

THE USE OF SOFT BANDAGE CONTACT LENSES IN THE MANAGEMENT OF PRIMARY (LOCALISED NON-FAMILIAL) CONJUNCTIVAL AMYLOIDOSIS: A CASE REPORT

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SUMMARY

We present a case of primary (localised non-familial) conjunctival amyloidosis, which is an uncommon condition, characterised by amyloid deposition within the substantia propria of the conjunctiva. Soft bandage contact lenses were fitted in order to protect the cornea from the mechanical abrasion of the irregular surface of the palpebral conjunctiva. We used non-ionic high water content (70%) soft bandage lenses from Filcon 4A with a Dk of 40. The lenses were well tolerated and gave significant relief of the symptoms.

SAMENVATTING

Wij beschrijven een patiënt met primaire (gelocaliseerde niet-familiale) conjunctivale amyloidose. Deze zeldzame aandoening wordt gekarakteriseerd door amyloid neerslag in de substantia propria van de conjunctiva. Zachte verbandlenzen werden aangepast om de cornea te beschermen tegen de mechanische irritatie van het onregelmatige oppervlak van de palpe-

brale conjunctiva. We maakten gebruik van zachte niet-ionische verbandlenzen, gemaakt van Filcon 4A, met een hoog watergehalte (70 %) en een Dk van 40. De lenzen werden goed verdragen en gaven een duidelijke verbetering van het comfort.

RÉSUMÉ

Nous présentons un cas d'amyloïdose conjunctivale primitive (localisée, non-familiale), qui est une affection rare, caractérisée par le dépôt d'amyloïde dans le tissu interstitiel de la conjonctive. Des lentilles de contact souples de bandage ont été posées pour protéger la cornée de l'irritation mécanique due à la surface irrégulière de la conjonctive palpébrale. Nous avons utilisé des lentilles de contact souples non-ioniques de Filcon 4A, ayant une teneur d'eau élevée (70%) et un Dk de 40. Les lentilles ont été bien tolérées avec une nette amélioration du confort.

KEY-WORDS

primary conjunctival amyloidosis, localised non-familial conjunctival amyloidosis, bandage contact lens.

MOTS-CLÉS

amyloïdose conjunctivale primitive, amyloïdose conjunctivale localisée non-familiale, lentilles de contact de bandage

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INTRODUCTION

Amyloidosis is a process characterized by the deposition of amorphous hyaline material in various body tissues. Amyloidosis can be classified as systemic (primary and secondary), localized (primary and secondary), familial and senile (table 1). A variety of proteins are present in amyloidosis: they differ between different forms of the disease. The most common type is protein AL, which is associated with primary systemic amyloidosis and primary and secondary localised amyloidosis. This protein has light chain immunoglobulins as precursors. Multiple myeloma should always be ruled out, because Bence Jones protein may be modified to form protein AL.

The major amyloid deposition in secondary systemic amyloidosis is protein AA. This is a degradation product of a serum acute phase reactant (apoSAA) which is produced by hepatocytes in the presence of underlying chronic infection and inflammation.

Other proteins that have been isolated include protein AF in familial amyloidosis, and protein AS in senile amyloidosis, both related to prealbumin (9,12). When we look at the classification of amyloidosis (table 1), we see two cate-

gories that are of particular relevance to the ophthalmologist:

PRIMARY SYSTEMIC AMYLOIDOSIS is unassociated with predisposing diseases other than multiple myeloma. It involves muscles, skin, nerves and blood vessels. The eye can be affected, causing amyloidosis of the orbit, vitreous, eyelids, nerves and lacrimal gland (24).

PRIMARY LOCALISED AMYLOIDOSIS is characterized by amyloid deposition within a specific tissue or organ. Localised conjunctival amyloidosis is a condition which is caused by amyloid deposition within the substantia propria of the conjunctiva. It is characterized by thickening of the conjunctiva and the eyelids, blepharoptosis and spontaneous eyelid ecchymoses. The palpebral and bulbar conjunctiva get a yellow and waxy appearance.

The primary form of localised conjunctival amyloidosis has no known infectious, traumatic or heredofamilial cause. Herbert (8, 17) was the first to describe a case of "colloid degeneration" of the conjunctiva, which appears to have been an example of primary localised conjunctival amyloidosis. Later on, different case reports were published, describing the disorder as a generally local process, though systemic amyloidosis (3, 5, 6, 14, 21) and especially

Table 1: *Classification of amyloidosis (Adapted from references 9 and 12)*

Classification	Characteristics	Location	Eye involvement	Amyloid Type	
1. Systemic amyloidosis	a. Primary systemic amyloidosis	No evidence of preceding or coexisting disease (except multiple myeloma)	Vessels and muscles of heart, tongue, GI tract, skin, nerve	Vitreous, muscles, pupils, eyelids, lacrimal gland	AL
	b. Secondary systemic amyloidosis	Coexistence of other conditions such as chronic infection, rheumatoid arthritis, lues, TB,...	Spleen, kidney, adrenals, liver	Rare	AA
2. Localised amyloidosis	a. Primary localised amyloidosis	Involvement of a single organ without generalized involvement and without antecedents	Skin, larynx, heart, uterus, tracheobronchial tree, GI tract	Cornea, lids, conjunctiva, orbit	AL
	b. Secondary localised amyloidosis	Involvement of a single organ without generalized involvement, after previous infection, inflammation ,...			
3. Familial amyloidosis					AF
4. Senile amyloidosis					AS



Fig 1. Clinical features of conjunctival amyloidosis after multiple debulking procedures and cryocoagulations. (a) Right eye: characteristic yellow, waxy amyloid deposits in the conjunctiva of the right upper lid. There are multiple conjunctival and subconjunctival scars. (a) Left eye: same features, the typical nodular infiltration of the conjunctiva is even more noticeable.

plasma cell disease should be ruled out. Eye involvement in secondary systemic, familial or senile amyloidosis is very rare. Treatment of conjunctival amyloidosis with various eye drops, cryotherapy, and surgical interventions can be unsatisfactory (10, 13, 19). We believe that the use of soft contact lenses can play an important role in the management of advanced cases.

CASE REPORT

A 67-year-old male presented in June 1994 with a spontaneous hematoma of the right upper lid. He complained about irritation and persistent foreign body sensation at both eyes, especially the right one. Ocular examination revealed a yellow and waxy appearance of the palpebral conjunctiva of the upper eyelids, associated with papillary hypertrophy.

The inferior cul-de-sac of each eye had the same yellow rubbery appearance with folding and redundancy of the conjunctiva. Further ocular examination showed no evidence of previous infection or inflammation. The patient was managed conservatively with tear lubricants, six times daily. In April 1995 he presented with a spontaneous subconjunctival hemorrhage of the left eye. The upper and lower eyelids of both eyes were more thickened and indurated than at the first examination. Fleshy yellow exophytic tumefactions of the palpebral conjunctiva were seen.

Biopsy of the right and left forniceal conjunctiva showed amorphous amyloid deposits within the substantia propria, which demonstrated green birefringence to polarised light following Congo red staining. A light infiltrate of chronic inflammatory cells with predominance of eosinophils was present in the stroma surrounding the amyloid deposits. The specimens demonstrated green fluorescence with Thioflavin staining.

Systemic investigations including FBC, ESR, LFTs, serum and urinary protein electrophoresis and chest radiograph were normal. A rectal biopsy, performed to exclude systemic amyloidosis was negative. The tentative diagnosis of primary localised non-familial conjunctival amyloidosis was confirmed.

During the following years, the conjunctival amyloid deposits of the upper lids of both eyes were debulked several times in order to obtain a more regular surface. Long-term local steroids and lubricants were given. Treatment of the most prominent nodules with cryocoagulation was performed several times.

Even after those surgical procedures and different topical treatments the patient still had a persistent burning sensation and soreness of both eyes.

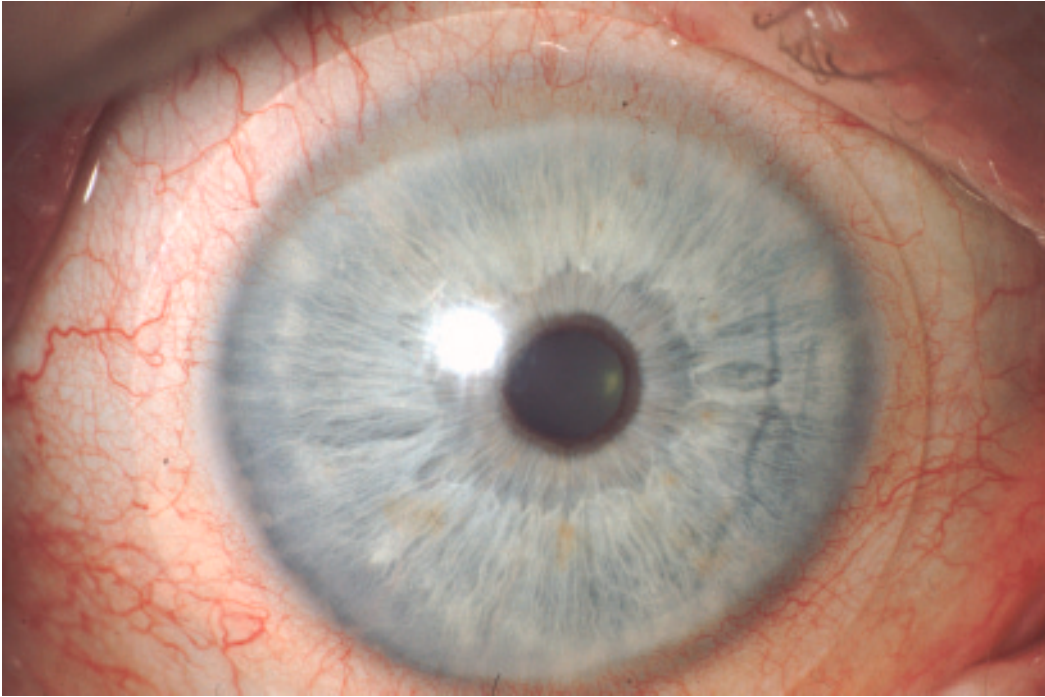
Soft bandage contact lenses were fitted in December 1996 in order to protect the cornea from the mechanical abrasion of the irregular surface of the palpebral conjunctiva. We used non-ionic lenses from Filcon 4A (Exel 70[®], Oculenti Contact Lens Practice, Netherlands) on an extended wear basis: FDA group 4 A, high water content (70%), with a Dk of 40.

The lenses had a base-curve of 8.70 and a large diameter of 18 mm. Every 6 weeks, lenses were manually cleaned and disinfected using a daily cleaning solution. They were rinsed and reinserted with physiological salt solution 0.9 %. After 3 months (or earlier when dirty) lenses were discarded. Up till now, the lenses are well tolerated and give significant relief of the symptoms. The patient is now being treated with hypromellose drops when necessary.

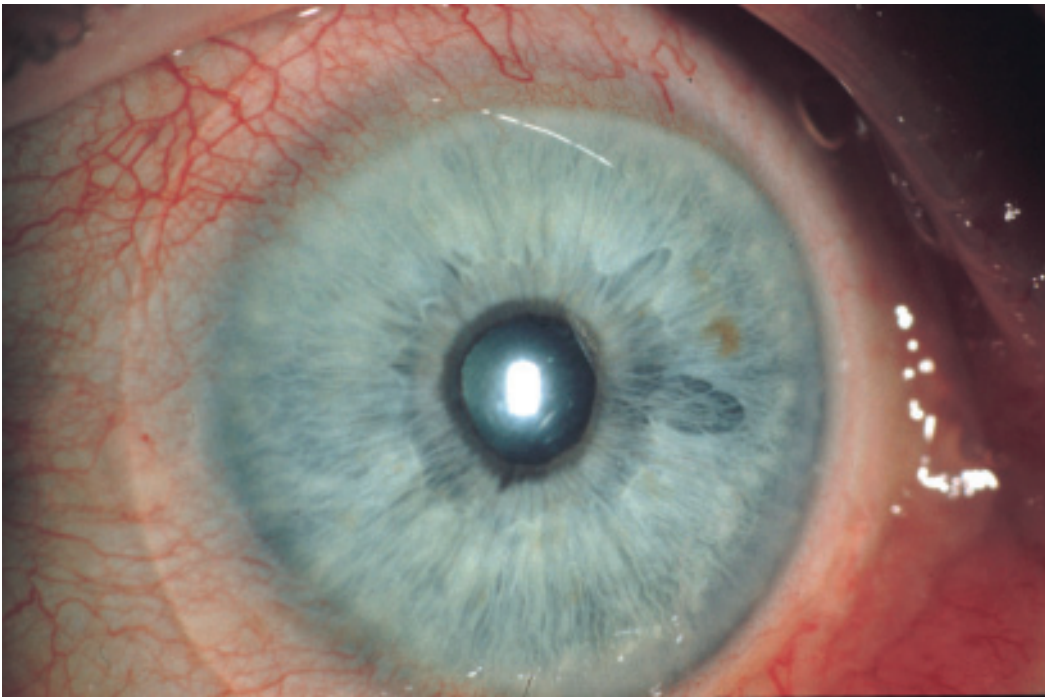
CONCLUSION

The most important therapeutic uses of soft bandage contact lenses include promotion of healing of corneal epithelial defects (1, 23), reduction of pain associated with corneal pathologies (15, 20) and closure of small corneal wounds (2, 11, 22). But they can also be used efficiently to protect the cornea from mechanical trauma such as entropion, trichiasis, and conjunctival abnormalities (4, 7, 16, 18).

In primary (localised non-familial) conjunctival amyloidosis, patients typically complain of a swelling of the eyelids and a burning sensation. Spontaneous subconjunctival hemorrhages and hematomas of the lids occur, and are probably a result of amyloid deposition in the walls of the blood vessels. As the deep conjunctival connective tissue layers become more infiltrated, the eyelids become thickened, giving a feeling of soreness. But the mechanical



a.



b.

*Fig 2. Soft bandage contact lenses (base-curve 8.70 and diameter of 18 mm) were fitted. The borders of these large contact lenses are clearly visible.
(a) Right eye (b) Left eye*

abrasion of the cornea, caused by the irregular palpebral conjunctiva itself, also seems to play an important role in the mechanism, which leads to the complaints the patients have. Therefore, surgical procedures, topical treatment and the use of soft contact lenses can be combined in order to relieve symptoms.

As in this case, a bandage lens efficiently protects the corneal surface from the shearing action of the lids, enhancing the comfort of our patient. Regular check-up is necessary in order to prevent complications.

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