European Journal of Ophthalmology / Vol. 16 no. 6, 2006 / pp. 895-897

Primary localized orbital amyloidosis: A case report

R. DI BARI[1], S. GUERRIERO[1], G. GIANCIPOLI[1], A. CANTATORE[1], G. SBORGIA[1], D. PISCITELLI[2]

[1]Department of Ophthalmology and ORL, Division of Ophthalmology
[2]Department of Pathological Anatomy, University of Bari, Bari - Italy

PURPOSE. *Amyloidosis refers to a heterogeneous group of disorders associated with deposition of chemically distinct fibril proteins. Isolated orbital amyloidosis is a rare condition and requires systemic examination. The authors report a case of amyloid deposit in the orbit whose systemic investigation has been negative.*
METHODS. *A 64-year-old woman presented to the eye clinic with left-sided orbital mass, mild exophthalmos with downward ocular displacement, and ptosis. The patient presented also visual acuity loss and ocular hypertension. No systemic involvement was noted by systemic workup and it confirmed the primary orbital amyloidosis.*
RESULTS. *The patient was subject to full clinical examination, laboratory examinations, orbital echography, magnetic resonance imaging and total body computed tomography scans, rectal mucosa, and temporal artery biopsies. It was necessary to exclude systemic amyloidosis. The presence of amyloid deposits was confirmed by biopsy of orbital mass. The chemical nature of deposit was characterized using light microscopy, immunohistochemistry, and electron microscopy.*
CONCLUSIONS. *Orbital amyloidosis is a very rare disease. It should be considered in the diagnosis of patients with ptosis and exophthalmos. The treatment usually consists of surgical removal of the amyloid mass and follow-up for a likely local recurrence. In our case, mass excision determined the decrease of exophthalmos and intraocular pressure. (Eur J Ophthalmol 2006; 16: 895-7)*

KEY WORDS. *Amyloidosis, Echography, Exophthalmos, Mass histopathology, Orbital MRI, Ptosis*

Accepted: May 23, 2005

INTRODUCTION

Amyloidosis is a group of disorders characterized by extracellular deposition of abnormal insoluble protein which is generally called amyloid. Amyloidosis is a rare disease (1).

The classification of amyloidosis is based on the tissue distribution of amyloid deposits (local or systemic amyloidosis), the absence or presence of preexisting disease (primary or secondary amyloidosis), and the chemical type of amyloid protein fibril. The major forms of this disease are primary systemic, secondary, and familial or hereditary amyloidosis.

Many types of amyloidosis have been identified. Each one is associated with deposits of a different kind of protein. In spite of this, all forms of amyloid present common light microscopy properties (eosinophilia, PAS +, Congo red +, apple green birefringence under polarized light) and a fibrillar ultrastructure. Amyloid protein accumulates mostly in interstitial area and around blood vessels.

Amyloidosis can involve any organ or system in the body. The heart, kidneys, gastrointestinal system, and nervous system are affected most often. Therefore, amyloid deposition can cause organ failure or death.

Orbital amyloidosis is a very rare disease. It usual-

 1120-6721/895-03$15.00/0

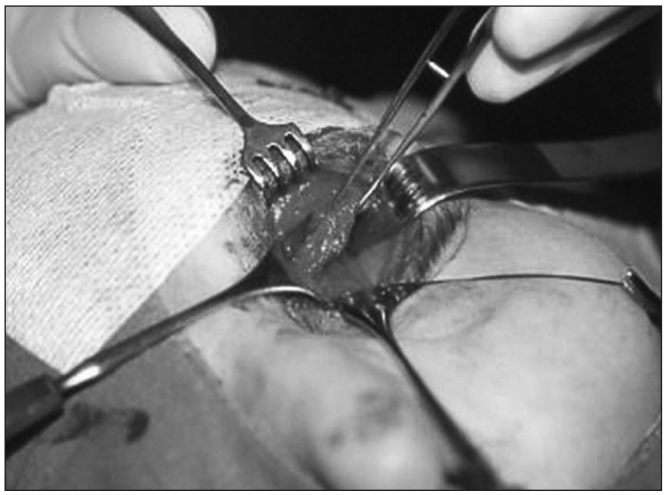

Fig. 1 - *Surgical isolation and individuation of mass under orbital roof.*

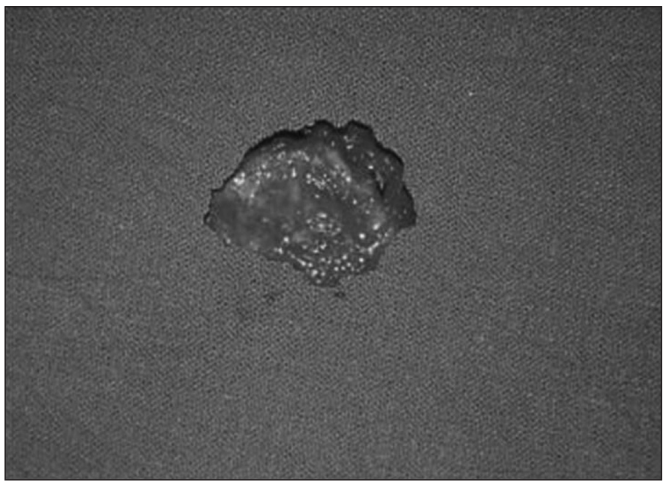

Fig. 2 - *Intraoperative or macroscopic feature of orbital mass removed with surgical resection.*

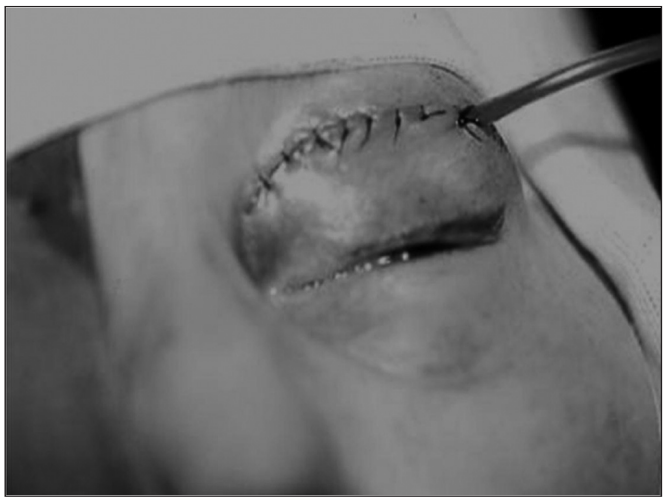

Fig. 3 - *Applications of deep layers and skin sutures and drainage tube.*

ly occurs in primary type, not as part of a systemic disease. Amyloidosis can involve eye and its adnexa. Although amyloidosis prefers eyelid (2) it can involve all ocular structures including anterior segment (3), vitreous (4), retina, choroid (5), optic nerve (6), blood vessel (7), orbit (8), conjunctiva (9), lacrimal gland (10), and extraocular muscles (11).

The most common presenting symptoms of primary localized orbital amyloidosis are proptosis (12), ptosis (13), and global displacement with no visual impairment.

Case report

A 64-year-old woman presented to us in September 2000 with a 3 months' history of orbital mass, proptosis, and ptosis. There was also a severe visual acuity loss of a few days duration in the left eye.

External examination revealed a mild proptosis with eyeball displaced anteroinferiorly and orbital firm mass in the upper portion of left orbit. The lesion was covered by hyperemic skin; it was also fixed on tissues below and it was of elastic-hard consistency.

The rest of the ocular examination revealed ocular hypertension (30 mm Hg) not responsive to medical therapy (14) and visual loss. The woman presented with a best-corrected visual acuity of 6/30 in the left eye. Fundus examination showed an edematous and pallid optic disc. Fluorescein angiography confirmed the acute ischemia of optic nerve head.

Orbital echography showed an extraconal firm mass, extended in retrobulbar area following superior rectus muscle. It was of middle-low reflectivity.

Orbital magnetic resonance imaging (MRI) revealed a soft hypointensity mass on T2, hypointensity in T1, and homogeneous contrast enhancement.

The treatment was surgical removal of mass. Surgical procedure was as follows:

1) Anterior orbitotomy
2) Isolation and individuation of mass under orbital roof (Fig. 1)
3) Removal of mass (Fig. 2)
4) Applications of deep layers and skin sutures and drainage tube (Fig. 3)

The biopsy of the orbital mass showed the presence of amyloid deposition in interstitial area and maximally around blood vessels. There was also infiltration of plasma cells admixed with granulomatous foreign body response. The excised material showed ex-

tensive amorphous eosinophilic material that stained by Congo red, which imparts an orange pink tint to amyloid deposit. When viewed by polarization microscopy it showed an apple green birefringence. The immunohistochemistry method using anti-human amyloid component monoclonal antibody was performed as final marker of amyloid.

Once the diagnosis of orbital amyloidosis was confirmed, additional laboratory tests and imaging procedures were performed to determine which organs or systems were affected. Full clinical examination, blood and urine tests, total body computed tomography scan, rectal mucosa, and temporal artery biopsies were normal.

DISCUSSION

In this case, our patient developed ocular hypertension because orbital mass obstructed aqueous humor outflow in episcleral veins. There were also local mechanical compressions that may explain ptosis and proptosis with ocular displacement.

Ischemic distress of optic nerve head was produced by deposition of amyloid in orbital arteries walls (especially short posterior ciliary arteries) that determined a marked reduction of blood flow.

The treatment was surgical removal of mass.

The absence of a capsule may determine the infiltration of surrounding tissues.

Total excision is usually impossible and surgery should be directed to the excision of the major part of the mass. For this reason the patient with primary localized orbital amyloidosis must be periodically controlled.

Orbital MRI was repeated after 20 days and showed a nearly complete removal of the mass. In this case excision of the mass determined the decrease of proptosis, ocular hypertension, and ocular soreness.

No author has a proprietary interest.

Reprint requests to:
Roberto Di Bari, MD
via Pietrocola 15
70124 Bari, Italy
robertodibari79@libero.it

REFERENCES

1. Murdoch IE, Sullivan TJ, Mosely I, et al. Primary localized amyloidosis of the orbit. Br J Ophthalmol 1996; 80: 1083-6.
2. Stack RR, Vote BJ, Evans JL, Elder MJ. Bilateral ptosis caused by localized superficial eyelid amyloidosis. Ophthal Plast Reconstr Surg 2003; 19: 239-40.
3. Aldave AJ, Principe AH, Lin DY, Yellore VS, Small KW. Lattice dystrophy-like localized amyloidosis of the cornea secondary to trichiasis. Cornea 2005; 24: 112-5.
4. Sandgren O. Ocular amyloidosis, with special reference to the hereditary forms with vitreous involvement. Surv Ophthalmol 1995; 40: 173-96.
5. Campos EC, Melato M, Manconi R, Autonutto G. Pathology of ocular tissues in amyloidosis. Ophthalmologica 1980; 181: 31-40.
6. Finlay KR, Rootman J, Dimmick J. Optic neuropathy in primary orbital amyloidosis. Can J Ophthalmol 1980; 15: 189-92.
7. Ing EB, Islon Z, Woolf MD. Systemic amyloidosis with temporal artery involvement mimicking temporal arteritis. Ophthalmic Surg Lasers 1997; 28: 328-31.
8. Levine MR, Buckman G. Primary localized orbital amyloidosis. Ann Ophthalmol 1986; 18: 165-7.
9. Lee Hm, Naor J, DeAngelis D, Rootman DS. Primary localized conjunctival amyloidosis presenting with recurrence of subconjunctival hemorrhage. Am J Ophthalmol 2000; 129: 245-7.
10. Conlon MR, Chapman WB, Burt WL, Larocque BJ, Hearn SA. Primary localized amyloidosis of the lacrimal glands. Ophthalmology 1991; 98: 1556-9.
11. Banerjee S, Bogman J, Reuser TT. Amyloid deposition in the extraocular muscles. Orbit 1999; 18: 105-6.
12. Martinez del Valle F, Selva O'Callaghan A, Solans Laque R, et al. Proptosis and orbital tumor in an 80-year-old woman. Primary orbital amyloidosis. Rev Clin Esp 2001; 201: 95-6.
13. Dinakaran S, Singh AD, et al. Orbital amyloidosis presenting as ptosis. Eye 2005; 19: 110-2.
14. Nelson GA, Edward DP, Wilensky JT. Ocular amyloidosis and secondary glaucoma. Ophthalmology 1999; 106: 1363-6.