Pseudophakic Phacoanaphylactic Endophthalmitis

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ABSTRACT
Two cases of pseudophakic phacoanaphylactic endophthalmitis are reported. Both responded favorably to a total posterior capsulotomy performed in addition to removal of the intraocular lens and remaining cortical material. Current theories on phacoanaphylactic endophthalmitis are discussed. Excision of the entire posterior capsule with anterior capsular flaps and entrapped cortex is emphasized as an integral part of the treatment of this condition.

Phacoanaphylactic endophthalmitis is a rare complication of extracapsular cataract extraction which often has a devastating outcome. With the recent rebirth of extracapsular cataract surgery and the marked decrease in the use of intracapsular techniques, this complication is being recognized with increasing frequency. Although this entity has been clearly defined, the treatment for it has not yet been clarified in the literature. Current recommendations for treatment of pseudophakic phacoanaphylactic endophthalmitis include removal of the intraocular lens and irrigation of the anterior chamber with removal of loose cortical material.1,2 We are reporting two patients who responded dramatically to intraocular lens removal combined with complete removal of the posterior capsule, anterior capsular flaps and all residual cortical material.

CASE REPORTS
CASE 1: A 74-year-old man with bilateral posterior subcapsular cataracts underwent phacoemulsification of the right lens on 8/13/82 (ORK). The vision was 6/6-3 one day postoperatively, and the postoperative course was uneventful for the next two months. At that time, the patient complained of blurred vision and a red eye without any discomfort. Examination showed multiple keratitis precipitates, normal intraocular pressure and a small piece of cortical material. He was treated with topical Pred Forte four times a day, and over the next two months developed a hypopyon associated with vitreous opacities. The patient was referred for consultation, and on 1/13/83 an anterior chamber paracentesis was performed (ACL). The pathology of this specimen was read as being consistent with phacoanaphylactic endophthalmitis.

In spite of treatment with topical and systemic antibiotics and steroids, the patient then developed an exudative vitreous reaction. A diagnostic vitrectomy was performed on 1/28/83 (ACL) and the patient was given intravitreal and subTenon's antibiotics. He was also treated with intravenous antibiotics. Cultures from this procedure were negative for bacteria and fungi.

Two months later the patient developed an inferior retinal detachment which was successfully repaired (ACL).
The patient continued to have multiple hypopyons unresponsive to topical, subconjunctival, and oral steroids. On 4/28/83 the intraocular lens was removed completely (ORK, ACL) including both lens loops, but the parade of hypopyons continued. The patient's vision deteriorated to finger counting at one foot.

Finally, on 2/10/84, the patient underwent a complete posterior capsulectomy with alpha chymotrypsin,


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removing all remaining cortical material, capsular flaps and posterior capsule (ORK, ACL). Pathologic examination of the posterior capsule demonstrated a reaction consistent with phacoanaphylaxis (Figure). The inflammation subsided dramatically and no topical steroids have been required since June 1984.

The vision in the right eye improved to 6/18. Fluorescein angiography demonstrated chronic CME and epiretinal membrane formation in the right eye.

An intracapsular cataract extraction was performed on the left eye with an uneventful recovery and visual acuity of 6/7.5.

CASE 2: A 58-year-old woman underwent phacoemulsification of the left lens with insertion of a posterior chamber intraocular lens (BDG). Her vision one day postoperatively was 6/9. Two weeks later she developed pain in the left eye. Examination revealed a vision OS of 6/21 with 2+ flare and cells in the anterior chamber, whitish opacity on the anterior lens surface and anterior synchiae. She was treated with high doses of Pred Forte, Homatropine, and Chloroptic. Over the next year, the inflammation waxed and waned while the patient continued treatment with oral, subconjunctival and topical steroids.

On 9/21/84 the patient underwent removal of the intraocular lens, complete capsulectomy with alpha chymotrypsin and insertion of an anterior chamber intraocular lens (BDG, ORK). Pathologic examination of the capsule was interpreted as being consistent with phacoanaphylactic endophthalmitis.

Postoperatively, a fibrinous membrane covered the intraocular lens which gradually disappeared. The steroids were tapered. The final visual acuity is reduced to 6/30 due to CME.

DISCUSSION

Significant postoperative uveitis is an unusual complication of modern extracapsular cataract extraction. A study by Kline from 1974-1980 of 1400 cataract extractions (some of which were intracapsular cataract extractions) included eight cases of postoperative uveitis as a complication of intraocular lens implantation. More recently, a large Food and Drug Administration study documented only a 1% incidence of postoperative uveitis in a year. Woodhams reported a 2% incidence of persistent uveitis in a study of 1147 cataract extractions.

In the early days of intraocular lens implantation the acute form of sterile postoperative uveitis was often attributed to the "toxic lens syndrome." Sterilization techniques that were used at that time for the implants probably provided the major source of this inflammation. Much speculation has also been made of the role of the intraocular lenses themselves causing an inflammatory response.

Fortunately, awareness of this problem early on has led to improved manufacturing techniques so that intraocular lenses today rarely contribute to postoperative uveitis. Apple has pointed out in a recent article that many cases of postoperative sterile uveitis occurring today probably represent phacoanaphylactic endophthalmitis.

Phacoanaphylactic endophthalmitis was first recognized by Straub in 1919 and later clarified by Verhoeff and Lemoine in their classic discussion in 1922. Phacoanaphylaxis is a chronic granulomatous inflammation with a characteristic histologic picture which follows lenticular trauma or surgery. Early on, investigators argued whether the reaction was due to an actual lens toxicity or to an allergic phenomenon. The latter view is held today. However, Rahi and Garner point out that often no eosinophils are seen, and the response is not IgE mediated, so that this entity is not an allergy in the true sense. More precisely, it is an inflammation secondary to auto-sensitization to one's own lens proteins which have been previously sequestered from the immune system. Marek et al., in an experiment of sensitizing rabbits to lens protein, has shown that it is probably an immune complex reaction.

Clinically, the onset of the disease takes place between 1-14 days following lens injury or surgery. However, in our experience, it may occur up to three months following extracapsular cataract extraction, and has been reported occurring as late as one year postoperatively. It has been suggested that an anaphylactic reaction may occur against proliferating lens cells, as in Elschon's pearls, and this probably accounts for the delayed reaction seen in these cases.

The patient presentation is quite variable. Although patients have been reported in the literature presenting with a violent uveitis, occasionally the only clinical findings are decreased vision and a hypopyon in the anterior chamber.

The classic histological picture of phacoanaphylaxis is that of a zonal inflammation, around the lens or lens remnants. The center of this is a polymorphonuclear response, surrounded concentrically by a granulomatous inflammation including epithelioid and giant

FIGURE: Pathologic specimen of posterior capsule with anterior capsular flaps and cortical material.
cells along with other cells of subacute and chronic inflammation.\textsuperscript{13}

Rarely, phacoanaphylaxis may occur bilaterally.\textsuperscript{14} Because phacoanaphylactic endophthalmitis is seen concurrently with sympathetic ophthalmia in 25\% of cases of sympathetic ophthalmia, the first eye of a bilateral phacoanaphylactic endophthalmitis is often automatically called the exciting eye and the second, the sympathizing eye.\textsuperscript{15} This nomenclature adds to the confusion between bilateral phacoanaphylaxis and sympathetic ophthalmia. In fact, Eason and Zimmerman have demonstrated that in many cases where a presumptive diagnosis of sympathetic ophthalmia was made in the second, unoperated eye the actual histologic picture was of a true phacoanaphylactic endophthalmitis.\textsuperscript{16}

The outcome of phacoanaphylaxis is often devastating. Without treatment, the complications of phacoanaphylaxis are those of chronic uveitis, including secondary glaucoma, cyclitic membrane formation, hypotony, retinal detachment and phthisis bulbi.\textsuperscript{1} Treated cases often fare little better. Interestingly, the principles of treatment have changed little since the early days of extracapsular surgery when phacoanaphylaxis was not an uncommon complication of cataract extraction. The recommendation for treatment at that time was for removing remaining lens material thoroughly by irrigation.\textsuperscript{7} More recently, Apple has reported a case of phacoanaphylactic endophthalmitis which was treated with removal of the intraocular lens and anterior vitrectomy. The patient lost all sight and eventually the eye was enucleated.\textsuperscript{2} A similar case was reported by Smith in which the patient developed phacoanaphylactic endophthalmitis 11 weeks after a cataract extraction, was treated with irrigation and aspiration of the anterior chamber, and ended with hand movements vision and a quiet eye.\textsuperscript{17} Other cases have been reported recently in the literature with similarly disappointing results.

The first cases of phacoanaphylactic endophthalmitis with favorable outcomes were reported in 1965 by Riise. The first patient had the onset of uveitis one year after extracapsular cataract extraction with no response to topical and systemic steroid administration. However, the uveitis cleared "as if by magic" after a complete posterior capsulotomy was performed. Riise then went on to perform the same on another patient with similar results.\textsuperscript{12}

We have also shown a dramatic response to posterior capsulotomy in the two cases of pseudophakic phacoanaphylactic endophthalmitis reported in this paper. The first was in a patient who incurred a "parade of hypopyons" which cleared quickly following posterior capsulotomy. In fact, the initial treatment of our first patient did entail removal of the intraocular lens. Notably, during the procedure, the lens loops were free and did not have to be left behind. Had it been necessary for us to sever the prolene loops and remove only the optic from the eye, we might have blamed the prolene for the continued inflammatory reaction. The diagnosis of pseudophakic phacoanaphylactic endophthalmitis could have been delayed.

One of the basic principles of treatment in phacoanaphylactic endophthalmitis is that removal of all lenticular material is necessary to quell the inflammatory response. We feel that the cortical material is intimately attached to the capsular remnants and, therefore, its removal can only be completely accomplished by a total capsulectomy. This includes removal of the posterior capsule, anterior capsular flaps along with any residual cortical material. In addition, Apple stresses the fact that the inflammation may worsen if only the implant is removed.\textsuperscript{6}

The technique of posterior capsulectomy was developed in the early days of modern extracapsular surgery. At that time it was routinely performed by many well-known surgeons, before the full benefits of maintaining the integrity of the posterior capsule were appreciated.\textsuperscript{18} A Flieringa ring is placed around the globe and secured in four quadrants. The previous cataract wound is opened and the intraocular lens is removed. If possible, this should be done without severing the lens loops. Next, alpha chymotrypsin is injected beneath the iris. After waiting three minutes, the iris is retracted and the capsule is removed manually with forceps in a hand-over-hand technique.\textsuperscript{19} It is possible, if done very carefully, to remove the posterior capsule, anterior capsule flaps, and all entrapped cortex in one piece. At this point, if no difficulties have been encountered during the surgery, an anterior chamber lens can be inserted (as was done in our second case).

In conclusion, we feel that pseudophakic phacoanaphylactic endophthalmitis is an important cause of postoperative uveitis following extracapsular cataract extraction. The prognosis for vision without aggressive removal of all lenticular material is uniformly poor. We stress the importance of performing a complete posterior capsulectomy when this disorder is suspected. The posterior capsule should then be submitted for pathologic examination to confirm the diagnosis. Prompt recognition of pseudophakic phacoanaphylactic endophthalmitis and treatment in this manner should lead to an improved prognosis.

\textbf{REFERENCES}


