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Choroidal Osteoma

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Choroidal osteomas are benign, juxtapapillary, choroidal tumors that mandate no treatment. They occur predominantly in young (median age, 20 years) females (90%). While ophthalmoscopy and fluorescein angiography reveal suggestive ophthalmic findings, echography, plain radiography, and especially computed tomography (CT) are important in establishing the correct diagnosis. These lesions may be clinically confused with various dystrophic lesions, atypical or amelanotic malignant choroidal melanoma, metastatic carcinoma, leukemic or lymphomatous infiltrates, choridal hemangioma, choroidal scars of the macula, and organized subretinal hemorrhage. The exclusion of the differential consideration of malignant melanoma is most important since radiation and enucleation are therapeutic possibilities. Review of the literature shows that all nine reported cases with CT scans had clearly demonstrable calcific densities in the posterior pole of the affected eye. Four other cases are reported here with a summary of the typical clinical, radiologic, and pathologic findings. The role of CT in the evaluation of choroidal osteoma and the distinctiveness of the CT findings are stressed.

Choroidal osteoma was first reported by Gass et al. [1] in 1978 as a benign choroidal tumor that produced visual symptoms in three young women and one girl. The opthalmic literature now contains at least 26 case reports delineating characteristic clinical and pathologic features of this entity. We present four more cases with particular attention to the radiologic findings of this lesion, which are both distinctive and helpful in establishing the correct diagnosis.

Case Reports

Case 1

A 14-year-old girl had marked reduction of visual acuity in her left eye and an established scar from "chorioretinitis." The right eye was normal. At age 19 she underwent an extensive but unrevealing uveitis survey. At age 28, her corrected vision was 20/20 in her normal right eye (OD) and 20/400 in the left eye (OS). The ophthalmic examination of the left eye was normal except for "an old scar from a healed chorioretinitis."

At age 35, a routine examination again identified a normal right eye and 30/400 visual acuity OS. Except for left afferent pupillary defect, the anterior segment examination was normal. Ophthalmoscopy revealed a pink white tumefaction extending from about the two o'clock to the ten o'clock meridian of the disk and extending as far as four disk diameters away. Some secondary hyperpigmentation was present at the borders of the lesion, suggesting previous subretinal blood or fluid. Small "crow's-feet" similar to haversian canals were identified within the choroidal tumor. Contact B-scan echography and standardized A-scan echography demonstrated a sonically dense lesion suggesting calcification (fig. 1A). The 5-mm-thick axial computed tomographic (CT) scan without contrast injection showed a subtle, but definite 5-mm-wide arcuate calcification inferolateral to the optic disk on the left. The CT numbers averaged 175 Hounsfield units (H) (fig. 1B).

Case 2

A 15-year-old girl was seen after 4 weeks of "itching" of both eyes treated by her personal physician for "allergy." During an opthalmologic examination, a "tumefaction" was identified in the left posterior pole.

A subsequent consultation revealed corrected visual acuity of 20/15 OD and 20/20 OS, with about 2 diopters of hypermetropia OS. The right eye was otherwise normal. Ophthalmoscopy of the left eye identified a shallow secondary retinal detachment overlying a large, rosy white plaquelike choroidal tumor filling the macular area (fig. 2A). Contact A-scan and B-scan echography showed a highly reflective, sonically dense, sound-absorbing, plaquelike lesion involving the posterior pole, temporal to the optic nerve.

Follow-up examination at 1 year revealed some flattening of the detachment and secondary pigment epithelial changes over the tumor, but visual acuity was preserved at the 20/25 level. The 5-mm-thick axial CT scan without contrast enhancement showed a 2–3 mm calcified lesion inferolateral to the optic disk with CT numbers averaging 258 H (fig. 2B).

Case 3

A 33-year-old woman had documented excellent visual acuity in each eye as a young adult. However, routine physical examination at that age revealed an unusual white "scar" in the left posterior pole. Several ophthalmic evaluations were obtained with no specific diagnosis.

At age 35, her visual acuity was 20/15 OD and 20/20 OS. The right fundus was normal. Ophthalmoscopy of the left fundus revealed a broad zone of choroidal hyperpigmentation involving the posterior one-third of the globe. Within this choroidal nevus and along the inferior and temporal pole of the optic disk was a creamy white choroidal mass with small choroidal vascular tufts within it. Contact echography showed a highly reflective, dense mass corresponding to the region just below the disk, with profound sonic shadowing behind it. Follow-up examination at 1 year revealed no

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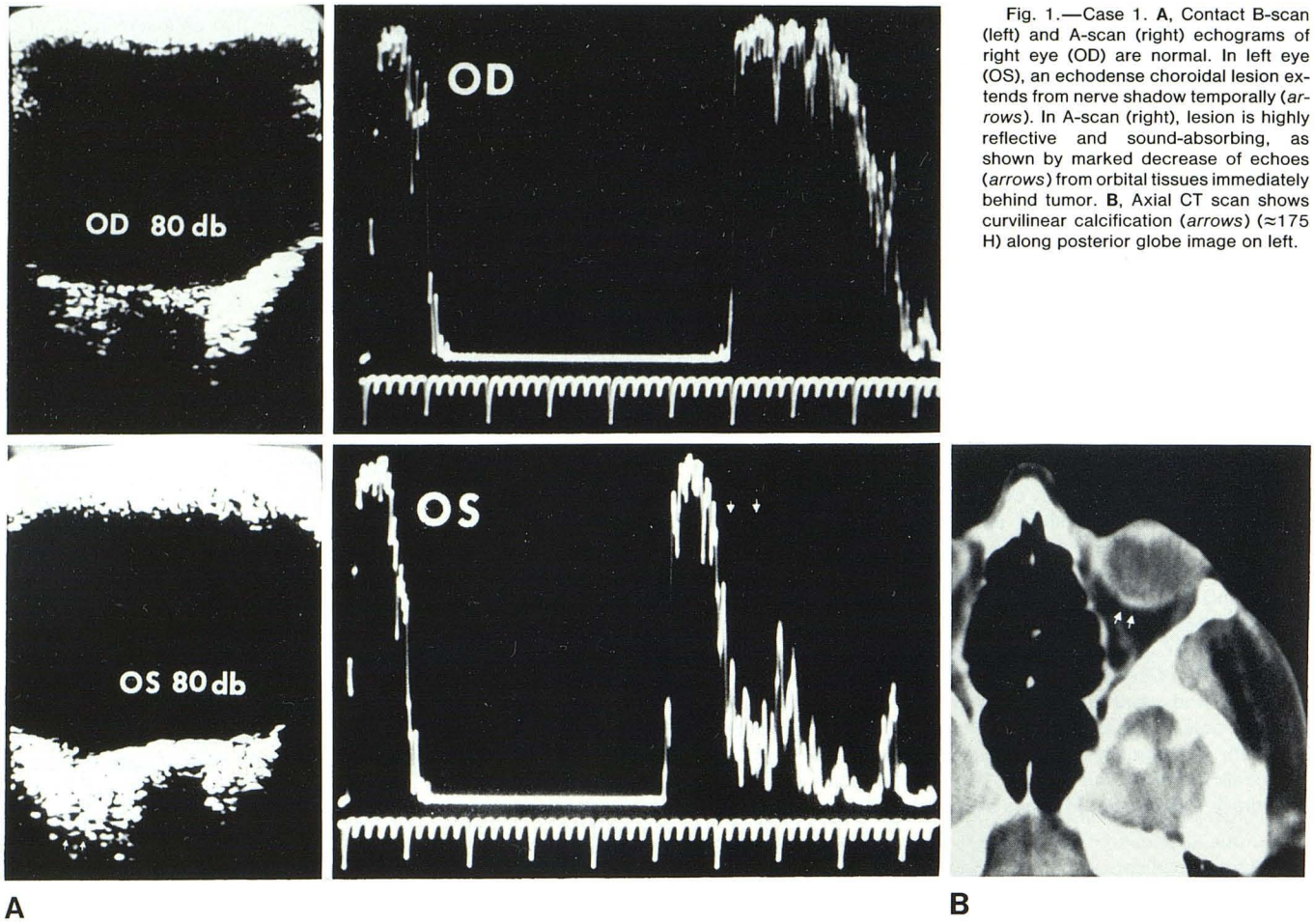

Fig. 1.—Case 1. **A**, Contact B-scan (left) and A-scan (right) echograms of right eye (OD) are normal. In left eye (OS), an echodense choroidal lesion extends from nerve shadow temporally (*arrows*). In A-scan (right), lesion is highly reflective and sound-absorbing, as shown by marked decrease of echoes (*arrows*) from orbital tissues immediately behind tumor. **B**, Axial CT scan shows curvilinear calcification (*arrows*) (≈175 H) along posterior globe image on left.

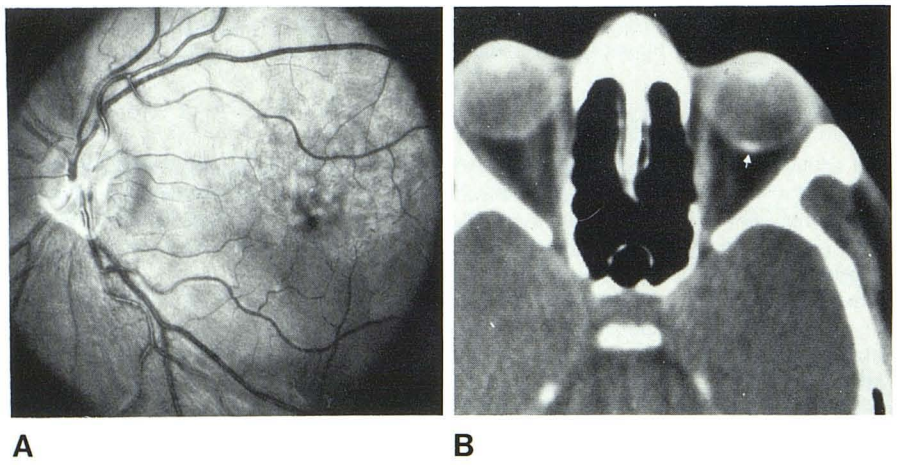

Fig. 2.—Case 2. **A**, Monochromatic reproduction of fundus photograph. A slightly elevated, fleshy white tumefaction is next to disk and underlying entire macular area. Mild disruption and clumping of pigment on top of tumor. Note its sharp demarcation. **B**, Axial CT scan shows 6-mm-wide calcification (*arrow*) (≈258 H) just temporal to left optic disk.

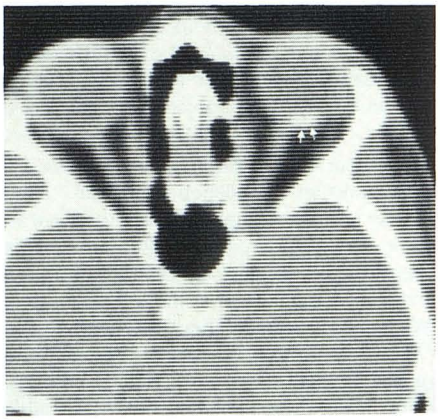

Fig. 3.—Case 3. Axial CT scan shows 5-mm-wide calcification (*arrows*) (≈284 H) just inferior and temporal to left optic disk.

change in either visual acuity or the appearance of the lesion. The 5-mm-thick axial scan without contrast enhancement again showed a 3–4-mm-wide calcification below the optic disk with CT numbers averaging 284 H (fig. 3).

Case 4

A 5-year-old girl was seen with an acquired esotropia, blurred vision, and the sensation of flashing lights. Her visual acuity was 20/400 OD and 20/20 OS. The left eye was otherwise normal. Ophthalmoscopy revealed a minimally elevated choroidal yellow white lesion extending around the superior temporal border of the optic disk and the macular area and downward to the inferior temporal vascular arcade. There was a shallow serous retinal detachment with extensive intraretinal cystoid macular edema.

Standardized contact echography revealed a choroidal mass lesion that was sonically dense but not specifically compatible with calcification. The follow-up examination a few years later showed an increase in the sonic density and shadowing behind the lesion, evidence of progressive calcification. CT scans at age 5 and 7 years did not show definite calcification. A presumptive diagnosis of choroidal osteoma was made and it was anticipated that calcification would appear on future scans.

Discussion

Choroidal osteomas are benign juxtapapillary choroidal ossified tumors. Typically, affected patients are young and otherwise healthy women [2]. Of the 30 cases reported to date (including ours), only three (10%) have been males and only two were reported to be black [3]. In about 75% of reported patients, the tumor is unilateral; it is not clear, however, if this represents a skewed reporting of bilateral cases [4, 5]. Most patients present with blurred vision, metamorphopsia, or central scotomata. The majority of patients have visual field defects smaller than would be expected from the size of the tumor. At the initial examination, about four of five patients will have visual acuity better than or equal to 20/30. However, only one-half of affected eyes will maintain this visual acuity. Initially, fewer than 10% of patients will have visual acuity of 20/200 or worse, but about 25% will ultimately have vision in this range, because of superimposed diskiform macular degeneration.

Ophthalmoscopic examination reveals a round or oval, irregularly elevated, rosy to yellow white tumor in the choroid either near the optic disk or in the macular area. There may also be irregular orange or gray pigment clumps on the tumor surface. The borders are well demarcated with an irregular scalloped or geographic outline; pseudopodlike projections have also been described. Serous retinal detachments, presumably secondary either to decompensation of the retinal pigment epithelium or to subretinal neovascularization, have been described in about one-third of affected eyes and may be the basis for eventual decrease in visual acuity. The presence of multiple short branching vascular trunks on the surface of the tumor is another typical feature that distinguishes osteomas from choroidal hemangiomas and metastatic tumors. Tumor size ranges from 1.5 × 2 to 9 × 15 disk diameters. After a follow-up period of 0.5–9 years, tumor growth is reported in about one-half of affected eyes [2].

Fluorescein angiography reveals early mottled hyperfluorescence, particularly in the more hypopigmented areas. This appears to result from an accentuation of the normal choroidal lobular perfusion due to the vascular supply of the cancellous bone. The small vascular tufts may also be evident. In the late phases, there is diffuse staining where marrow spaces occur within the tumor.

Histopathologic examination [1, 6] revealed normal mature cancellous bone with dense trabeculae separating numerous interconnecting marrow spaces. Large numbers of osteocytes and occasional osteoclasts are associated with the trabeculae. The marrow spaces are filled with loose connective tissue, cavernous blood-filled spaces, and thin-walled capillary-type vessels. Hematopoietic activity has not been observed. There are focal areas where the pigment epithelium is atrophic and depigmented while in other areas the pigment epithelium appears intact.

Echography characteristically demonstrates a highly reflective, acoustically dense, irregularly elevated mass with marked acoustic shadowing behind the globe. When the system sensitivity is reduced to suppress other scleral echoes, the choroidal mass is still readily apparent.

Of the cases reported to date, conventional radiographs, either routine orbital views or polytomograms, were obtained on 18 patients. Demonstration of increased radiodensity corresponding to the choroidal osteoma was achieved in 18 (85.7%) of the 21 affected eyes. However, Gass [2] reported that the initial reading of plain films did not mention the radiodensity in 10 of 15 affected eyes, either because it was missed or because it was not evident on plain films. In seven eyes, polytomography was needed to unequivocally demonstrate the calcification. Plain films were obtained in our case 4 only, and calcium was not evident.

CT, on the other hand, has been highly effective in documenting the presence of the tumor. In the prior CT reports of nine affected eyes, the calcific tumor was easily seen. CT defines a lesion of bone density in the posterior pole of the affected eye, commonly on the temporal side of the topic nerve. Three of our four cases had obvious calcification while the one that did not remains a presumptive diagnosis based on both ophthalmoscopy and echography suggesting subtle, progressive calicification. The radiographic differential diagnosis of intraocular calcification is limited. It includes retinoblastoma, phthisis bulbi, calcified drusen of the optic disk, dystrophic calcification after trauma, hemorrhage or infection, and, theoretically, metastatic calcification in hyperparathyroidism [7, 8]. Each of these lesions can be differentiated clinically from choroidal osteoma.

The clinical differential diagnosis of choroidal osteoma includes amelanotic or atypical malignant melanoma, metastatic carcinoma, leukemic or lymphomatous infiltrates, choroidal hemangioma, a variety of macular choroidal scars, and resolving subretinal hemorrhage. It is claimed that ^{32}P testing usually permits differentiation between benign and malignant lesions. However, since bone rapidly accumulates radioactive phosphorus, a false-positive ^{32}P test is obtained in choroidal osteoma and has been the basis for enucleation on at least one occasion [2]. Ophthalmic echography and CT should characterize the lesions adequately since no other clinical possibility is associated with calcification. Thus the combination of characteristic features by ophthalmoscopy, echography, and CT should allow the proper diagnosis of the patients with choroidal osteoma.

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