Scleral melting in a patient with conjunctival rhinosporidiosis

R M L De Doncker, R J W de Keizer, J A Osterhuis, A Maes

Abstract
A remarkable case of a scleral melting reaction in association with conjunctival rhinosporidiosis is presented and its surgical treatment with a scleral graft described.

Rhinosporidiosis is a granulomatous disease of the mucous membranes, caused by Rhinosporidium seeberi, an organism of uncertain taxonomic position, most probably a fungus. It usually infects the mucous membranes of the nose and nasopharynx but may infect all other mucous membranes and even the skin.1 Very rare cases of systemic infections of the liver and bones have been described.

The disease is worldwide but endemic in Africa and Asia, and most frequent in India and Sri Lanka. It occurs sporadically in other countries.2 4 Inoculations of the eye and related structures are grouped as oculosporidiosis.2 Most frequently the lacrimal sac is affected by extension from the nasal mucosa. Next most frequently the conjunctiva is subject to a primary infection, which may spread to the orbital tissue, the sclera, and the eyelids. A scleral defect associated with conjunctival rhinosporidiosis is rare, only two cases having been described. A third case is presented here.

Case report
A healthy 23-year-old white male agriculturist was referred to the ophthalmic outpatient department by his optician because he had noticed a bluish tumour on the temporal sclera of the right eye when fitting contact lenses. According to the patient this tumour had developed over one year and did not cause any trouble. There was no history of ocular disorder or trauma. The corrected visual acuity was 1-2 with +3-00 D.

Biomicroscopic examination of the cornea and the anterior chamber showed no abnormalities. In the inferior temporal part of the eye was a bluish circular swelling of the sclera about 4 mm in diameter. The overlying conjunctiva was movable over the scleral lesion. More temporal and under the margin of the scleral lesion were dilated conjunctival capillaries and scattered grey-white subepithelial corpuscular bodies of less than 1 mm diameter (Fig 1). The scleral lesion showed dim transillumination. It was decided to keep the lesion under observation.

Several months later the patient was examined because of an adenovirus keratoconjunctivitis. The scleral lesion had increased in size, and a strongly protruding scleral ectasia had developed, with considerable transillumination (Fig 2). Fundus examination at the site of the scleral lesion with the Goldmann contact lens revealed no retinal abnormalities. Diascral B scan ultrasonography provided an acoustic-free scleral image at that site without signs of choroidal lesions. On fluorescein angiography no abnormal conjunctival or scleral vessels were seen; the scleral lesion was delineated by hypo-fluorescence (Fig 3). The results of these investigations pointed to a scleral defect.

Because of impending perforation of the globe it was decided to perform a homograft of preserved human sclera over the scleral staphyloma. First an argon-laser coagulation of the retina round the lesion was performed. To obtain maximal reduction of the intraocular pressure during surgery mannitol was administered intravenously, and paracentesis and a large canthotomy were performed. The abnormal part of the conjunctiva was resected; Tenon’s capsule was kept intact. A 10 mm diameter scleral homograft was inserted over the 8 mm diameter defect with interrupted 6-0 silk sutures (Fig 4). In addition a running suture of 10-0 silk was placed over the margin of the graft (Fig 5). During a two-year follow-up the conjunctival...
infection showed no relapse, nor were there signs of graft failure.

HISTOPATHOLOGY
Light and electron microscopic examination of the conjunctival lesions showed a typical appearance. Below a normal epithelium many double walled cysts were observed in the conjunctival stroma, with an inner layer staining strongly positive with periodic acid Schiff and an outer Alcian blue positive layer. The cysts contained many vacuoles and eosinophilic granular material (Figs 6, 7, 8). These cysts were typical of rhinosporidiosis spores in different stages of maturity.

Discussion
Conjunctival rhinosporidiosis is a well known ocular infection. Clinically it causes vascularised, dark red, polypoid masses. The surface of the polyps is covered with distinct grey-white spherules (sporangia). The granulomatous masses can grow to resemble mucosal tumours.

The ratio of nose to eye infections is 4/6/1. In contrast to conjunctival sporotrichosis and coccidioidomycosis Parinaud's oculeoconjunctival syndrome never occurs. The mode of infection is unknown; experimental inoculation in animals has been unsuccessful, and cultures of the organism have failed to grow.

Transmission from animals to humans has never been seen. It is mainly children and young male adults who are affected. In all studies a male-female ratio of 4:3:1 is found. Epidemiological studies showed that conjunctival rhinosporidiosis has a predilection for agricultural workers, so that contact with contaminated soil may be a causative factor. Light and electronmicroscopic studies have provided most information on the organism. A young trophocyte undergoes nuclear division until it contains over 4000 nuclei. The inner layer of the trophocyte membrane differentiates, and a mature sporangium containing thousands of endospores is formed. The free spores can be isolated in the tears of an affected person.

Scleral melting in association with conjunctival rhinosporidiosis is very rare, having until now been described only twice. In contrast to the normal clinical picture the granulomatous reaction of the conjunctiva in our patient was less obvious; a scleral defect with scattered grey-white subepithelial granules on the margin was the main symptom.

The clinical diagnosis of this type of scleromalacia is based on the concomitant conjunctival findings, but light and electronmicroscopic examinations were required for the confirmation of the diagnosis. A differential diagnosis from other non-inflammatory scleral melting syndromes must be made (Table 1). Scleromalacia perforans is more frequent than rhinosporidiosis and affects the eyes of persons with seropositive rheumatoid arthritis. Spontaneous intercalary perforation is unrelated to rheumatoid arthritis and affects persons of a younger age.

The only effective treatment is radical surgical excision followed by thermal and chemical coagulation. Nevertheless, relapse is the rule rather than the exception.
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Figure 7  One cyst in detail contains granular eosinophilic material and lipid vacuoles. (Haematoxylin-eosin, ×53.)

Table 1  A differential diagnosis between the various conditions with scleral melting*

<table>
<thead>
<tr>
<th>Scleral melting</th>
<th>Spontaneous interciliary plaques</th>
<th>Hyaline plaques</th>
<th>Rhinosporidiosis scleral melting, 3 cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>50-75</td>
<td>25-50</td>
<td>60-75 &lt;25</td>
</tr>
<tr>
<td>Sex</td>
<td>Females</td>
<td>Either</td>
<td>Either</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>Constant</td>
<td>Unrelated</td>
<td>Unrelated</td>
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<tr>
<td>Biocularity</td>
<td>Bilateral</td>
<td>Single</td>
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<tr>
<td>Distribution</td>
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<td>Nil</td>
</tr>
<tr>
<td>Inflammation</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>Pathology</td>
<td>Necrosis</td>
<td>Degeneration</td>
<td>Unknown</td>
</tr>
<tr>
<td>Pain</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil (atrophy?)</td>
</tr>
<tr>
<td>Evolution</td>
<td>Progressive</td>
<td>Stationary</td>
<td>Progressive</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Bad</td>
<td>Good</td>
<td>Good</td>
</tr>
</tbody>
</table>


No medical treatment has proved to be effective in the systemic and mutilating conditions. Griseofulvin and amphotericin have been used without success. In cases of scleral ectasia a homograft of preserved human sclera gives good results.

The pathological basis of the scleral melting reaction in the immediate envirom of the conjunctival infection is obscure. The absence of direct contact between conjunctival lesions and the scleral defect suggests that an immunological or enzymatic process could be the cause. Further investigation has so far been impossible because attempts to culture the spore have been without success.

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