Phacogenic Uveitis

Parul Jain MS, FICO, Pallavi Dokania MBBS, Richa Aggarwal MBBS, Pooja Jain MS, Aditi Manudhane MS, J.L. Goyal MD, Ritu Arora MD

Guru Nanak Eye Centre, New Delhi.

Phacogenic or lens induced uveitis is defined as anterior uveitis with or without vitreous inflammation directed against released or retained lens protein, occurring days to weeks after surgical or traumatic disruption of lens capsule, or spontaneously due to a hypermature cataract. Terms like phacoanaphylactic and phacotoxic uveitis have been used to describe inflammation that occurs after surgical or traumatic disruption of the lens capsule. It is now generally agreed upon that both these terms are not appropriate with respect to the cause of lens induced uveitis. Anaphylaxis is mediated by IgE, basophils and mast cells, none of which have been shown to play a role in lens induced uveitis. There is no evidence to support the hypothesis that the lens proteins are directly “toxic” to ocular tissues.

Epidemiology
Relatively rare disorder, accounting for less than 1% of all uveitis cases.

Common in elderly population, with peak incidence in 6th to 7th decade, with the exception of traumatic cataract which is common in younger population.

However, there is no racial or sex predilection.

Pathophysiology
Lens proteins are immunologically privileged because they are isolated from fetal circulation early in embryonic life (sequestered) and they may initiate an immunological sensitization only after entering the aqueous humour. After a break in the lens capsule and sensitization to lens protein, an immune complex mediated phenomenon develops.

Clinical presentation
Symptoms may include severe light sensitivity, epiphora, pain, floaters, decreased vision and redness. Vision may range from 6/6 to no perception of light.

Signs
Acute case may occur within 24 hours of cataract surgery in a patient who has previously undergone cataract extraction in the fellow eye.

The patient usually has the following findings on examination:
- Large number of cells in anterior chamber and vitreous.
- Mutton fat keratic precipitates (granulomatous inflammatory response).
- Posterior synechiae.

Figure 1: Showing granulomatous uveitis with mutton fat keratic precipitates, posterior synechiae with traumatic cataract with ruptured anterior lens capsule.

Ref: Middle East African Journal Of Ophthalmology (MEAJO).
• Pseudohypopyon.
• Fragments of lens matter in anterior chamber or vitreous.
• Raised intraocular pressure due to trabeculitis, lens debris in angle or posterior synechiae.
• There may be an associated vitritis but typically there is no involvement of the retina, choroid or the optic nerve.

Sub acute or chronic cases occur within 2-3 weeks of lens capsule disruption (surgical or traumatic) and is less severe. The patient usually has the following findings on examination:
• Cells in anterior chamber (non granulomatous inflammatory response).
• Flare in anterior chamber.
• Keratic precipitates.
• Posterior synechiae.
• Epiretinal membrane.
• Macular edema.

Pathology
Cytopathological examination of aqueous humour in phacogenic uveitis may reveal a zonal granulomatous inflammatory reaction, with polymorphonuclear leucocytes, multinucleated giant cells, mononuclear cells (containing eosinophils, plasma cells and histiocytes), surrounding a nidus of damaged or retained lens matter.6

Diagnosis
Diagnosis is mainly based on history and clinical examination, however complimentary examination may be useful to rule out other entities and complications.

High resolution B scan ultrasonography and OCT of anterior segment can be done to rule out IOL related uveitis.

Posterior segment B scan ultrasonography to detect complications like vitritis, retinal detachment, choroidal detachment, retained lens matter and intraocular foreign body.

CT scan to rule out retained intraocular foreign body in post traumatic cases.

Treatment
If there is minimal residual lens material present, the inflammation will in most cases eventually resolve with prolonged corticosteroid therapy. Surgical removal of the offending antigen, the lens material, has to be performed in patients with persisting inflammation, significant amount of retained lens matter and uncontrolled elevation of intraocular pressure.

Course and outcome
There is often a good outcome with prompt and aggressive management.

Complications
• **Cystoid macular edema:** The incidence of cystoid macular edema after complicated cataract surgery with retained lens material has been reported to be approximately 7%.
• **Secondary Glaucoma:**
  o Leakage of lens proteins through the injured lens capsule with or without leakage of serum proteins from uveal blood vessels in lens-induced uveitis may block the trabecular outflow causing secondary glaucoma.
  o Trabecular meshwork obstruction may occur with the accumulation of white blood cells (macrophages and activated T lymphocytes) or their aggregations. These may cause peripheral anterior synechiae and subsequent closed-angle glaucoma.
  o Obstruction may arise from inflammatory debris (e.g. proteins, fibrin, high molecular weight proteins) and from lens particles. These proteins increase the aqueous viscosity, which may contribute to increased intraocular pressure.

• **Corneal endothelial loss:** occurs due to inflammation.

Phacolytic Glaucoma
Phacolytic glaucoma results due to lens protein leakage through an intact lens capsule in a hypermature cataract.7 Proteins that leak into the anterior chamber are engulfed by macrophages, which in turn cause blockage of the trabecular meshwork resulting in the elevation of intraocular pressure.

Epidemiology
Common in elderly population, with peak incidence in 6th to 7th decade. There is no racial or sex predilection.

**Clinical Features:** Phacolytic glaucoma occurs in patients who have a hypermature cataract and presents as an acute inflammation with marked flare and large cells in the anterior chamber. The cornea is usually edematous and generally without any evidence of keratic precipitates. Some of these patients show evidence of anterior chamber angle recession or rupture of the posterior lens capsule. In general, the lens capsule surrounding the hypermature cataractous lens is intact, although it is markedly thinned. Some patients with phacolytic glaucoma exhibit conjunctival injection.

Phacogenic nongranulomatous uveitis can be easily mistaken for phacolytic glaucoma, because both of these entities can result in increased intraocular pressure and
inflammatory cells in the anterior chamber. However, phacolytic glaucoma occurs only in patients with a mature or hypermature cataract who have no evidence of traumatic or surgical disruption of the lens capsule.

Pathology

It typically exhibits a non-granulomatous inflammation around the disrupted lens. This cellular infiltrate consists of lymphocytes, histiocytes, and some polymorphonuclear leukocytes; epithelioid cells, giant cells, and the zonal pattern are not seen in this entity. The macrophages rarely contain eosinophilic granular material, which is commonly noted in cases of phacolytic glaucoma.

Diagnosis: Diagnosis is mainly based on history and clinical examination, however complimentary examination may be useful to rule out other entities and complications, which include gonioscopy, ultrasound bscan, etc.

Treatment: The treatment of phacolytic glaucoma consists of reduction of intraocular pressure by acetazolamide and/or hyperosmotic agents, followed by surgical removal of the entire lens. Most patients respond well to this approach, but the patient with angle recession combined with phacolytic glaucoma may require long-term topical or systemic antiglaucoma therapy following extraction of the lens.

IOL Related Uveitis

IOL related uveitis (though not strictly a subset of phacogenic uveitis) may be the cause of chronic postoperative inflammation in patients who have undergone cataract extraction. A number of factors can result in IOL-related uveitis, including lens-iris or lens-ciliary body contact, or complement activation in the aqueous by certain types of lens materials.

Chronic uveitis may also be seen in patients with low grade postoperative endophthalmitis caused by agents like Propionibacterium acnes, Staphylococcus epidermidis and Candida parapsilosis.

Table: Differential diagnosis of phacogenic uveitis

<table>
<thead>
<tr>
<th>Condition</th>
<th>Differentiating features</th>
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<tbody>
<tr>
<td>ACUTE/EARLY (within first week of injury or trauma)</td>
<td>Rapidly progressive, positive stain and culture</td>
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<tr>
<td>1. Acute infectious endophthalmitis (Bacteria usually aerobes)</td>
<td></td>
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<tr>
<td>2. TASS (Toxic anterior segment syndrome)</td>
<td>within 1st day after surgery, marked corneal oedema, nil post segment inflammation</td>
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<tr>
<td>Subacute/Chronic (weeks to months)</td>
<td></td>
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<tr>
<td>1. Low grade endophthalmitis</td>
<td>positive culture</td>
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<tr>
<td>2. IOL related uveitis</td>
<td>poorly placed iol in uveal tissue, iris transillumination defect</td>
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<td>3. Retained intraocular foreign body</td>
<td>B scan, CT scan shows the presence of FB</td>
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References