

Fahr Syndrome: Pure Dementia Presentation

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Abbreviations

MMSE: Mini-Mental State Examination; MoCA: Montreal Cognitive Assessment

Clinical Image

Fahr syndrome is defined as the accumulation of calcium deposits throughout the basal nuclei structures, which has an estimated prevalence of 1/1000,000 [1-2]. Calcinosis of the central nervous system can be an incidental finding in a large percentage of patients and can become unnoticed without being related to a particular disease [3-4]. Here, we present the case of a 74-year-old male patient with a three-year history of insidious cognitive decline, characterized by memory impairment, difficulty in planning complex activities, changes in personality, and unremarkable medical history. Neuropsychological assessment revealed multidomain cognitive impairment with alterations in working memory, episodic memory, semantic memory, and executive functions; MMSE: 21/30, MoCA: 19/30. Brain computed tomography revealed extensive bilateral symmetrical calcifications, located mainly in the basal and periventricular regions. Laboratory workup showed abnormal parathyroid hormone levels: 6.88 pg/mL (normal: 15-65 pg/ml), and the rest of laboratory test was unremarkable; an idiopathic hypoparathyroidism diagnosis was made (Figure 1).

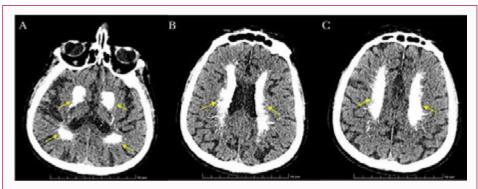


Figure 1: Brain Computed Tomography.

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> Received Date: 31 Jul 2023 Accepted Date: 11 Aug 2023 Published Date: 16 Aug 2023

Citation:

Flores Lazo SL. Fahr Syndrome: Pure Dementia Presentation. Ann Clin Case Rep. 2023; 8: 2455. ISSN: 2474-1655.

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