

Benign orbital pseudolymphoma. Case report

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SUMMARY: Benign orbital pseudolymphoma. Case report.

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Orbital pseudolymphomas are uncommon benign tumors composed of reactive lymphocytes. The typical clinical presentation is painless onset of ptosis, proptosis, diplopia, or eyelid swelling. Virtually any adnexal structure can be affected, but lacrimal gland, orbital soft tissue, or extraocular muscles are the usual sites of involvement. Historically, treatment has been with oral corticosteroids or localized radiotherapy. We present a case of benign orbital pseudolymphoma.

RIASSUNTO: Pseudo-linfoma orbitario. Caso clinico.

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Le lesioni pseudo-linfomatose dell'orbita sono neoplasie benigne piuttosto rare. Clinicamente caratterizzata da una sintomatologia costituita prevalentemente da ptosi, proptosi e diplopia, la lesione può interessare tutte le strutture contenute nella cavità orbitaria con possibile coinvolgimento dei coni muscolari. Alla chirurgia segue un trattamento farmacologico steroideo e una possibile radioterapia locale. Si presenta un caso giunto alla nostra osservazione.

KEY WORDS: Lymphoma - Orbit - Surgery.
Linfoma - Orbita - Chirurgia.

Introduction

Pseudolymphoma is not a specific disease but rather an inflammatory response to known or unknown stimuli that results in a lymphomatous-appearing but benign accumulation of inflammatory cells (1). Resemblance to lymphoma is usually most apparent histologically, but some examples may also mimic lymphoma clinically. When known, the inciting agent should be included within the diagnosis.

The term pseudolymphoma without istological alterations should be reserved for idiopathic cases. To distinguish a benign from malignant lymphoid lesion elsewhere in the body, the integrity of the architecture of lymph node is an important criterion (2). In the orbit,

however, lymph nodes are not present and histological distinction, between lymphocytic infiltration from benign and malignant lymphatic tumours is often difficult.

Case report

A 33 years male presented with gradually progressive proptosis of right eye lasting four months. Ocular examination shows a firm irregular and nontender palpable mass above and to the lateral side of the eyeball, displacing it downwards and slightly outwards; the movements were equal on both sides. Systemic and haematological examination revealed no abnormality. CT exam revealed a pathological structure in the superior eyelid extending medially (Fig. 1).

An anterior orbitotomy was performed; the histopathological lesion consisted of lymphoid tissue, mixed with fibrovascular septa and adipous areas (Fig. 2 A). The lacrimal gland (Fig. 2 B) was also involved. Polyclonal B areas, with plasma cells positive to both kappa (Fig. 2 C) and lambda (Fig. 2 D) immunoglobulin heavy chains were easily recognizable. The lesion was diagnosed as lymphoid hyperplasia.

The postoperative course was uneventful and the patient was discharged in third day with a therapy with prednisolone without evidence of recurrence after 10 month follow-up.

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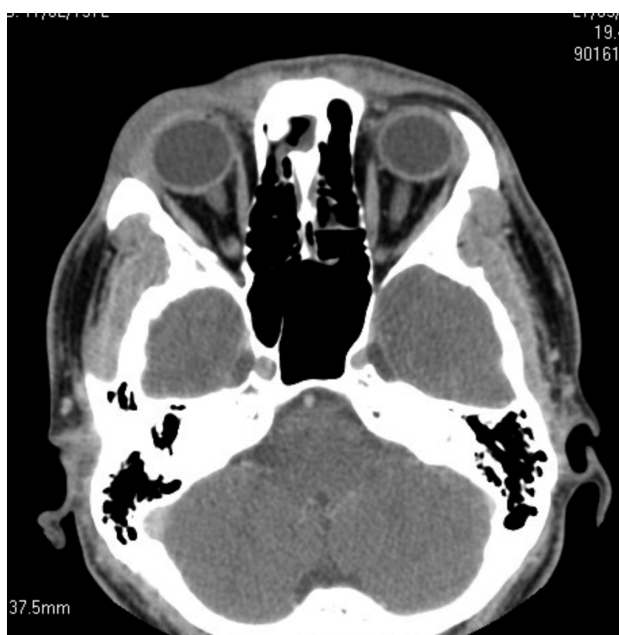


Fig. 1 - CT scan shows a right mass in the superior eyelid and lacrimal gland extending medially and posteriorly.

Discussion

Inflammatory or reactive lesions clinically and pathologically can mimic malignant tumors and are described by a variety of terms, including inflammatory pseudotumor or pseudolymphoma. Inflammatory pseudotumors usually present abruptly and are painful, whereas pseudolymphomas are usually painless. Pseudolymphomas are rare nonneoplastic processes that are pathologically composed of reactive B-cell follicles in a background infiltrate of T lymphocytes, plasma cells, and varying degrees of fibrosis (3). They have been reported in various nodal or extranodal sites, such as skin, breast, lung, pancreas, brain, and liver.

Orbital pseudolymphoma is a partially descriptive term meaning collections of lymphocytes, often accompanied by occasional plasma cells, that accumulate within particular adnexal locations around the eye (4). A popular question for the pathologist is: "Is it lymphoma or pseudolymphoma?"; the answer is not always easy, except in the typical cases.

The diagnostic criteria for the malignant lymphomas are well established and need not be reiterated here. It is the pseudolymphomas which merit our recognition and consideration. Commonly the pseudolymphomas are extranodal. The gross appearance is not usually helpful. Microscopically there is generally preservation of a follicular pattern with the presence of germinal centres, although in some instances this may be largely obscured; there is a background suggestive of granulation tis-

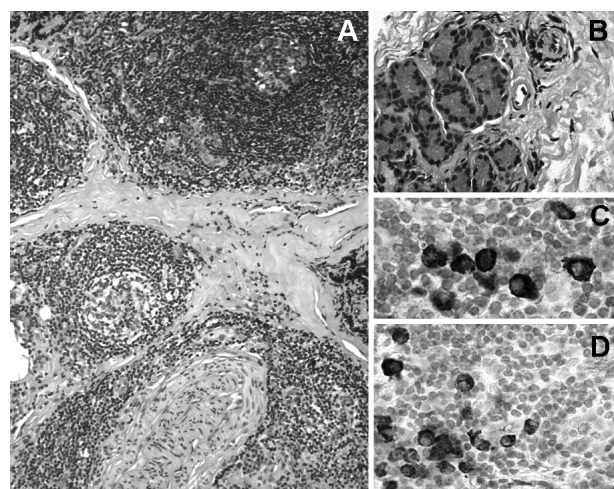


Fig. 2 - Follicles with germinal centres inside lymphoid tissue, incompletely subdivided by fibrovascular septa (A). Lacrimal gland with focal lymphocytes and abundant fibrous tissue (B). Plasma cells positive to kappa (C), and lambda (D) immunoglobulin heavy chains. (A, B, haematoxylin and eosin; original magnification: A, x100. B, x200. C, D, immunohistochemistry, diaminobenzidine; original magnification, x400).

sue and a polymorphism of cells but no Reed-Sternberg cells. There may be discrete or focal granulomas; and fibroblastic or fibrocytic proliferation. Giant cells of the foreign body or Langhans type may be encountered (5).

Patients usually present with a painless mass in the lacrimal gland that produces ptosis, proptosis, diplopia, generalized eye swelling, or disturbance of vision. The mass, if anterior enough, can be seen as a typical salmon patch on the conjunctiva or, if deeper, can be imaged with either computed tomography (CT) or magnetic resonance imaging (MRI).

Orbital pseudolymphomas are usually idiopathic and are distributed in the lacrimal gland, conjunctiva, eyelid, and uvea, in that order of frequency (6). It is not possible to differentiate a benign from a malignant orbital mass on physical examination or by CT or MRI. Biopsies should be performed on all masses using an excisional technique to differentiate pseudolymphoma from lymphoma. Biopsy specimens typically reveal a chronic inflammatory process composed of numerous hyperplastic lymphoid follicles in a background infiltrate of T lymphocytes, polytypical plasma cells, and fibrosis.

Treatment of pseudolymphomas has traditionally been with corticosteroids or external beam radiation therapy (EBRT). These treatments are often initially effective, but patients frequently experience relapse after corticosteroid therapy has been tapered or require long-term therapy that is associated with unacceptable adverse effects (7, 8). Although relapse is less likely with EBRT, dry eye, cataract, and radiation retinopathy are possible adverse effects.

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