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Case Report

Surgical treatment of a rare case of bilateral ptosis due to localized ocular amyloidosis

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Abstract

We describe a rare case of a 31-year old woman with bilateral ptosis due to localized amyloidosis. She referred a nine-year history of ptosis and surgical treatment with frontalis suspension three years previously. Following complete ophthalmological examination and evaluation of the ptosis we carried out tarsal and fornix biopsy, which revealed accumulation of a weakly eosinophilic amyloid positive substance. We performed surgical correction using the levator aponeurosis–Müller's muscle complex re-adaptation technique and amyloid substance debulking in all the palpebral layers in the left eye. The material obtained was stained with hematoxylin–eosin, Congo Red, PAS and alpha-actin, which confirmed amyloid deposition. Successively, the right eye was operated in the same manner and entropion was managed by dissection and removal of amyloid from subconjunctival layers. Five years following surgery, the corrective procedure for ptosis was still effective. Surgical treatment of ptosis is very complex and requires precise indications. Appropriate management depends on the etiopathogenesis, accurate diagnosis, and clinical findings.

Keywords: Ptosis, Pseudoptosis, Amyloidosis, levator aponeurosis–Müller's muscle complex re-adaptation

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Introduction

Amyloidosis is a condition characterized by the deposition of amorphous proteinaceous material in tissues. It is divided into systemic or localized forms.[1,2] Ocular and palpebral amyloidosis is a rare event and amyloid can be deposited in any part of the orbit, globe, or adnexa.[3–5]

Clinical presentation of periocular or orbital amyloidosis is variable depending on the deposited quantity and the tissues involved. A common clinical sign is ptosis (54%) where the degree of eyelid drooping is due to combination of ptosis and pseudoptosis,[3–7] the later being defined as a condition caused by a group of disorders that simulate ptosis without the malfunction of the levator muscle.[8]

We report a rare case of bilateral localized amyloidosis with ptosis treated using the levator aponeurosis–Müller's muscle complex re-adaptation technique[9] and amyloid substance debulking in all the palpebral layers.

Case report

A 31-year-old woman presented with severe bilateral ptosis (Fig. 1) and she referred onset of the pathology nine years before with gradual worsening accompanied by general

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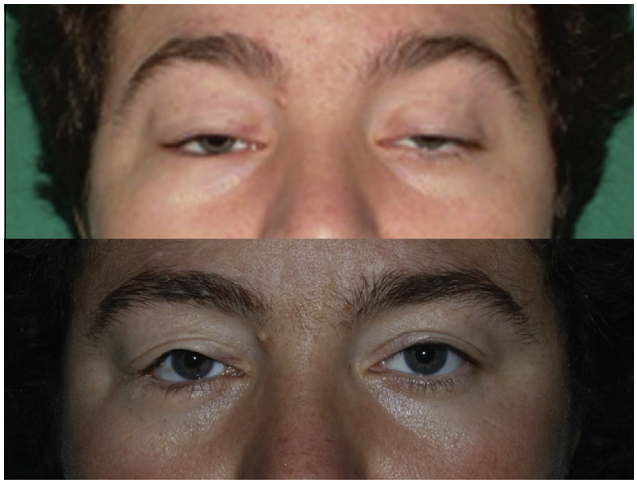

Figure 1. Ptosis due to amyloidosis and results after surgery. Upper image: frontal view before treatment; lower image: frontal view five years after levator aponeurosis–Müller's muscle complex advancement treatment.

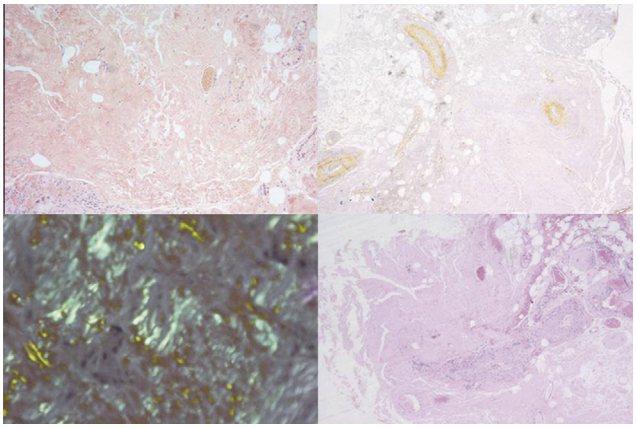

Figure 2. Histological staining of amyloid. Superior right: Congo-Red staining detecting amyloid deposition; Inferior right: Congo-Red showing apple-green birefringence in polarized light. Superior left: α-actin staining. Inferior left: PAS staining showing not abnormal deposition of glycogen.

fatigue. Diagnosis of myasthenia had been excluded by means of blood tests and instrumental examination. The medical history of the patient was negative for multiple myeloma or lymphoproliferative disorders. The family history was negative. The patient referred frontalis suspension surgery with silicone rods three years before. On presentation to our unit, nine years after initial presentation, the patient presented eyelid and subcutaneous swelling and severe ptosis with a palpebral fissure measurement of 4 mm and 3 mm in the right and left eye, respectively. There was entropion of the right eye. Levator muscle function was almost absent. Lid eversion showed a conjunctival component of amyloidosis. Slit lamp examination did not show other ocular alterations and dilated fundus examination with ophthalmoscopy was unremarkable in both eyes.

A tarsal and fornix biopsy in the left eye revealed accumulation of a weakly eosinophilic amyloid positive substance. Systemic investigations, including serum and urine protein immunoelectrophoresis, showed no abnormality. The ptosis in the left eye was surgically treated with local anesthesia; the silicon rods were removed then through a cutaneous incision approximately 8 mm from the eyelid margin, an ellipse of skin, subcutaneous and orbicularis oculi muscle was removed, the levator aponeurosis–Müller's muscle complex was isolated and dissected, eight millimeters of the isolated complex were then inserted into the anterior aspect of the tarsal plate (measuring 4 mm over the latter), based on the preoperative degree of ptosis and levator function. Meticulous debulking of the semi-gelatinous substance, revealed during dissection of each layer was performed (i.e. muscle, subconjunctiva, subcutis). The material obtained was stained with hematoxylin-eosin, Congo Red, PAS and alpha-actin. Congo Red staining showed green birefringence, and amyloid deposition was confirmed (Fig. 2).

Two months later, the ptosis and lower-lid entropion in the right eye worsened. The same technique was used to correct the ptosis. Entropion was managed by dissection and removal of the amyloid substance from the subconjunctival layer without lateral canthoplasty.[10]

In the examinations following surgery the down-up test in both eyes showed a palpebral fissure measurement of 7 mm in upward gaze, 6 mm in the primary position and 2 mm in the downward gaze. Five years following surgery, the corrective procedure for ptosis was still effective (Fig. 1).

Discussion

Amyloidosis is a disease characterized by extracellular deposition of a fibrillar proteinaceous insoluble material due to abnormal production or a deficit of elimination of these proteins.[2,3]

Local amyloidosis can be caused by deposition of monoclonal immunoglobulin light chains (κ or λ) usually by benign B-cell or plasma cell clones.[3–6] Periocular and ocular amyloidosis is rare and unilateral forms are more frequent. The largest case series of ocular and periocular amyloidosis, composed of 24 patients, was recently reported by Liebovitch, where the mean age of patients was 57 ± 17 years with a female to male ratio of 15:9.[5]

According to Liebovitch, ocular and periocular amyloidosis can be presented as a subconjunctival or subcutaneous mass (95.8%), eyelid ptosis (54.2%), pain (25%), exophthalmos (21%), alteration in muscular activity (16.6%) and recurrent hemorrhage (12.5%).[5]

The symptoms vary according to extent of the disease and the structures involved.

The time between onset and diagnosis of the disease has been reported up to 11 years.[5] Although the 31 year-old female patient described herein was younger than patients reported previously, and nine years had passed before diagnosis was made. Pseudoptosis was reported for the first time by Uchida in 1962[11] and classified as a subgroup of ptosis by Beard in 1969[8]; it is a condition caused by a group of disorders that simulate ptosis with the lowering of the eyelid but without the malfunction of the levator muscle.[8,11] In the present case drooping of superior eyelid was due to the accumulation of amyloid substance in all eyelid layers: subcutaneous, subconjunctival, interstitial space of muscle, and Whitnall's ligament, thus, increasing the weight of the eyelid and creating a greater effort for the aponeurotic muscle complex.

Moreover, accumulation of amyloid substance weakened the muscle and created adherence within Whitnall's ligament and the levator muscle aponeurosis exercising mechanical check. Therefore, this case could be classified as ''true ptosis'' as apposed to ''pseudoptosis'' since it was caused by both mechanical accumulation of amyloid in the lid lamella and malfunction of the levator muscle.

Management of ptosis can be a challenge. Myasthenia must be ruled out. The paucity of local specific symptoms in rare cases can lead to incorrect diagnosis. Magnetic resonance imaging and computed tomography can be important to localize orbital structures that are involved and to evaluate the presence of amyloid material in muscular tissue.[5,12] Furthermore, surgical treatment is very complex and requires precise indications. Appropriate management depends on the etiopathogenesis, accurate diagnosis, and clinical findings. Frontalis suspension is indicated in cases of absent levator muscle functionality,[13,14] and although it was not indicated in this case of ptosis it had been performed as the diagnosis of amyloidosis had not been made. Indeed, management of ptosis in these cases is based on removal of localized amyloid,[4,5] levator muscle advancement, levator resection,[3] the Fasanella technique and, more rarely, tarsal and conjunctival resection, combined with reconstruction of the tarsus with conchal cartilage and levator muscle complex reinsertion.

In the case reported herein two factors required consideration: the mechanical component of the ptosis linked to the presence of amorphous material in the lid lamella which can interfere with a correct evaluation of levator muscle functionality, and involvement of the levator muscle complex. Residual levator muscle functionality and the possibility of tissue examination during surgery led us to choose the levator aponeurosis–Müller's muscle complex re-adaptation technique and amyloid substance debulking. Indeed, the previous suspension technique, where tissues cannot be examined during surgery, had not been successful. The choice of the appropriate surgical procedure was, thus, confirmed by evaluation of levator functionality during surgery performed in local anesthesia, and post-surgical testing.

Conclusions

In the present case the correction of ptosis using levator aponeurosis–Müller's muscle complex advancement and reinsertion into the tarsus plate, yielded a good degree of correction by providing improved levator muscle function afforded by its release from the septal ligaments and its advancement. Meticulous debulking of the amyloid substance that weakened the muscle and created adherences within Whitnall's ligament and the levator muscle aponeurosis contributed to the good functional and esthetic results obtained. Appropriate management of ptosis depends on the etiopathogenesis, an accurate diagnosis, and clinical findings. Amyloidosis of the levator muscle may be a precursor to orbital amyloidosis; therefore, good surgical results should not lead to complacency for careful surveillance and regular follow-up of patients over time.

Conflict of interest

The authors declared that there is no conflict of interest.

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