Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/survophthal

Clinical challenges

An old disease in an atypical place

Laura Pelegrín, MD, PhD[a,*], *Alfredo Adán, MD, PhD*[a], *Armando López-Guillermo, MD*[b], *Antonio Martinez, MD, PhD*[c], *Carol L. Shields, MD*[d]

[a] *Department of Ophthalmology, Hospital Clínic de Barcelona, Barcelona, Spain*
[b] *Department of Hematology, Hospital Clínic de Barcelona, Barcelona, Spain*
[c] *Department of Pathologic Anatomy, Hospital Clínic de Barcelona, Barcelona, Spain*
[d] *Ocular Oncology Service, Wills Eye Hospital, Philadelphia, PA, USA*

ARTICLE INFO

Article history:
Received 16 January 2014
Accepted 21 January 2014
Available online 28 May 2014
Peter Savino and
Helen Danesh-Meyer, Editors

(In keeping with the format of a clinical pathological conference, the abstract and key words appear at the end of the article.)

1. Case report

A 72-year-old man presented with 6 months of progressive decreasing visual acuity in his left eye (LE), and complaining of redness which persisted despite topical steroidal therapy. He also developed increased intraocular pressure which responded to treatment with fixed-combination brimonidine 0.2%–timolol 0.5% twice a day.

His medical history included osteoporosis, being treated with risedronate sodium and calcium, and prostatic hypertrophy. Three of his brothers died from colon cancer.

Best corrected visual acuity was 20/25 in his right eye (RE) and 20/100 in LE. Adnexae and ocular motility were normal. Slit lamp examination revealed 1+ cells in the anterior chamber and granulomatous keratic precipitates in LE. The intraocular pressures were 12 mm Hg RE and 24 mm Hg LE. Fundoscopy revealed trace vitreous cells and a choroidal infiltrate with demarcation lines in the mid-peripheral (Fig. 1A). The right eye was normal.

What is your initial formulation of this patient?

What tests would you perform?

1.1. *Comments by Carol L. Shields, MD*

The clinical features in this older patient include unilateral vision loss, anterior chamber reaction, elevated intraocular pressure, and choroidal infiltrate. The combination of these features are suggestive of an inflammatory, infectious, or neoplastic condition. The presence of temporal macular choroidal folds could represent hypotony, hyperopia, compression from orbital tumor, or choroidal tumor/inflammatory infiltration.

To better understand the anterior segment findings, ultrasound biomicroscopy or anterior segment optical coherence tomography (OCT) could delineate the angle and indicate if there is angle closure or a solid mass. The choroidal infiltrate could be imaged with fluorescein angiography to ascertain the vascular pattern of the choroidal findings and detect any related retinal vascular leakage. Enhanced depth imaging OCT would be particularly beneficial to appreciate the specific pattern of infiltration and related retinal findings. Metastatic tumors tend to show a slightly "lumpy bumpy" topography, whereas lymphoid tumors show a more "rippled or seasick"

* Corresponding author: Laura Pelegrín, MD, PhD, Institut Clínic d'Oftalmologia (ICOF), Hospital Clínic i Provincial de Barcelona, C/Sabino Arana n° 1, 08028 Barcelona, Spain.
E-mail address: laurapelegrin79@hotmail.com (L. Pelegrín).
0039-6257/$ – see front matter
http://dx.doi.org/10.1016/j.survophthal.2014.01.011

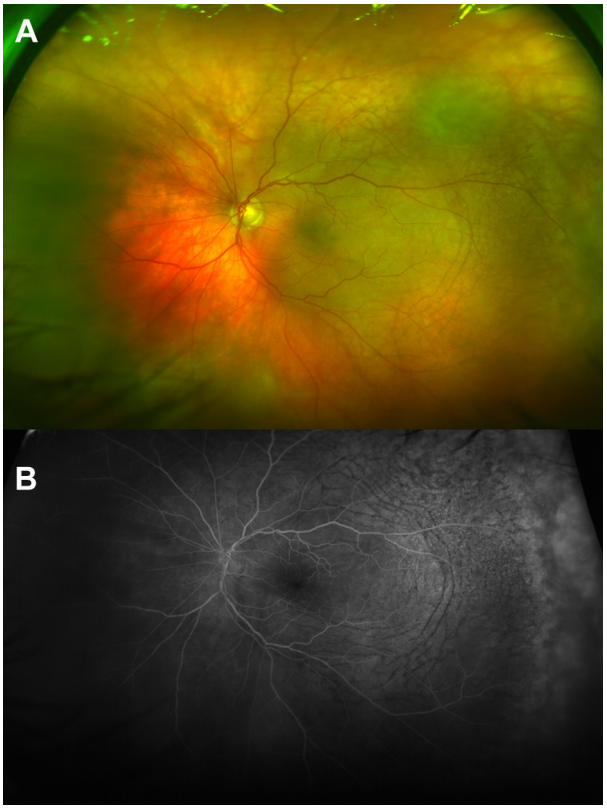

Fig. 1 – Clinical findings. A: Fundus photograph shows a choroidal infiltrate with signs of chronicity such as demarcation lines in the mid-periphery of the retina. B: Intravenous fluorescein angiogram shows early hypofluorescence with moderate, late hyperfluorescence.

topography. Equally important, standard B-scan ultrasonography could assist in identifying extent of mass and presence of extraocular component. Magnetic resonance imaging could be useful to appreciate deeper soft tissue orbital infiltration.

Laboratory testing would be helpful to evaluate for tuberculosis (PPD, quantiferon gold), sarcoidosis (chest radiography and ACE), and systemic infection (blood culture, complete blood count).

2. Case report (continued)

Intravenous fluorescein angiography showed early hypofluorescence with moderate late hyperfluorescence (Fig. 1B). B-scan demonstrated a choroidal lesion with a low internal acoustic reflectivity and no retinal detachment (Fig. 2A). OCT showed irregular changes at the retinal pigment epithelial and choroidal level.

Complete blood count, renal and liver function tests, and urinary sediment were normal. Angiotensin converting enzyme was elevated (91.9 U/L), and quantiferon test was normal. Computerized tomography (CT) of the thorax, abdomen, and pelvis showed lymph node enlargement. CT scan of the head and orbits confirmed choroidal thickening of 3 mm with contrast uptake (Fig. 2B). The ocular muscles and lacrimal gland were normal, and there was no intracranial mass. Oral prednisone at 0.5 mg/kg/day was initiated with a weekly taper.

One month later, he was taking 5 mg/day of prednisone; visual acuities were 20/20 RE and 20/50 LE. Intraocular pressures were 15 mm Hg in both eyes. Slit-lamp examination LE revealed sectorial conjunctival hyperemia, trace cell in the anterior chamber, and a nodular iris thickening for two clock hours (Fig. 3). In addition an abnormal dilated iris vessel accompanied the iris thickening. The choroidal infiltrate remained unchanged.

What would you do now?

3. Comments (continued)

He did not respond to a fairly low dose of oral corticosteroids, implying that this was not sarcoidosis or other inflammatory processes. The elevated ACE level might suggest sarcoidosis, but the fundus pattern is not typical of the classic multifocal choroidal infiltration found with this disease. The clinical features are more strongly suggestive of a neoplastic

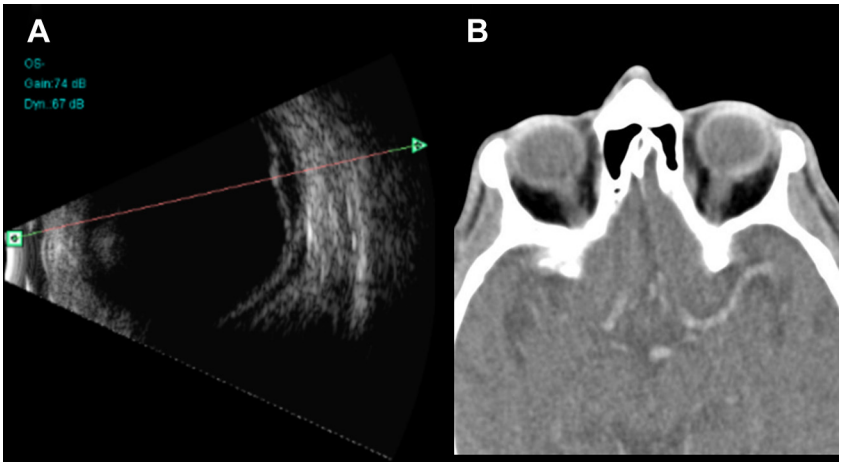

Fig. 2 – Ancillary tests. A: B-scan of the choroidal lesion with low internal acoustic reflectivity and no retinal detachment B: CT scan of the head and orbits confirmed a choroidal thickening of 3 mm with contrast uptake.