The Value of Differential Diagnosis of Fahr's Disease by Radiology

Y Elshimali

Citation

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Abstract

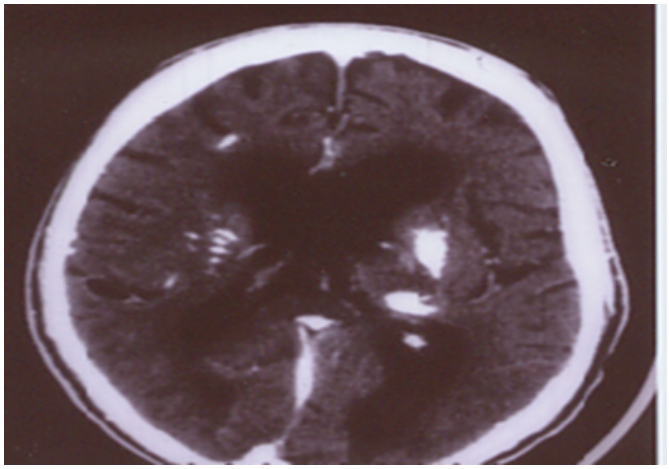
This is a case report of Fahr's disease or Bilateral StriatoPallidoDentate Calcinosis (BSPDC) which is a neurological entity characterized clinically by Parkinsonism, chorea, dystonia, ataxia, mental deterioration, seizures and neuropsychiatric features. And pathologically (1) by massive bilateral calcification of the basal ganglia, sulcal depths of the cerebral cortex, dentate nuclei of the cerebellum, internal capsule and lateral parts of the thalamus. It is very important to differentiate Fahr's disease from Parkinson's disease and other neurological identities upon interpretation of the imaging data.

CASE REPORT

This was a 70-year-old white male, with history of Parkinsonism, tardive dyskinesia, dysphasia and hypertension. Patient presented with cachexia and dehydration. Computed tomogram of the head (Fig. 1) showed bilateral, symmetrical, calcified deposits of the brain.

Figure 1

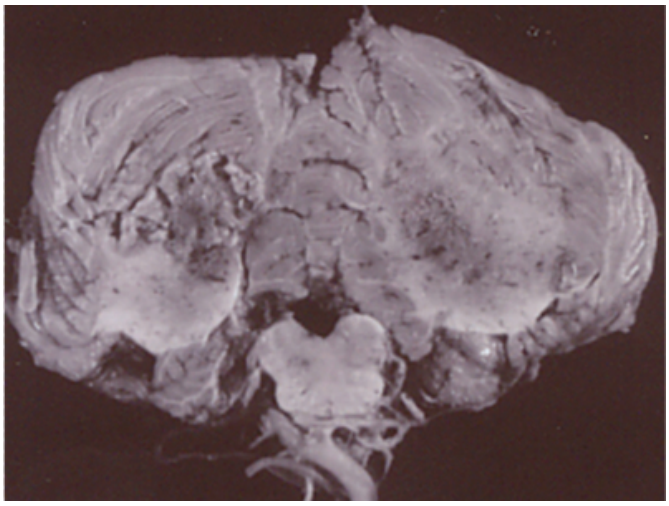
Figure 1: Computed Tomography of Fahr's Disease showing bilateral calcifications of the Basal Ganglia.



The patient's condition deteriorated gradually and he expired with acute bronchopneumonia due to Fahr's disease. Autopsy was performed and showed moderate to severe arterioscleroses of blood vessels at the base of the brain. Multiple coronal sections of the cerebral hemispheres and deep nuclear structures showed symmetrical heavy calcifications of bilateral globus pallidus, and pulvinar areas measuring approximately 1.0 x 2.0 cm on each side. Cerebellar nuclei are densely calcified bilaterally and symmetrically (Fig 2).

Figure 2

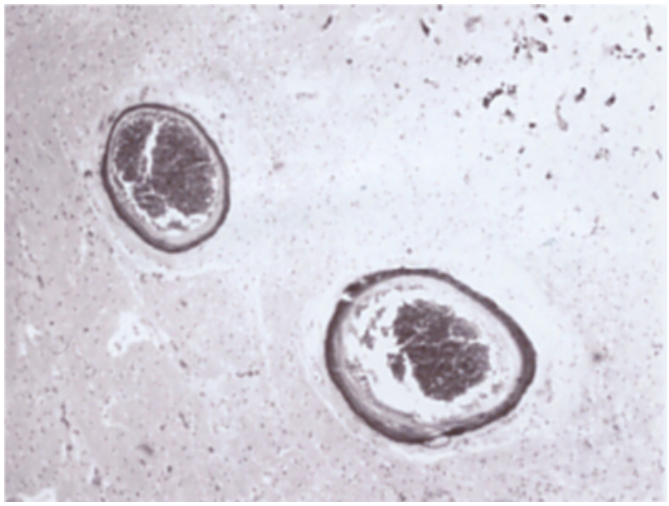
Figure 2: Grosse section of the cerebellum in Fahr's disease showing marked calcifications of dentate nuclei.



Pathological examination of the brain showed bilateral and symmetrical calcium deposits in the walls of the small vessels, capillaries and white matter of the basal ganglia-globus pallidus, dentate nucleus and corpus striatum-(Fig 3), cerebellum, thalamus, hypothalamus, hippocampus, and cerebrum.

Figure 3

Figure 3: Microscopic section from dentate nucleus of cerebellum showing extensive calcifications of small blood vessel wall. X4.



DISCUSSION

The calcium deposits in the brain may occur before the onset of the symptoms, usually in the third decade of life. Although it may also be evident in childhood ([2,5]) and with advancing age the amount of calcification increases. In Fahr's disease the mineral deposits tend to be selective for small capillaries and small vessels of white matter, which is different from that in atherosclerosis ([7]). The calcification may include endothelial and stromal vascular cells as well as the interstitium. However, the local circulatory disturbances such as regional ischemia ([1]) have been regarded as the primary event precipitating the deposition of calcium as well as other minerals. Other contributed factors are abnormality in the ([3]) calcium metabolism or local inflammatory process ([6]). Also, the calcification could be a primary event occurring without preceding circulatory dysfunction, since a significant familial type suggests either autosomal recessive or dominant inheritance ([2]). Brain calcification without symptoms such as the small calcifications in the basal ganglia, and less commonly in the dentate nucleus of the cerebellum can occur in elderly patients.

CONCLUSION

Radiological diagnosis could be the starting point to guide the clinician for possibility of Fahr's disease. The differential diagnosis includes but not limited to; ([4]) Parkinson's disease, Huntington's disease, progressive supranuclear palsy, Wilson's disease, spasmodic torticollis, oligodendroglioma, low-grade astrocytoma ([1]), and arteriovenous malformation. Therefore Fahr's Disease or BSPDC is a diagnosis of exclusion.

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Author Information

Yahya I. Elshimali, M.D., FASCP
Assistant Professor of Pathology, Olive View-UCLA Medical Center