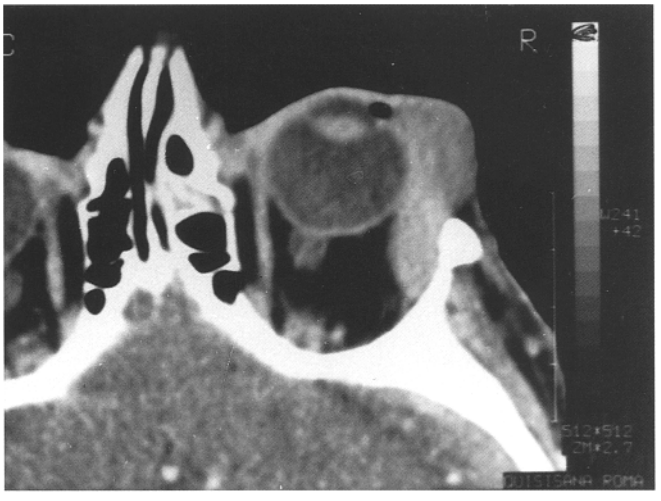

Fig. 2 Right orbit computerized tomography: tumefaction of solid density was present at the level of the orbital canthus region, in contact posteriorly with the region of attachment of the external rectus muscle and extending anteriorly to the soft eyelid parts

Histological examination of the tumefaction (by needle aspiration) revealed densely packed cells composed of lymphocytes, elements of a lymphoplasmacytoid nature and scattered macrophages. Clear atypias were evident. Cytological analysis confirmed the infiltration of lymphoplasmocytoid elements.

Computerized tomography (CT) of both cranial orbits showed a neoplasm originating from the lacrimal gland (Fig. 2).

The patient was given a series of general examinations with the following results.

Haemoanalysis showed reduced RBC ($304\times10^6/mm^3$) and Hb (80 g/dl), reduced prothrombin activity (43%), partial prothrombin time (35–40 s) and fibrinogen level (165 mg/ml), and increased ESR (150 mm/h); electrophoresis showed a double molecular chain in the gamma cathode zone; and immunofixation revealed increasing light K-chain IgM gammopathy.

Examination of a specimen of bone marrow from the chest showed diffuse infiltration of a monomorphic population, composed of small lymphocytes with a plasmacytoid tendency.

Ultrasonography demonstrated increased size with regular echo structure and no focal alterations in both the liver and the spleen.

Both renal ultrasonography and chest CT showed an increase in the dimensions of the respective lymph nodes.

On the basis of the findings Waldenström's disease with early and rare multiple ocular impairment (lacrimal gland and retina) was diagnosed.

Treatment

The patient received therapy with melphalan (10 mg/day, along with 50 mg of prednisone per day for 7 days a month). This treatment was continued for a total of nine cycles until total regression of the tumefaction and of the retinal vessel alterations was found in both eyes.

Discussion

We encountered a rare case of Waldenström's macroglobulinaemia which was characterized both by bilateral infiltration of the lacrimal gland with elements of a lymphoplasmacytoid nature and by bilateral retinal involvement. This consisted in a "hyperviscosity retinopathy" very similar to that of non-Hodgkin's lymphomas and frequently associated with Waldenström's disease [1, 3–7, 15].

Although a few cases of deposits in the cornea and one case of secondary glaucoma related to disease have been described [8, 10, 12], in our patient we observed no pathological changes in the cornea or in intraocular pressure.

In pathologies such as lymphoma, hepatosplenomegaly and lymphoadenopathy associated with the involvement of retinal vessels are frequently present [13], and these were the findings in our patient.

The lacrimal gland may be the site of numerous neoplastic processes: while its involvement is possible, albeit rare, in plasmacytoma, neoplastic infiltration in Waldenström's macroglobulinaemia is quite exceptional [13, 16, 18, 22].

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