Fahr’s disease (Cerebrovascular Ferrocalcinosis)

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ABSTRACT
Intracranial calcification may be physiological or pathological. Fahr’s disease or cerebral calcinosis is a rare disorder characterized by the presence of abnormal calcium deposits and associated cell loss in certain areas of brain. The condition is also called idiopathic basal ganglia calcification. Local circulatory ischaemia have been regarded as the event in the calcium deposition. CT is the best modality to evaluate intracranial calcifications.

KEY WORDS:
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CASE
73 years old man presented with the history of altered consciousness and slurred speech for 2 days. He was having changing moods for few years. No other significant complaints as described by his family member. Patient was normotensive. No history of seizure disorders or other significant history suggesting intracranial disorder in the past. Muscle tone and power was symmetrical. Patient was referred to radiology department from emergency for computed tomography of head with the provisional diagnosis of cerebro-vascular accident. Axial computed tomography of head without of intravenous contrast media was performed.

CT FINDINGS
Bilateral symmetrical extensive calcification of basal ganglia, thalami and cerebellum/dentate nuclei.

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Fahr’s disease is a rare neurological disorder of unknown etiology characterized by extensive symmetrical calcification of the basal ganglia and dentate nuclei in the cerebellum. Fahr’s disease is most widely used term for this illness but it has various nomenclature like; cerebral calcinosis, idiopathic basal ganglia calcification, cerebrovascular ferrocalcinosis, striopallidodentate calcification etc.

Some of the intracranial structures calcify and considered physiological with aging, are pineal gland 60%, habenular comissure 30%, choroids plexus 10%, duramater 7%, petroclenoid / interclenoid ligaments 12%, pituitary gland and carotid arteries. There are various causes of pathological calcifications in brain parenchyma, they may be infective, traumatic, metabolic and congenital conditions. Common causes of pathological calcification of basal ganglia include hypoparathyroidism, pseudohypoparathyroidism, mitochondria cytopathy and most of the cases are idiopathic or some cases are familial.

Fahr’s disease (cerebral calcnosis or idiopathic bilateral basal ganglia calcification) is a rare familial neurological disorder characterized by idiopathic massive calcification in basal ganglia and brain parenchyma. These cases mostly present with progressive movement disorders such as parkinsonian features, chorea, tremor, dystonia, dysarthria, dementia, and paresis or speech impairment associated with progressive calcnosis of the brain parenchyma. And some of the cases may present with seizure, syncope or stroke like events. Intracranial calcifications may sometimes be observed on plain radiographs and frequently revealed by CT in normal older people. CT is the best modality to evaluate intracranial calcifications. Regarding this case the patient’s age, presenting symptoms, location and volume of calcification is so extensive is suggestive to diagnose the Fahr’s disease according to criteria described in various literatures. In cases with nonclassical CT findings, radiological diagnosis could be the starting point to guide the clinician for possibility of Fahr’s disease. The differential diagnosis of this disease include (but not limited to) Parkinson’s disease, Huntingstone’s disease, Wilson’s disease, oligodendrogliaoma and AVM. So exclusion of these conditions is necessary to diagnose the idiopathic bilateral basal ganglia calcification. In Fahr’s disease deposition of calcium occur selectively in small vessels and capillaries of cerebral white matter, which is different from that in atherosclerosis. Local circulatory ischaemia have been regarded as the primary event precipitating the calcium deposition. Brain calcifications without symptoms or nonpathological situation can occur in the basal ganglia and less commonly in the dentate nucleus of the cerebellum calcification in elderly patients.

REFERENCES: