SUPERIOR LIMBIC KERATOCONJUNCTIVITIS

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All about the Eye

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A 34-year-old lady presents with chronic inflammation and irritation of superior limbus. What is the most
likely diagnosis, pathogenesis and management of the disease? 2+3+5

Introduction

- Superior limbic keratoconjunctivitis (SLK) is a relatively uncommon chronic disease of the superior limbus and the superior bulbar and tarsal conjunctiva.
- It affects one or both eyes of middle-aged women, approximately 50% of whom have abnormal thyroid function (usually hyperthyroidism); approximately 3% of patients with thyroid eye disease have SLK.
- The condition is probably under-diagnosed because symptoms are typically more severe than signs.
- The course can be prolonged over years although remission eventually occurs spontaneously.

Differential diagnosis

- There are similarities to mechanically induced papillary conjunctivitis, and a comparable clinical picture has been described with contact lens wear and following upper lid surgery or trauma.

Pathogenesis

- The pathogenesis of superior limbic keratoconjunctivitis (SLK) has not been established, but it is thought to result from mechanical trauma transmitted from the upper eyelid to the superior bulbar and tarsal conjunctiva.
- The condition is believed to be the result of blink-related trauma between the upper lid and the superior bulbar conjunctiva, precipitated in many cases by tear film insufficiency and an excess of lax conjunctival tissue.
- With increased conjunctival movement there is mechanical damage to the tarsal and bulbar conjunctival surfaces, the resultant inflammatory response leading to increasing conjunctival oedema and redundancy, with the creation of a self-perpetuating cycle.
- It may be analogous to conjunctivochalasis affecting the lower bulbar conjunctiva.

Diagnosis

- **History**- enquiry should be made about contact lens wear, and previous eyelid surgery or trauma.
- **Age, sex, onset**-
  - Condition develops in women 20–70 years of age and may recur over a period of 1–10 years.
  - Usually resolves spontaneously.
  - It is often bilateral; however, 1 eye may be more severely affected than the other.
  - SLK can be associated with ATD or blepharospasm.
- **Symptoms**
  - Foreign body sensation, ocular irritation, redness
  - Burning,
  - Mild photophobia,
  - Mucoid discharge
  - Frequent blinking, and are often intermittent.
- **Signs**-

1. Conjunctiva

  - A fine papillary reaction on the superior tarsal conjunctiva

- Injection and thickening of the superior bulbar conjunctiva
- Papillary hypertrophy of the superior tarsal plate, often having a diffuse velvety appearance
- Hyperemia of a radial band of the superior bulbar conjunctiva that stains with rose Bengal and may be best seen macroscopically.
- Light downward pressure on the upper lid results in a fold of redundant conjunctiva crossing the upper limbus
- Petechial hemorrhages may be present.
- Keratinization can be demonstrated on biopsy or impression cytology.

2. Limbus

- Limbal papillary hypertrophy, hypertrophy of the superior limbus
- Limbal palisades may be lost superiorly.

3. Cornea

- Superior punctate corneal epithelial erosions are common and are often separated from the limbus by a zone of normal epithelium.
- Superior filamentary keratitis develops in about one-third of cases.
- Mild superior pannus resembling arcus senilis may be seen in long-standing disease.
- Keratoconjunctivitis sicca is present in only about 50%.
- Fine punctate fluorescein and Rose Bengal staining of the superior bulbar conjunctiva above the limbus and the superior cornea just below the limbus

- **Investigations-** The condition can often be diagnosed by clinical signs.

1. Histopathology- Biopsy or impression cytology may reveal keratinization of the superior bulbar conjunctiva.

- Hyperproliferation, acanthosis, loss of goblet cells, and keratinization are seen in **histologic** sections of the superior bulbar conjunctiva.
- However, scrapings or **impression cytology** of the superior bulbar conjunctiva showing characteristic features of nuclear pyknosis with "snake nuclei," increased epithelial cytoplasm–nucleus ratio, loss of goblet cells, or keratinization may be helpful in diagnosing mild or confusing cases.

2. Thyroid function test- Patients with SLK should undergo thyroid function tests, including

- Free thyroxine (T4),
- Thyroid-stimulating hormone (TSH),
- Thyroid antibody levels.

Treatment

- **Topical**

1. Lubricants (preservative-free may be preferred) to reduce friction between the tarsal and bulbar conjunctiva should be used regularly and frequently.
2. Acetylcysteine 5% or 10% four times daily to break down filaments and provide lubrication.
3. Mast cell stabilizers and steroids to address any inflammatory component; steroids may be best used in short intensive courses with rapid tapering, and should be reserved for severe cases.
4. Promising results have been reported with topical rebamipide.
5. Ciclosporin 0.05% twice daily as primary or adjunctive therapy, particularly in the presence of coexisting keratoconjunctivitis sicca.
6. Retinoic acid to retard keratinization.

7. Autologous serum 20% drops can be beneficial but may require instillation up to 10 times a day.

- **Contact lens**
  - Soft contact lenses, which intervene between the lid and the superior conjunctiva, are effective in some cases.
  - Large-diameter bandage contact
  - Interestingly, a unilateral lens may provide bilateral relief.
- **Supratarsal steroid injection.**
  - 0.1 ml of triamcinolone 40 mg/ml may break the inflammatory cycle.
- **Surgical management**
  - Resection of the bulbar conjunctiva superior to the limbus/superior limbal conjunctiva, either in a zone extending 2 mm from the superior limbus or of the area staining with rose Bengal, is often effective in resistant disease.
  - Lax conjunctiva is removed, with regrowth tending to be firmly anchored.
  - There is no consensus as to whether underlying Tenon capsule should be excised.
  - Amniotic membrane transplant and conjunctival fixation sutures.
  - Conjunctival ablation by applying silver nitrate 0.5% (not cautery sticks) or thermocauterization of the superior bulbar conjunctiva
  - Treatment of associated thyroid dysfunction may improve SLK
  - Temporary superior and/or inferior punctual occlusion.

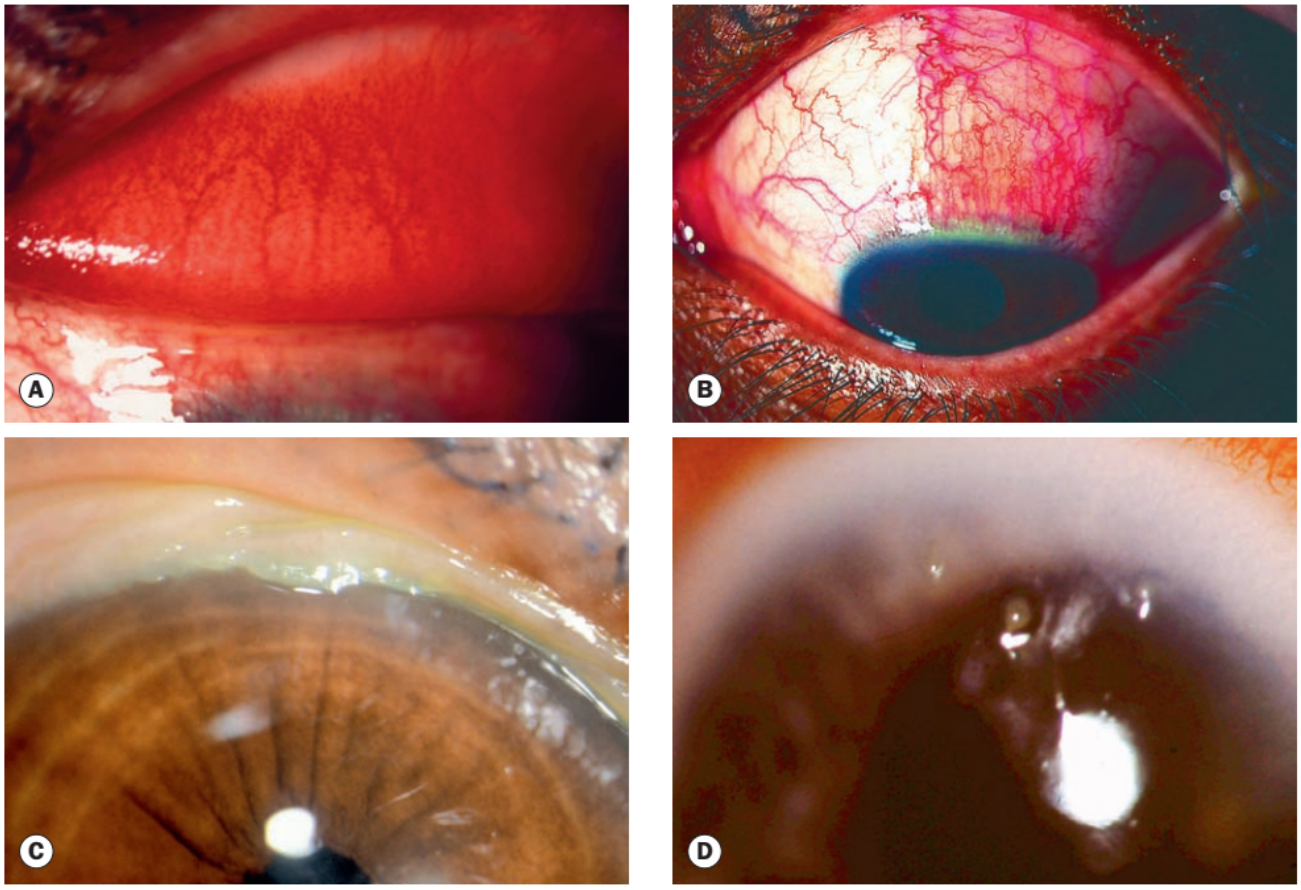
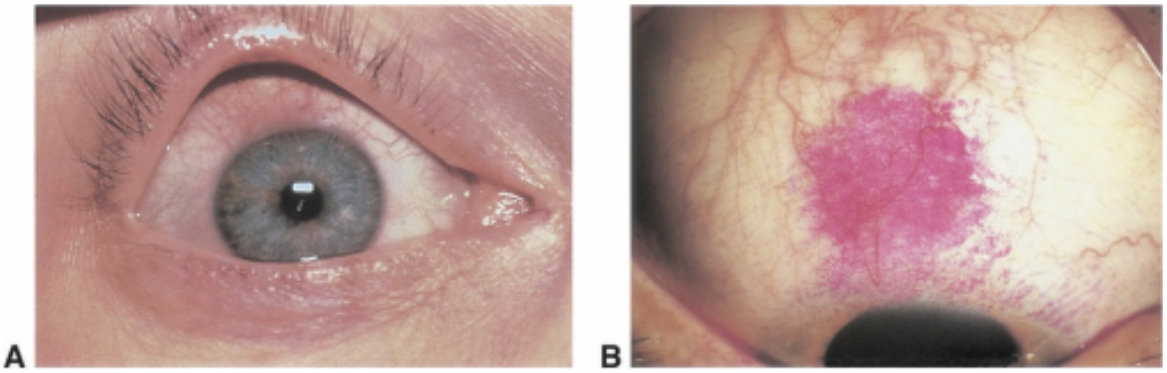


Fig. 5.26 Superior limbic keratoconjunctivitis. **(A)** Diffuse velvety papillary hypertrophy; **(B)** hyperaemic band of superior bulbar conjunctiva with limbal papillae, stained with rose Bengal; **(C)** fold of redundant conjunctiva; **(D)** superior corneal filaments



A. Superior limbic keratoconjunctivitis. B. Rose Bengal dye staining pattern in SLK