

Incidental finding of Fahr's disease in an elderly person

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Section: Neuroradiology

Area of Interest: Neuroradiology brain

Procedure: Imaging sequences

Imaging Technique: CT

Special Focus: Calcifications / Calculi Case Type:

Clinical Cases

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Patient: 74 years, male

Clinical History:

A 74-year-old man with known lung carcinoma and prostate carcinoma, on chemotherapy, presented with the recent onset of headache and blurred vision with clinical suspicion of cerebral metastasis. There was no significant biochemical derangement. No known history of any neuropsychiatric illness or neurological deficit.

Imaging Findings:

The unenhanced CT images of the brain demonstrate extensive bilateral symmetrical calcifications in cerebellar hemispheres (dominantly affecting the dentate nucleus), caudate nucleus, lentiform nucleus, thalamus, corona radiata, subcortical frontal cortex and occipital lobes. The unenhanced and contrast-enhanced images did not reveal any focal oedema or enhancing parenchymal lesions to suggest metastasis.

Discussion:

Fahr's disease is a rare, mostly inherited neurodegenerative condition, with usual presentation in fourth or fifth decade of life [1]. The most common initial clinical manifestations are movement disorders with features of parkinsonism, chorea and tremors. The other clinical features include cognitive impairment, progressive neuropsychiatric symptoms, seizures and headache [2, 3]. Fahr's disease is also known as idiopathic basal ganglia calcification and familial cerebral ferrocalcinosis. The most typical features of Fahr's disease include extensive symmetrical calcifications in globus pallidus, putamen, caudate nucleus, dentate nucleus, thalami and subcortical white matter. There is usually no direct link of the symptoms to the severity of calcifications [2-4].

The differentials of Fahr's disease include the other causes of pathological calcifications of basal ganglia; metabolic conditions like hypoparathyroidism, pseudohypoparathyroidism, hyperparathyroidism, infective conditions like HIV infection, neurocysticercosis, toxoplasmosis. The other conditions include senile ageing, toxic conditions like carbon monoxide poisoning and lead poisoning and inherited diseases like Cockayne syndrome and mitochondrial diseases. There is no specific treatment as the disease is usually progressive [4].

The CT imaging remains the modality of choice for diagnosis [1-5], to confirm and assess the severity of calcifications. The susceptibility-weighted imaging (SWI) is the most sensitive technique of MR Imaging to demonstrate calcium and blood products. Inclusion of susceptibility-weighted imaging (3D gradient-echo sequence) at routine MR Imaging is of immense value, considering the fact that MRI may be the first imaging modality for investigating the patients with neuropsychiatric manifestations [5].

Presence of extensive bilateral symmetrical basal ganglia, cerebellar and subcortical calcifications at CT imaging,

absence of metabolic and other infective conditions, are the most diagnostic features of Fahr's disease. To conclude, the patients with Fahr's disease can remain asymptomatic and be diagnosed incidentally at neuroimaging.

Differential Diagnosis List: Fahr's disease, Hypoparathyroidism, Pseudohypoparathyroidism

Final Diagnosis: Fahr's disease

References:

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2. Saleem S, Aslkam HM, Anwar M Anwar S, Saleem M, Saleem A, Rehmani MA. (2013) Fahr's syndrome: literature review of current evidence. Orphanet J Rare Dis 2013;8:156 (PMID: [24098952](#))
3. Alam ST, Aswani Y, Anandpara KM , Hira P (2015) CT findings in Fahr's disease. BMJ Case Rep 2015 Mar20;2015 (PMID: [25795750](#))
4. Hegde AN, Mohan S, Lath N, Lim CC (2011) Differential diagnosis for bilateral abnormalities of the basal ganglia and thalamus. Radiographics 2011;31:5–30. (PMID: [21257930](#))
5. Sahin N, Solak A, Genc B , Kulu U. (2015) Fahr disease: use of susceptibility-weighted imaging for diagnostic dilemma with magnetic resonance imaging. Quant Imaging Med Surg 2015; 5(4): 628–632. (PMID: [26435928](#))

Figure 1

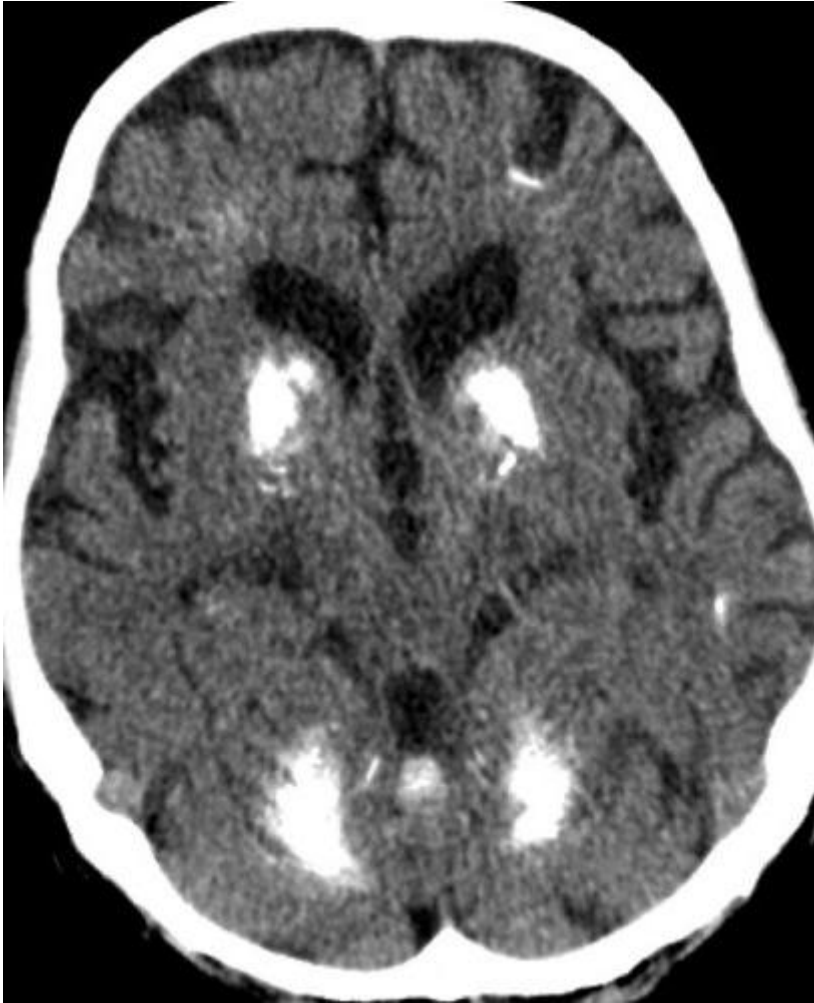
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Description: Bilateral extensive dentate nuclei calcification **Origin:** Lahiri AK, Department of Radiology, Worcestershire Royal Hospital, Worcester, UK.

Figure 2

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Description: Bilateral basal ganglia and cerebellar calcifications **Origin:** Lahiri AK, Department of Radiology, Worcestershire Royal Hospital, Worcester, UK.

Figure 3

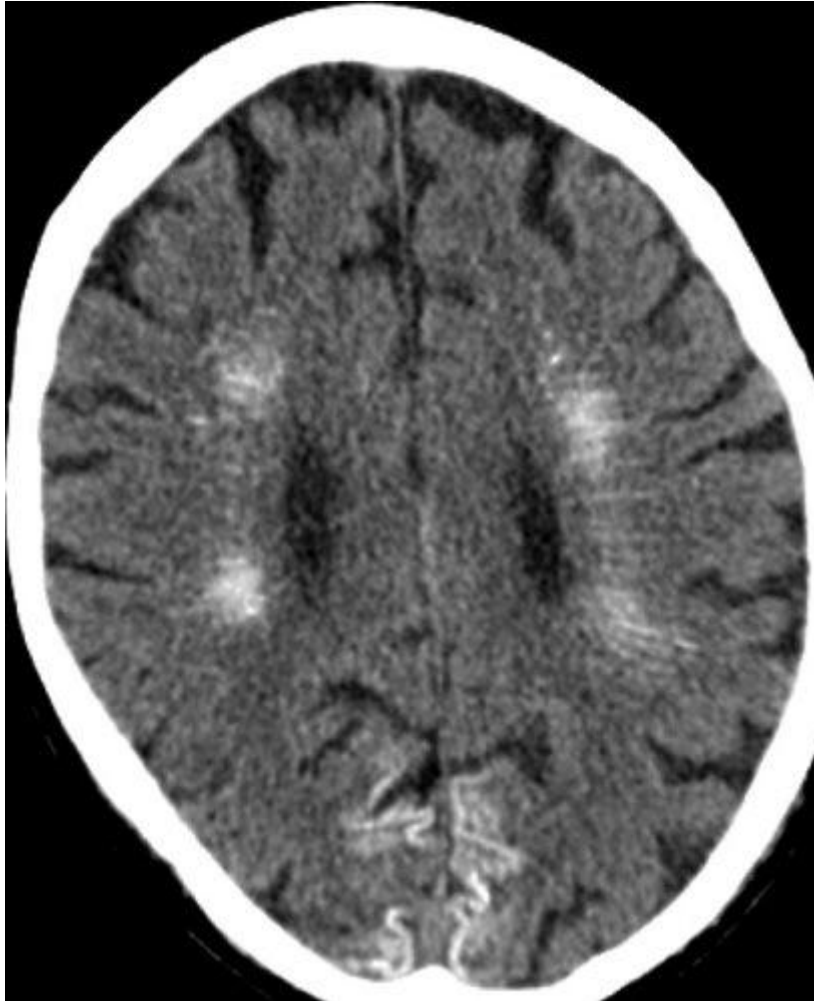
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Description: Bilateral symmetric thalamic calcifications. **Origin:** Lahiri AK, Department of Radiology, Worcestershire Royal Hospital, Worcester, UK.

Figure 4

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Description: Bilateral cerebral calcifications. **Origin:** Lahiri AK, Department of Radiology, Worcestershire Royal Hospital, Worcester, UK.