A hidden neurologic disease uncovered in the trauma bay

Dear Editor,

A 60-year-old male was brought in as a trauma alert by ambulance after being found lying on the ground by bystanders. The patient was reported to have been confused with slurred speech, gaze preference, and weakness of his lower extremities at the scene. After arrival, his gaze preference was no longer evident; however, he continued to exhibit confusion as well as significant dysarthria. The patient was not found to have any significant traumatic injury during his primary and secondary survey. Given the patient’s presentation, the most possible causes for his altered mental status included intracranial hemorrhage, stroke, or seizure. A computed tomography (CT) head was ordered which showed diffuse calcium deposits throughout the brain, most notably over the basal ganglia and thalamus [Figure 1].

Fahr’s syndrome is a rare, genetically dominant, inherited neurological disease characterized by abnormal bilateral deposits of calcium in motor centers of the brain, most notably being the basal ganglia.\[1\] The exact incidence of this disorder is unknown and is likely underreported as it is often uncovered incidentally.\[2\] In some cases, these calcifications may be asymptomatic.\[3\] However, when present, clinical symptoms vary widely and may include a decline in motor function, dementia, neuropsychiatric disorders, seizures, dysarthria, headache, vertigo, and eye impairment.\[1,2\] This syndrome can also present with Parkinsonian-like symptoms such as muscle rigidity, pill-rolling tremor, and shuffling gait although these tend to manifest themselves later in the course of disease development.\[1\] The most typical presentation of this
disorder includes a combination of seizures, rigidity, and dementia.[2] Classically, the age of syndrome onset is 40–50 s; however, it has also been reported to have occurred in children.[1]

 Diagnostic criteria for Fahr’s disease have been modified based on previous evidence and consist of bilateral calcification of the basal ganglia, progressive neurological dysfunction, absence of biochemical abnormalities, absence of infectious, traumatic or toxic cause, and review of family history.[3] Imaging modalities for diagnosis are typically brain CT or magnetic resonance imaging (MRI).[2] Hydroxyapatite is a mineral in calcium, seen in bone and teeth. This mineral is responsible for the hyperdensity seen on CT scan. Calcium deposition is not as easily seen on MRI imaging and best visualized on CT studies. Fahr’s disease has many causes and therefore difficult to treat. Classical and recent anatomical studies about the role of the basal ganglia in movement coordination, subcortical region in cognitive association and mood determination, and hypoxia in epileptic activity point to correlation between clinical features and calcification site and possibly related to hypoperfused areas.[4]

The differential diagnosis for intracranial calcification is diverse, including both benign and pathological etiologies. Other possible causes of basal ganglia and cerebellum calcifications include neoplasms, arteriovenous malformations, dystrophic calcifications in chronic infarctions, chronic vasculitis, parasitic infections, neurofibromatosis, tuberous sclerosis, Sturge-Weber syndrome, and metabolic disorders.[2] Other uncommon causes include postanoxic and toxic exposure to carbon monoxide, chemotherapy, radiation therapy, and lead intoxication. Recognition of the intracranial calcification early in a patient’s disease course may be beneficial for finding cause and therefore more expedited treatment.

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