loma in high myopia has been described recently.4

In the present case, we believe the healing of the posterior filtering site was caused by mechanical action and by the formation of a fibrotic scar secondary to the external silicone buckle that was fitted to cover the scleral defect.

Although ocular hypotony caused by spontaneous rupture of thin sclera forming the floor of a chorioretinal coloboma is an uncommon event, an external scleral buckling technique was beneficial in this case and provides a therapeutic option.

Francesco Viola, MD
Francesco Morescalchi, MD
Enrico Gandolfo, MD
Giovanni Staurenghi, MD

Correspondence: Dr Staurenghi, Department of Ophthalmology, University of Brescia, Via Tiraboschi 8, 20135 Milan, Italy (giovanni.staurenghi@unimi.it).


Juvenile Xanthogranuloma
With Presumed Involvement of the Optic Disc and Retina

Juvenile xanthogranuloma (JXG) is a rare idiopathic granulomatous disorder of early childhood. Ocular involvement of the anterior segment, notably the iris, is well recognized. Involvement of the optic nerve disc is exceptionally rare and is associated with loss of vision. We are aware of only 2 previously published reports of optic disc involvement, one proven on histologic examination1 and the other presumed.2 We describe herein a third child who was initially identified by screening and in whom it was possible to detect early optic disc involvement. As far as we know, we describe for the first time the prospective clinical management of this vision-threatening condition during more than 2 years of follow-up. This case illustrates the natural history of this condition and demonstrates that vision can be partly preserved with early detection and treatment.

Report of a Case. A previously healthy 11-month-old white girl was referred to the pediatric dermatology department with a 5-month history of a progressive yellowish papular rash on her face and eyelids (Figure 1 and Figure 2). A skin biopsy at age 12 months confirmed classic JXG composed of plump histiocytic cells intermingled with spindle cells and numerous multinucleate Touton giant cells, the histopathologic hallmark of JXG (Figure 3). Immunostaining for factor XIIIa was positive, while CD1a staining was negative and S100 staining showed occasional positive staining of dendritic cells. A diagnosis of JXG was made. It was decided to

![Figure 1. Facial distribution of the yellowish papular rash involving the eyelids at 11 months of age.](image1)

![Figure 2. Close-up view of a juvenile xanthogranuloma papule.](image2)
observe the skin lesions because of no systemic involvement and because cutaneous JXG frequently resolves spontaneously. Because of “blank spells,” a referral was made at age 13 months to a pediatric neurologist who did not find any neurological abnormality. Results of a brain magnetic resonance imaging scan with gadolinium and an electroencephalogram were also found to be normal.

Initial routine screening in the pediatric ophthalmology clinic at age 17 months revealed hypermetropia (4.50 diopter spheres [DS] OU) but otherwise healthy-looking eyes with a visual acuity of 20/32 OU, no squint, and unremarkable optic nerve discs. Between 17 and 24 months of age, she developed anisohypermetropia (4.00 DS OD and 7.50 DS OS) and intermittent exotropia in her left eye. Her left optic disc margin was noted to be slightly blunted at 21 months of age. However, pattern reversal visual evoked potentials showed no significant interocular difference. Patching was initiated and glasses prescribed. On review at age 27 months, an infiltrating mass of the left optic nerve papilla was seen, which extended to the macula (Figure 4). Within 6 days, her visual acuity deteriorated from 20/60 to 20/400 OS.

She had had chickenpox at age 19 months but was otherwise healthy. No other ocular abnormalities, including the anterior segment, were seen. Her physical examination findings were unremarkable, and she has remained otherwise healthy throughout her follow-up. No axillary freckling or cafe-au-lait spots were found. A chest radiograph showed no abnormality.

Pretreatment laboratory values (followed by reference ranges) included the following: white blood cells, 9.9 × 10^3/µL (5.0-15.0 × 10^3/µL); lymphocytes, 6.5 × 10^3/µL (2.0-9.5 × 10^3/µL); monocytes, 0.7 × 10^3/µL (0.3-1.5 × 10^3/µL); eosinophils, 0.2 × 10^3/µL (0.3-0.8 × 10^3/µL); basophils, 0.1 × 10^3/µL (0-0.2 × 10^3/µL); red blood cells, 5.0 × 10^12/µL (3.7-5.3 × 10^12/µL); hemoglobin, 13.2 g/dL (10.5-13.5 g/dL); platelets, 701 × 10^3/µL (150-450 × 10^3/µL); erythrocyte sedimentation rate, 5 mm/h (0-10 mm/h); C-reactive protein, 0.9 mg/dL (0-2.0 mg/dL); glucose, 92 mg/dL (5.1 mmol/L) (63-99 mg/dL [3.5-5.5 mmol/L]); sodium, 142 mEq/L (135-145 mEq/L); potassium, 4.4 mEq/L (3.5-5.5 mEq/L); serum urea nitrogen, 12 mg/dL (4.4 mmol/L) (9-19 mg/dL [3.3-6.6 mmol/L]); creatinine, 0.4 mg/dL (32 µmol/L) (0-0.6 mg/dL [0-56 µmol/L]); calcium, 10.4 mg/dL (2.60 mmol/L) (9.1-10.6 mg/dL [2.28-2.64 mmol/L]); magnesium, 2.1 mg/dL (0.88 mmol/L) (1.8-2.4 mg/dL [0.74-1.00 mmol/L]); phosphate, 5.9 mg/dL (1.89 mmol/L) (4.0-5.5 mg/dL [1.29-1.78 mmol/L]); albumin, 4.5 g/dL (3.5-5.5 g/dL); total bilirubin, 0.5 mg/dL (9 µmol/L) (0-1.1 mg/dL [0-18 µmol/L]); alkaline phosphatase, 210 U/L (110-350 U/L); and ala-

**Figure 3.** Typical Touton-type giant cells showing the classic multinucleate cell with nuclei in a circumferential arrangement surrounded by small fat droplets are seen on an eyelid skin biopsy specimen (hematoxylin-eosin, original magnification approximately ×160).

**Figure 4.** Evolution of the yellowish-white lesion extending from the left optic disc to the macula at age 27 months (A), 39 months (B), and 47 months (C).
nine transaminase, 19 U/L (5-45 U/L). Further serologic test results were positive for IgG antibodies against varicella zoster virus and measles virus and were negative for cytomegalovirus antibody, Epstein-Barr virus IgG, and herpes simplex viruses 1 and 2 IgG. The angiotensin-converting enzyme level was 11 U/L (20-90 U/L), and the Toxocara antibody test result was negative at age 2 years 8 months.

B-mode ultrasound of her left globe revealed a raised solid lesion overlying the optic disc and adjacent macula. Magnetic resonance imaging demonstrated an optic disc lesion without further optic nerve, orbital, or intracranial extension. Electrophysiologic findings are shown in Figure 5.

She received 2 once-daily pulses of intravenous methylprednisolone succinate (500 mg/d, or 30 mg/kg of body weight at each pulse) followed by oral prednisolone sodium phosphate at a dosage of up to 3 mg/kg per day. Corticosteroid treatment was well tolerated, and her visual acuity rapidly recovered from 20/400 OS to 20/80 OS. Her corticosteroid dosage was therefore slowly tapered for 3 months. Within 2 weeks of stopping the corticosteroid treatment, however, her left vision deteriorated again owing to vitritis and progression of the optic disc lesion. Oral prednisolone treatment was restarted at 3 mg/kg. She did well for the subsequent 16 months on a slow corticosteroid taper. With patching and glasses, vision in the left eye improved to as much as 20/50.

Twenty months into her treatment, she developed a large peripapillary tractional retinal detachment with vitreous hemorrhage. No clinical evidence of rebound inflammation was seen. Despite vitrectomy and membrane delamination, the macula remained severely puckered because of persistent or recurrent traction. Intraoperative biopsy of the vitreous and posterior hyaloid confirmed an inflammatory membrane but showed no evidence of active xanthogranuloma. Her vision 3½ months following her surgery was 20/200 OS and 20/20 OD while receiving 1.5 mg/d of maintenance prednisolone. The relationship between visual acuity, corticosteroid treatment, and patching is shown in Figure 6.

**Comment.** Involvement of the optic disc is a rare complication of JXG. In the 2 cases reported previously, the affected eyes were already blind at initial examination. It is therefore not known how best to treat optic disc and macular involvement. To our knowledge,
we herein report for the first time the prospective long-term response to early treatment and the electro-physiologic findings in such a case.

Loss of vision may be due to different mechanisms. Serial pattern reversal visual evoked potentials demonstrated attenuation and increased latency in the left eye of our patient. Attenuation was also found in the left pattern electroretinogram, indicating left macular pathway dysfunction. Moreover, the left flash electroretinogram showed a persistent relative delay in B-wave latency to all intensities compared with the right eye, indicating additional involvement of the inner retina, probably due to early retinal vascular compromise or the direct effect of infiltration. We observed tractional retinal detachment of the macula due to fibrosis as a late complication and as a further mechanism of visual loss in posterior segment disease. We emphasize that, apart from the direct pathologic effects of JXG disease and its treatment, special consideration should be given to the recognition of amblyopia and its management to maximize the remaining visual potential in the affected eye.

We believe that corticosteroid treatment was effective in treating the optic disc and macular lesion during the inflammatory phase, as shown by the marked initial improvement of inflammatory activity and vision while receiving systemic corticosteroids, the clinical deterioration on stopping treatment, and the clinical improvement on starting corticosteroid regimens again. Once the inflammation was controlled, vision improved further with treatment for amblyopia (Figure 6). Glasses were prescribed and updated throughout follow-up. Visual acuities were usually obtained by the same senior orthoptist (C.T.) who also supervised the occlusion therapy. The final visual deterioration at age 3 years 11 months followed 20 months of systemic corticosteroid treatment and was caused by tractional retinal detachment thought to be due to fibrosis rather than rebound inflammatory activity.

Should children with cutaneous JXG routinely be screened by ophthalmologists? Ocular involvement subsequent to initial cutaneous presentation is potentially vision threatening but uncommon. Chang et al.\(^4\) reported a survey incidence of ocular complications in children with cutaneous JXG of 0.3% and a literature incidence of 0.4%. Children with newly diagnosed JXG, multiple skin lesions, and onset at 2 years or younger were found to be at greatest risk and should therefore be targeted for surveillance.\(^3\) Chang\(^4\) suggested annual or semiannual ophthalmic screening in these high-risk groups. As a mini-
mum, all parents should be instructed to seek immediate specialist ophthalmological advice in case of any new eye pain, redness, squint, or visual complaint in a child with recognized cutaneous JXG. If intraocular involvement is found, we recommend close follow-up and early intervention, as clinical deterioration may be precipitous and difficult to control.

Göran Darius Hildebrand, MD, MRCOphth, MRCS(Edin)
Chris Timms, DBO(T)
Dorothy A. Thompson, PhD
David J. Atherton, FRCP
Marian Malone, FRCPath
Gill Levitt, FRCP
D. Alistair H. Laidlaw, FRCOphth
Isabelle Russell-Eggitt, FRCOphth
David S. I. Taylor, FRCP, FRCS, FRCOphth

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Correspondence: Dr Taylor, Department of Paediatric Ophthalmology, Great Ormond Street Hospital for Children, London WC1N 3JH, England (DSIT@btinternet.com).


Bilateral Optic Neuropathy Associated With Voluntary Globe Luxation and Floppy Eyelid Syndrome

Report of a Case. An obese 35-year-old man was examined because of loss of vision in his right eye and gradually decreasing vision in his left eye. He also complained of redness, irritation, and a foreign-body sensation bilaterally. His medical history and his vision had previously been excellent, according to his medical records at the company at which he was employed as a computer engineer. However, during the past 4 years he had had an obsessive-compulsive disorder, which was treated with risperidone. His parents reported that he had a peculiar habit of luxating his eyes several times a day. When he was asked to demonstrate this, he easily everted his upper eyelids (Figure 1) and luxated his globes with his finger (Figure 2). He then

Figure 1. Voluntary eversion of the floppy upper eyelid.

Figure 2. A, Luxation of the globe with the patient’s finger. B, Lateral view of the luxated globe. C, Luxated globe as seen from above the patient.