Orbit

The International Journal on Orbital Disorders, Oculoplastic and Lacrimal Surgery

ISSN: 0167-6830 (Print) 1744-5108 (Online) Journal homepage: www.tandfonline.com/journals/iorb20

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To cite this article: Zachary J. Berbos, Michael S. Lee, Renzo A. Zaldivar, Stefan Pambuccian & Andrew R. Harrison (2010) Intravascular Lymphoma Presenting as an Orbital Mass Lesion: A Case Report, Orbit, 29:2, 91-93, DOI: 10.3109/01676830903336288

To link to this article: https://doi.org/10.3109/01676830903336288

Published online: 16 Apr 2010.

Orbit, 29(2), 91–93, 2010

ISSN: 0167-6830 print/ 1744-5108 online
DOI: 10.3109/01676830903336288

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ORIGINAL ARTICLE

Intravascular Lymphoma Presenting as an Orbital Mass Lesion: A Case Report

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ABSTRACT

We describe a case of intravascular lymphoma that presented as an orbital mass lesion. A 77-year-old female presented with longstanding fatigue and one-month of diplopia. Magnetic resonance imaging (MRI) revealed a frontal lobe brain lesion and a right orbital mass. Brain biopsy was interpreted as anaplastic oligodendroglioma. Orbital biopsy revealed intravascular lymphoma. On review of brain histopathology, the diagnosis was revised to CNS intravascular lymphoma. To the best of our knowledge, this case represents the first report of intravascular lymphoma presenting as an orbital mass lesion.

KEYWORDS: Orbit; Lymphoma; Intravascular

INTRODUCTION

Intravascular lymphoma (IVL) is a rare, large B-cell non-Hodgkin's lymphoma (Pfleger and Tappeiner, 1959; Wick et al., 1986) characterized by growth of neoplastic cells within lumina of small blood vessels. Intravascular lymphoma commonly presents with cutaneous and/or central nervous system symptoms. A previous case series described intraocular involvement presenting with reduced visual acuity (Elner et al., 1986). We report a case of IVL presenting as an orbital mass.

Case Report

A 77-year-old woman complained of chronic malaise and diplopia for 4 weeks. Medical history was significant for breast cancer 34 years prior, and a 4-month history of pituitary adenoma. One month prior to our evaluation she underwent brain biopsy of a right fronto-temporal lesion at an outside institution that was interpreted as anaplastic oligodendroglioma (Figure 1).

Examination revealed visual acuities of 20/40 OD and 20/50 OS. The right pupil was 1mm larger than the left and poorly reactive with an afferent papillary defect. Slit-lamp examination revealed marked nuclear sclerosis. External examination showed ptosis of the right upper lid. One millimeter of proptosis was noted OD.

Magnetic resonance imaging and computed tomography scans demonstrated a right superotemporal intraconal mass between the superior and lateral rectus muscles that enhanced with gadolinium (Figure 2). There was no bony destruction and the lesion molded to the globe.

Superior transconjunctival orbital exploration was performed under general anesthesia. Dissection into sub-Tenon's space revealed a large intraconal mass. Multiple biopsies were obtained and sent for pathologic evaluation. The orbital fat and lacrimal gland

Received 29 December 2008; revised 17 June 2009; accepted 27 June 2009

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appeared histologically unremarkable. A small focus of vascular prominence contained intraluminal cells with high nuclear to cytoplasmic ratios and occasional prominent nucleoli (Figure 3). A panel of immunoperoxidase stains confirmed the histologic suspicion of IVL. The atypical intravascular cells expressed CD45, CD20, and CD5, but not CD3 or CD10 or CD30. CD34 and CD31 stains highlighted the endothelial cells confirming the intravascular location of the neoplastic cells (Figure 4). On review of the brain histopathology from an outside institution, the diagnosis of this lesion was revised to CNS intravascular lymphoma.

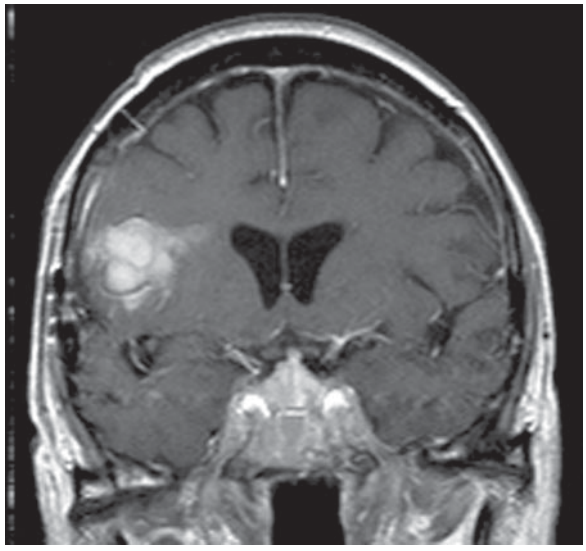

FIGURE 1

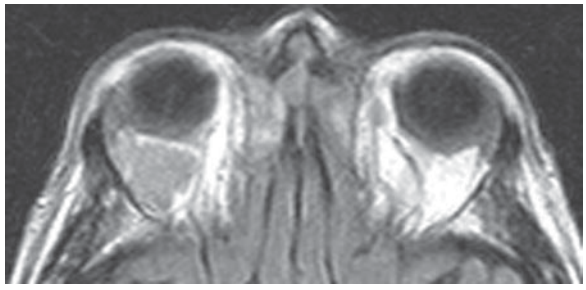

FIGURE 2

COMMENT

Intravascular lymphoma is a rare extranodal large B-cell non-Hodgkin's lymphoma, characterized by widespread intravascular proliferation of neoplastic cells. The tumor predominantly affects middle-aged to older patients, without predilection for either sex (Ferreri et al., 2004). Clinical presentation is heterogeneous, making diagnosis elusive. Half of IVL cases are diagnosed at autopsy, where involvement of the brain is followed in frequency by involvement of the bone marrow, adrenal, prostate, and kidney. Symptoms result from ischemic end-organ dysfunction secondary to microvascular occlusion. The most common presentations are cutaneous lesions involving the trunk and lower legs and/or non-specific neurological manifestations. Diagnosis is histological, based on biopsy of presenting lesions (Gill et al., 2003). The reason for confinement of neoplastic cells

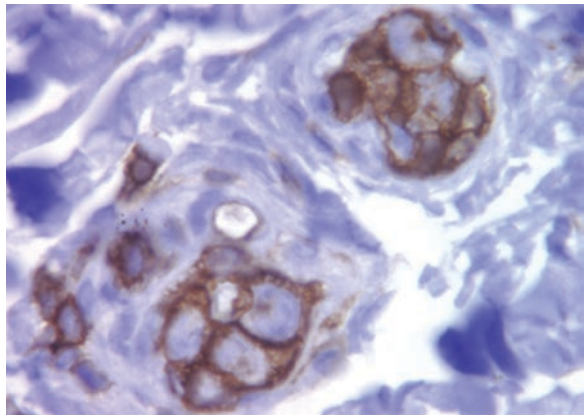

FIGURE 4

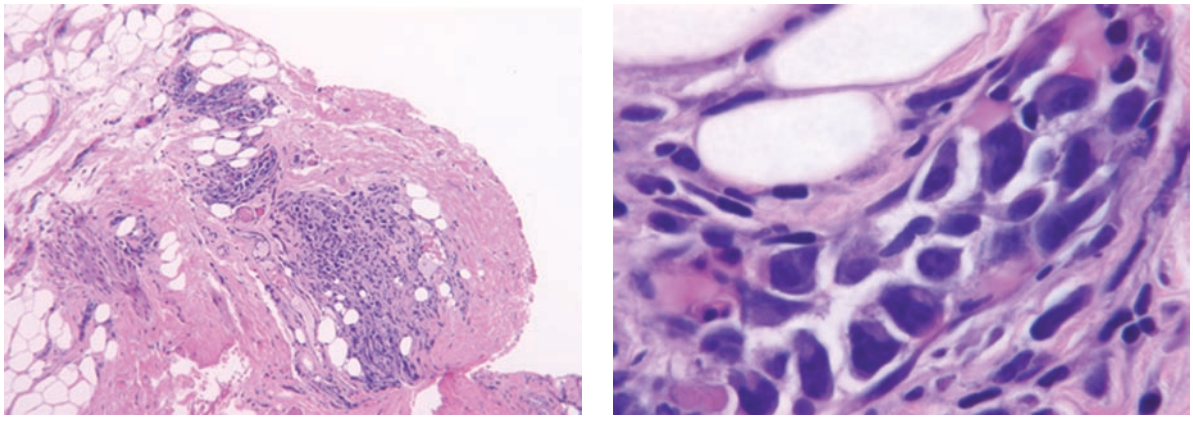

FIGURE 3

within vascular lumina is unknown but may involve abnormalities of adhesion molecule expression (Jalkanen et al., 1989; Kanda et al., 1999; Ponzoni et al., 2000).

Histologically, intravascular lymphoma is composed of neoplastic lymphocytes with large vesicular nuclei, prominent nucleoli, and scant basophilic cytoplasm. Immunohistochemical analysis typically identifies the lymphocytes as B-cells. T-cell, histiocytic, and NK-cell variants have been described, but are exceedingly rare (Wu et al., 2005). The cells are found embedded within a network of fibrin, forming thrombi in lumina of small vessels.

Treatment of intravascular lymphoma consists of anthracycline-based (CHOP) chemotherapy with or without radiotherapy. Despite aggressive therapy, prognosis is poor. Following the diagnosis of IVL, our patient was offered a palliative regimen of radiation and rituximab. Unfortunately, the patient deteriorated rapidly and died less than one month following diagnosis.

Although lymphomas comprise 6 to 8% of all orbital tumors (Bardenstein, 2005) and almost all histologic types of lymphoma have been described in the orbit, we could not find any previous reports of orbital IVL.

***Declaration of interest*:** Supported in part by Research to Prevent Blindness (NY, NY) and the Lions and Lionesses of Minnesota. The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper

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