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Josef D. Williams, MD *

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Case Report

Fahr Syndrome in Acute Trauma and Neuro oncology Settings: Two Illustrative Cases and A Narrative Review

Josef D. Williams, MD [1]; Vagif Kazimli, BS [1], Thabasya Veeramani, BS [1]; Mckenzie Daffin, BS [1]; John Page BS [1]; Sandra Gad BS [2]; Carson Caddy BS [1]; Bashar Dawoud, MD [1]; Alejandro N. Santos, MD, PhD [1]; Joacir Graciolli Cordeiro, MD, PhD [1]

[1] Department of Neurosurgery, Miller School of Medicine, University of Miami, Miami, USA

[2] St. George University School of Medicine, True Blue Bay, Grenada

***Corresponding Author:** Josef D. Williams, MD, Department of Neurosurgery, Miller School of Medicine, University of Miami, 1095 NW 14 Terrace, Suite 222, Miami, FL 33136, USA.

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Abstract:

***Background*:** Fahr syndrome denotes acquired, typically bilateral and symmetrical intracranial calcifications with predilection for the basal ganglia, subcortical white matter, and cerebellum. Primary Familial Brain Calcification, formerly called Fahr disease, refers to genetic forms, whereas Fahr syndrome usually describes secondary calcifications in the context of disturbances of calcium phosphate homeostasis, mitochondrial disease, chronic kidney disease, infections, or other systemic conditions. The radiologic appearance can be mistaken for acute hemorrhage or traumatic injury in emergency settings, which may trigger inappropriate escalation of care.

Objective: To describe two illustrative cases that presented in acute care environments, one in a trauma context and one in neuro oncology, and to propose a pragmatic imaging and laboratory approach that helps clinicians distinguish calcification from blood products and identify likely etiologies of secondary calcification.

Methods and overview: We present structured case narratives focusing on imaging sequences, Hounsfield unit sampling, and clinical decision points, integrated with a concise narrative review to offer a stepwise diagnostic strategy suitable for emergency departments and neurosurgical services.

Findings: In both cases, systematic Hounsfield unit sampling with values of 150 HU or higher together with persistence on bone windows supported the interpretation of mineralized tissue rather than acute blood. In the trauma case, this averted an unnecessary escalation toward surgical intervention. In the neuro oncology case, recognition of calcifications coexisting with hemorrhagic metastases allowed appropriate oncologic coordination and symptom control without emergency neurosurgery.

Conclusions: A structured head CT review that combines targeted Hounsfield unit sampling with deliberate use of bone windows, followed by focused laboratory testing of calcium, phosphate, magnesium, parathyroid hormone, and renal function, improves diagnostic precision in patients with suspected Fahr type calcifications and helps avoid unnecessary procedures.

Key words: Fahr syndrome; primary familial brain calcification; brain calcification; basal ganglia; trauma; neuro oncology; computed tomography

Introduction

Fahr syndrome encompasses acquired intracranial calcifications that are most often bilateral and relatively symmetric with a predilection for the deep gray nuclei, subcortical white matter, and the cerebellar dentate region [1]. Primary Familial Brain Calcification is usually a genetic entity with autosomal dominant inheritance and mutations in genes involved in phosphate transport and microvascular integrity. Clinical manifestations

range from parkinsonian features and dystonia to ataxia, cognitive and psychiatric changes, seizures, and autonomic symptoms [2]. Some individuals remain asymptomatic and come to attention only when head imaging is obtained for unrelated reasons. Non-contrast head CT is the most practical modality to detect intracranial calcifications, but accurate interpretation requires attention to pattern, symmetry, attenuation, and window settings.

Distinguishing calcification from hemorrhage matters in the emergency department. Misinterpreting calcifications as acute bleeding can lead to unnecessary hospitalization, reversal of antithrombotic therapy, or unwarranted neurosurgical consultation. Conversely, overlooking acute blood among calcified foci may delay critical interventions. Useful discriminators are the anatomic distribution and symmetry of lesions, typical attenuation values on CT, and verification on bone windows. Adjunctive MRI, especially susceptibility weighted imaging and where available quantitative susceptibility mapping, helps differentiate diamagnetic mineralization from paramagnetic blood breakdown products.

Secondary causes should be considered early because several are actionable. Disturbances of calcium phosphate metabolism such as hypoparathyroidism and advanced chronic kidney disease are classic [3]. Additional associations include mitochondrial disorders and inflammatory or infectious conditions. An efficient initial laboratory panel includes calcium, phosphate, magnesium, parathyroid hormone, and renal function with estimated glomerular filtration rate. Vitamin D status may be informative in selected cases. Even when the underlying driver cannot be fully corrected, clarifying the cause informs prognosis and longitudinal management [2,3].

Case Presentation

Case 1 trauma setting

A 60-year-old woman was brought to the trauma bay after a motor vehicle collision. She was hemodynamically stable and reported a mild headache without nausea or vomiting. Neurologic examination showed no focal deficits. Non-contrast head CT obtained as part of trauma evaluation demonstrated multiple dense foci within the deep gray nuclei that were bilateral and nearly mirror symmetric, most conspicuous within the globus pallidus and caudate head. An extra axial hyperdense crescent along the left convexity raised concern for a small acute subdural hematoma, creating tension between a potential acute extra axial bleed and deep intraparenchymal hyperdensities. **Figure 1 displays CT findings from the initial non-contrast head CT.**

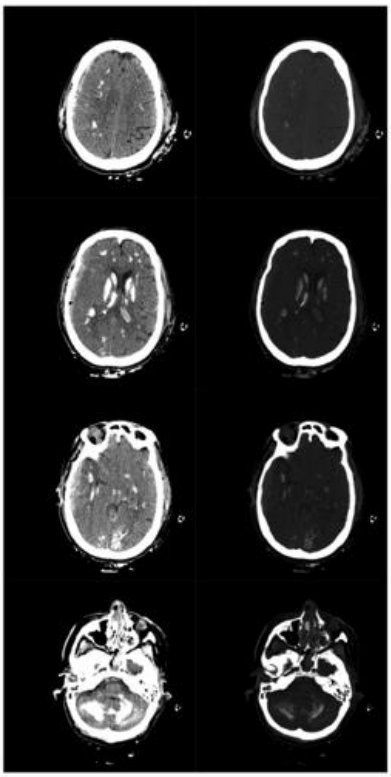

Figure 1 Case 1 CT scan demonstrating wide spread intracranial calcifications. The left side represents the standard brain window, while the right side is presented in the bone window.

Targeted Hounsfield unit measurements resolved the ambiguity. The extra axial crescent measured approximately 50 HU, compatible with acute blood, whereas the deep gray foci measured 150 HU or higher. On bone windows, the deep foci remained strikingly conspicuous, supporting mineralized tissue. The distribution was strictly bilateral and symmetric and matched the typical basal ganglia pattern of Fahr type calcifications rather than the irregular asymmetric pattern of traumatic contusions. No mass effect or edema was associated with the deep hyperdensities. The patient remained neurologically stable. The team concluded that the intraparenchymal findings represented Fahr syndrome and managed the small suspected subdural component conservatively with observation and repeat checks. No emergent neurosurgical intervention was pursued.

Case 2 neuro oncology setting

A 49-year-old woman with a history of ischemic stroke, chronic kidney disease, and biopsy proven lung adenocarcinoma with previously treated brain metastases presented with acute right arm pain and progressive weakness over several hours. She had received carboplatin and pemetrexed with durvalumab maintenance and had undergone hippocampal sparing brain radiotherapy months earlier. A stroke code was activated. Non contrast head CT and CT angiography were obtained immediately.

The non contrast CT revealed numerous supra and infratentorial lesions with mixed density. Several hemispheric lesions displayed hyperdense components suggestive of intralesional hemorrhage, and the largest measured approximately 3 centimeters. Beyond these tumor related findings, there were also diffuse subcortical and cerebellar hyperdense foci in a pattern consistent with mineralization. Hounsfield unit sampling showed values of 150 to 200 HU in the calcified appearing foci, whereas surrounding parenchyma was 50 HU or less. Calcified foci persisted on bone windows. CT angiography did not show large vessel occlusion. **Figure 2 displays multiple slices from this non-contrast CT.**

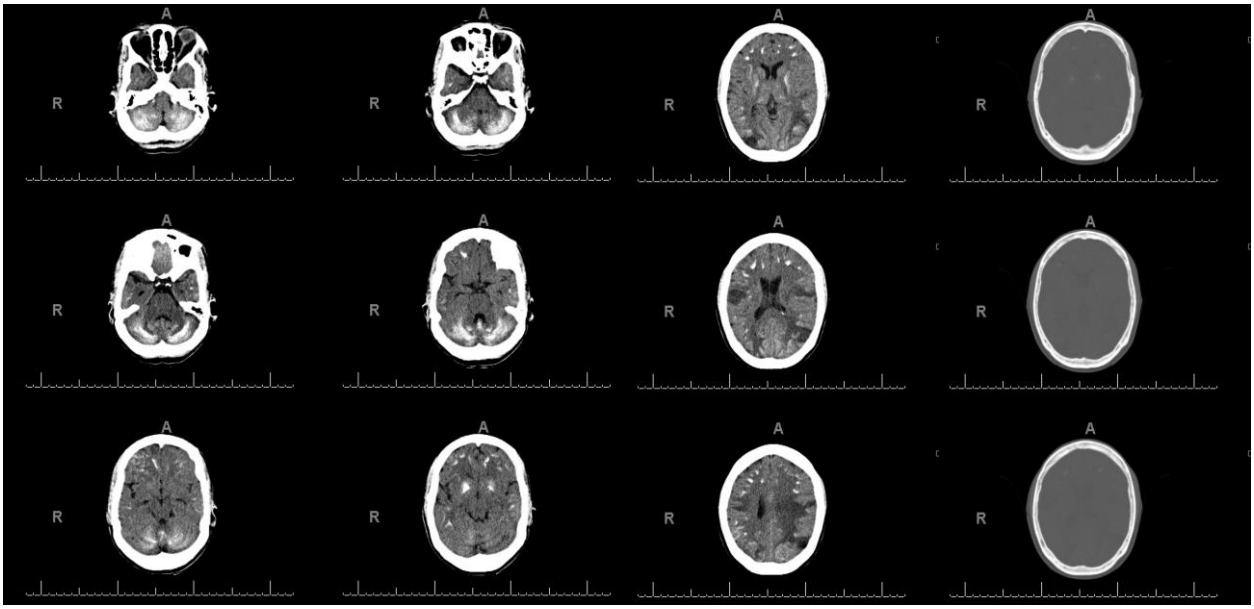

Figure 2: Case 1 CT scan demonstrating wide spread intracranial calcifications. Images from left to right display ascending slices in the standard brain window, while the right side is presented in the bone window.

MRI with and without contrast confirmed multiple ring enhancing lesions compatible with metastatic disease and demonstrated susceptibility effects within several lesions consistent with intratumoral hemorrhage. There was mild vasogenic edema with approximately 5 millimeters of rightward midline shift but no impending herniation. Accurate recognition that diffuse calcifications coexisted with hemorrhagic metastases was crucial. The patient was co-managed with radiation oncology and medical oncology. Dexamethasone was started for edema control and levetiracetam for seizure prophylaxis. With no mass effect requiring decompression and no large vessel occlusion, no emergent neurosurgical or endovascular procedure was indicated.

Discussion

Terminology and pathophysiology:

Primary Familial Brain Calcification represents a genetic entity, whereas secondary forms are commonly referred to as Fahr syndrome. Shared histopathologic themes include perivascular mineral deposition, endothelial dysfunction, and microangiopathy. In secondary presentations, derangements of calcium phosphate balance, for example hypoparathyroidism and chronic kidney disease, create a biochemical milieu that favors ectopic intracranial mineralization.4 Mitochondrial disorders and inflammatory or infectious processes have also been linked. Clinical expression is heterogeneous and ranges from parkinsonian features and dystonia to ataxia, cognitive and psychiatric changes, seizures, and autonomic symptoms. A notable minority remains asymptomatic despite substantial calcific burden, which underscores the need for clinicoradiologic correlation rather than reflexive intervention [4,5].

Imaging approach and practical discriminators:

Non contrast head CT is the workhorse for detection, but interpretation should be structured. First, assess pattern and symmetry. Bilateral near mirror involvement of basal ganglia, subcortical U fibers, and the cerebellar dentate region favors calcification over hemorrhage. Second, sample Hounsfield units. Acute intraparenchymal blood commonly measures about 30 to 70 HU, whereas calcifications are typically 150 HU or higher and may exceed 200 HU. Third, check bone windows. Mineralized foci remain conspicuous, while soft tissue hemorrhage is less apparent. When available, MRI with susceptibility weighted imaging and, where applicable, quantitative susceptibility mapping can help distinguish diamagnetic calcium from paramagnetic blood breakdown products and adjudicate ambiguous foci [5,6].

Differential diagnosis in trauma and oncology:

In trauma, alternatives include cortical contusions, diffuse axonal injury, and hemorrhagic shear lesions, which are usually asymmetric, cortically based, and accompanied by edema. This contrasts with the fixed symmetric deep gray pattern typical of calcification. In oncology, hemorrhagic metastases, radiation related change, treatment related mineralization, and chronic entities such as old cavernous malformations

or treated neurocysticercosis may coexist. Systematic attention to anatomic distribution, HU values, and persistence on bone windows prevents over attribution of every hyperdense focus to blood and helps triage which lesions are acutely dangerous versus chronic background.[7,8]

***Stepwise diagnostic strategy for acute care*:**

Step 1 identify whether hyperdense foci are bilateral and symmetric in typical locations such as basal ganglia, subcortical white matter, and the cerebellar dentate. Step 2 sample Hounsfield units in representative foci and in any suspected blood products. Values of 150 HU or higher support calcification, whereas values around 30 to 70 HU support acute blood. Step 3 review deliberately in bone windows to confirm mineralization. Step 4 use MRI, including susceptibility weighted sequences, when anatomy is complex due to tumor or prior radiotherapy or when CT features remain equivocal. Step 5 order focused laboratory tests to screen for common secondary causes such as calcium, phosphate, magnesium, parathyroid hormone, and renal function, with vitamin D as clinically indicated. Step 6 manage symptoms and underlying causes with seizure prophylaxis when appropriate, edema control for mass effect, and early consultation with endocrinology and nephrology for metabolic drivers and with neuro oncology and radiation oncology for tumor related issues [6-8].

Implications for neurosurgical decision making:

For acute neurosurgical services, the pivotal question is whether a hyperdense finding represents an emergent surgical problem. Integrating symmetry, Hounsfield unit sampling, and bone window verification reduces false alarms and avoids unnecessary procedures. In trauma, correctly labeling symmetric basal ganglia calcifications as background findings enables conservative management of small extra axial bleeds when clinically appropriate. In oncology, separating benign mineralization from truly hemorrhagic space occupying lesions focuses intervention on findings that drive current symptoms.

Limitations and generalizability:

These are illustrative cases designed to sharpen diagnostic reasoning rather than to quantify test characteristics. Not all calcifications reach 150 HU and motion, partial volume effects, or beam hardening can blur attenuation distinctions. Likewise, not all acute hemorrhage falls within a narrow range. The proposed approach is therefore intentionally multimodal and combines pattern recognition with Hounsfield unit sampling, bone window review, and selective MRI. The suggested laboratory panel is a pragmatic screen that should be adapted to clinical context and resource availability. Despite these caveats, the sequence is feasible in high throughput emergency settings and is likely to reduce misclassification.

***Future directions*:**

Prospective work that links quantitative imaging metrics with metabolic profiles and longitudinal clinical outcomes could refine thresholds for Hounsfield unit-based decision making, clarify when MRI adds decisive value, and identify which secondary etiologies most strongly predict progression, symptoms, or response to targeted metabolic management.

Conclusion

A focused CT routine that checks symmetry, samples Hounsfield units, and reviews bone windows reliably distinguishes Fahr type calcifications from acute hemorrhage at the point of care. Paired with a brief metabolic screen and, when needed, susceptibility weighted MRI, this approach improves confidence, avoids unnecessary interventions, and aligns care quickly with the true cause of symptoms in trauma and neuro oncology.

Conflict of interest and disclosure of funding statement:

The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

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