

Headache with transient neurological deficit: a case report with hemispheric hypoperfusion and without lymphocytosis

Elena Martínez Velasco¹  · Patricia Mulero¹ · María Pedraza¹ · Ana I. Calleja¹ · Angel Luis Guerrero¹

Received: 10 February 2016 / Accepted: 15 April 2016
© Springer-Verlag Italia 2016

Dear Editor,

Transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) is an uncommon disorder described in 1981 by Bartleson et al. Current diagnostic criteria (ICHD-III beta version 7.3.5) consider episodes of moderate or severe headache, lasting hours and associating cerebrospinal fluid (CSF) pleocytosis with lymphocytic predominance [1]. Neuroimaging studies and CSF culture are required to be normal.

Neurological deficits [2] more commonly reported are sensory symptoms (78 % of cases), aphasia (66 %), and motor weakness (41 %). In addition to these symptoms, other features as elevation of CSF proteins [2] have also been described.

To draw attention to this potentially misdiagnosed disorder, our aim is to report a case of headache and transient neurological deficits with a severe cerebral perfusion disturbance and the absence of CSF lymphocytosis.

A 15-year-old male with a prior history of migraine was admitted to emergency department due to a non-fluent aphasia and sensory and motor deficit in right limbs of 40 min of evolution. He described a severe stabbing headache starting in right fronto-parietal scalp associating phonophobia and photophobia. A cerebral unenhanced and perfusion computed tomography (CT) was performed, while headache and a mild sensory disturbance in his right limbs were still present. Unenhanced CT was normal, but perfusion CT revealed an increased mean transit time (MTT) and a diminished cerebral blood flow (CBF)

throughout the entire left cerebral hemisphere not confined to a particular vascular territory (Fig. 1). Cerebral blood volume (CBV) was normal. Findings were consistent with hypoperfusion throughout all left cerebral hemisphere.

Neurological deficits lasted less than 1 h. A diffusion-weighted brain magnetic resonance imaging (DWI-MRI) was performed within 24 h from clinical onset with no restricted diffusion lesions and with normal signal intensity of the brain parenchyma. An ultra-early carotid ultrasound imaging and transcranial Duplex showed a generalized increase in distal vascular resistance and segmental accelerations in both middle cerebral arteries with the absence of arterial occlusion. The angiography TC showed no alterations. The existence of a complete hemispheric perfusion/diffusion mismatch, together with the clinical course including the presence of headache, led to a presumed diagnosis of a syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL), so a lumbar puncture was performed 3 days after admission without pleocytosis or hyperproteinorachia. A new CT perfusion 5 days after clinical onset showed normal cerebral flow with normal CBV, MTT, and CBF in left cerebral hemisphere. Carotid ultrasound imaging and transcranial Duplex study was repeated with normalization of distal resistance and flow accelerations. Cerebral SPECT 10 days after clinical onset showed areas of decreased radionuclide uptake suggesting decreased blood flow in left temporal–parietal region (Fig. 2).

CSF microbiological studies and extensive immunological determinations were negative. The patient suffered no new episodes and neurological exam was normal at hospital discharge. Control cerebral SPECT was repeated 2 months later with blood flow normalization (Fig. 2b).

The presentation of sudden-onset neurological deficits at emergency department always leads neurologist to be

✉ Elena Martínez Velasco
emartinezvelasco@gmail.com

¹ Neurology Department, University Hospital, Avda. Ramón y Cajal, 3, 47005 Valladolid, Spain

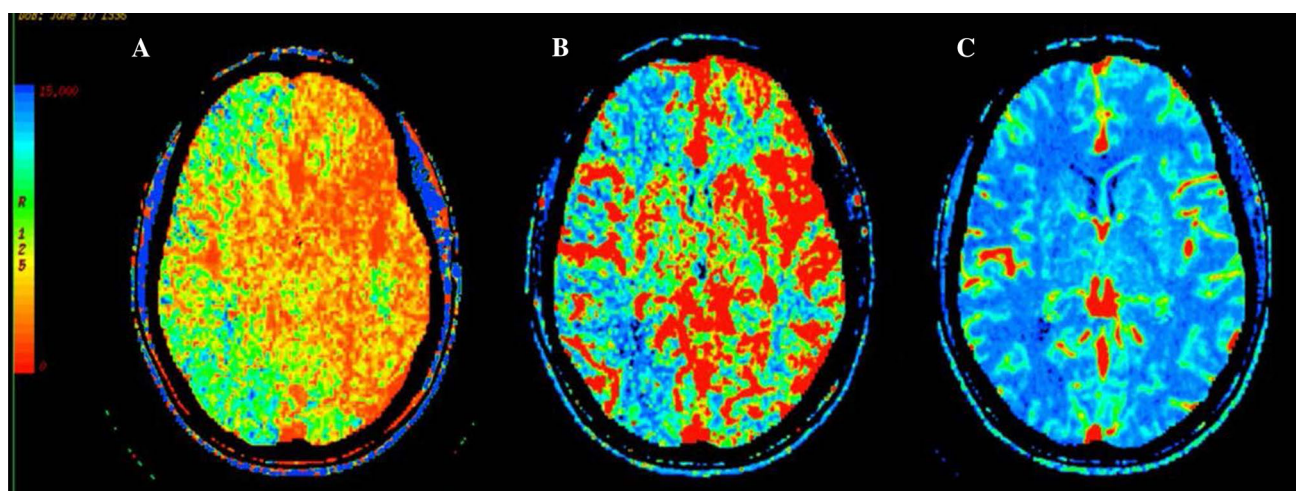


Fig. 1 Perfusion computed tomography at emergency department revealed an increased MTT (a) and a diminished CBF (b) with normal CBV (c) throughout the entire left cerebral hemisphere

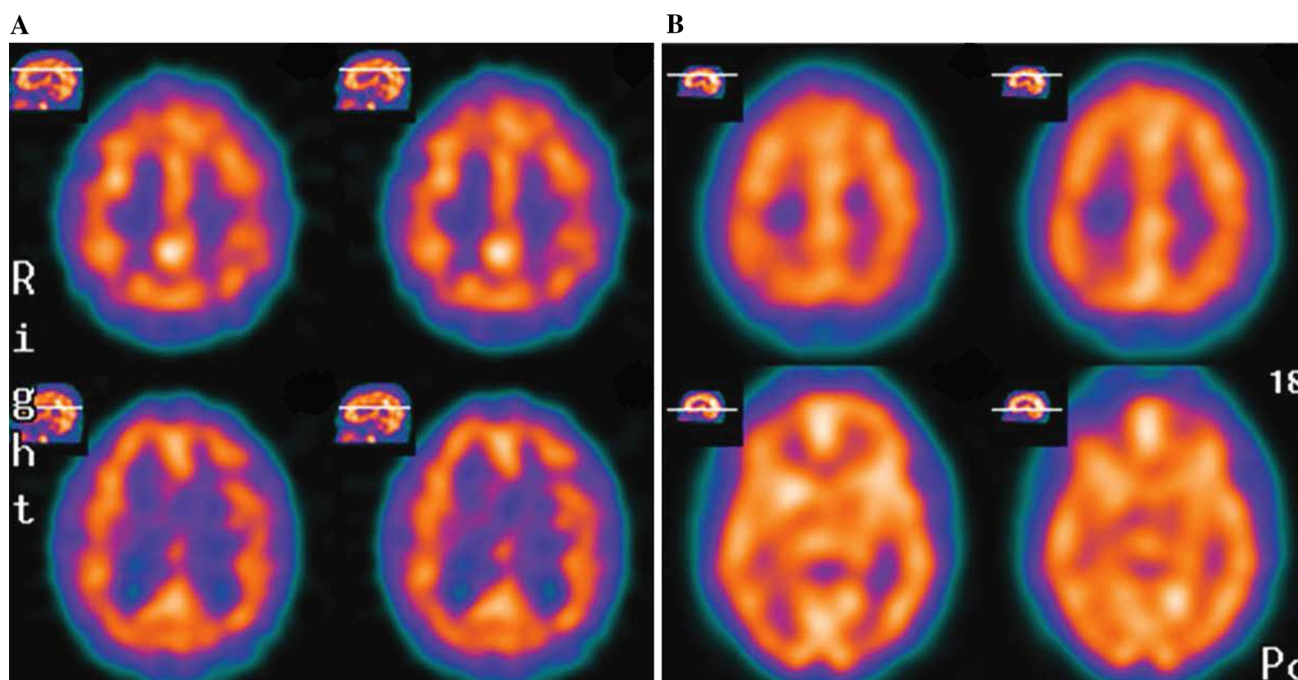


Fig. 2 First cerebral SPECT during acute phase a showed areas of decreased blood flow in the left temporal–parietal region. Two months after clinical onset a new cerebral SPECT, b showed blood flow normalization

aware of the possibility of a stroke. Entities such as HaNDL should be considered in the differential diagnosis of stroke particularly in young patients [3]. Existence of hypoperfusion in the entire left cerebral hemisphere in an area not restricted to a vascular territory, and the absence of a lesion restricted in DWI with a complete perfusion/diffusion mismatch rule out an acute stroke [4]. This pattern in a whole hemisphere with patterns of benign oligemia rather than ischemia has been previously described during a HaNDL attack [4, 5].

Pathophysiology of HaNDL remains unknown. It has been suggested a vascular disorder mediated by an inflammation in the meningeal vascular system, resembling cortical spreading depression-like mechanism similar to that proposed for migraine with aura. Reversible hypoperfusion observed in our case in cerebral CT perfusion, and brain SPECT could support this vascular theory.

Cerebrospinal fluid lymphocytosis is a consistent finding in the literature, and therefore, it was included among HaNDL diagnostic criteria [1]. In spite of the absence of

lymphocytosis in our patient, clinical characteristics as well as CT perfusion and SPECT reversible changes support, in our opinion, the diagnosis of this syndrome. The absence of pleocytosis in our patient could be explained by a delay in performing lumbar puncture; nevertheless, in the same way that SPECT or electroencephalographic anomalies may be absent in this syndrome, we can hypothesize that there might be an HaNDL variant with vascular involvement and no CSF inflammation.

In conclusion, in this case, as shown in previous reports [5], we have observed cerebral hypoperfusion during the acute state of an incomplete form of HaNDL, a rare but probably under-recognized syndrome. This information and the possible absence of lymphocytosis might be considered in the future revision of ICHD. Our description shows the important role that advanced imaging techniques, as CT perfusion, can play in ruling out an acute stroke, and in unnecessary and non-innocuous therapies as endovenous thrombolysis.

Compliance with ethical standards

Conflict of interest None.

References

1. Headache Classification Subcommittee of the International Headache Society (2013) The International Classification of Headache Disorders: 3rd edition (beta version). *Cephalalgia* 33(9):629–808
2. Gómez-Aranda F, Cañadillas F, Martí-Massó JF et al (1997) Pseudomigraine with temporary neurological symptoms and lymphocytic pleocytosis. A report of 50 cases. *Brain* 120:1105–1113
3. Krause T, Nolte CH (2012) The syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) as an acute ischemic stroke mimic leading to systemic thrombolysis: a case report. *Clin Neurol Neurosurg* 114:689–690
4. Pettersen JA, Aviv RI, Black SE, Fox AJ, Lim A, Murray BJ (2008) Global hemispheric CT hypoperfusion may differentiate headache with associated neurological deficits and lymphocytosis from acute stroke. *Stroke* 39:492–493
5. Barragán NG, Guillán M, Zarza B et al (2012) Focal CT hypoperfusion in HaNDL. *J Neurol* 259:1755–1757