Fahr’s disease and stroke

Andreia Machado Ribeiro, Madalena Paulino, Ana Pidal, Ana Palricas, Socorro Piñeiro, and Alexandre Amaral e Silva

Background: Fahr’s disease (FD) is a rare neurodegenerative condition characterized by symmetric intracranial calcifications, mainly in basal ganglia and dentate nuclei of the cerebellum. Clinical manifestations may include movement disorders, dementia and behavioural symptoms.

Case report: A 53-year-old female with FD presented to emergency department with acute onset of speech disturbance. Neurological examination showed aphasia, right central facial palsy and right upper limb paresis, National Institutes of Health Stroke Scale (NIHSS)=4. She was admitted at our Stroke Unit and treated with Clopidogrel 75mg and Atorvastatin 20mg. Laboratory tests were normal, except for LDL 111 mg/dL. Electrocardiogram showed sinus rhythm. Cranial computed tomography demonstrated left cortical-subcortical frontotemporal hypodensity, suggesting acute ischemic vascular injury in the peripheral territory of the middle cerebral artery, and exuberant calcifications involving basal ganglia and thalamus (in accordance with diagnosis of FD). Carotid, vertebral and transcranial doppler ultrasound were normal, as well as transthoracic echocardiogram. At discharge she maintained minor right central facial palsy and gait imbalance, NIHSS=2, modified Rankin Scale=3. She was referred to continue her rehabilitation process in a rehabilitation centre. No other etiology was found for stroke in this patient, beyond the possible association to FD.

Conclusion: Some studies have proposed an association between young-onset ischemic stroke and FD. The calcium deposition in the intracranial blood vessels wall leads to a reduction of vessel elastance, thereby predisposing to ischemia. The evidence for the proposed association between FD and stroke is still scarce and further studies are needed.