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LETTER TO THE EDITORS

Video head impulse test contributes to Susac syndrome diagnosis

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Dear Sirs,

Susac syndrome (SuS) is a rare autoimmune endotheliopathy classically affecting young women, causing varying degrees of encephalopathy, as well as visual and cochleo-vestibular dysfunction [1]. Although its pathophysiology remains uncertain, some authors believe that the pre-capillary arterioles of the brain, retina and inner ear are most affected, causing multiple microinfarctions in these areas [2, 3].

SuS diagnosis is challenging because the clinical triad is present in less than 30% of cases. Optimization of complementary testing is therefore important, to reach earlier diagnosis and potentially avoid severe or irreversible relapses in untreated patients [4]. Central corpus callosum microinfarctions called *snowballs*, as well as linear infarcts and upper corpus callosum lesions named *spokes* and *icicles*, have been observed on brain MRI. In addition, internal capsule infarctions in a *string of beads* pattern are frequently present. Fluorescein angiography will show endovascular hyperfluorescence and retinal artery branch occlusions. Tonal audiometry reveals unilateral involvement predominantly affecting low tones, but bilateral or complete and sudden hypoacusis may occur, secondary to cochlear infarcts. Associated symptoms include tinnitus, vertigo and nausea [3]. Even though recent diagnostic criteria mention audiograms, vestibular caloric testing and evoked myogenic responses as the most useful tests to evaluate inner ear function, use of the video head impulse test (vHIT) has not been studied in these patients [4]. vHIT is a new, rapid and well-tolerated vestibular function test which identifies the function of all six semicircular canals, and can provide evidence of dysfunction, even in the emergency room setting [5, 6].

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We report a case in which vHIT use was useful to establish SuS diagnosis and support early patient management.

A 19-year-old female with no relevant past medical history presented to the emergency department complaining of intense headache, somnolence and confusion. On clinical examination, disorientation, mild cognitive impairment and brisk deep tendon reflexes were observed. Brain MRI (Fig. 1) showed multiple small vessel infarcts with predominant involvement of the corpus callosum. MR angiography was normal. SuS was suspected. CSF biochemistry showed eight leukocytes/mm3, normal glucose and a slightly elevated protein level (67 mg/dl). Patient referred no visual or cochleo-vestibular symptoms.

To better examine the inner ear, a vHIT was performed, showing right-sided posterior semicircular canal dysfunction with decreased gain and presence of overt saccades, suggesting involvement of the inferior branch of the right vestibular nerve (Fig. 2). Caloric testing was not performed nor were evoked myogenic responses evaluated.

Given patient signs and symptoms at presentation, MRI lesions found, CSF results (PCR and cultures) and normal echocardiogram, multiple sclerosis, primary angiitis of the central nervous system, acute encephalitis and paradoxical embolism were ruled out as differential diagnoses.

Because the patient was suspected of having SuS, preemptive treatment with methylprednisolone pulses (5 g in 5 days) and intravenous immunoglobulins (2 g/kg) was administered during the acute phase. Once stabilized, mofetil mycophenolate, oral steroids (1 mg/kg) and monthly IVIG were prescribed as maintenance treatment and the patient was discharged. Rituximab was added 6 months later after a clinical relapse characterized as encephalopathy, unilateral hearing loss and headache. Three months later due to intense headache, vertigo, multiple microinfarctions on MRI and endovascular hyperfluorescence in FA, cyclophosphamide was added, achieving disease control.

Definitive SuS was confirmed on an outpatient basis based on typical brain MRI findings, multiple BRAO and endovascular hyperfluorescence on fluorescein angiography and subtle neurosensorial left hypoacusis on tonal audiometry.

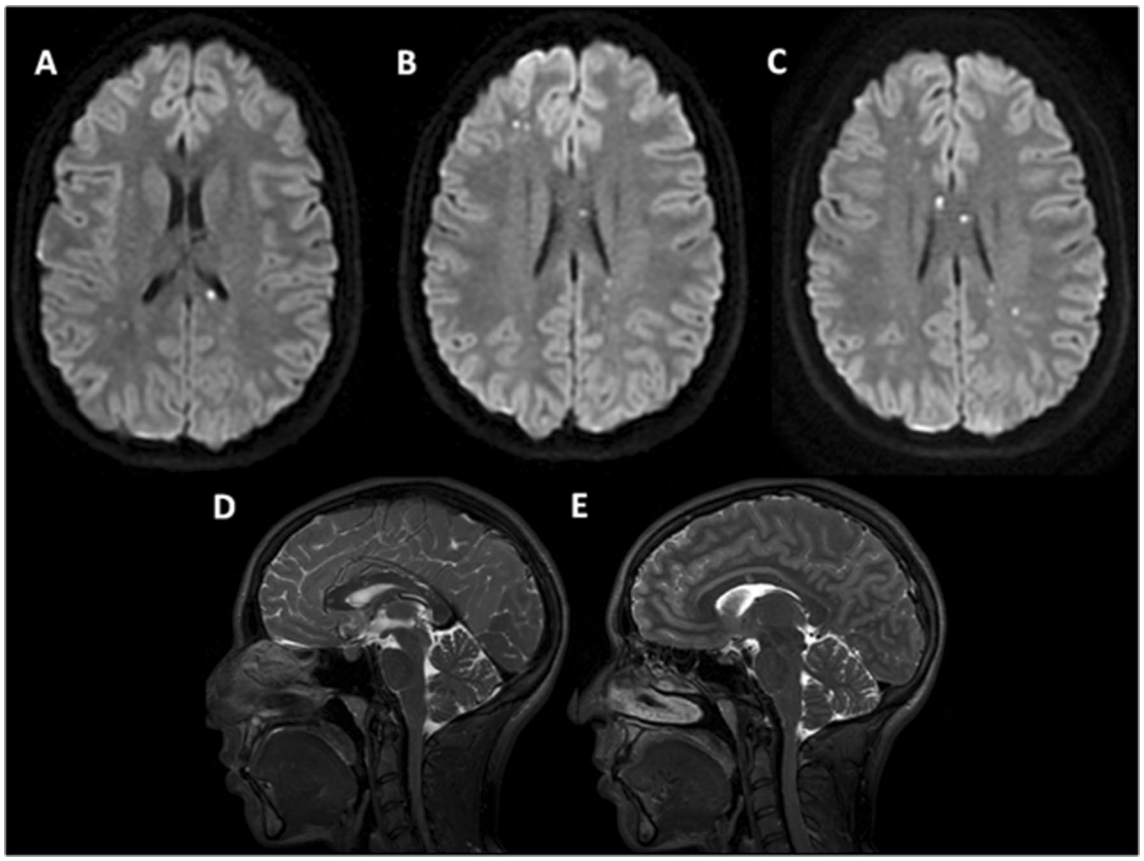

Fig. 1 Brain magnetic resonance imaging. **a** Corpus callosum splenium infarct. **b** Subcortical and corpus callosum microinfarction. **c** Genu, body and subcortical infarcts. **d** Central fibers *snowball* lesions in corpus callosum genu and body. **e** Snowball lesion of the body of the corpus callosum

As mentioned, SuS could not be confirmed at disease onset, as patient presented only encephalopathy and brain small vessel infarctions. To detect subclinical cochleovestíbular symptoms, a vHIT was performed, which showed decreased gains in the right posterior semicircular canal with overt saccades, reinforcing a SuS diagnosis.

To the best of our knowledge, there are no other clinical reports or case series mentioning use of vHIT in Susac syndrome patients. vHIT is a simple, fast and cost-effective test which evaluates eye movements to brief, passive, unpredictable, small, head turns in planes oriented along the horizontal, as well as the left anterior/right posterior canals and the right anterior/left posterior canals, using a head-mounted video camera. To objectively measure semicircular canal function during head turns, this method measures gains in vestibulo-ocular reflex quantified by an automatized software [6, 7]. VOR gain is the ratio of eye to head velocity and should ideally be about 1.0. In practice, normal healthy subjects typically show VOR gains below 1.0 (around 0.8–0.9), measuring adequacy of the semicircular canal response: i.e., vestibular function. Vestibular injury will reduce vestibulo-ocular reflex gains; therefore, eye movement will not be proportional to head rotation velocity, resulting in reduced gain and corrective saccades. "Overt saccades" can be observed once the head has stopped moving and the affected side identified. Some patients present catch-up saccades during the head impulse, which are not detectable by the clinician, also known as "covert" saccades. Unlike caloric testing, which evaluates only the horizontal semicircular canals, and is time consuming and not well tolerated by patients, vHIT is well tolerated (even in cases of acute vestibular syndrome), quick to complete (takes around 10–15 min) and provides objective evaluation of physiological function of all six semicircular canals [5, 6]. Likewise, vHIT was chosen acutely instead of caloric testing or evoked myogenic responses because those ancillary methods are uncomfortable or are technically difficult to obtain in the inpatient setting, respectively.

Even though anterior inferior cerebellar artery occlusion can cause hearing loss and isolated posterior semicircular canal or cochleo-vestibular dysfunction, we still considered that this patient´s deficit pattern was due to microvascular ischemia, and found unequivocal SuS findings on MRI with

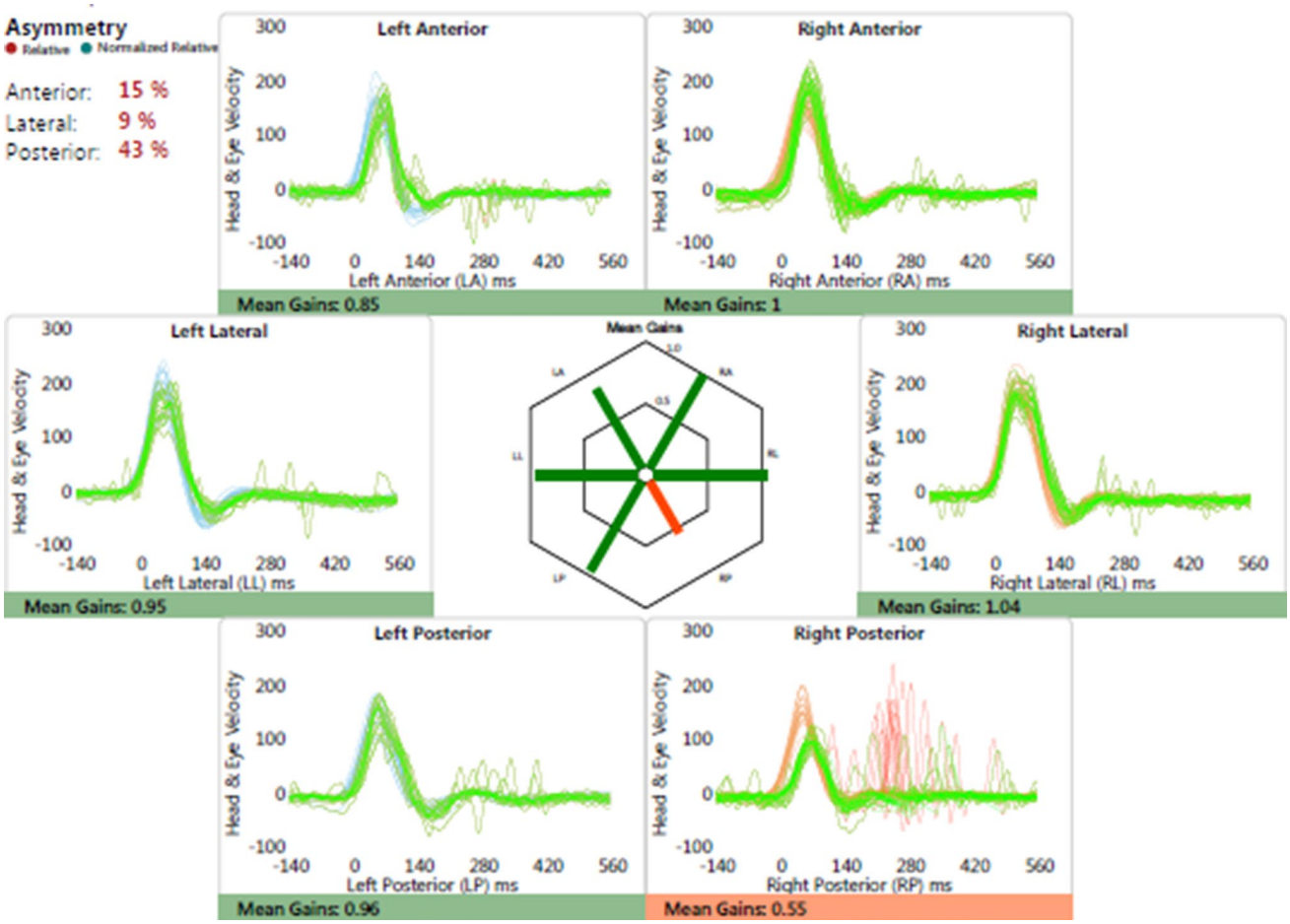

Fig. 2 Video head impulse test. HEX representation showing reduced gains of the posterior semicircular canal on the right side (black arrow) and overt saccades (white arrow). In the center: low gain on the right posterior semicircular canal (red line). Normal gains for the other semicircular canals (green lines)

normal MR angiography and characteristic vascular lesions on fluorescein angiography [8].

vHIT should be considered as a confirmatory test of vestibulo-cochlear dysfunction in patients in whom SuS is suspected, as it may detect subclinical involvement and support intense early treatment [9].

Compliance with ethical standards

Conflicts of interest The authors have no disclosures to report related to this article.

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