Cataract surgery in juvenile xanthogranuloma: Case report and a brief review of literature

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There is limited literature on the management of cataracts in juvenile xanthogranuloma (JXG). A 2-month-old girl presented to us with hyphema, secondary glaucoma OU and skin nodules suggestive of JXG. She developed bilateral cataracts during her follow-up and was treated successfully with cataract surgery and aphakic rehabilitation.

Key words: Cataract surgery, juvenile xanthogranuloma, non-langerhans histiocytic proliferation

Cataracts have been reported in juvenile xanthogranuloma (JXG), but there is limited experience with regards to management and long-term outcome.[1] Early treatment is essential to prevent amblyopia. We report successful management of bilateral cataracts in a patient with JXG.

Case Report

A 2-month-old girl was brought to us with haziness of both eyes for the past 20 days. She was a full-term baby delivered by cesarean section with a birth weight of 2.75 kg. Her antenatal period was uneventful. The parents denied any history of trauma and the child was otherwise healthy. On examination (under general anesthesia), she was noted to have conjunctival hyperemia, increased corneal diameter (13.5 mm OD, 11 mm OS), corneal haze with small blood clots in the anterior chamber of both eyes. No iris nodules were evident [Fig. 1]. Intraocular pressure (IOP) was noted to be 20 mm Hg OD and 8 mm Hg OS by Perkin’s tonometer. A B-scan ultrasound of the posterior segment was normal OU. A few yellowish-orange skin nodules were evident [Fig. 1]. Intraocular pressure (IOP) was noted to be 20 mm Hg OD and 8 mm Hg OS by Perkin’s tonometer. A B-scan ultrasound of the posterior segment was normal OU. A few yellowish-orange skin nodules were noted on the forehead and neck [Fig. 2], that progressively increased on follow-up. Remainder of her systemic and neurological examination was normal. Her hemogram and coagulation profile were within the normal limits. Excision biopsy of a forehead skin lesion was performed under general anesthesia and histopathological examination of the specimen showed non-langerhans histiocytic proliferation [Fig. 3]. Immunohistochemistry was negative for CD1a and cytokeratin; positive for CD68. With the above clinical picture and laboratory finding, a diagnosis of JXG was made.

She was started on a topical antibiotic steroid combination, cycloplegic and dorzalamide OU. On her review visits, she developed posterior synechiae and a few iris nodules in the left eye, which resolved with intensive topical steroids. Her IOP and fundus examination were within the normal limits. The steroids were tapered off after 2 months. She maintained a steady central fixation throughout her treatment and was also on regular review with a pediatrician. Four months after her first presentation, she developed bilateral anterior subcapsular cataract, which gradually progressed. She underwent cataract surgery in the left eye under general anesthesia. Topical prednisolone acetate 4 times a day was added to topical antibiotics a day before surgery. A superior bridle suture was placed. After peritomy, the bleeders were cauterized and a limbal groove made with a blade breaker. The anterior chamber was entered by deepening the groove and trypan blue

Figure 1: From top left, corneal edema right eye; top right: Hyphema after resolution of corneal edema left eye; bottom left: Development of cataract; bottom right: After cataract extraction

Figure 2: Resolution of skin lesion over the period of 1 year
was injected under an air bubble. Intracameral adrenaline was used to achieve pupillary dilatation and reduce intraoperative bleeding. Subsequently, 2% hydroxypropyl methylcellulose was injected into the anterior chamber. Iridocapsular adhesions were gently lysed by sweeping a cyclodialysis spatula under the iris. The anterior capsule was punctured by a bent 26 G needle and capsulorhexis done with an Utrata forceps. The lens matter was aspirated by a Simcoe cannula. The bottle height was raised to increase pressure in the anterior chamber and tamponade any bleeding. Subsequently, the posterior capsule was punctured by a bent 26 G needle in a sideways fashion and capsulorhexis completed by an Utrata forceps. Sufficient anterior vitrectomy and a peripheral iridotommy were done by an automated vitrector. An intraocular lens (IOL) was not implanted given the age of the patient (<1 year). Tight wound closure with 10-0 monofilament nylon suture was ensured. After surgery, she was started on topical steroid antibiotic combination and cycloplegics, which were tapered over 6 weeks. The right eye cataract surgery was done 6 weeks after the left eye surgery and a similar procedure was followed. Her perioperative period was uneventful. On her last follow-up, 1 year after surgery, the child was comfortable with aphakic correction and the visual axis was clear. The IOP was monitored with Pulsair tonometer. The IOP was normal on all occasions and no antiglaucoma medications were needed. Her perioperative period was uneventful. The skin lesions appeared to be regressing.

**Discussion**

JXG is a rare benign disorder that presents with skin lesions during infancy. The skin lesions are discrete yellow/orange nodules on the face and back. They resolve spontaneously and do not require any specific treatment.[5] Histopathologically it is characterized by a non-langerhans cell histiocytosis. A reactive granuloma with foreign body giant cells and Touton giant cells is characteristic.[1]

![Image 1](image1.png)

**Figure 3: Histopathology of skin lesion. Low power (×100): Structure of acanthotic skin with sub epidermal infiltration of xanthomatous cells (arrow) and inflammatory cells with encircling adnexal structure and extending into subcutaneous zone. High power (×1000): Sheets of foamy histiocytes (arrow) with interspersed multinucleated giant cells (block arrow)**

Ocular involvement in JXG is in the form of diffuse or discrete vascular iris nodules, which may bleed, resulting in spontaneous hyphema and secondary glaucoma. The condition may also involve the ciliary body, anterior choroid, cornea, lids, limbus, orbit, retina, optic nerve and optic nerve, but intraocular involvement is rare (<1% of cases).[2,3] Presentation is typically unilateral, though bilateral cases like ours have been reported. Indeed JXG should be included in the differential diagnosis of spontaneous hyphema in this age group and one should make a careful evaluation to detect skin nodules, which can be difficult to detect early in dark skinned individuals. The hyphema usually responds to conservative treatment.[5,4]

Cataracts may develop and management can be difficult. It is believed that any ocular surgery in JXG may be complicated by hyphema.[6] The perioperative period was uneventful in our patient. This could be because patient was taken up for surgery once the iris nodules and hyphema had resolved and patient was continued on topical steroids. It is likely that the use of intracameral adrenaline, raising the bottle height and a tight wound closure reduced the risk of bleeding in our patient. In general, bimanual anterior limbal vitrectomy is preferred over a pars plana vitrectomy after primary posterior capsulotomy in pediatric cataract surgery.[6]

This is because the technique is simpler and most cataract surgeons are comfortable with an anterior approach. An IOL was not implanted in our patient considering the anterior segment inflammation and the bilaterality. Furthermore, the safety and benefits of IOL implantation in infancy is not well-established.[7,9]

We could not find any other reports detailing on cataract surgery in JXG. Management as detailed above may reduce the risk of bleeding and improve the visual outcome.

**References**