Sectoral Keratitis and Uveitis

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Case Presentation

A 26year old healthy woman, with one year history of recurrent idiopathic uveitis in both eyes was referred in January, 1995. Initial evaluation showed a best -corrected visual acuity of 20/20 in both eyes. Slit lamp examination (SLE) revealed 1+ cells in the anterior chamber, left eye. The intraocular pressure was normal in both eyes and the posterior pole was within normal limits bilaterally. Initial treatment with Pred Forte every two hours was given. Six months later the patient reported ocular symptoms of pain and photophobia in the left eye. SLE showed 1+ conjunctival injection and 2+ cells in the anterior chamber. The patient was treated with topical steroids with good response. Two years later, she had recurrence of the ocular symptoms in both eyes. SLE showed 1+ corneal injection, 2+ cells in the anterior chamber and microcystic edema inferiorly in the right eye. A microcystic edema was limited to the superotemporal corneal quadrant, with associated mutton fat keratic precipitates and 1+ cells in the anterior chamber. The IOP was 50 in the right eye and 20 in the left. Based on the clinical findings, entities such as sarcoid, herpes simplex, herpes zoster, tuberculosis and syphilis were included in the differential. The work up showed positive serum titers for herpes simplex virus type 1- IgM antibodies. Secondary glaucoma was controlled with medical treatment in the left eye, while trabeculectomy was required for the right. Acyclovir 800/mg per day was initiated with subsequent improvement of the ocular findings. However, improvement of the ocular manifestations was slow and development of a superonasal sectoral scarring was observed in the left eye. The patient has been able to maintain a visual acuity of 20/40 in the right eye and 20/25 in the left. Treatment with Acyclovir 800mg/day, topical treatment with steroids and antiglaucoma medications has been continued.

I. Introduction

Sectoral keratitis is a term that has been used to describe an inflammatory corneal stromal infiltrate that is well circumscribed to a quadrant or a sector of the cornea. The association between sectoral keratitis and uveitis is not common. However, we have observed some cases in our service in which these clinical findings are present. Additionally, some other similar cases have been reported in the literature. The most common entity related to sectoral keratitis and uveitis, as in the case presented, is herpes simplex virus. However, the entities included in the differential are diverse. We are presenting this case because we believe that it is important to be familiar with the presentation and the differential diagnosis in these cases, in order to make the correct diagnosis and to choose an appropriate treatment.

II. Clinical characteristics

In the Duke Elder's System of Ophthalmology text, in 1965, the name of keratitis profunda or deep keratitis was applied to a type of deep interstitial keratitis, localized centrally and sometimes in the periphery, associated with anterior uveitis. (1) The association of keratic precipitates, corneal opacity and anterior uveitis characterized the clinical course of this entity. The clinical manifestations and course of the disease seen in our patients are similar to those observed in the cases described of keratitis profunda.

Characteristic clinical signs and symptoms:

1. Mostly in adults

2. Usually unilateral ocular involvement
3. Acute onset with symptoms of blurred vision, photophobia, redness, pain and lacrimation.
4. Initial clinical findings are: edema of the corneal epithelium, deep interstitial opacity localized to a sector of the corneal periphery (usually)
5. Complete resolution of symptoms and clinical signs are observed in the majority of cases.
6. Common complications:

- Secondary glaucoma
- Secondary corneal scarring

III. Differential Diagnosis

The limited number of entities reported for the differential diagnosis of patients with sectoral keratitis and uveitis include:

1.Herpes simplex virus

2. Herpes zoster virus

3. Cytomegalovirus

4. Leprosy

5. Syphilis

6. Tuberculosis

7. Sarcoidosis

8. Relapsing fever

9. Lymphogranuloma venereum

10. Chronic myelomonocytic leukemia

Mondino reported 4 cases with herpes zoster ophthalmicus who developed a dense pannus limited to a sector of the superior cornea. (2) The pannus was associated with peripheral infiltrates in a subepithelial or anterior stromal location, and was not associated with disciform edema, interstitial keratitis or scleritis. A mild anterior uveitis was also present. Additionally, a case of CMV keratitis limited to a corneal quadrant and associated with uveitis in a patient with AIDS has been reported. (3)

Infiltration of the cornea in cases of leprosy is commonly observed. In one study from India, stromal infiltrates or avascular keratitis was documented in 21 eyes of 89 patients. (4) One of 2 distinct types was characterized by a corneal infiltrate localized in the anterior stroma, close to the limbus, in the superonasal, superotemporal or at the 6 o'clock position. Iritis was seen in an acute state in one case, and healed iridocyclitis were found in 11 patients.

A case of chronic myelomonocytic leukemia, in which the patient presented with sectoral corneal infiltrates, uveitis and secondary glaucoma, was reported. (5) No organisms were isolated from either the cornea or the conjunctiva, so corneal infiltrate was thought to be secondary to leukemic infiltration of the peripheral cornea. In cases of lymphogranuloma venereum, one of the three

different patterns of corneal involvement that has been described shares similar characteristics with the ocular findings observed in our patients. (6) Roy and Alvarez, in1885, suggested the occurrence of tuberculosis interstitial keratitis, for first time, by the finding of M. tuberculosis in the cornea in one case. The infiltration occurs in the middle or deeper layers of the cornea and is frequently limited to a sector. The vascularization is similarly localized and it is often unilateral. (6)

Interstitial keratitis in acquired syphilis is usually uniocular, limited to a sector-shaped area of the cornea. The pathological changes affect the posterior layers of the cornea most severely. (6) Relapsing fever is an acute infectious disease characterized by recurrent paroxysmal febrile episodes associated with considerable prostration caused by the Borrelia recurrentis. An interstitial keratitis in the superficial layers of the stroma has been recorded. (6, 7) This, usually occurring late in the disease, is transient and not associated with extensive neovascularization, resembling the clinical picture of a mild keratitis of syphilitic origin.

IV. Pathophysiology

The specific compromise of the peripheral cornea in some immunologic and inflammatory diseases may be explained in part by some immunologic differences that exist between the peripheral and central cornea. (8, 9) Because the peripheral cornea is located closer to the conjunctiva, which has blood vessels, lymphatic vessels and immunoreactive cells including lymphocytes and plasma cells, antigens in the peripheral cornea may elicit an immune response more easily than the central cornea.

The three most significant immunologic differences between the peripheral and central cornea are related to Langerhan's cells, the complement system, and the immunoglobulins. The Langerhan's cells are more abundant in the periphery than in the central part of the cornea.

The ratio of C1, which is the recognition unit of the classic pathway of complement, in the peripheral-to-central cornea, is 5:1. Therefore, antigen-antibody complexes may activate complement more effectively in the periphery. Also, the concentration of IgM has been found higher in the periphery.

In herpes virus keratitis peripheral infiltrates may represent an immune response to viral antigens. (The finding that the infiltrates and vessels were limited to a sector of the cornea suggests viral involvement of branches of the corresponding nerves.

It is not known yet whether the antigen represents live virus, residual viral antigens, or altered host antigens capable of eliciting autoimmune responses. (10,11,12)

V. Treatment

Choosing of treatment is based on the underlying associated entity. In cases of herpes virus infection our patients shown a good response to treatment with Acyclovir 800 mg per day, local and topical steroids and antiglaucoma medications. However, in contrast to herpes virus infection affecting the center of the cornea, these cases limited to a corneal sector in the periphery show a more prolonged course of the disease and slow response to medical treatment. Therefore, a close follow up is recommended. (12, 13)

VI. Summary

The entities related with sectoral keratitis and uveitis are diverse. Since these entities are different and some of the diseases are not only visual but also life threatening, it is important for the ophthalmologist to be familiar with the differential to be able to make the pertinent diagnostic

investigation, accurate diagnosis and appropriate management. Close follow up for these patients is recommended because peripheral involvement may have a longer course and may be more difficult to treat than central corneal lesions.

In addition, some patients may have diminished corneal sensation and they may not be aware of symptoms, so complications such as melting, epithelial defects, and superinfection are not uncommon.

VII. References

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