INTRAOCULAR LENS EXPLANTATION IN A PATIENT WITH JUVENILE IDIOPATHIC ARTHRITIS-ASSOCIATED UVEITIS

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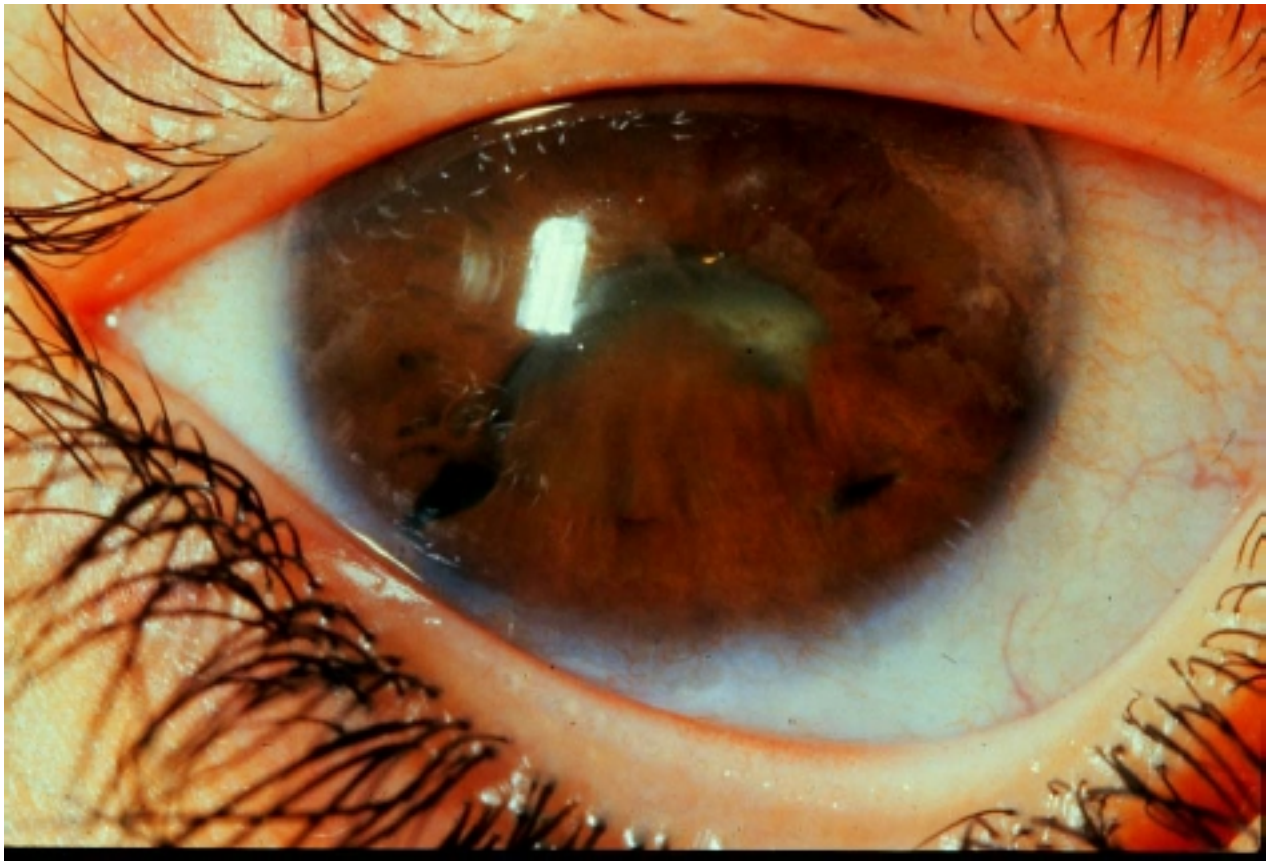
CASE

The patient was a 7 year old girl from Israel, with a history of Juvenile Idiopathic Arthritis (JIA) of pauciarticular onset at age 2. She had had a long history of anterior uveitis of the left eye, which was treated with topical steroids. She developed band keratopathy and cataract. Her cataract was removed by lensectomy and vitrectomy. The posterior lens capsule was not removed and a PC IOL was placed. Postoperatively, the patient had recurrent anterior uveitis, complicated by formation of a retrolenticular membrane. The IOL was progressively pulled superiorly and anteriorly by contraction of this membrane.



On presentation her vision was 20/30 OD and hand movements OS. The left eye had band keratopathy and trace cells in the anterior chamber. The IOL was displaced superiorly and anteriorly. A thick retrolenticular membrane was noted. Ultrasonography showed vitreous debris; the retina was attached.

The patient underwent pars plana vitrectomy, IOL explantation and cyclitic membranectomy. She was treated pre-operatively with prednisolone sodium phosphate 1% every 2 hours, and oral prednisone for three days. Post-operatively she was continued on this regimen for 4 weeks. Intra-operative and post-operatively fundus exams showed severe macular edema with macular cyst and a macular hole. The patient's visual acuity in the left eye remained at hand movements. According to her Ophthalmologist in Israel, the patient's vision is now 20/30 OD and counting fingers OS, and the left eye has been without inflammation since removal of IOL.

DISCUSSION

Juvenile Idiopathic Arthritis (JIA) occurs in about 0.16 to 1.13 individuals per 1000. It is more

common in girls, with a female to male ratio of 3 to 2. There are 3 types of onset-systemic onset which comprises about 20% of all cases of JIA, polyarticular onset, which comprises another 20% and pauciarticular onset comprising the remaining 60%. There are strong associations of pauciarticular onset and antinuclear antibody positivity with uveitis. Ninety percent of JIA-associated uveitis patients have pauciarticular arthritis, and 90% of these are ANA positive. However, uveitis does occur in patients with polyarticular arthritis and in patients who are ANA negative. Therefore routine regular screening examinations are necessary even in patients with polyarticular arthritis and who are ANA negative.

JIA associated uveitis is typically insidious, chronic, and usually asymptomatic. It often goes undetected until the patient is referred to an ophthalmologist. Because of poor vision secondary to damage produced by this asymptomatic inflammation, it is therefore recommended that patients with pauciarticular JIA should be examined every two to three months, patients with polyarticular JIA should be examined every 6 months, and patients with systemic JIA should be examined once a year. Patients with a history of uveitis should be followed every 8 weeks.

Cataract formation is common in patients with JIA-associated uveitis, as a result of chronic inflammation and corticosteroid treatment. Kanski reported an incidence of 46%.1

Cataract surgery is especially challenging in JIA patients. Band keratopathy and posterior synechiae make visualization difficult. The eyes of JIA-associated iridocyclitis patients typically develop secondary pupillary membranes and cyclitic membranes, which require further surgical removal. The post-operative inflammation can be exuberant and difficult to control. Since intraocular lens implantation is imprudent in this patient population, visual rehabilitation can be difficult in young children if there are problems with contact lens tolerance.

The early reports of intracapsular cataract extraction, needling and aspiration of JIA iridocyclitis-associated cataract showed poor results, with a high incidence of phthisis. In Smiley's report,2 66% of the eyes were completely blind 5 years after surgery, and only 15% of the eyes achieved a 20/40 or better vision. In the report by Key and Kimura,3 only 15% of the eyes had vision better than 20/40, and more than 50% of eyes saw less than 20/200. Through trial-and-error experience during the past 20 years from multiple centers a consensus has developed that extracapsular cataract extraction or phacoemulsification combined with total vitrectomy carries the greatest likelihood of successful visual rehabilitation for JIA patients. Intact posterior capsule and anterior vitreous may act as a scaffold for cyclitic membrane formation. Contraction of the cyclitic membrane typically leads to ciliary body detachment, resulting in hypotony and phthisis bulbi.

The procedures of choice then, are either combined phacoemulsification and pars plana vitrectomy, with complete removal of the lens, capsular fragments, zonules and anterior vitreous, or pars plana lensectomy and vitrectomy. By using the pars plana lensectomy-vitrectomy technique, Kanski and Shun-Shin4 operated on 101 eyes. Fifty five percent achieved a vision of 6/18 or better. (Follow-up 6 months to 9 years, with an average of 7months) Flynn5 reported vision of 20/40 or better in 70% of his patients employing this approach. (10 eyes, 6-62 months) Both of these reports claim a low incidence of cyclitic membrane formation, ranging from 0 to 3%.

Surgical complications can include early postoperative hypotony, which occurs in about 40% of cases, glaucoma (15-25%), and macular edema (20-31%). Other complications include choroidal detachment, vitreous hemorrhage, pupillary membrane formation, phthisis, retinal detachment and tear, and hyphema.

Fox and associates6 studied the causes of reduced visual acuity on long-term follow-up after cataract extraction in JIA-associated iridocyclitis patients. They followed 16 eyes for an average of 51 months, ranging 12 months to ten years. They found that post-operatively 81% saw better than 20/70; 69% saw better than 20/40. However with longer follow-up, 56% maintained 20/70 or better vision, and only 38% maintained a vision better than 20/40. The loss of visual acuity with time after surgery was a result of macular pathology or glaucoma. It is important to point out that as many as 62% of the eyes had some degree of inflammation pre-operatively. It is not clear in the report the degree or duration of inflammation post-operatively, but the authors did comment that many eyes continued to have low-grade intraocular cellular reaction. The importance of complete elimination of any intraocular inflammation, pre and post operatively cannot be over-emphasized.

We have advocated a stepladder approach in aggressiveness for the control of inflammation in

JIA patients, which begins with topical corticosteroid and mydriatics. This is followed by regional corticosteroid injections as needed. If the inflammation persists after 90 days of treatment or recurs with steroid withdraw, a systemic non-steroidal anti-inflammatory drug is added. The next step would be systemic corticosteroids for no longer than 3 months, unless inflammation is controlled on every third day low dose prednisone. If the inflammation still persists, systemic immunosuppressive chemotherapy should be administered. The first choice is methotrexate, followed by azathioprine, cyclosporine, infliximab, chlorambucil, and cyclophosphamide. Foster7 reported 100 eyes with JIA associated uveitis being treated from 1976 to 1992. During this period, 18% of the eyes developed significant cataract. This number is much lower than the previously reported 46% incidence rate, and probably should be accredited to the use of immunosuppressive agents that are steroid sparing. The cataracts were removed by the combined phacoemulsification and pars plana vitrectomy technique. The average postoperative vision was 20/40. At an average of 4.3-year follow-up (range 1-15 years), 75% of eyes maintained a vision of 20/40 or better.

Recently Holland and associates8 published a report of IOL implantation in 8 eyes of 7 patients with JIA uveitis. IOL was placed following phacoemulsification without vitrectomy. The authors reported 20/40 vision in all eyes with an average of 17.5 months follow-up. Complications included posterior synechiae formation in 2 eyes, posterior capsular opacification in 5 eyes, and new onset glaucoma in 2 eyes. This is a very limited study with a small number of patients, followed for a relatively short time. Four were adult patients with long-standing, burned-out uveitis. Of the 3 children, one 9 year old post-operatively developed posterior synechiae and a pupillary membrane, which required surgical removal. In the same issue of the AJO, an editorial9 emphasized that the data from this series cannot be used to justify placement of intraocular lens in all JIA patients, especially in children.

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