CASE REPORT

The child with spontaneous recurrent bleeding in the eye

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Accepted 25 May 2014

SUMMARY

Juvenile xanthogranuloma is a self-limiting dermatological condition characterised by spontaneous hyphaema due to uveal involvement causing neovascularisation. Recurrent hyphaema in a child should alert the clinician towards this rare diagnosis. Appropriate investigations and treatment should be directed towards treating this possible diagnosis associated with neovascular glaucoma where routine filtering surgeries may be associated with several complications.

BACKGROUND

Juvenile xanthogranuloma represents a spectrum of histiocytic disorders including benign adult xanthogranuloma and progressive nodular histiocytosis. This rare condition affects children mostly and is commonly seen as elevated papulonodules in the skin and eye or may present as neovascular glaucoma rarely.^{1–5} The nodules are self-limiting or may resolve with systemic steroids, while ocular complications require surgery to prevent permanent visual loss. A high index of suspicion is required to diagnose this rare entity and investigations should be tailored to diagnose this condition. Surgical management of such a case is a challenge since routine filtering surgery may be associated with high risk of failure and a high rate of vision threatening complications.

CASE PRESENTATION

A 10-year-old female presented with repeated sudden spontaneous painless loss of vision in the left eye (3 episodes in 1 month) and a baseline intraocular pressure (IOP) of 12 mm Hg and

48 mm Hg in the right and left eye, respectively. On examination, her best corrected visual acuity was 20/20, N6 in the right eye and projection of rays in the left eye. Anterior chamber showed dispersing hyphaema filling the whole anterior chamber in the left eye with no view of anterior segment (figure 1A). Slit lamp evaluation was within normal limits in the right eye.

Review history did not reveal any history of trauma. Gonioscopic details were hazy in the left eye while angle was wide open in all quadrants in the right eye. She was started on maximum medical treatment in the left eye with topical brimonidine, dorzolamide along with loteprednol, cycloplegics, systemic acetazolamide inhibitors and intravenous mannitol 1 g/kg body weight under supervision.

INVESTIGATIONS

Possible causes of spontaneous hyphaema in a young child include leukaemia, secondary malignancy, juvenile xanthogranuloma, medulloepithelioma, neurofibromatosis and rarely viral keratouveitis. Ultrasound examination revealed a normal posterior segment while ultrasound biomicroscopy (UBM) showed focal iris thickening in the inferior and nasal quadrants in the left eye. There was no obvious mass lesion involving the iris or ciliary body.

Laboratory tests ruled out bleeding diathesis and normal cell count while systemic workup was negative for any primary lesion elsewhere in the body. An anterior chamber (AC) tap revealed only plasmid aqueous with no tumour cells.

Paediatric and dermatological evaluation did not reveal any other systemic or haematological abnormalities with no primary lesions elsewhere in the

Figure 1 Clinical photograph showing anterior chamber filled with hyphaema (A) with no view of anterior segment details. (B) Postoperative photograph showing resolution of hyphaema with iris mamillations on the iris surface (arrows).

To cite: Rao A, Padhy D. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-203925

body. She had two repeated episodes of fresh bleeding within the anterior chamber 2 days after AC tap.

DIFFERENTIAL DIAGNOSIS

A negative systemic workup and no focal lesion on UBM ruled out leukaemia or intraocular tumours. AC tap did not reveal any viral aetiology. Systemic workup for neurofibromatosis was also negative with no obvious skin lesions.

TREATMENT

Her IOP failed to get controlled despite maximum medical treatment and measured 58 mm Hg 2 days after AC tap with two recurrent episodes after the procedure. Keeping a possibility of Juvenile xanthogranuloma (JXG) with neovascular glaucoma with UBM showing focal iris thickening in the nasal and inferior quadrants, she was taken up for glaucoma drainage implant (FP8 paediatric Ahmed glaucoma valve) surgery with position of the plate in the superonasal quadrant with AC wash. Intraoperatively, the iris showed presence of neovascularisation with thickened iris with altered architecture prominent in the inferior and nasal quadrants after washing the anterior chamber. The iris in the same quadrants was found to be very fragile, which was taken for biopsy and histopathological evaluation. The tube was placed in the superonasal quadrant with no rebleed seen until the end of the procedure.

Postoperatively, her IOP was well controlled on no medication while new vessels were seen over the iris with interspersed iris mamillations (figure 1B), both of which disappeared with postoperative steroids tapered over 2 months.

OUTCOME AND FOLLOW-UP

Dermatological evaluation was sought again to rule out systemic juvenile xanthogranuloma and neurofibromatosis, which was however negative. Iris biopsy showed normal iris tissue interspersed with fibrotic tissue and no tumour cells.

She regained full functional vision of 20/25 despite extensive ghost vessels and complicated posterior subcapsular cataract (figure 2, white arrow) and regressing new vessels (red arrows, inset) nasally with normal IOP of 12 mm Hg at 1 year final follow-up.

At her final follow-up at 15 months, there was evidence of recurrence of iris nodules inferiorly with partial refilling of the regressed (supplementary file) vessels clinching the clinical diagnosis of ocular juvenile xanthogranuloma, for which she is being

kept under very close follow-up to look for recurrence of bleeding. While a repeat iris biopsy would help in tissue diagnosis of the same, a good visual acuity of 20/25 precludes any unnecessary surgical intervention which may cause a rebleed with consequent visual loss. A review dermatological examination with clinical photographs has confirmed the diagnosis of ocular juvenile xanthogranuloma without skin involvement. She is under close dermatological follow-up for a close watch of any dermatological lesions which may mandate skin biopsy for diagnosis.

DISCUSSION

Juvenile xanthogranuloma (JXG) is a self-limiting dermatological condition characterised by spontaneous hyphaema due to uveal involvement causing neovascularisation of iris (NVI) rarely.^{1 2} Neovascular glaucoma can cause intractable glaucoma as seen in our case necessitating surgery.³

This entity represents a spectrum of histiocytic disorders including benign adult xanthogranuloma and progressive nodular histiocytosis. It affects children mostly and is commonly seen as elevated papulonodules in the skin and eye or as neovascular glaucoma as in our case.²⁻⁵ The nodules are usually self-limiting but rarely may require systemic steroids. Ocular sight threatening complications require surgery to prevent permanent visual loss. Iris involvement is commonly seen in the form of iris nodules,⁶ spontaneous hyphaema, uveitis, heterochromia or secondary glaucoma due to iris neovascularisation.⁶⁻¹⁰ Other ocular tissue involvement is less frequent and orbital tumours are very rare. This has to be differentiated from other cases of hyphaema in children like trauma, herpes simplex viral uveitis or ocular tumours like medulloepithelioma or iris melanoma.⁸ Other associations like neurofibromatosis are rarely reported.⁶ In our case, all these were ruled out prior to deciding definitive surgery.

The skin is involved in only half of the cases which is diagnosed by skin biopsy of associated skin lesions.¹ Though skin lesions are associated features of juvenile xanthogranuloma, ophthalmological involvement is seen in 0.5–10% of cutaneous JXG cases. Lesions contain polyhedral histiocytes with Touton cells being present in 85% of the cases.^{4 5} Iris biopsy in our case did not reveal any abnormality since the area of biopsy may not have contained the nodules which were seen interspersed superiorly in the postoperative period.

While most lesions resolve spontaneously or with systemic, oral or topical steroids, our case had recurrent bleeding due to new iris vessels and resultant neovascular glaucoma. Since these new vessels were restricted to the anterior segment, functional recovery was possible with IOP control after surgery, albeit with posterior subcapsular cataract, which may require future surgery.

In case of a total hyphaema with no visible iris or anterior segment details with intractable glaucoma, management

Figure 2 Clinical photograph showing ghost vessels in the anterior chamber (white arrow) and regressing new vessels (red arrow) at 3-month follow-up.

Patient's perspective

This child with spontaneous hyphaema underwent tremendous pain and photophobia for almost 10 days while investigations were being carried out for ruling out other disorders. Daily intravenous mannitol and pain with a 'red' eye had severely disabled her and she was under constant agony before she was taken up for definitive surgery. Her good visual outcome has helped her resume school and is now happily pursuing her studies at school and she is comfortable despite the presence of significant subcapsular cataract, since she is free of pain now.

Learning points

- ▶ Spontaneous hyphaema in a child should be appropriately investigated to rule out all possible causes.
- ▶ Recurrent spontaneous hyphaema with raised intraocular pressure (IOP) should alert the clinician to the possibility of intraocular tumours or juvenile xanthogranuloma with neovascular glaucoma.
- ▶ A valved drainage implant may be useful for IOP control in cases with hyphaema with intractable glaucoma which may reduce changes of postoperative hypotony or failure.
- ▶ Placement of the plate has to be based on the supposed quadrant free of iris lesions as evidenced by preoperative ultrasound biomicroscopy.
- ▶ Good visual outcomes are possible if appropriate tailored investigations and surgical modifications are decided by thorough preoperative workup.

decisions have to be based on the consideration of the best or worst case scenario.³ In our case, routine trabeculectomy would have been disastrous in terms of high risk of failure due to NVI or risk of extensive bleed due to severe hypotony in the post-operative period. So we chose a valved glaucoma drainage device which helps in adequate control of IOP without hypotony as also reducing chances of failure in neovascular glaucoma. Placement of the device was planned in the superonasal quadrant as opposed to the conventional superotemporal quadrant

keeping in mind the focal thickening and fragility of the iris in the inferonasal quadrant which was taken up for biopsy. Such tailored decisions helped achieve good visual recovery and resolution of NVI postoperatively and good vision in our case despite central cataract.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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