

**Figure 3.** 18F-fluoro-2-deoxyglucose positron emission tomography scan showing 2 isolated hypermetabolic foci in the left upper pulmonary lobe (top arrow) and the left hilum (bottom arrow).

MRI, and bone scintigraphy. A whole-body 18F-fluoro-2-deoxyglucose PET (FDG-PET) scan revealed 2 isolated hypermetabolic foci in the left upper pulmonary lobe and the left hilum, while the eye lesion was not detected (**Figure 3**). The patient received a short course of radiotherapy to the right eye. An upper-left lung lobectomy was performed and confirmed a primary adenocarcinoma (2 cm diameter; T1 N2 M1, stage IV). A chemotherapy of cisplatin and gemcitabine was initiated. At 6 months' follow-up, the choroidal lesion was no longer present and visual acuity has stabilized.

**Comment.** Positron emission tomography scanning is able to identify a previously unknown occult primary tumor in 20% to 50% of patients after unsuccessful conventional diagnostic investigations.<sup>2</sup> No data are available concerning the smallest size of a lesion that can be detected; detection depends on the lesion's metabolic activity. Furthermore, FDG-PET contributes to detection of tumor dissemination, and to the patient's therapeutic management.<sup>3,4</sup> The improvement of primary tumor detection rates is of great prognostic and therapeutic value, especially for patients with 1 solitary

visceral metastasis, as an option for curative treatment is not unlikely.<sup>3,4</sup> In our particular case, FDG-PET proved to be a very useful method for diagnosis and therapeutic management.

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### Ocular Hypotony Secondary to Spontaneously Ruptured Sclera in Choroidal Coloboma

We describe a patient who developed unilateral optic disc edema due to ocular hypotony secondary to spontaneous rupture of the thin sclera forming the floor of a chorioretinal coloboma. B-scan ultrasonography was useful in confirming the diagnosis of a posterior filtering site. The scleral defect was successfully treated with implantation of a meridional silicone buckle.

**Report of a Case.** A 63-year-old white woman complained of a 3-month history of painless blurred vision in her left eye. Her medical and ocular history was unremarkable. Her best-corrected visual acuity was 20/20 OD and 20/50 OS, with intraocular pressure of 15 mm Hg OD and 4 mm Hg OS. Biomicroscopic fundus examination of the patient's left eye was remarkable for

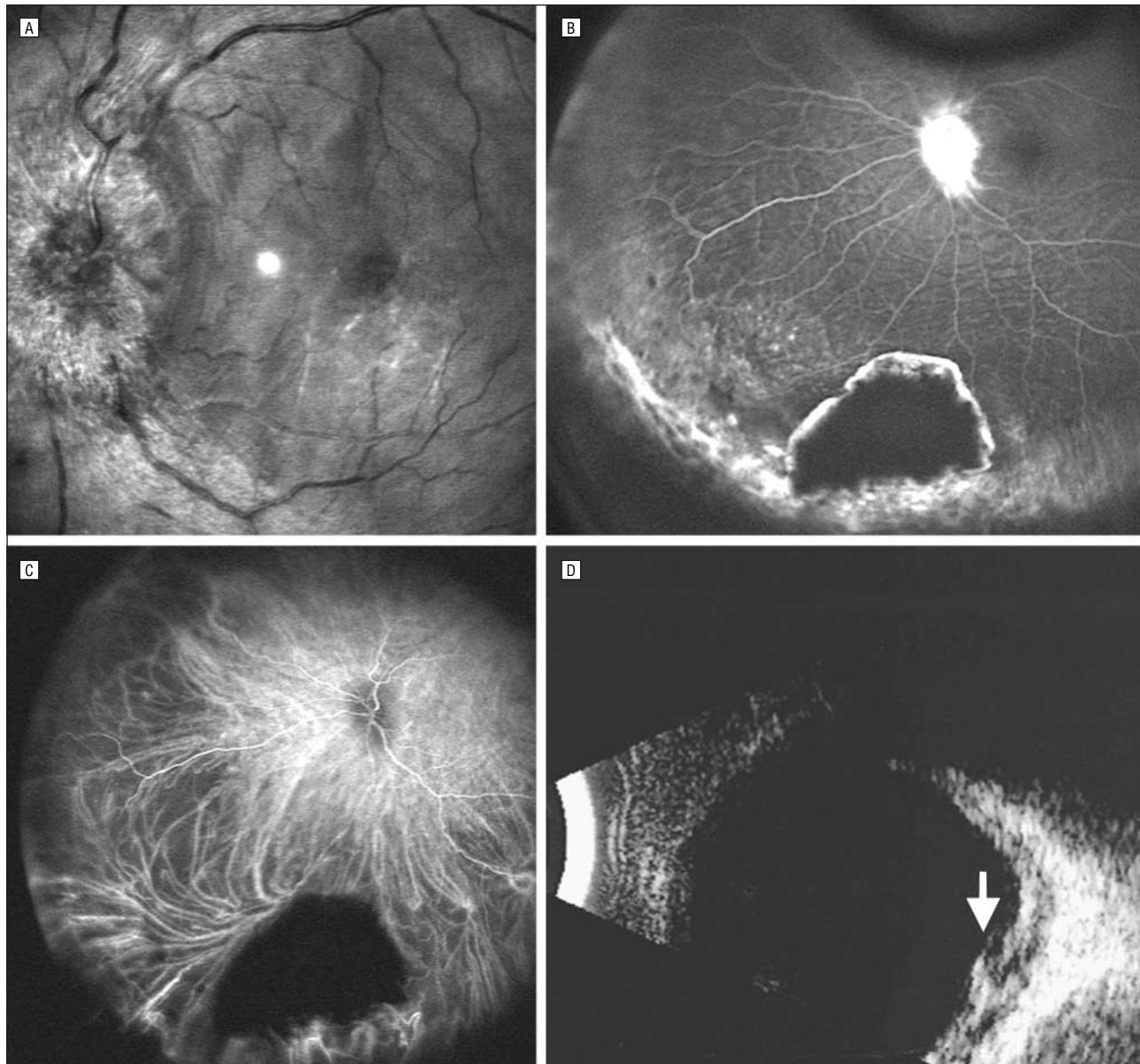
fully developed optic disc edema with gross elevation of the optic nerve head, blurred disc margins, and choroidal folds consistent with hypotony (**Figure, A**). The peripheral fundus examination showed a chorioretinal coloboma in the inferonasal equatorial retina, with an oval retinal break and a subclinical retinal detachment restricted to the colobomatous area. The size of the coloboma was about 6 disc diameters, while the size of the break was 1.5 disc diameters. The disc and the macula were not involved in the coloboma. Results of examination of the right eye were normal.

Fluorescein angiography showed dye leakage from the optic nerve in the left eye, staining at the edge of the coloboma, and a dark area caused by the absence of the choroid and of the retinal pigment epithelium (**Figure, B**). Indocyanine green angiography confirmed the absence of choroid (**Figure, C**). B-scan ultrasound examination showed a diffuse swollen choroid and a hypoechoic region posterior to the sclerochoroidal defect in the markedly thin and deformed sclera that formed the floor of the coloboma, consistent with fluid (**Figure, D**).

On the basis of findings from the clinical examination and supporting imaging studies, a diagnosis of ocular hypotony secondary to spontaneous posterior rupture of the thin sclera, forming the floor of the coloboma, was made.

Treatment options were evaluated; the day before surgery, the retinal break was treated with laser photocoagulation along the border of the coloboma as a prophylactic procedure. During the surgery, limited prolapsed vitreous was excised, 2 vertical mattress sutures were placed between the scleral coloboma to create a deep buckle, and a meridional solid silicone buckle was fitted to cover the scleral defect, with the goal of sealing the posterior filtering site.

One year postoperatively, examination of the left eye was remarkable for a visual acuity of 20/25 and intraocular pressure of 13 mm Hg; fundus view showed the retina attached with a mildly swollen optic nerve head and some choroidal folds still present.



A, Infrared image of the left fundus with a swollen disc and choroidal folds consistent with ocular hypotony. B, Wide-field fluorescein angiogram showing leakage from the optic nerve in the left eye, staining at the edge of the coloboma, and a dark area caused by the absence of the choroid and of the retinal pigment epithelium. C, Wide-field indocyanine green angiogram showing the absence of choroid as a dark area. D, B-scan ultrasound scan showing a diffuse swollen choroidal layer and a full-thickness passage between the posterior chamber and the retrobulbar tissue (arrow).

**Comment.** Typical coloboma is a congenital defect caused by the improper closure of the embryonal fissure. The reported incidence of retinochoroidal coloboma is 0.14%, and in 40% of these patients rhegmatogenous retinal detachment may develop sometime during their lifetime.<sup>1</sup>

Unilateral optic disc edema secondary to ocular hypotony after a spontaneous rupture of the thin sclera, forming the floor of the posterior chorioretinal coloboma, has not yet been described, to our knowledge. We preoperatively iden-

tified an oval retinal break that occurred within the intercalary membrane of the coloboma, with a subclinical retinal detachment restricted to the colobomatous area (type IIA according to the classification by Gopal et al<sup>1</sup>). No vitreal traction or operculum could be identified in front of the break, suggesting the atrophic origin of the break within the atavistic retinal tissue. This type of break, occurring in the retinal tissue within the coloboma, corresponds to the second type of breaks identified by Gopal et al.<sup>1</sup>

Most likely the asymptomatic

retinal detachment, restricted to the colobomatous area, was caused by the break in the diaphanous tissue that is continuous anatomically with the neurosensory retina. The posterior bulging of the sclera in the affected area, forming a staphyloma, has been reported in the literature.<sup>2,3</sup> The retinal break and the asymptomatic retinal detachment could have been present for a long time, and the spontaneous rupture of the thin sclera could have occurred afterward. A similar case of ocular hypotony secondary to spontaneous ruptured posterior staphy-

loma in high myopia has been described recently.<sup>4</sup>

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.

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### 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 examination<sup>1</sup> and the other presumed.<sup>2</sup> We describe herein a third child who was initially identified by screening and in whom it was possible to detect early optic disc involve-

ment. 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.



Figure 2. Close-up view of a juvenile xanthogranuloma papule.