Brit. J. Ophthal. (1966) **50**, 663

CASE NOTES

JUVENILE XANTHOGRANULOMA TREATED WITH LOCAL STEROIDS*

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JUVENILE xanthogranuloma is an iris tumour associated with a skin lesion commonly occurring in the first year of life. Haemorrhage and secondary glaucoma frequently accompany the ocular lesion, but it is not generally regarded as neoplastic. This case is described because it responded satisfactorily to local steroids.

Case Report

A 5½-month-old female infant was admitted to St. Paul's Eye Hospital, Liverpool, on August 2, 1964, because the mother had noticed a grey swelling of the left iris 2 weeks previously; she had also noticed that the eye suddenly "went brown" 2 days before admission (? was this due to a spontaneous hyphaema). She was sure that both eyes were normal and had blue irides at birth. There was no family history of eye disease. The child had two siblings; one had died of atelectasis at one week of age, and the other was 4 years and 9 months old and alive and well. The birth was a full-term normal delivery.

Examination.—A mass was noted in the anterior chamber of the left eye with diffuse hyphaema. Apart from the eye, a small yellowish lesion was noted on the right cheek about ¾″ × ¼″ in size, which had been present since the child was 3 months old. Physical examination revealed no other significant abnormality. The child's weight was 17 lb. 4 oz.

An examination under general anaesthetic was undertaken on August 8, 1964. Anaesthesia was induced with nitrous oxide and maintained with Halothane. The findings were as follows: corneal diameter 11·5 (left). Left eye brown iris tumour of the lower segment associated with diffuse hyphaema. Tension (Schiötz weighted) 14·6 mm. Hg in the right eye and 38·9 mm. Hg in the left.

The right eye was normal. A sternal marrow puncture was taken.

Treatment.—A provisional diagnosis of xanthogranuloma was made and treatment was started with gutt. Betnesol-N 2-hrly and gutt. atropine 1 per cent. three times daily to the left eye and o.d. to the right.

Laboratory Investigations: Hb 71 per cent., white cell count 9,500, polymorphs 38 per cent., small lymphocytes, 57 per cent., monocytes 4 per cent., eosinophils 1 per cent. Sternal marrow count normal. Chest *x* ray normal.

The patient was seen by a plastic surgeon who stated that the face lesion should not be biopsied unless absolutely necessary, because of the risk of cosmetic deformity.

Progress.—The patient remained in hospital for a further 6 weeks until her discharge on September 26, 1964. During this time a further five examinations under general anaesthesia were carried out. Treatment was continued with steroids and atropine locally to the left eye, and Diamox 65 mg. daily was started on August 13. The intra-ocular pressure initially remained between 35 and 45 mm. Hg but gradually subsided, and by the time of discharge was 28 mm. Hg with Diamox therapy. The corneal diameter enlarged a little and was 12·5 mm. on discharge.

The hyphaema was slow to clear but had gone by the end of 4 weeks. The tumour gradually subsided; after 3 weeks of treatment it could hardly be seen, and after 4 weeks was no longer visible.

* Received for publication June 16, 1965.

Follow-up.—The patient was examined under general anaesthesia at monthly intervals. The pressure remained controlled and Diamox was stopped after one month's out-patient treatment. The corneal diameter did not continue to enlarge and there was no recurrence of tumour or hyphaema.

On February 11, 1965, the lower iris over the site of the initial tumour was atrophic and drawn forwards to a broadly-based peripheral anterior synechia below. There was also some posterior synechia below. At first this suggested the appearance of tumour, but examination with the binocular microscope showed only distortion of the iris plane due to peripheral anterior synechiae. There was some lens opacity below, but a good view was obtained of the fundus, and the disc, macula, and vessels were normal. Gonioscopy showed a light brown generalized discoloration of the angle.

Since then the eye has remained quiet and the mother says the child sees well with the eye. There is no evidence of strabismus.

The right eye has always been found to be normal.

Discussion

The condition of naevo-xantho-endothelioma was first described in the dermatological literature by McDonaugh (1909). The first case with ocular involvement was described by Blank, Eglick, and Beerman (1949). Since then the diagnosis has been made with increasing frequency and Sanders (1962) described twenty cases, seventeen of them in the first year of life. The classical picture of spontaneous hyphaema associated with iris tumour and raised tension in the first year of life should suggest the diagnosis of juvenile xanthogranuloma.

Howard (1962) pointed out the necessity for bilateral fundus examination to exclude certain of the differential diagnoses: retrolental fibroplasia, retinoschisis, persistent primary hyperplastic vitreous, retinoblastoma, and blood dyscrasias. Of these, retrolental fibroplasia, retinoschisis, and retinoblastoma can often be excluded by bilateral fundus examination.

The importance of making the diagnosis is clear, because in the past many of these eyes have been needlessly removed because of suspected malignant tumours. The diagnosis in our case was made on the basis of the typical features, there being no evidence of any other condition. Repeated examination of the other eye showed it to be normal and helped to exclude some of the differential diagnoses suggested above. Blood-film and marrow-puncture examinations excluded blood dyscrasias.

Suggested therapy for juvenile xanthogranuloma has involved local and systemic treatment with steroids, radiotherapy, and excision. Radiotherapy was advocated by Maumenee and Longfellow (1960), the suggested dose being not more than 200 r in any one treatment and 500 r in all. Excision was part of the treatment carried out in the only other case described in the English literature by Moore and Harry (1965). Gass (1964) suggested that, if the case presented with iris tumour and glaucoma, then treatment should be initially with systemic steroids and irradiation.

The treatment of the present case is interesting in that the condition appears to have responded to local steroids and Diamox alone. The use of systemic steroids was considered and rejected, since these carry great dangers, especially in young infants. It is now generally accepted that retardation of normal growth occurs in children given systemic steroids as well as all the unpleasant side-effects seen in adults. Radiotherapy carries dangers of radiation cataract, xerosis, and corneal damage. By contrast, it would appear that a preliminary trial with local steroids is worthwhile;

these carry few risks apart from possible steroid glaucoma for which a careful watch should be kept.

Summary

A case of juvenile xanthogranuloma was interesting in that it appeared to have responded to treatment with local steroids and Diamox alone.

My thanks are due to Mrs. Dorothy Barton and Mr. T. D. H. Gray for their permission to publish this case and for their advice.

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