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MR Imaging in Pseudoxanthoma Elasticum

Grönblad-Strandberg syndrome known as pseudoxanthoma elasticum (PXE) is a rare connective tissue disorder, the inheritance of which shows both autosomal and recessive characteristics [1]. The disease most commonly involves the skin, eyes, cardiovascular system, and gastrointestinal tract [2]. The vascular connective tissue supporting the retina, known as the Bruch membrane, tends to calcify [3]. Arterial calcifications may result in a variety of clinical manifestations including hypertension; angina; and neurologic deficits caused by ischemia, infarction, or hemorrhage [4, 5].

Case Report

A 43-year-old man was referred to us in September 1987 for consultation and evaluation of a variety of neurologic disturbances. He had a 3-year history of progressive visual loss, right-sided pulsatile tinnitus, and paroxysmal dizzy spells on rapid head movements. He had experienced a right retinal hemorrhage in 1984 and a left retinal hemorrhage in 1985. The presence of cutaneous papular lesions had led eventually to histopathologic confirmation of PXE by axillary biopsy. Our initial neurologic evaluation was unremarkable with the exception of poor visual acuity. Otologic examination revealed spontaneous and posturally induced nystagmus and right-sided recruitment on caloric testing, consistent with a CNS lesion. MR imaging (1.5-T) of the head was performed (Fig. 1).

Discussion

CT findings in PXE have been described as ischemic infarctions, generalized atrophy, and subarachnoid hemorrhage [3–6]. The English-language MR literature contains no reports of characteristic features describing brain lesions of PXE. The only report of such a case is in the Japanese literature: Maeda et al. [5] described MR of the brain revealing high-intensity lesions in a patient with PXE. The lesions corresponded to low-attenuation areas on the patient's brain CT, and angiography proved that the lesions were caused by infarctions.

The long list of differential diagnosis of increased signal on T2-weighted images include ischemia, infarction, infection, primary demyelinating disease, dementia, neoplasm, connective tissue disease, radiation injury, gliosis, and normal aging [7–9]. In our case, MR shows multiple small areas of increased signal intensity on T2-weighted images in the basal ganglia and deep white matter. In light

of our patient's age and clinical history, these findings probably are not caused by either the aging process or other etiologies such as infection, demyelinating disease, neoplasm, or exposure to radiation. We suggest that these findings are consistent with lacunar infarctions. The ischemic infarctions are probably due to occlusion of small penetrating arteries injured by connective tissue changes caused by PXE. Correlation with histopathology and future MR studies of other PXE patients are required to confirm this hypothesis.

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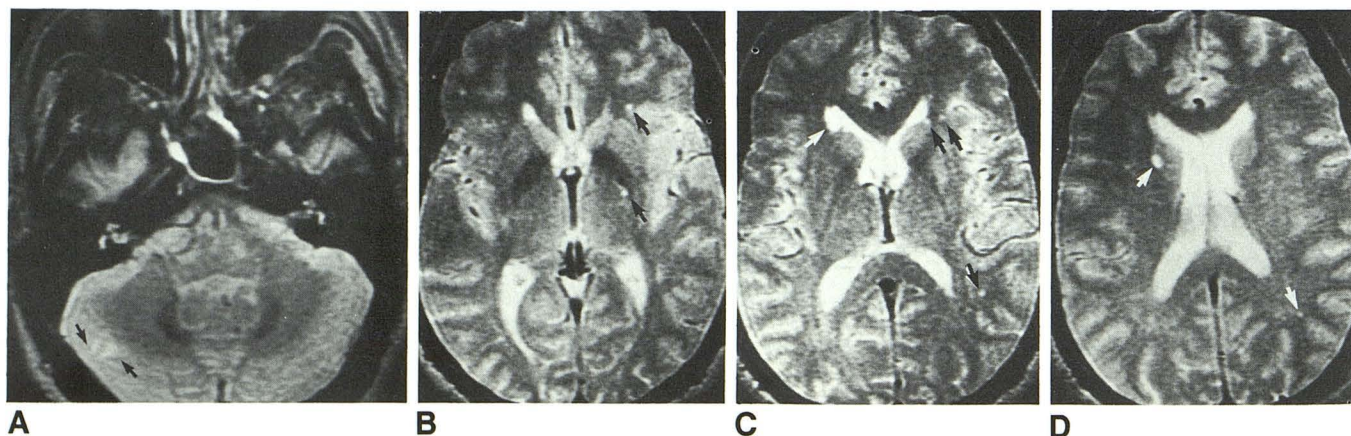


Fig. 1.—Abnormal high-intensity lesions in pseudoxanthoma elasticum.

A–D, Spin-echo axial MR images (2000/70) show multiple high intensities (arrows) in both basal ganglia, deep white matter, and right cerebellum.