Congenital Amblyopia, Scleral Calcification and Choroidal Melanoma

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Calcification of the sclera is occasionally seen in degenerating eyes. It may occur as an aging change, secondary to chronic ocular disease or, rarely, as a result of excessive vitamin D treatment. These calcifications usually are seen as small discrete patches which replace the scleral substance most typically just anterior to the insertions of horizontal extraocular muscles. Grossly they represent bluish spots which may sometimes be visible through the conjunctiva and may be mistaken for scleral defects. Although less common than the anterior form, calcification of the posterior sclera has been reported and may rarely even exhibit bone formation.

This is to report the unusual association of symmetrical calcification in the posterior sclera with malignant melanoma in an amblyopic eye. A case of symmetrical calcification in the anterior sclera is presented for comparison.

Case Reports

Case 1: This left eye of an 85-year-old male was removed on February 27, 1964, for blindness and pain in absolute glaucoma. Gross and histologic examination revealed absolute hemorrhagic glaucoma secondary to old occlusion of the central retinal vein. There was an acute corneal ulcer, bullous keratopathy, angle closure, senile cataract and atrophy of inner retina as well as optic nerve.

Two patches of scleral calcification measuring 2 x 2 mm. were observed anterior to the insertions of the horizontal rectus muscles (Fig. 1). These patches were symmetrical and sharply limited. They extended to the scleral surface, but did not involve the innermost layers. The calcification appeared to replace the scleral architecture as seen in hematoxylin and eosin stained sections.

Case 2: This 67-year-old female was seen in this Eye Clinic on November 6, 1967, complaining of difficulty with near vision. Her left eye had had poor vision since childhood. There was no his-

Fig. 1. (Case 1). Low power view of front part of the eye. Two symmetrical patches of calcification (arrows) are seen in the sclera directly anterior to the insertion of the horizontal eye muscles. (Paraffin section, H & E stain, X 50.)

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tory of previous eye disease or injury and the family eye history was negative.

Examination revealed best corrected vision of OD: 20/50 and OS: counts fingers at 3 feet. There was a small angle left esotropia. Applanation tension was OD: 13 and OS: 14. Funduscopic examination of the right eye revealed many macular drusen and absence of the foveal reflex. The left eye revealed an elevated, yellowish, solid lesion with distinct borders just above and temporal to the macula. The lesion was approximately three disc diameters in size.

A diagnosis of senile macular degeneration OD was made. Malignant choroidal melanoma and strabismic amblyopia were diagnosed in the left eye.

General physical examination was essentially unremarkable. Laboratory studies included a serum calcium of 9.7 mg.% and serum phosphorus of 3.8 mg.% The hemoglobin was 15.0 Gm%, the hematocrit 44.0 volumes% and the WBC 7,500. The urinalysis was normal with a PH of 5.0. X-rays of the orbits, spine and long bones were normal and showed no unusual soft tissue calcification.

Enucleation of the left eye was performed on November 8, 1967. Gross examination revealed a normal sized eyeball. Two small bluish spots were noted in the horizontal on the posterior aspect of the sclera. A choroidal tumor of light brown color measuring 8 x 3 x 9 mm. was found superior and temporal to the foveal region when the eye was opened.

Histologic study showed a normal anterior segment except for diffuse infiltration with lymphocytes and plasma cells in the iris and ciliary body. There were degenerative changes of the vitreous and cystic changes of the peripheral retina as well as sclerosis of its blood vessels. The retina was also degenerated in the region of a protruding choroidal tumor (Fig. 2). This tumor was composed of spindle-shaped cells with very little pigmentation. Most of its cellular nuclei were small and without a nucleolus. However, some cells with nucleolated nuclei were found. No mitoses were seen. The tumor had not broken through Bruch’s membrane nor had it invaded the sclera. A patch of calcification was observed in the sclera next to the center of the choroidal tumor (Fig. 2). Two more smaller patches were seen in the sclera near the posterior pole of the tumor (Fig. 2).

Serial sections showed the central patch of calcification to be continuous with an area of calcification in the horizontal and next to the insertion of the inferior oblique muscle (Fig. 3). However, sections through the horizontal also showed another very similar and symmetrical cal-

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Fig. 2. (Case 2). Choroidal melanoma in the eye. Cystic degeneration of the retina on the tumor and patches of scleral calcification (arrows) next to the tumor are seen. The insertion of the inferior oblique is seen on the outside of the sclera. (Paraffin section, H & E stain, X 150.)

Fig. 3. (Case 2). Horizontal section of the eye seen at low power showing the two symmetrical patches of scleral calcification (arrows). The melanoma (Fig. 2) was found adjacent to extensions of the calcified patch seen on the right (temporal) side. (Paraffin section, H & E stain, X 50.)
cified area nasal to the disk (Fig. 3). All calcified patches in this case involved the inner portions of the sclera, but they spared the outermost layers (Figs. 2 and 3). The sclera was otherwise normal and the optic nerve was also normal.

The histopathologic diagnosis was: malignant melanoma of the choroid of a spindle A cell type without extraocular extensions. The tumor was found next to one of two symmetrical patches of calcification in the posterior sclera.

Discussion

Symmetrical patches of calcification in the anterior sclera are relatively common (Case 1), while such symmetrical patches in the posterior sclera (Case 2) are a rarity. The association of the rare posterior type of this symmetrical degenerative change with strabismic amblyopia adds to the interest in the second case. However, the fact that a malignant choroidal melanoma was found adjacent to one of the posterior calcified patches makes this a unique observation.

The tumor in Case 2 is of the slow-growing spindle A cell variety. One could speculate that the calcification in the sclera underlying the choroidal tumor could have occurred secondary to ischemia and necrosis caused by the slow-growing tumor. Such a view, however, would not explain the presence of the other very similar patch of calcium that was found in the horizontal of the nasal aspect of the posterior sclera. This nasal patch of calcification had no relationship to the choroidal tumor. One could also speculate that a longstanding patch of calcification in the sclera next to the choroid could have caused the choroidal melanoma by chronic low-grade irritation. This is a most fascinating line of thought, indeed. It leads to the fact that malignant tumors in other tissues are well known to occur following chronic irritation—either from premalignant tumor or from normal tissue components. It also leads to the fact that the cause and origin of malignant uveal melanomas are obscure in most cases (comp. Wolter and Bryson)4 and that all interesting associations should, therefore, be recorded.

It remains that the present case (Case 2) exhibits a most unusual and thought-provoking association of ocular changes. The question whether these associations are significant—or just coincidental cannot as yet be answered.

Summary

A malignant choroidal melanoma of the spindle A cell type was found next to the temporal patch of symmetrical calcification of the posterior sclera in an eye with strabismic amblyopia. A more typical case of anterior symmetrical scleral calcification in an eye with absolute glaucoma is presented for comparison.

References


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