Diagnosis and Management of Sympathetic Ophthalmia

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Like so many diseases we see in our patients, the clinical presentation of sympathetic ophthalmia (S.O.) may vary considerably from text book descriptions. Accurate diagnosis is not easy for clinically suspected S.O. is pathologically confirmed less than half of the time. The diagnosis is often missed, as 15% of the pathologically diagnosed cases are not suspected clinically.

CLINICAL FEATURES

The classical clinical features of S.O. described by MacKenzie in 1830 are well known and need not be repeated. The classical description is that of moderate to severe form of disease but S.O. like any inflammatory disease may present in a very mild form with the only signs in the sympathizing eye being a low grade, non-granulomatous anterior uveitis or alternately a minimal anterior vitritis or juxta-papillary choroiditis.

Additional clinical features include Dalen-Fuchs spots (which are best seen at the mid periphery of the fundus), infiltrates at the ora serrata and a peu d'orange appearance of the fundus. Later in the course of the disease chorioretinal scarring may be observed at the posterior pole as well as in the periphery where multiple small chorioretinal scars present a moth eaten appearance. The peripapillary inflammation is a useful feature in following the course of the disease.

DIAGNOSTIC TESTS

Clinically useful immunologic testing has not yet been developed. The uveal pigment skin test which has been employed in the past for the diagnosis of S.O. should no longer be performed. With our current understanding of transplantation biology it would be unreasonable to transplant cow tissue or even histoincompatible human tissue to one of our patients.

Fluorescein angiography is a valuable test for the diagnosis of S.O. The characteristic picture seen in S.O. is identical to Harada's disease. The picture consists of multiple drusen-like spots where the fluorescein persists. The dye may spread from these foci and collect in large pools in areas of exudative detachment.

Ultrasonography has been used to confirm thickening of the choroid.

DIFFERENTIAL DIAGNOSIS

Harada's disease, bilateral phacoanaphylactic endophthalmitis, sympathetic irritation or reactivation of a pre-existing uveitis are the major differential diagnoses to consider.

There are a number of cases we have collected which were diagnosed as Harada's disease which subsequently have been reclassified as S.O. when a penetrating wound was discovered on histopathologic exam. Although difficult to believe there are patients who repeatedly deny any history of the eye injury which was discovered on pathologic examination. The absence of a penetrating wound is the only reliable features which differentiates S.O. from Harada's disease. Both the reported clinical and histopathologic differences between S.O. and Harada's disease may largely reflect case selection. The signs and symptoms, clinical course complications, fluorescein angiographic findings and histopathology are remarkably similar. The variability between different studies of patients with Harada's disease is as large as the proposed differences between S.O. and Harada's disease.

Phacoanaphylactic endophthalmitis (P.E.) has been closely associated with S.O. but several studies find that in recent years the coincidence is only 5%. No correlation is found between the severity of S.O. and the incidence of P.E. Experimental studies show no correlation between the susceptability of a given strain of animals to experimental S.O. and their susceptability to experimental P.E. Although the association of S.O. and P.E. may be coincidental depending upon lens damage by the penetrating wound, bilateral P.E. presents a problem in differential diagnosis. Principal differentiating features are; evidence for rupture of the lens capsule, variation in the onset of inflammation in the two eyes and absence of choroidal thickening on ultrasonography.

Sympathetic irritation may present a problem in the differential diagnosis of mild inflammations. When the primary disturbance is removed the sympathetically irritated eye improves. Removing the exciting eye in S.O. has not been demonstrated to improve the inflammation in the sympathizing eye. Only a third of the patients with sympathetic irritation have cells in the anterior chamber and only a few cells are ever observed.

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The patient's history and evidence of chorioretinal scarring in the early stages of the inflammation suggest that the bilateral uveitis occuring after a penetrating injury may represent a reactivation of a pre-existing uveitis rather than S.O. The clinical chorioretinitis of the inflammation and the usual diagnostic tests performed in cases of uveitis may be helpful in differential diagnosis.

HISTOPATHOLOGY

Several variations from Fuchs' classical histopathologic description of S.O. have been recently reported. Retinal perivasculitis is found in 50% or more of the cases. Focal involvement of the choriocapillaris occurs in 40% of the cases. Disruption of the retinal pigment epithelium with retinal extension of the inflammation is observed in 20% of the cases. The severity of the choroidal inflammation and extent of the granulomatous response is related to differences in uveal pigmentation.

TREATMENT

If there is a potential for useful vision, then every attempt should be made to save an injured eye. If an eye is so severely injured that no useful vision can be expected then it should be enucleated within 2 weeks of injury. There is approximately a two week period after primary repair of a penetrating wound in which the physician may evaluate the prognosis for recovery of useful vision.

It is unreasonable to enucleate an eye for the purpose of preventing sympathetic ophthalmia much later than two weeks after injury. It is not justified to remove an injured eye which is potentially functional particularly after S.O. has developed. There is no evidence that this produces improvement in the sympathizing eye. After treatment the injured eye may end up with the best vision.

When the diagnosis of S.O. has been established, corticosteroids at a dose sufficient to control the inflammation should be given as early as possible. We ordinarily employ 100 to 200 mgm of oral prednisone daily for 3-5 days and then begin alternate day therapy with a gradual reduction to maintainence levels once control of the inflammation is achieved. Treatment should continue for six months after the inflammation has cleared. Systemic steroids may be supplemented with subtenons injections of soluble steroids as well as topical steroids.

In refractory patients or when medical problems or systemic complications prevent the use of large doses of steroids, combined therapy with immunosuppressive agents may be effective (e.g. 10 mg of prednisone plus 2 mg/kg azothioprine or 7mg/kg chlorambucil daily). The potential for both serious side effects or stimulation of neoplasms suggest that immunosuppressive treatment should be used only in coilaboration with an oncologist after careful discussion of the side effects with the patient.

Prophylactic steroids have not prevented the development of S.O. while immunosuppressive doses of steroids in a recent penetrating wound introduce an unwarranted risk of infection.

There are no reliable data on the complications of glaucoma surgery in S.O. patients however a number of S.O. patients have had cataract surgery during remissions without complications.

It is important to remember that relapses occur in most patients. The interval between relapses may be more than 10 years. Prompt treatment and careful life time follow-up can produce an encouraging prognosis for (20/60 or better) useful vision in nearly 70% of the patients with S.O.

Behcet, Vogt-Koyanagui-Harada e Esclero-Uveítes

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Entre as uveites difusas, ao lado da oftalmia simpática, devem ser discutidas a Síndrome de Behcet e a Síndrome de Vogt-Koyanagui-Harada. São uveítes acompanhadas geralmente de vários sinais extra-oculares e que costumam acometer ambos os olhos apresentando grande gravidade. A causa é desconhecida, talvez auto-imune ou viral e tem nítida predisposição racial; genética ou geográfica.

É importante lembrar que o prognóstico visual para estes pacientes é razoável, desde que **o diagnóstico feito precocemente seja acompanhado da terapêutica adequada já nas primeiras fases da doença.**

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