

Primary Amyloidosis of the Eyelid: A Case Report

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Abstract. *The Authors describe a case of primary localized amyloidosis of the eyelids, focusing on diagnosis and possible treatment. The involvement of several structures of the eyelid such as muscles, conjunctiva and puncta lacrimalia, as well as peculiar features of these proteinaceous deposits, suggest different combined procedures and precise timing in choosing the appropriated therapy.*

Amyloidosis is mainly a systemic disease with possible localization involving different organs. These deposits have a specific tropism for connective tissue and perivascular spaces and are characterized by an abnormal extracellular deposit of an amorphous proteic material with specific ultra structural features. Clinical classification includes five groups: primary systemic, primary localized, secondary to a chronic illnesses, secondary to multiple myeloma, and familiar type. The involvement of the oculo-palpebral region may occur only in the first two forms and may affect different structures such as the cornea, the vitreous body, the lacrimal glands, extraocular striated and smooth (Muller) muscles, the conjunctiva and eyelids (1-4). The clinical picture in conjunctival and eyelid amyloidosis localization may consist of eyelid oedema, papules and yellowish sub-conjunctival plaques (crumbly, easily bleeding and able to affect the entire surface of the palpebral conjunctiva), signs of conjunctival irritation, lumps or swellings and an eyelid ptosis. The ptosis may be due to a mass that dislocates the eyelid free margin or have a functional pathogenesis if the Muller muscle is affected by the disease (5-8).

The diagnosis of eyelid primary amyloidosis is based on symptoms, patient's anamnesis, clinical examination and finally a biopsy specimen (9, 10). Radiological examinations,

tomography or ultrasound are useful for the correct therapeutical approach and to exclude a possible systemic involvement of the disease. The medical treatment of symptomatic primary eyelid amyloidosis is based on the use of ascorbic acid to reduce the bleeding tendency of these lesions and anti-inflammatory drugs, locally or general. The surgical approach is based on the resection of the affected structures, with direct closure of the areas of loss in the initial localized forms, or with more complex and different reconstructive procedures in cases of greater extent of lesions (11). Other therapeutic options are electro-cauterization, cryotherapy, and selective debridement (12).

Case Report

A 30-year-old man affected by a primary amyloidosis bilaterally localized in the eyelids presented with a mild degree of ptosis on the left and a moderate degree on the right side associated with severe oedema of the lower lids (Figure 1). At the clinical examination, moderate swelling, 1 cm large, was apparent in the medial part of right upper eyelid, mobile to the overlying skin, which seemed to cause the downward displacement of the free eyelid margin and the disappearance of the superior orbital sulcus. On the inside of the eyelids, upper and lower, the patient had extensive bilateral yellowish conjunctival vegetations, easily bleeding, which also affected the conjunctival fornix, the internal canthus and puncta lacrimalia (Figures 2 and 3). The patient had routine preoperative laboratory tests, ophthalmological evaluation, palpebral ultrasound and orbital tomographic examinations. The specific tests showed hypoechogenicity of the lesions and the involvement of the surrounding structures (Figure 4). The patient underwent two sessions of a combined treatment with electrocauterization and CO₂ laser vaporization to reduce the conjunctival vegetations and a surgical resection of eyelid swelling, which, intraoperatively, had the appearance of a cyst with a colourless gelatinous content. In the postoperative period the patient had antibiotic therapy by oral (4 days) and topical (14g) administration. No complications were recorded and the patient was discharged on the third day. Follow-up at six months and after 1 year

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Figure 1. *Preoperative frontal view.*



Figure 2. *Preoperative focus on conjunctival deposits on the right eye.*

showed a permanent correction of the eyelid ptosis and a significant reduction of ocular symptoms despite limited residual lesions on the conjunctiva (Figures 5-8).

Discussion

Primary eyelid amyloidosis is a rare clinical entity that often leads to misdiagnosis. Differential diagnosis includes basal cell or squamous cell carcinoma, lachrymal gland carcinoma or lymphoma (13, 14). Ultrasonography and tomography are usually able to suggest the correct diagnosis but they are also mandatory to establish the relationships of the masses with the surrounding structures before surgery. Moreover, general clinical examination and ophthalmologic tests are also indicated to complete the preoperative evaluation which should lead to a correct therapeutic strategy, avoiding partial or multiple treatments. In fact, a surgical approach alone may often be associated with a high risk of incomplete excision and local recurrence, in the early or late postoperative period. Radical surgical excision, even in highly specialized oculoplastic centres, may cause severe impairment of both structure and function of the eyelid (15). For these reasons, other methods such as electro cauterization, cryotherapy, selective curettage and radiotherapy have already been described in addition to surgery as useful tools to remove subconjunctival amyloid vegetations (16).

Our experience confirmed that combined procedures are the gold standard treatment of these lesions. Localized masses should always be excised surgically since amyloid swelling does not present as encapsulated cyst and the deposits are often scattered inside the tissue, simply limited by an area of increased thickness. Particular care must be paid when the Muller muscle or the elevator muscle of the upper eyelid are involved in order to avoid a secondary



Figure 3. *Preoperative focus on the left eye.*

ptosis, as well as when the lesions surround the puncta lacrimalia. The treatment of this area, in fact, must be carried out with extreme caution to avoid iatrogenic injury to lachrymal ducts which can cause permanent epiphora.

Subconjunctival amyloid vegetations are instead to be treated with other methods. In our opinion, the use of CO₂ laser should be preferred due to a lower risk of pathological scarring; radiotherapy and other techniques may result in more severe side-effects and are not to be considered a first choice, especially for a benign disease. Specific attention must be paid to prevent the onset of postoperative

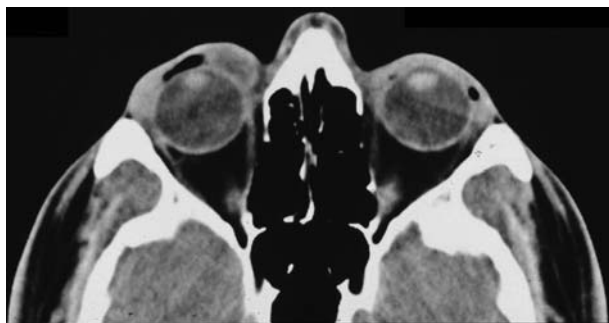


Figure 4. Preoperative CT scan.



Figure 7. Postoperative view of left conjunctiva.



Figure 5. Postoperative frontal view.

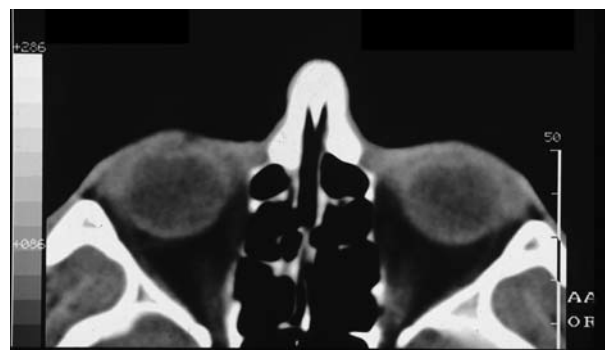


Figure 8. Postoperative CT scan.



Figure 6. Postoperative view of right conjunctiva and punctum lacrimale.

complications such as conjunctiva sinechiae that may occur in the case of mirror lesions. Therefore, amyloid deposits on conjunctival fornixes, whether palpebral or bulbar, should always be treated in multiple sessions in order to limit unfavourable healing (17).

Finally, the timing of surgical procedures should be made on the basis of the symptoms. The indication for a surgical

treatment comes mainly from the patient's clinical presentation. In any case, regarding the frequency of recurrences of this disease, less invasive treatments should be preferred.

Conclusion

The therapeutic issues involved in treating eyelid amyloidosis are still numerous and insoluble. The awareness that there is not any one radical treatment is certainly frustrating for the surgeon, but the benign nature of the disease demands a multidisciplinary approach in qualified centres.

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