Intracerebral Calcification in Sjögren's Syndrome

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KEY WORDS: intracerebral calcification, tenal tubular acidosis, Sjögren's syndrome (SS)

IMAJ 2023; 25: 351–351

PATIENT DESCRIPTION

The patient, a 32-year-old woman diagnosed with Sjögren's syndrome according to the 2016 European League Against Rheumatism (EULAR)/American College of Rheumatology (ACR) criteria, presented with paresthesia of her face and limbs. Extra glandular manifestations of her primary disease included severe Raynaud's phenomenon and chronic interstitial nephritis. There was no family history of neurologic diseases. Neurological examination was notable for symmetrical decreased sensation in the upper limbs distally. The rest of the neurological examination was unremarkable.

Laboratory study revealed creatinine of 1.4 mg/dl (eGFR 50 ml/min/1.73 m^2). calcium, phosphorus, vitamin D, and parathyroid hormone (PTH) were all within the normal range. Repeated blood gases analysis and urinalysis revealed normal anion gap metabolic acidosis along with alkaline urine, suggestive of distal renal tubular acidosis.

Non-contrast brain computed tomography (CT) demonstrated extensive symmetrical calcification in the lentiform [Figure 1A, arrows] and caudate nucleus as well as in the frontal subcortical area [Figure 1A, arrowhead]. On brain magnetic Resonance Imaging (MRI) and fluid attenuated inversion recovery (FLAIR) images supratentorial leukoariosis was seen by multiple hyperintense areas in the white matter were noted [Figure 1B]. Non-contrast chest and abdominal CT did not reveal vascular calcification, signs of nephrocalcinosis, or nephrolithiasis.

COMMENT

The common findings on brain MRI in Sjögren's syndrome include nonspecific T2-weighted periventricular and subcortical white matter hyperintense areas [1]. Intracerebral calcification has not been reported previously in patients with SS. However, cerebral calcification has been described in patients with system sclerosis, systemic lupus erythematosus, and sarcoidosis related to vascular damage and ongoing venous inflammation [2,3]. Distal renal tubular acidosis has also been associated with vascular calcification. Cerebral calcifications have been described in patients with distal renal tubular acidosis, particularly in patients with carbonic anhydrase II deficiency [4].

In our patient, the hyperintense lesions on MRI were consistent with previous reports in Sjögren's syndrome and may have been related to an autoimmune mechanism. However, as our patient also had severe Raynaud's phenomenon and renal tubular acidosis, her prominent intracerebral calcification may have been related to a combination of vascular, metabolic, and inflammatory factors.

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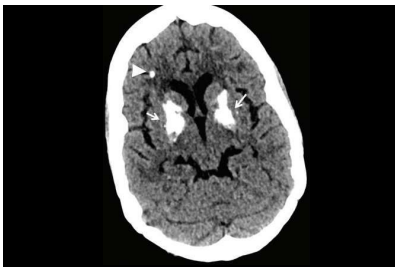
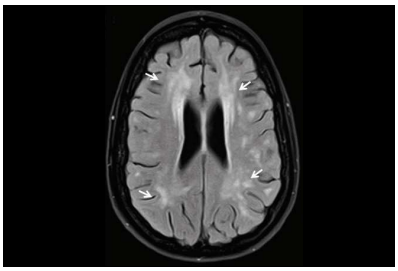
Figure 1. Brain computed tomography shows extensive symmetrical calcification in the lentiform (arrows in A) and caudate nucleus as well as in the frontal subcortical area (arrowhead) **[A].** A brain magnetic resonance image shows multiple hyperintense areas in the white matter (arrows) **[B]**

[A]



[B]



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