Bilateral juvenile xanthogranuloma of the iris

OSMOND BRUCE HADDEN
From the Department of Ophthalmology, Auckland Hospital,
Auckland, New Zealand

Juvenile xanthogranuloma is most commonly a self-limiting dermatological condition of infancy, but intraocular involvement has been well documented during the last 15 years. Correct diagnosis is especially important because of the possibility of successful eradication of the lesion and control of its complications. The present case report outlines one treatment regimen which proved satisfactory. The case is of interest also because it is only the third reported instance of bilateral juvenile xanthogranuloma of the iris.

Case report

A 4-month-old infant was referred with a diagnosis of glaucoma of the left eye. The only history obtainable was that the eyes were noted to be of different colours at birth. The infant's general health was good, and pregnancy and birth had been normal. Examination under general anaesthesia revealed an injected and enlarged left eye. The central five-sixths of the cornea was opaque, probably mainly due to blood-staining, but through the periphery of the cornea blood was seen in the anterior chamber (Fig. 1). The right eye was noted to have a moderate hyphaema but was otherwise normal. The transverse corneal diameters measured 11 mm on the right and 13 mm on the left. Intraocular pressures under anaesthesia were 19 mm Hg on the right and 12 mm Hg on the left using the Draeger applanation tonometer. Several days later without any treatment, the hyphaema in the right eye partially absorbed to reveal a characteristic whitish-yellow lesion on the iris (Fig. 2). At that time four raised pink nodules were noticed on the infant's scalp, the largest measuring about 3 mm in diameter (Fig. 3). Biopsy of two scalp nodules showed an infiltration of the dermis by foamy histiocytes, among which occasional lymphocytes were dispersed. No Touton giant cells were seen (Fig. 4).

Complete blood count, blood clotting factors, and blood biochemistry were normal, including serum lipid electrophoresis. A total x-ray skeletal survey was also normal.

Treatment was begun with oral prednisolone 20 mg per day; the dose was gradually reduced over a 2 mth period. At the same time, radiotherapy was given by the anterior route, using gamma rays from a linear accelerat-

Discussion

Juvenile xanthogranuloma has been reported as a rare cause of epibular tumours (Cogan, Kuwabara, and Park, 1958; Zimmerman, 1965; Nordenstof and Andersen, 1967), and of unilateral exophthalmos (Zimmerman, 1965; Sanders, 1966). However the most common and most damaging ocular manifestations occur with juvenile xanthogranuloma of the iris. Two patients with bilateral iris involvement have been previously reported (Radian, Radian, and Ostfeld, 1964; Smith and

FIG. 1 Left eye before therapy. Enlarged, opaque, and blood-stained cornea
Table  Summary of treated cases of juvenile xanthogranuloma of the iris

<table>
<thead>
<tr>
<th>Author</th>
<th>Date</th>
<th>Age at onset of symptoms</th>
<th>Raised intraocular pressure</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newell</td>
<td>1957</td>
<td>4 mth</td>
<td>Yes (37 mm Hg)</td>
<td>Goniotomy, cyclodialysis, trephine X-ray therapy (400 rad) over 10 days</td>
<td>Tension controlled after trephine Resolved in 1 mth</td>
</tr>
<tr>
<td>Hedges</td>
<td>1959</td>
<td>3 mth</td>
<td>Yes (46 mm Hg)</td>
<td>Topical steroids for 2 mth then x-ray therapy (600 rad) Pilocarpine, acetazolamide, goniopuncture, systemic steroids Lastly, x-ray therapy (500 rad)</td>
<td>Resolved 15 mth later Resolved 8 mth after irradiation</td>
</tr>
<tr>
<td>Maumenee and Longfellow</td>
<td>1960</td>
<td>3 yr</td>
<td>No</td>
<td>Right eye: X-ray therapy (400 rad) over 5 days Left eye: pilocarpine, steroids, 3 goniopunctures</td>
<td>Eye retained but blind Resolved after 14 mth Enucleated</td>
</tr>
<tr>
<td></td>
<td></td>
<td>11 mth</td>
<td>Yes (80 mm Hg)</td>
<td>Left eye: x-ray therapy (180 rad) No therapy. Absolute glaucoma Excision of iris lesion followed by systemic steroid and sulfisoxazole</td>
<td>No recurrence after 2½ yr</td>
</tr>
<tr>
<td>Cleasby</td>
<td>1961</td>
<td>6 mth</td>
<td>Yes (27 mm Hg)</td>
<td>Left eye: No therapy. Absolute glaucoma Excision of iris lesion Postoperative glaucoma Irradiation (400 rad)</td>
<td>Resolved in 10 days Resolved in 4 wk</td>
</tr>
<tr>
<td>Radian and others (bilateral case)</td>
<td>1964</td>
<td>3 mth</td>
<td>Right eye: buphthalmic</td>
<td>Topical steroids and atropine Acetazolamide</td>
<td>Resolved in 4 wk</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Left eye: no</td>
<td>Right eye: Topical atropine and chloramphenicol Systemic tetracycline Left eye: Acetazolamide, topical and systemic steroids</td>
<td>Right eye: enucleated Left eye: improved after 5 days</td>
</tr>
<tr>
<td>Gass</td>
<td>1964</td>
<td>4 mth</td>
<td>Buphthalmic</td>
<td>Right eye: X-ray therapy (180 rad) No therapy. Absolute glaucoma Excision of iris lesion Postoperative glaucoma Irradiation (400 rad)</td>
<td>No recurrence after 2½ yr</td>
</tr>
<tr>
<td></td>
<td></td>
<td>22 mth</td>
<td>No</td>
<td>Left eye: x-ray therapy (180 rad) No therapy. Absolute glaucoma Excision of iris lesion Postoperative glaucoma Irradiation (400 rad)</td>
<td>No recurrence after 2½ yr</td>
</tr>
<tr>
<td>Moore and Harry</td>
<td>1965</td>
<td>1 yr</td>
<td>No</td>
<td>Topical steroids and atropine Acetazolamide</td>
<td>Resolved in 4 wk</td>
</tr>
<tr>
<td>Clements</td>
<td>1966</td>
<td>5 mth</td>
<td>Yes (40 mm Hg)</td>
<td>Right eye: X-ray therapy (180 rad) No therapy. Absolute glaucoma Excision of iris lesion Postoperative glaucoma Irradiation (400 rad)</td>
<td>No recurrence after 2½ yr</td>
</tr>
<tr>
<td>Smith and Ingram (bilateral case)</td>
<td>1968</td>
<td>2 wk</td>
<td>Right eye: buphthalmic</td>
<td>Topical steroids and atropine Acetazolamide</td>
<td>Resolved in 4 wk</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Left eye: yes (36 mm Hg)</td>
<td>Right eye: Topical atropine and chloramphenicol Systemic tetracycline Left eye: Acetazolamide, topical and systemic steroids</td>
<td>Right eye: enucleated Left eye: improved after 5 days</td>
</tr>
</tbody>
</table>

FIG. 2 Right eye before therapy. Hyphaema has partially absorbed to reveal edges of yellow xanthogranuloma of iris (arrows)
Bilateral juvenile xanthogranuloma

FIG. 3 Raised, pink nodules on scalp, 3 mm diameter

FIG. 4 Histological view of dermis of excised skin nodule. Foamy histiocytes among which a few lymphocytes are dispersed. Haematoxylin and eosin. × 300
FIG. 5 Right eye after 3 wk of corticosteroid and radiation therapy. Hyphaema and iris lesion have resolved

FIG. 6 Left eye after 3 wk of therapy. Yellow xanthogranulomata now visible on iris (arrow)

Ingram, 1968). The patient of Radian and others closely resembles the present patient, in that the worse eye was buphthalmic and the better eye had a hyphaema. As in the present patient, the buphthalmic eye also did not respond to therapy but the better eye did. Radian and others treated both eyes with irradiation, and in addition the better eye was subjected to gonipuncture.

The Table summarizes the various published methods used to treat juvenile xanthogranuloma of the iris. In considering the efficacy of any treatment, it must be realized that the iris lesions like the skin lesions may be self-limiting. Resolution may not be related to the therapy. Gass (1964) discussed treatment in general, and suggested that the best course might be a trial of systemic steroids for a few weeks, and if this failed, excision or irradiation could be tried.

He considered excision to be impractical if the lesion involved more than one quadrant of the iris. If glaucoma was present initially, Gass suggested steroid and radiation therapy from the outset, as delay could mean the loss of the eye. Currently, the initial therapy of choice would seem to be systemic and local steroids if there is no glaucoma. But if glaucoma is present, then external irradiation should be combined with the steroids. All reported cases treated with steroids and irradiation have responded, except for two buphthalmic eyes—that of Radian and others and the left eye of the present case. Harris (1953) considered that gamma irradiation in doses exceeding 500 rad should be regarded as potentially cataractogenic.

Summary

Juvenile xanthogranuloma of the iris has only twice before been reported as occurring bilaterally. In this present bilateral case, treatment with steroids and irradiation was successful in eradicating the lesion from the lesser affected eye.

I wish to thank Dr C. C. Ring, Senior Ophthalmic Surgeon, Auckland Hospital, for permission to publish his case, and Dr D. H. Nicholson of Miami, Florida, for helpful advice.

References

GASS, J. D. M. (1964) Ibid., 71, 344
HAM, W. T. (1953) Ibid., 50, 618
HEDGES, C. C., JR. (1959) Amer. J. Ophthal., 47, 683