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Case Report

Cerebral Calcifications in Systemic Lupus Erythematosus

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Background

Cerebral calcifications of the brain parenchyma or vasculature are common neuroimaging findings. Etiologies include acquired or congenital infections, genetic syndromes, intracranial neoplasms, immunodeficiencies, metabolic disorders, metastatic lesions, toxic, traumatic, and vascular malformations. Calcifications may also result from chemotherapy and radiation treatment. The prevalence of cerebral calcifications on computed tomography (CT) scans increases with age, ranging from 1% in young individuals to 20% in the elderly [1].

Cerebral calcifications are better detected with CT than magnetic resonance imaging (MRI) [2]. Basal ganglia calcifications in the elderly are usually asymptomatic and may be incidental findings on CT scans performed for dementia, head trauma, seizures, or other indications [3]. Cerebral calcifications may also be associated with pathology (i.e., calcified aneurysms, Fahr's disease, neoplasms, neurocysticercosis, Herpes Simplex infection, toxoplasmosis, tuberculomas).

In this case report, we discuss a patient with systemic lupus erythematosus (SLE) who presented with one year of progressive right leg weakness and frequent falls. Her CT scan demonstrated extensive brain calcifications (Figures 1 and 2). She received a thorough evaluation to determine whether the brain calcifications were responsible for her symptoms.

Case

A 43-year-old African American female with SLE for 13 years presented with one year of progressive right leg weakness and urinary incontinence. For the last several months, she has had recurrent falls and uses a borrowed walker to ambulate. She had no constipation, bowel incontinence, or problems with memory or speech. She denied alcohol or illicit drug use.

Her SLE has been complicated by alopecia, arthralgias, cardiomyopathy, discoid lesions and panniculitis, hypertension, immune thrombocytopenia, oral ulcers, and avascular necrosis of the femur. She also had IgM nephropathy, an episode of optic nerve demyelination, and Vitamin B12 deficiency.

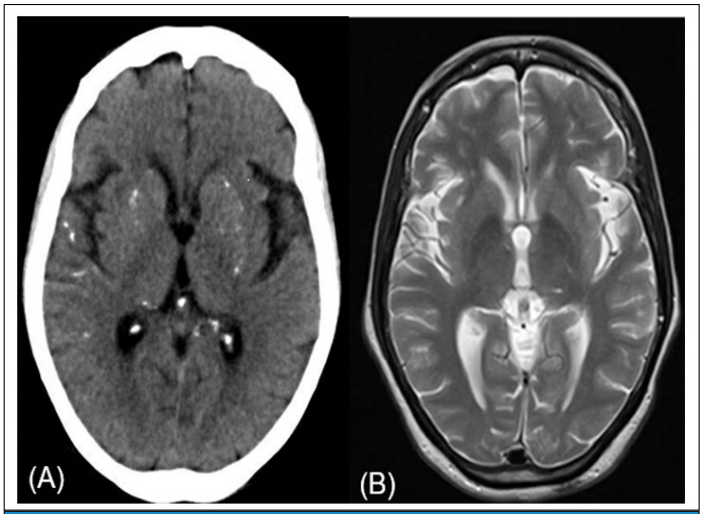

Figure 1: Calcifications in basal ganglia and right lemporal lobe. (A) Axial Brain CT, (B)Axial MRI T2 sequence. CT visualization superior to MRI

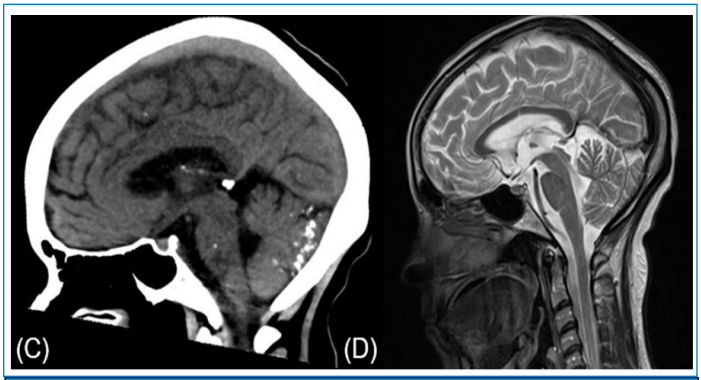

Figure 2: Calcifications seen in cerebellum. (C) Sagittal Brain CT. (D)Saggital MRI T2 sequence. CT visualization superior to MRI.

One year before presentation, she experienced neurologic events suggestive of seizures. For example, she would wake up on the floor without remembering an event. She never sought treatment for these episodes, and no diagnosis was made.

Treatment for SLE has included hydroxychloroquine, IVIG, methotrexate, mycophenolic acid, prednisone, and Rho (D) immune globulin. She was poorly compliant with her medications.

Neurologic exam revealed normal mental status and cranial nerves. Motor exam was normal in the upper extremities but remarkable for right leg weakness without a clear proximal, distal, or radicular pattern. Reflexes were 3+ in the upper extremities, 2+ in the knees, and absent in the ankles. Babinski signs were present bilaterally. Sensory exam was reduced to pinprick in the median nerve distribution of the right hand and pinprick and temperature in the right ankle and foot. Vibration and proprioception were normal. There was no sensory level. Coordination was normal in the upper extremities and difficult to assess in the legs due to weakness. She could not walk unassisted.

Neuroimaging

CT brain demonstrated bilateral curvilinear calcifications in the basal ganglia and nodular calcifications in the brainstem, cerebellum, and temporal lobe (Figures 1, 2). Brain MRI confirmed multiple punctate foci of increased susceptibility throughout bilateral cerebral and cerebellar convexities and basal ganglia consistent with calcifications. There was also superficial siderosis of the left frontal lobe, lacunar infarcts in the body of the corpus callosum and left periventricular white matter, and patchy ill-defined periventricular subcortical white matter hyperintensities suggestive of chronic small vessel disease.

MRI of the cervical, thoracic and lumbar spinal cord revealed no enhancement. There was atrophy from C7-T5, right greater than left. There was also a subtle hyperintense signal on T2 weighted images from T3-T5, right greater than left.

Laboratory (normal values in parentheses)

Complete blood count: White blood cells 3.7 (4,000-10,000/mcL), hemoglobin 11 (12-16 g/dL), hematocrit 34.3 (35-45%), platelets 241 (150,000-450,000/mcL).

Chemistries: sodium 144 (135-145 mmol/L), potassium 3.7 (3.6-5 mmol/L), chloride 114 (101-110 mmol/L), CO2 22 (21-31 mmol/L), BUN 10 (6-20 mg/dL), glucose 82 (70-100 mg/dL), creatinine 0.7 (0.5-1.2 mg/dL), calcium 8 (8.4-10.2 mg/dL), phosphorus 4.1 (2.5-4.6 mg/dL), AST 11 (15-46 unit/L), ALT 8 (10-60 unit/L).

Other: Copper 59 (80-158 mcg/dL), ESR 78 (0-20 mm/hr), folate 27.2 (3-20 mcg/L), magnesium 1.7 (1.8-2.5 mg/dL), parathyroid hormone 37 (15-65 pg/ml), serum aquaporin-4 antibody levels 18.2 and 11.9 (normal 0-3 unit/mL), Vitamin B12 197 (180-914

pg/mL), Vitamin E 8.3 (7-25.1 mg/L). HIV and RPR negative.

CSF

Lumbar puncture: WBC 25 (0-8/mm3), red blood cells 200 (0/mm3), lymphocytes 89 (40-80%), monocytes 11 (15-45%), protein 69 (15-45 mg/dL), glucose 29 (50-80 mg/dL). Cryptococcal antigen negative. CSF culture revealed skin flora. No malignant cells.

Discussion

Brain calcifications are uncommon in SLE except in the presence of cerebral lupus [4]. Our patient did not present with neuropsychiatric symptoms. However, the CSF profile revealed elevated protein, low glucose, and a lymphocytosis consistent with central nervous system (CNS) inflammation. The patient had a history of possible seizures, which may have been a symptom of CNS lupus. She had one episode of optic nerve demyelination 13 years ago. MRI revealed cervicothoracic atrophy and hyperintense signal in the upper thoracic region. Serum NMO antibodies were elevated on two occasions. The patient's episode of optic nerve demyelination, spinal cord findings, and positive aquaporin-4 antibodies were consistent with the diagnosis of NMO.

Involvement of the CNS in SLE is not rare, affecting up to 50% of lupus patients. In descending order, the most common features of CNS lupus are headaches, seizures, visual failure, fatigue, hemiparesis, memory impairment, confusion, personality change, and depression. Our patient may have had epileptic seizures, which occur in up to 42% of lupus patients [5].

Up to 30% of patients with cerebral lupus may have brain calcifications. Cerebral calcifications are bilaterally symmetric and most commonly found in the globus pallidus and cerebellum, as in our patient. Other commonly involved structures include the caudate head, centrum semiovale, globus pallidus, putamen, and thalamus [6].

Our patient had no symptoms such as bone pain, constipation, kidney stones, or psychiatric changes to suggest hypercalcemia. Her calcium, phosphorus, and parathyroid hormone levels were normal.

Fahr's disease, or primary familial brain calcification, is a rare syndrome characterized by symmetric calcifications involving the basal ganglia, centrum semiovale, dentate nucleus and thalamus (bilateral striopallidodentate calcinosis) [7]. Patients typically present in middle age, and the exact incidence is unknown. There are both hereditary and sporadic forms. Patients may exhibit a wide variety of neurologic symptoms, most commonly movement disorders. Ataxia, cognitive impairment, epileptic seizures, and psychiatric disorders may occur. Patients may also be asymptomatic [7]. Our patient had basal ganglia and cerebellar calcifications but no chorea, dystonia, cognitive or psychiatric symptoms. The leg weakness was her only motor symptom. Our patient did not have genetic testing for Fahr's disease. Cerebral calcifications associated with SLE seemed a more likely diagnosis than Fahr's disease.

MRI Imaging of the cervical, thoracic and lumbar spine revealed cervicothoracic atrophy and increased signal in the upper thoracic cord. We attributed the myelopathy to SLE, Vitamin B12, copper deficiency, and NMO.

Discharge medications included prednisone, mycophenolate mofetil, prednisone, and Vitamin B12.

Conclusions

This patient's right leg weakness and incontinence were related to multifactorial myelopathy and unrelated to the extensive cerebral calcifications. Physicians should be aware that CNS calcifications can occur in various brain regions, including the basal ganglia and cerebellum in up to 30% of patients with cerebral lupus. Calcifications are better visualized on CT than MRI. Cerebral calcifications in SLE may be an incidental finding, as in this case.

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