

# Ocular manifestations in a case of childhood cicatricial pemphigoid

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**PURPOSE.** *Cicatricial pemphigoid is a rare chronic autoimmune disease, characterized by progressive alterations on the skin and mucous membranes. Ocular lesions consists of chronic conjunctivitis, progressive subepithelial fibrosis on the conjunctival fornix together with symblepharon formation, obstruction of the Meibomian ducts, sicca syndrome, and occasionally, entropion associated with consequent trichiasis and corneal ulcers.*

**METHODS.** *A 9-year-old patient with cicatricial pemphigoid with severe eye involvement came to our observation. A complete anamnesis, ophthalmologic examination, and systemic evaluation, including serum antibody levels evaluation and biopsies of mouth, vulva, parotid, and intestine mucosa, were performed.*

**RESULTS.** *Ocular examination showed blepharospasm, conjunctival hyperemia, symblepharon, total cicatricial corneal leucoma, severe dryness, trichiasis, and eyelid edema. The results of medical and surgical treatment are reported.*

**CONCLUSIONS.** *Given the serious clinical picture, possibly due to a late diagnosis, it was not possible to avoid relapse and appearance of new cicatricial adherences for which we considered wait and see the most appropriate approach, protecting the anterior segment of both eyes, sacrificing their function. Further follow-up was not possible as the patient died. An early diagnosis would have had a significant influence on the clinical course and on the response to therapy. (Eur J Ophthalmol 2008; 18: 636-8)*

**KEY WORDS.** *Cicatricial pemphigoid, Corneal leucoma, Symblepharon*

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## INTRODUCTION

Cicatricial pemphigoid is a rare chronic pathology, of autoimmune etiology, characterized by progressive alterations on the skin and mucous membranes (1).

Many different parts of the body may be affected by cicatricial pemphigoid: oral and/or nasal mucosa, pharynx, larynx, esophagus, genitalia, small and large intestine, anus, urinary tract, and the eye (2, 3).

Patients with ocular involvement may present chronic conjunctivitis, progressive subepithelial fibrosis on the conjunctival fornix together with symblepharon, obstruction of the Meibomian ducts, sicca syndrome, and occasionally, entropion associated with conse-

quent trichiasis and corneal ulcers (1, 4).

Immunofluorescence has made a major contribution to the comprehension of cicatricial pemphigoid: antibodies and complement bound in a linear band at the epidermal-dermal junction were identified. Further studies demonstrated that the pathogenesis is attributable to the binding between circulating antibodies and specific antigens of the basement membrane (5); as reported by Razaque et al, other factors such as macrophage migration inhibitory factor, connective tissue growth factor, and interleuchine-4 play an important role in the pathogenesis of cicatricial pemphigoid (6, 7).

Cicatricial pemphigoid usually manifests around the



Fig. 1 - Our patient before surgery.

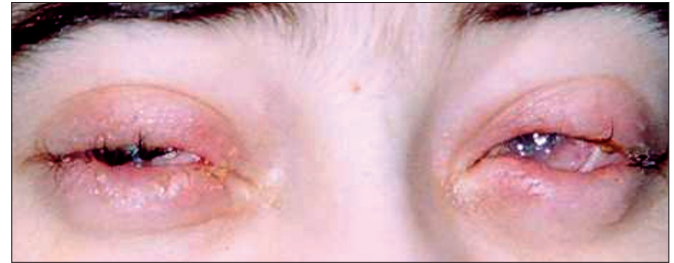


Fig. 2 - Our patient after surgery.

sixth or seventh decade of life; it is rarely observed in subjects under 20 years of age. Young patients present more severe and less easily manageable ophthalmologic symptoms than older patients (1, 4).

## Case report

A 9-year-old patient with cicatricial pemphigoid with severe eye involvement came to our observation at the Department of Ophthalmological Sciences of the Medical School, Federico II University, Naples.

Anamnesis showed that 5 years previously the patient had had cutaneous lesions diagnosed as atopic eczema and had been treated with topical and systemic steroids. After about 1 year, the patient presented alopecia and episodes of conjunctivitis. The parents reported that the patient had had recurrent episodes of cicatricial conjunctivitis for about 3 years.

At hospital admission serum autoantibodies levels (antinuclear antibody, anti-double-stranded DNA, anti-ds-DNA, anti-extractable nuclear antigen) were within normal limits. Biopsies of mouth, vulva, parotid, and intestine mucosa were performed. Histopathology revealed a rich infiltrate of lymphocytes, plasma cells, and eosinophils. Direct immunofluorescence showed IgG and C3 deposits along the basal membrane.

Systemic evaluation showed mouth ulcers, periungual infections, and perianal and vaginal erosions with the presence of scar tissue on the outer labia, bilateral choanal atresia, and laryngeal stenosis due to the presence of scar tissue.

Ocular examination showed blepharospasm, symblepharon with keratinization of the lower bulbar conjunctiva and scarring of the caruncle and puncta,

severe dry eye, corneal leucoma, trichiasis previously treated with electrolysis, eyelid edema, and conjunctival hyperemia (Fig. 1).

The anamnesis, clinical findings, histopathology, and direct immunofluorescence confirmed the diagnosis of cicatricial pemphigoid with ocular involvement. Our first priority was to give 25 mg of prednisone daily in order to relieve the symptomatology. The severity of the clinical condition prompted us to intervene surgically, to which cyclosporine therapy was associated. Surgery was carried out in order to resolve the choanal atresia and vulvar and vaginal lesions.

Ocular plastic surgery was performed to reconstruct the anatomy of the fornices. Silicone shapers were positioned in both eyes. Electrolysis and plastic surgery of the eyelid margins were also carried out.

No significant improvement was obtained from cyclosporine therapy and thus 5 mg daily tacrolimus was introduced. With this therapeutic regime a marked improvement of the oral, vulvar, ocular, and nasal lesions was obtained, thus avoiding worsening of the laryngeal stenosis.

Eye surgery avoided symblepharon formation and maintained good vision (Fig. 2).

Despite the presence of eyelid shapers, trichiasis due to eyelash regrowth caused bilateral corneal perforation; an autologous fibrin glue was utilized in an attempt to repair the corneal lesion and the two eyelid shapers were removed.

After a few weeks, fornices foreshortening reappeared and significantly worse systemic symptomatology was observed.

At this point we considered wait and see the most appropriate approach, protecting the anterior segment of both eyes, sacrificing their function. Further follow-up was not possible as the patient died.

## DISCUSSION

The serious clinical ophthalmologic condition observed in our patient may have derived from the delay in diagnosing cicatricial pemphigoid; in fact, early diagnosis followed by prompt and efficient therapy are usually sufficient to prevent the ophthalmologic complications.

Our patient's case history shows that the advanced stage of the disease is responsible for the irreversible ophthalmologic complications that occurred despite the introduction of immunosuppressive drugs in therapy. In agreement with the relevant literature, we confirm that the ocular symptoms in childhood cicatricial pemphigoid are more severe and less manageable than in adult pemphigoid (1, 4).

In our patient 5 mg tacrolimus daily was more therapeutically efficient than cyclosporine therapy. Oph-

thalmic surgery produced positive psychological benefits by significantly improving vision.

Given the worsening of systemic clinical conditions, we considered wait and see the most appropriate approach, protecting the anterior segment of both eyes, sacrificing their function.

Early diagnosis would have had a significant influence on the clinical course and on the response to therapy.

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