copy of these vacuoles suggests secondary lysosomes. It is unknown exactly how the impaired lysosomal function results in the clinical features, but it is likely that retained storage material in the retina along with other, yet-to-be-elucidated mechanisms results in widespread RPE dysfunction and photoreceptor death.

In conclusion, the 2 patients presented herein and those described by Springer et al demonstrate that, in addition to corneal and lenticular opacity, retinal dystrophy may develop in patients with α-mannosidosis. Consideration of retinal function, either through ERG or detailed fundus examination, is warranted prior to planning any surgical correction of corneal or lenticular opacity. Further study is required to determine the frequency with which it occurs and the rate of progression.

Robert Jackson Courtney, MD
Mark E. Pennesi, MD, PhD

Author Affiliations: Casey Eye Institute, Department of Ophthalmology, Oregon Health and Science University, Portland, Oregon.

Correspondence: Dr Pennesi, Casey Eye Institute, Oregon Health and Science University, 3375 Terwilliger Blvd, Portland, OR 97239 (pennesim@ohsu.edu).

Financial Disclosure: None reported.

Funding/Support: This study was supported by grants C-GE-0706-0365-OHSU01 and from Foundation Fighting Blindness; Collins Medical Trust; and Research to Prevent Blindness.


Spontaneous Regression of Small Melanocytic Choroidal Tumor

The management of small melanocytic choroidal tumors is controversial because the natural course and metastatic potential of these lesions are not clearly defined. Factors predictive of growth into melanoma include a tumor thickness greater than 2 mm; the posterior margin touching the optic disc; presence of symptoms (flashing, floaters, and blurred vision), orange pigment, and subretinal fluid; ultrasonographic hollowness; absence of halo; and absence of drusen.

The purpose of our report is to describe the rare behavior of 2 cases of small melanocytic choroidal tumors with several risk factors for growth into melanoma that exhibited spontaneous regression during follow-up. To the best of our knowledge, both cases represent the first description of spontaneous regression of small melanocytic choroidal tumors with risk factors for growth into melanoma.

Report of Cases. Case 1. A healthy 23-year-old woman was referred to our ophthalmology department after complaining of distorted vision (metamorphopsia) and blurred vision in the right eye for more than 1 month. Her visual acuity was 20/25 in the right eye and 20/20 in the left eye. During a fundus examination of the right eye, she was found to have an oval melanotic juxtafoveal choroidal lesion with clumps of orange pigment spreading over the surface. Furthermore, it had overlying subretinal fluid extending into the macula, and neurosensory macular detachment was demonstrated by optical coherence tomographic scans. B-scan ultrasonography revealed a choroidal mass 6.49 mm in width and 2.27 mm in thickness.
A-scan ultrasonography revealed high internal reflectivity, a regular structure, and no angle kappa.

Owing to the risk factors for growth, we decided to monitor the lesion closely. The patient underwent a complete ophthalmic evaluation, retinography, optical coherence tomography, and ultrasonography every 3 months. Six months after her initial visit, an optical coherence tomographic scan revealed spontaneous resolution of subretinal fluid, improving her visual acuity to 20/20 and without apparent change in its height. Moreover, after a 4-year follow-up period, the ophthalmoscopic and ultrasonographic appearance of the choroidal lesion resulted in almost complete regression to 1.38 mm height, yielding flat pigmentation with overlaying fibrotic metaplastic areas (Figure 2). The patient was not pregnant during the follow-up period.

Case 2. A healthy 71-year-old woman presented to our department to be evaluated for the onset of orange pigmentation on an asymptomatic choroidal nevus noted on routine fundoscopy 2 years prior in the right eye (case 2). During examination, visual acuity was determined to be 20/20 in both eyes. Evaluation of the right eye revealed an oval melanotic choroidal lesion located in the inferotemporal quadrant with small overlying clumps of orange pigment. Neither subretinal fluid nor drusen were observed. Ocular ultrasonography revealed a choroidal lesion measuring 6.5 × 7.0 mm in basal diameter and 2.2 mm in thickness. The tumor margins to the optic nerve and the fovea were 3 mm and 4 mm, respectively (Figure 3).

Our diagnosis was choroidal nevus with risk factors for growth into melanoma. Close observation every 3 months was advised. One year after the initial presentation, fundus examination and B-scan ultrasonography revealed a flat lesion. After a 6-year follow-up period, we observed a progressive involution of the pigmented areas with exposure
of the underlying sclera (Figure 4). The white area in the center of the tumor clinically corresponded to a depression without tumoral tissue with the same aspect as that of a choroidal coloboma (congenital or surgical coloboma). We did not find any ultrasound signal that could correspond to a fibrous metaplasia. During the follow-up period, fluid or exudates over the lesion were not evident clinically or by use of optical coherence tomography.

Comment. There is clinical and histopathological evidence suggesting that choroidal melanoma arises from preexisting choroidal nevi. It seems likely that, at some stage in its development, a choroidal melanoma will potentially resemble a choroidal nevus and be harbored within the “nevus” population until its ophthalmoscopic or ultrasonographic features give warning of its malignant nature, and it ceases to be classed as a nevus. Thus, most of these lesions are named as small melanocytic choroidal tumors until growth is demonstrated. For Yanoff and Zimmerman,2 this term is used until the identification of several risk factors for growth, at which point the tumors are considered as uveal melanomas.

We sought to establish a correlation between the spontaneous regression of cutaneous and choroidal lesions. Spontaneous involution is a phenomenon present in a variety of cutaneous lesions, including cutaneous melanocytic nevi and cutaneous melanomas. The phenomenon has also been observed in limited cases of uveal melanoma.3 However, it has not been reported in small melanocytic choroidal lesions with risk factors for growth.

The underlying pathogenesis is not yet fully understood, but cell-mediated immunity (cytotoxic T lymphocytes) is thought to play a key role in the progressive destruction of nevus cells.4 It has been demonstrated in the murine intraocular melanoma model that spontaneous regression can occur in immunologically intact hosts, as determined through the use of cytotoxic T lymphocytes and delayed-type hypersensitivity responses directed against melanoma cells.5 To our knowledge, both previously reported cases represent the first descriptions of spontaneous regression with no recurrence over an extended follow-up period of small melanocytic choroidal tumor with risk factors for growth into melanoma.

Jose M. Caminal, PhD
Maravillas Abia, MD
Daniel Lorenzo, MD
Luis Arias, PhD
Juan C. Mesa, PhD

Author Affiliations: Department of Ophthalmology, Bellvitge University Hospital, L’Hospitalet de Llobregat, Barcelona, Spain (Drs Caminal, Abia, Lorenzo, Arias, and Mesa). Correspondence: Dr Caminal, Department of Ophthalmology, Bellvitge University Hospital, Hospital de Llobregat, Barcelona 08062, Spain (jmcaminal@bellvitgehospital.cat).

Financial Disclosure: None reported.