Peripheral Uveitis in Three Children of One Family

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The familial occurrence of peripheral uveitis is rare. In 1963 Kimura and Hogan were the first to observe this disorder in two siblings, a brother and sister. Since their publication, no additional families have been reported. In this paper we will describe peripheral uveal inflammatory disease in three siblings.

CASE REPORTS

Patient No. 1

The first family member examined was seen on January 7, 1975, at age 7. She was referred by a local optometrist with a diagnosis of "bilateral cataracts." Five months prior to examination, the patient had passed a school vision exam without difficulty. In 1973, she had been examined by an optometrist who reported 20/20 vision in each eye. The youngster's health was good and there was no family history of eye disease.

Examination showed an uncorrected vision of right eye 20/70 and Jaeger O; left eye 20/100 and Jaeger 7. External and motility examinations were normal. Cycloplegic refraction showed: right eye +1.00 sphere equals 20/80; left eye, plano equals 20/200. The pupils dilated poorly. Slit-lamp examination showed two plus cells and flare in each eye and clear lenses. Indirect ophthalmoscopy revealed marked central vitreous opacities, clearing towards the periphery. The patient would not tolerate scleral depression; no peripheral retinal or ora serrata exudates were seen. Complete blood count and estimated sedimentation rates were negative. A presumptive diagnosis of peripheral uveitis was made and the patient was started on prednisolone acetate 1% drops qid.

On this regimen, the vision cleared to 20/60 in each eye but the cells and flare remained. On a subsequent examination, it was finally possible to see pars plana exudate, located inferotemporally in each eye. Because further improvement did not occur, on February 24, 1975, 1 cc of Depo-Medrol (80 mg/cc) was injected inferotemporally beneath Tenon's capsule in each eye. Vision improved in two weeks to 20/25.

By June of 1975, the vision had gradually declined to its preinjection level of right eye 20/60 and left eye 20/100. In July both eyes were reinjected using 0.5 cc of Depo-Medrol (80 mgs/cc) inferotemporally. This again produced considerable reaction of the right conjunctiva including aseptic necrosis with drainage of a portion of the injected material. The vision improved to right eye, 20/30 and left eye, 20/40 though the anterior chamber reaction persisted.

In October, the vision in the right eye was 20/300 and in the left eye 20/100. On November 4, 1976, an injection of Kenalog 1 cc (40 mgs/cc) inferotemporally was given in each eye. One week later, the vision was right eye, hand movements and left eye, 20/200. Again there was considerable reaction at the injection site. The posterior subcapsular lens opacities became more apparent and some hemorrhages developed in the right eye in the area of the pars plana. However, the vision gradually improved and in January 1977, all medications were stopped.

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On July 21, 1978, SC was reinjected with Kenalog, 40 mg, in each eye because of a recurrence of inflammation in the left eye which reduced the vision to 20/400. She had +1 flare and cells on the right and a +2 on the left. In addition, there was some hemorrhage inferiorly on the exudate. Following injection of the steroid, the vision gradually cleared to 20/70. On January 30, 1979, examination showed a diffuse, though mild, vitreous hemorrhage in the right eye through which no details could be seen. The vision was counts fingers. The left eye, at this time, was correctable to 20/50. By February 23, 1979, the vitreous hemorrhage cleared resulting in 20/60 vision in each eye. The application pressures were normal. Indirect ophthalmoscopy showed persistent pars plana exudate mingled with hemorrhage inferiorly, particularly on the left.

Comments

This case illustrates several points of interest. The first of these is the apparent clearing of the peripheral uvea inflammation with the use of local corticosteroids. There are a number of patients in whom the therapeutic response to steroids is seen without the use of either periocular or systemic administration of the drug. Although the local application of drops is generally not the route of choice in patients receiving treatment for this disorder, it has proven effective in an occasional patient.

Periocular corticosteroid injections are the most valuable and consistent treatment of this disorder. Unfortunately, as with the use of corticosteroids by any route, significant complications have frequently been noted. In this case, aseptic necrosis of the conjunctiva developed at the site of the injection. While this complication is relatively uncommon with single injections, it occurs with some regularity in patients receiving multiple doses. Treatment includes the use of local antibiotics in order to prevent infection. Spontaneous resolution of the necrotic process occurs in one to three weeks.

The third point of interest is the presence of significant hemorrhages in the area of the pars plana in this patient. Although hemorrhagic changes in the pars plana and hemorrhage into the vitreous are uncommon findings in patients with peripheral uveitis, a small percentage of patients with this disorder will exhibit these findings.

Patient No. 2

The second child affected in this family, SC, the older brother of Patient No. 1, was first seen in October 1975, at age 9. He complained about the absence of the lower half of his vision in the left eye upon first awakening in the morning. Vision returned after 45 minutes. His health was otherwise good.

Examination showed a vision of 20/20 and Jaeger 0 in each eye. Slit-lamp examination showed a quiet anterior segment. His retina and ora serrata were normal to indirect ophthalmoscopic examination with scleral depression.

On November 3, 1976, he returned after failing a school vision test. He reported difficulty seeing with the left eye which was now blurred all of the time. Examination showed a vision in the right eye of 20/25 and Jaeger 0 and in the left eye 20/100 and Jaeger 14. There was a black fundus reflex on the left with the retinoscope and mild cells and flare in each anterior chamber more marked on the left. Anterior vitreous opacities were present on the left. Indirect ophthalmoscopy with scleral depression showed inferior peripheral inflammatory changes in both eyes, most marked nasally.

Kenalog, 1 cc (40 mg/cc) was injected inferonasally in subtenon’s space in both eyes on November 4, 1976. The anterior chambers rapidly quieted and the vision cleared to right eye 20/25, left eye 20/30 by December 24, 1976. The right anterior chamber cleared completely; the left retained a slight reaction until January 1977. When re-examined on February 13, 1977, the vision was right eye 20/20 and left eye 20/30. On June 24, 1977, he had quiet anterior chambers and a vision of 20/30 in each eye. He had early posterior subcapsular cataracts in each eye, but still showed pars plana exudates.

On March 23, 1978, his right vision was reduced to 20/70 with increased vitreous haze. On March 24, 1978, 40 mg of Kenalog was injected beneath Tenon’s capsule, inferiorly. His vision improved to 20/30 in both eyes by June 27, 1978. Because of another reduction in vision (20/60), on September 13, 1978, Kenalog 40 mg, was also injected in the left eye. The vision improved slowly to 20/40, last recorded on February 23, 1979. He has shown no bleeding in association with his inflammation. There are some irregularities of his left macula, however.

Comments

Cases seen in large referral centers tend to reflect the worst end of the spectrum of peripheral uveitis. There are, however, large numbers of young patients managed in outpatient clinical settings who readily responded to minimal treatment regimens. This second younger illustrates this with his dramatic and long-term (17 months) response to a single injection of periocular steroids. An improvement of vision from 20/100 to 20/30 occurred following one periocular injection of Kenalog. It can be anticipated, however, that as in this case, most patients will require periodic injections to control the inflammation.

Patient No. 3

The third and last child in the family is the youngest brother of Patient No. 1. AC was first given a routine examination at age 5 on July 25, 1975. The youngster was asymptomatic. His eyes were entirely normal, although, lack of cooperation prohibited indirect ophthalmoscopy of the far retinal periphery.

This youngster returned on November 12, 1976, with a complaint of poor vision from the right eye. The vision was right eye, +1.00 equals 20/30 and left eye
+1.25 +0.75 x 15 equals 20/40. Slit-lamp examination revealed a quiet anterior chamber but there existed cells and flare in the anterior vitreous on the right. Indirect ophthalmoscopy showed a cellular vitreous reaction more nasally than temporally and more inferiorly than superiorly. Scleral depression was still not possible in this youngster. No change in his status was noted on December 12, 1976. On June 24, 1977, the anterior chamber showed a +2 flare in the right eye, +1 on the left, and a moderate amount of cells in the vitreous. The vision was 20/30 in each eye. No treatment was started. Scleral depression was not possible.

Minimal activity persisted until February 4, 1979. At that time, his vision dropped to 20/70 in the right eye and remained 20/30 OS. Although no treatment was instituted, his vision improved to 20/60 OD when examined in April 24, 1979.

Comments
In this final patient, although signs of the inflammation persisted, the vision remained better than 20/40 in each eye. The vision in the right eye has recently fallen to 20/60 and treatment may soon be required. Nevertheless this case demonstrates the opportunity to withhold treatment in patients with a minimal decrease in vision while still preserving visual function.

DISCUSSION

This report of a second family with peripheral uveitis suggests that the occurrence of this disorder should be considered in all family members when one presents with the problem. The etiology of this disorder in this family as well as in most patients presenting with the clinical syndrome, remains obscure. An etiologic evaluation in this family consisting of tuberculin skin testing, serum protein electrophoresis, and ELISA (Enzyme-linked immunosorbent assay) failed to show a relationship with any of the known diseases related to the peripheral uveitis syndrome. Since all of these youngsters were exposed to the same environment, the presentation of this disorder might be infectious or may merely represent a genetic predisposition within the family.

The clinical pattern followed by the youngsters demonstrates the variable course of the disease. Two of the youngsters required local and periocular corticosteroid treatment while the third youngster did not receive medication.

It is our belief that unless the patient is significantly bothered by vitreous floaters and the vision is less than 20/40, treatment should not be initiated. Macular changes, papilledema, exudative retinal separation are other indications for initiating treatment. A short trial of local corticosteroids may be attempted when there is significant anterior chamber inflammation accompanying the peripheral uveal tract disorder. In most patients, however, periocular injections of corticosteroids are the treatment of choice. General anesthetic is required for youngsters under the age of 12 in all except the most unusually cooperative patients. Systemic corticosteroids should be considered only after periocular delivery of corticosteroids has failed to improve the patient’s clinical status. The use of antimetabolite agents should not be considered unless a marked decrease of vision is found in both eyes.

SUMMARY

Peripheral uveitis presenting in three siblings is described. The course and response to treatment in each youngster is detailed. Periocular and local corticosteroid therapy was effective. Etiologic evaluation and immunologic surveys proved unrewarding. Ophthalmologic evaluation of families seems warranted following the detection of peripheral uveitis in any of its members.

REFERENCES