

Bilateral juvenile xanthogranuloma of the iris

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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 accelera-

tor. A total of 500 rad was given to the right eye and 650 rad to the left eye in 100 rad doses once a wk to both eyes over a 6 wk period. After 3 wk of corticosteroid and radiation therapy, the right iris lesion had gone and the hyphaema had cleared (Fig. 5). The cornea of the left eye had cleared just enough to permit yellow lesions to be seen on that iris (Fig. 6). After 4 wk of therapy, a fresh crop of small subcutaneous nodules was noticed on the scalp at the vertex, but there was no relapse of the ocular condition. On successive examinations under anaesthesia during the following 4 mth, the pressure in the left eye became very high and a left trabeculectomy was performed. Unfortunately, 5 mth after surgery, the left eye had not improved; extensive granulomata remained on the iris, and the pressure was still high. The right eye, however, remained well with no recurrence of the iris lesion and with a normal pressure and clear lens. No further treatment was given to try to improve the left eye.

Discussion

Juvenile xanthogranuloma has been reported as a rare cause of epibulbar tumours (Cogan, Kuwabara, and Park, 1958; Zimmerman, 1965; Norden-toft 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

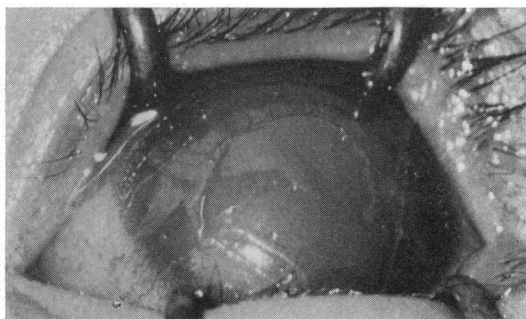


FIG. 1 *Left eye before therapy. Enlarged, opaque, and blood-stained cornea*

Table Summary of treated cases of juvenile xanthogranuloma of the iris

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

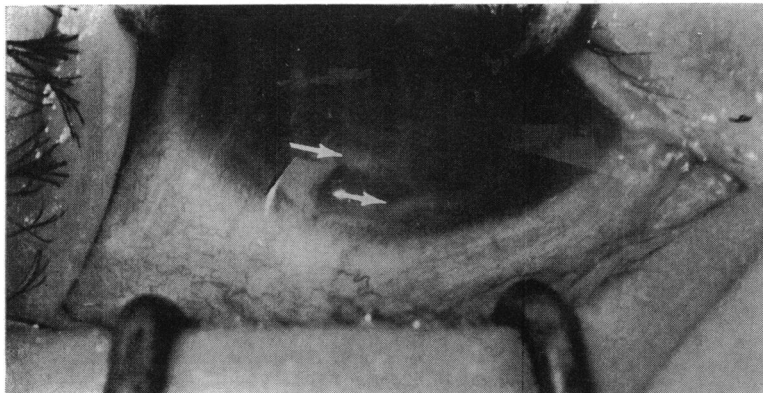


FIG. 2 Right eye before therapy. Hyphaema has partially absorbed to reveal edges of yellow xanthogranuloma of iris (arrows)



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

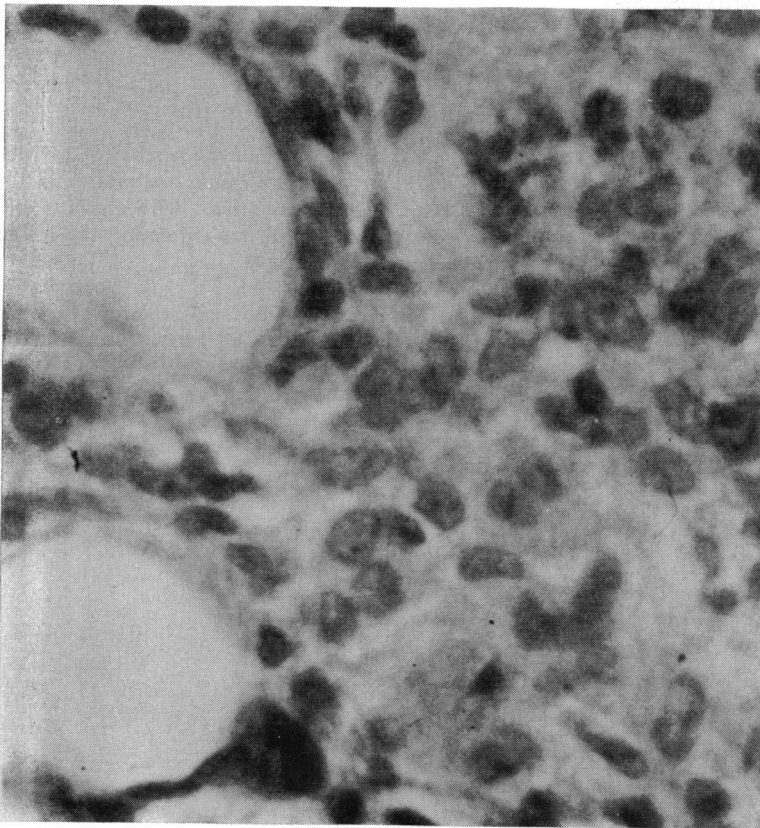


FIG. 4 *Histological view of dermis of excised skin nodule. Foamy histiocytes among which a few lymphocytes are dispersed. Haematoxylin and eosin. $\times 300$*

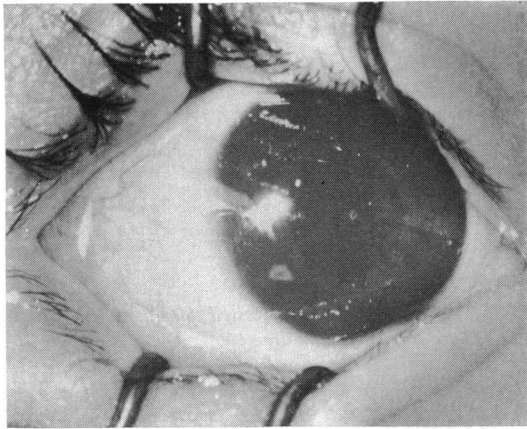


FIG. 5 Right eye after 3 wk of corticosteroid and radiation therapy. Hyphaema and iris lesion have resolved

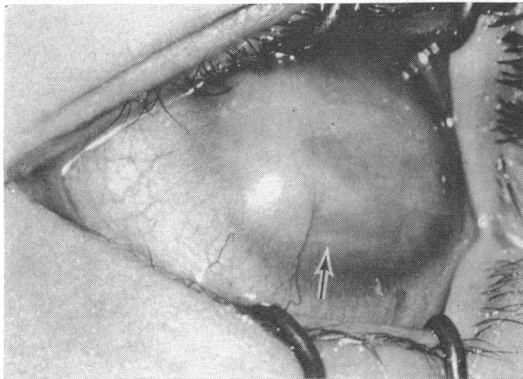


FIG. 6 Left eye after 3 wk of therapy. Yellow xanthogranulomata now visible on iris (arrow)

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 buphthal-

mic 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 goniopuncture.

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. Ham (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.

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