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CT of Drusen Bodies and Other Calcific Lesions of the Optic Nerve: Case Report and Differential Diagnosis

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One of the major causes of misdiagnosis of papilledema is buried drusen of the optic nerve head [1, 2]. Optic nerve drusen are calcified concretions of unknown etiology, located within the optic nerve head just anterior to the lamina cribrosa. As these concretions enlarge they may become visible ophthalmoscopically. When buried in the nerve head, they may cause difficulties in the ophthalmoscopic differentiation from true papilledema. With the advent of high-resolution computed tomographic (CT) scanning, appropriate axial and coronal cuts can demonstrate the calcified optic nerve head drusen and save the patient from further neuroradiologic procedures [2].

Case Report

A 22-year-old woman in previously good health was first evaluated in 1970 for complaint of headache. Physical examination at that time revealed bilateral elevated discs, interpreted as papilledema. Fluorescein angiography, skull radiographs, brain scan, bilateral carotid angiograms, and ventriculogram were all normal. The possibility of optic disk drusen was then raised. The patient was followed for the next 10 years with this diagnosis. Because of continuing headaches and decreasing vision in the left eye, the patient was referred for neuroophthalmologic consultation in September 1981. Optic nerve head drusen were evident ophthalmoscopically (figs. 1A and 1B). Visual field testing showed typical although marked visual field defects (fig. 1C). To be certain that there was not a superimposed unrelated basis for visual loss, CT in both coronal and axial projections was then done, using the Pfizer 0450/AS&E scanner with high-resolution pack in 3 mm slice thickness. Coronal and axial scans revealed high density, well defined punctate lesions in both optic nerve heads. The location of the drusen bodies was compatible with the patient's field defects (figs. 1D-1F).

Discussion

Drusen of the optic nerve head are histologically basophilic-staining, laminated, acellular bodies of various sizes and shapes. They range in size from 20 to 1,200 μm with an average size of 320 μm . They are found in 0.3%–2% of autopsied eyes, bilaterally in 9%–15%, and are composed of various components including a mucoprotein matrix, a carbohydrate moiety, and cyclic amino acids. Calcium is almost always present [3]. Of all diagnostic imaging methods, CT is the most sensitive for detection of small amounts of calcium. Compared with conventional radiography, CT has been demonstrated to show calcium completely unsuspected on plain films. The Pfizer/AS&E scanner is capable of resolving high-density lesions of 600 μm in diameter and detecting a single calcific density of about 300 μm in diameter. It is, therefore, reasonable to assume that the modern high-resolution CT scanners will be able to detect early calcified drusen of the nerve head.

Many etiologies have been suggested over the years, culminating in the suggestion by Spencer [4] that drusen represent a familial type of optic neuropathy in which the normal physiologic damming of orthograde axoplasmic flow anterior to the lamina cribrosa is aggravated by local factors. A model recently developed by Tso [5] proposed abnormal axonal metabolism that leads to the deposition of calcium crystals in mitochondria of intact axons. These axons are disrupted and calcium is further deposited around the extruded mitochondria. Small bodies that are created initially in that way may then coalesce to form the larger bodies seen ophthalmoscopically as drusen, often in association with the development of optic atrophy and visual field loss, usually peripheral and not severe [2, 5]. Peripapillary sub-

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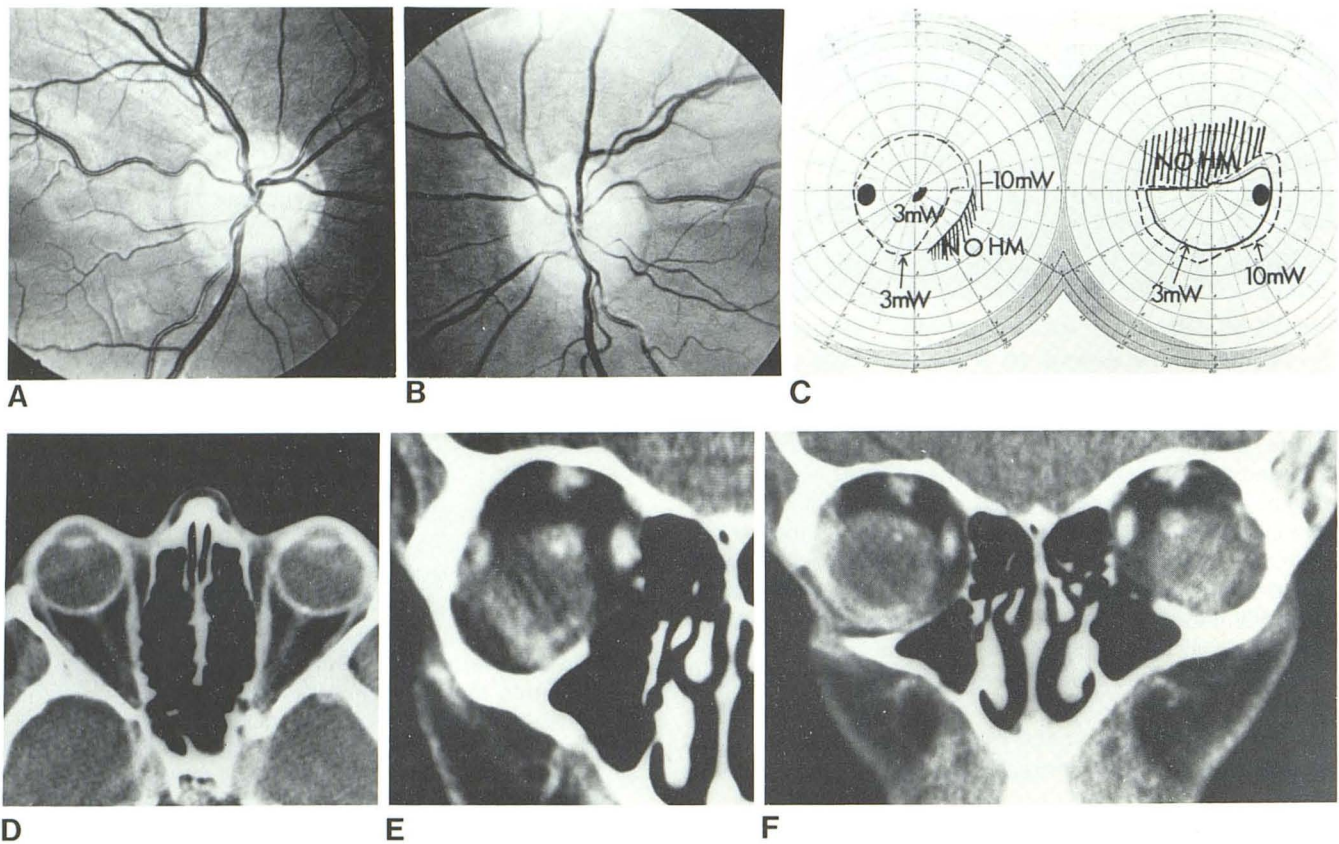


Fig. 1.—Right (A) and left (B) eyes. Prominent optic nerve heads with drusen bodies at disk margins. Evident loss of nerve fiber layer (residual nerve fibers best seen superotemporally, right eye). C, Visual fields showing dense upper altitudinal defect in right eye (right) and general depression with inferior nasal step and paracentral scotoma in left eye (left). D, Axial view.

Well defined high-density punctate lesions in both optic nerve heads. Coronal views of right (E) and left (F) eyes. Drusen bodies located inferiorly in right optic nerve head corresponding to dense upper altitudinal visual field defect in C.

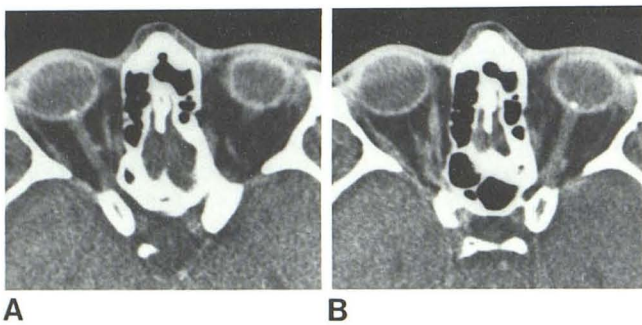


Fig. 2.—Axial CT demonstrates typical appearance of bilateral drusen bodies in optic nerve heads (confirmed ophthalmoscopically). A, Right eye. B, Left eye.

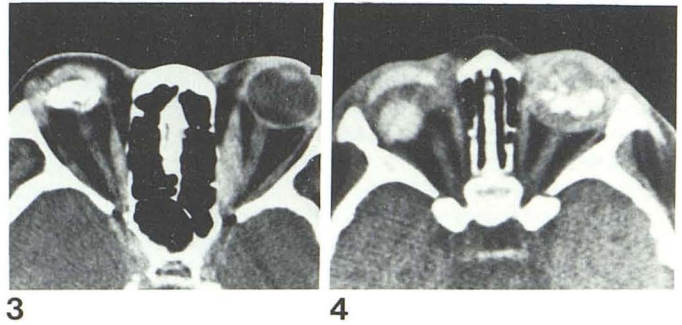


Fig. 3.—Axial CT of patient with Graves ophthalmopathy (enlarged left medial rectus muscle) demonstrates posterior globe calcification in small phthisical right eye.

Fig. 4.—Axial CT shows partly calcified retinoblastoma. Calcifications located anterior to optic disk margin. Prosthesis and shell in other orbit following enucleation for retinoblastoma.

retinal and superficial hemorrhages may further complicate the clinical diagnosis [6]. Fluorescein angiography may be helpful in the distinction from papilledema by showing early autofluorescence and late staining of the drusen. Sonography may show strong reflection at a low decibel level.

Examination of other family members can be helpful since there may be a familial occurrence.

CT is also of value in the diagnosis of buried drusen bodies. The characteristic CT appearance is one of well defined punctate lesions of high density in one or both

scleral canals. Figure 2 clearly demonstrates 1×1 mm bilateral calcified drusen in another patient. Buried drusen not ophthalmoscopically evident have been shown on CT [2]. It remains uncertain at what age the disk elevation develops and at what stage the presence of the calcification can be detected by CT. Since papilledema can be superimposed on buried drusen, the absence of retrobulbar optic nerve sheath distention seen with elevated intracranial pressure is important for further corroboration [7].

Differential Diagnosis

Other lesions that can cause calcification in the posterior part of the globe include phthisis bulbi, neoplasm (optic nerve glioma, meningioma, and retinoblastoma), hamartoma (tuberous sclerosis), and systemic disease with hypercal-

cemia [8]. Phthisis bulbi (fig. 3) is an end stage of diffuse ocular disease including trauma. There is marked atrophy of the globe with shrinkage and disorganization of the intraocular structures. Calcium may be deposited within a cataractous lens, sclera, uvea, and gliotic retina. Retinoblastoma is the most common intraocular neoplasm in children. Necrosis is usually present and calcification is a frequent and important diagnostic feature [8]. Figure 4 shows an example of partially calcified retinoblastoma in the region of the posterior globe. It can be differentiated from optic disk drusen by the floccular calcifications projecting into the vitreous body not confined to the optic nerve. Primary intraorbital meningiomas are occasionally calcified. Figure 5 demonstrates calcification surrounding the retrobulbar part of the optic nerve in a patient with biopsy-proven optic nerve meningioma. This entity can easily be differentiated from drusen bodies, which are confined to the optic nerve head. Giant drusen of the optic nerve (anterior to the lamina cribrosa) are astrocytic hamartomas usually associated with tuberous sclerosis. Histologically, most of them contain calcium. Figure 6 demonstrates such an astrocytic hamartoma. The CT appearance of optic nerve drusen and the glial hamartoma of tuberous sclerosis may be similar [10]. The ophthalmoscopic examination, in addition to other symptoms and signs of tuberous sclerosis, may help in the distinction between the two entities. Another lesion that causes calcification in the posterior portion of the globe is choroidal osteoma (figs. 7A and 7B). This newly recognized entity may be mistaken ophthalmoscopically for choroidal hemangioma, leukemia, metastatic carcinoma, or amelanotic melanoma [11]. CT is diagnostic by showing calcification occupying the posterior choroid and not confined to the optic nerve head (fig. 7C). Optic nerve gliomas usually present on CT as enlarged optic nerve; they can occasionally show calcification (e.g., in the posterior globe) [8].

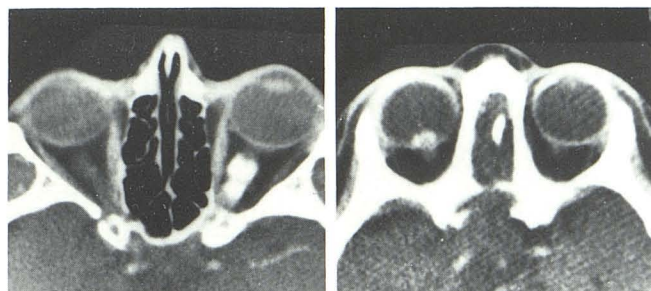


Fig. 5.—Partly calcified optic nerve sheath meningioma posterior to lamina cribrosa.
Fig. 6.—Inhomogeneous calcification in optic nerve head and adjacent retina in patient with tuberous sclerosis.

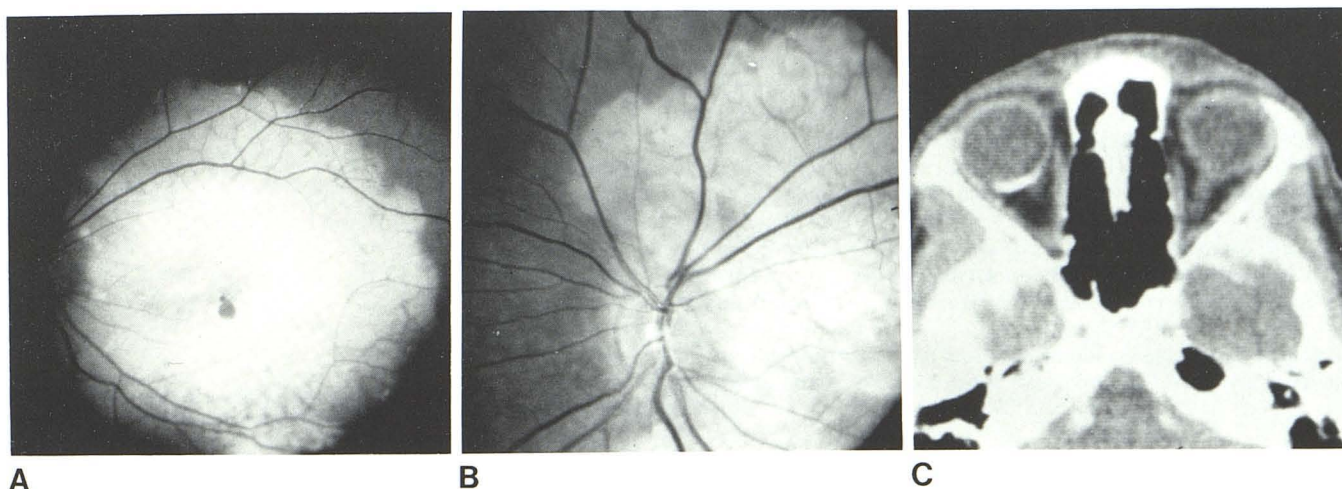


Fig. 7.—A, Wide-angle photograph of left fundus shows whitish, well circumscribed choroidal osteoma. Lesion is denser in center (around fovea). B, Higher magnification of same fundus. Left optic disk partly surrounded by

choroidal osteoma. C, Axial CT. Densely calcified choroidal osteoma seen ophthalmoscopically. Calcified lesion not confined to optic nerve head.

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