

Fahr disease

V Wadhwa, A Patel

Citation

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Abstract

A 33yr old male came to our dept with history of dementia, gait disorder & focal seizures. On plain ct brain following findings were seen-bilateral symmetrical basal ganglia , internal capsule, thalami, sub cortical white matter and cerebellar grey matter calcifications – suggestive of fahr disease

SYNONYMS

Fahr disease/Cerebrovascular ferrocalcinosis/Idiopathic non arteriosclerotic cerebral calcifications /Bilateral strio pallidodentate calcinosis/ Idiopathic basal ganglia calcification

PRESENTATION

It is a rare degenerative neurological disorder characterized by extensive bilateral basal ganglia calcifications that can lead to progressive dystonia,parkinsonism,and neuropsychiatric manifestations.

Usually asymptomatic in first two decades of life,despite presence of multiple brain calcifications.

Bimodal pattern of clinical onset-

Early adulthood(schizophrenic like psychosis)

- Sixth decade(extrapryramidal syndrome, subcortical dementia)
- Neurological manifestations vary,but movement disorders are most common.
- Parkinsonism most common, usually permanent and progressive.

Childhood transient parkinsonism also reported.

- Paroxysmal dystonic chorioathetosis.
- Seizures

Most common sign/symptoms

- Neuropsychiatric disturbance

- Cognitive impairment(subcortical dementia)
- Extrapryramidal movement disorders.

Age-onset of clinical symptoms is typically 30-60yr.

-an infantile form also described.

Gender-no predominance

Imaging findings-

General features-

Best diagnostic clue-bilateral symmetric basal ganglia ca+2 on CT

Location- -globus pallidum-most commom site of ca+2

Lateral pallidum more affected than medial pallidum.

Additional areas of involvement may include-

Putamen, caudate nuclei, thalami.

Cerebellum(especially dentate nuclei)

Cerebral white matter,internal capsule.

Morphology-variable extent, dense ca+2 often confirms to outline of basal ganglia.

Radiographic findings-Heavy bilateral /symmetrical basal ganglia ca+2 may be detectable on plain skull radiology.

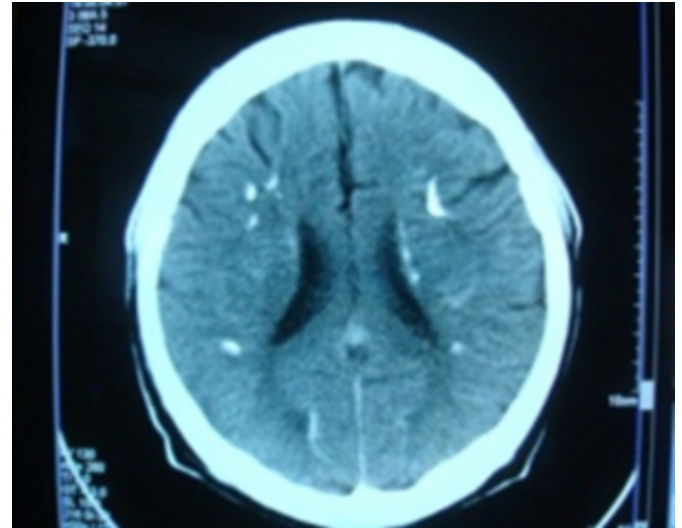
CT FINDINGS

NECT-bilateral, symmetrical calcium in basal ganglia, cerebral white matter, dentate nuclei, cerebellum.

Calcifications seen in-

Figure 1

Figure a: Bilateral Basal Ganglia, internal capsule

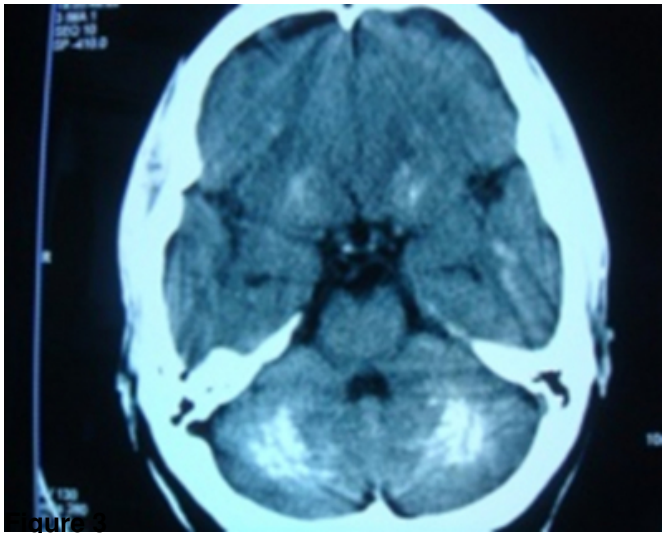


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Figure 2

Figure b: Cerebellar Grey Matter



References

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Figure c: sub cortical white matter

Author Information

Vineet Wadhwa

Dept of radiodiagnosis, R.L.Jalappa hospital and research centre ,Sri deva raj urs medical college

Akshay Patel

Rajiv Gandhi University Of Health Sciences