

Case report

Selective aphasia and focal hypoperfusion in a bilingual patient with HaNDL syndrome

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ABSTRACT

The syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) is a rare disease (1) characterized by a benign, self-limited headache syndrome accompanied by neurological deficits (isolated aphasia can be seen in nearly 22% of these patients (2)). Differential diagnosis between acute ischemic stroke and HaNDL syndrome has to be made in order to decide whether to perform a lumbar puncture or start reperfusion treatment early. CT perfusion have proved to be useful for differential diagnosis (3). We present a case of a HaNDL patient referred to the Emergency Department as a stroke in the context of acute onset of selective aphasia (Spanish) in a bilingual patient (French-Spanish). Urgent CT perfusion during the episode revealed increased mean transit time (MTT) with normal Cerebral Blood Flow (CBF) in posterior language areas. The case provides information on a HaNDL attack and its pathophysiology with hemodynamic changes in the acute period during the episode and the benign condition of the illness.

1. Case report

A previously healthy 35-year-old right-handed and late bilingual man presented with selective aphasia in Spanish (native speaking in French and learned Spanish in his early 20s, living in Spain since then). No prior history of migraine or headache disorder was recorded. He was referred to the emergency room with initial clinical diagnosis of stroke. On examination he was afebrile, oriented in time and place. No precipitating factors or viral illness were recorded. The funduscopy was normal and neurological examination revealed a moderate selective aphasia in Spanish (second language) but he was able to communicate in French (native language). Urgent CT perfusion-angiogram performed 2 h after the onset, revealed increased MTT and TTP with a normal CBF in the MCA territory, confined to parietal and temporal regions (image 1-A). CT angiogram was normal, basal CT was normal. He had a National Institutes of Health Stroke Scale Score of 6 (scored for questions, commands and language items, without impairment of motor, visual, extinction or neglect items), explored in Spanish, with no deficits in native French (language assessed by Neurologist A.C.C., bilingual in French and Spanish). He made paraphasic mistakes and his speech was difficult in Spanish. His abilities to name, repeat, understand, and write were also impaired in Spanish but could communicate in perfect spoken French without fluency or comprehensive mistakes. The patient was a late bilingual subject (learned as an adult with formal

education), living and working in Spain for more than 15 years, working in a high qualified job. All serum laboratory testings were within normal reference range. During the assessment in the ER department the patient experienced a moderate headache lateralized to the left side. He was not treated with thrombolytic therapy and was admitted in the Stroke Unit during 24 h with analgesic treatment. On reassessment 24 h after onset of symptoms, aphasia in Spanish was mild, only with minor fluency changes (6 words in one minute that begin with the letter F on the Montreal Cognitive Assessment Test, MoCA). No other cognitive domains were affected on MoCA. A lumbar puncture was performed, 24 h after initial assessment. Opening pressure was normal (12 cm H₂O) and cerebrospinal fluid studies revealed an elevated protein count of 2.1 g/L (reference range 0.15–0.45 g/L) and normal glucose of 56 mg/dl (serum glucose 91 mg/dl). A pleocytosis with a cell count of $128 \times 10^6/L$ with 88% lymphocytes was detected. PCR for Herpes and Varicella virus were negative and CSF cultures were negative with a GRAM stain without microorganisms. 48 h after admission, language was normal and CT perfusion changes had resolved (image 1-B). MRI, performed 72 h after initial onset, (T1 with contrast, T2, FLAIR and DWI) was normal. Routine chest X-rays, blood count, and an extensive biochemical work-up including liver, kidney, thyroid function tests, coagulation studies and serological tests for collagen, rheumatic, venereal, infectious, and parasitic disease were either normal or negative. The patient showed an excellent functional

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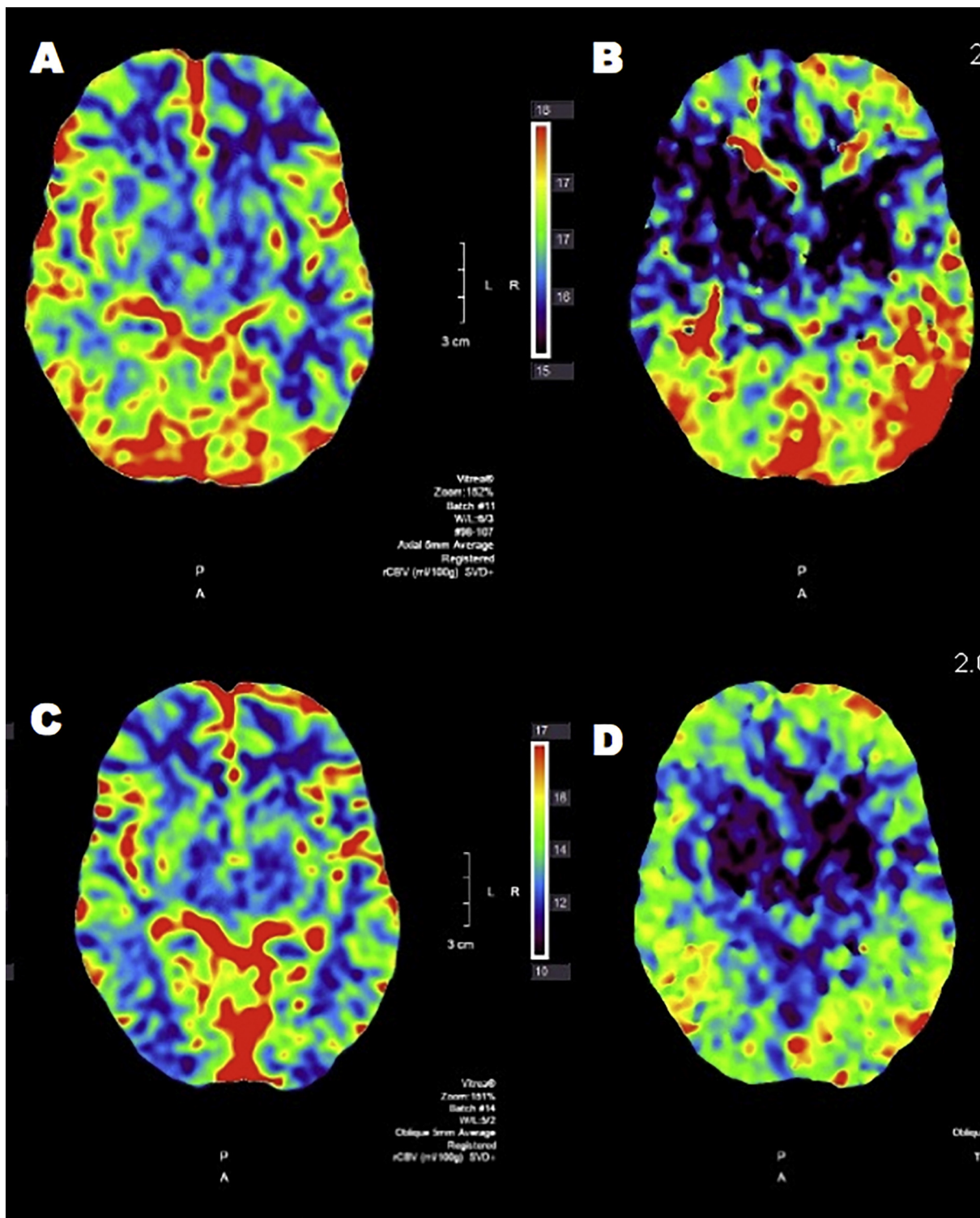


Fig. 1. CT perfusion changes: Axial CT perfusion imaging reveals focal hypoperfusion in the left hemisphere in language areas (A: Cerebral Blood Volume, B: Mean Transit Time) performed 2 hours after the onset. 48 h after the first assessment, symptoms were absent and CT perfusion showed complete normalization of both parameters (C, D)

recovery and was asymptomatic at discharge at day 5.

2. Discussion

The case presenting with selective aphasia for late language only, has a clinical history and CSF profile that meet diagnostic criteria for HaNDL [1]. The diagnosis of this syndrome is mainly an exclusion

diagnosis. Differential diagnosis should consider ischemic stroke, acute meningoencephalitis or aseptic meningitis. CT perfusion imaging performed during this acute episode of HaNDL revealed left temporal and parietal hypoperfusion without vessel occlusion in language areas. Clinical reports [4–6] support the differential localization pattern in bilingual patients where two languages occupy different but overlapping areas in the dominant hemisphere and there is evidence that

second languages may be represented and preferentially processed in temporo-parietal regions (posterior) maybe because native languages regions (frontal or anterior) become highly specialized and resist neural recruitment by a non native-language [6]. Focal hypoperfusion rather than global hypoperfusion, similar to the pattern in middle cerebral artery territory infarction can be observed frequently in HaNDL patients, when CT perfusion imaging is performed in the acute period [2-3]. The normalization of perfusion changes (full reversal in less than 48 h) shows the clinical and imaging relevance of posterior language areas in late bilingual patients, [5] like our patient. To our knowledge, this is the first reported HaNDL patient with selective aphasia and illustrates that temporo-parietal regions in the dominant hemisphere are involved in processing second languages and native language can be preserved (Fig. 1).

Authors contributions

Antonio Cruz Culebras: design and acquisition of data.

Rocio Vera: study supervision.

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