slowed by her difficulty organising her thoughts. She has had to delegate all administrative tasks to her daughter, needs to carry a planner at all times, and is unable to return to her job (jewellery shop assistant) because of difficulties counting money and following clients' conversations.

Her first valid neuropsychological examination, completed once psychiatric symptoms had stabilised, yielded a score of 28/30 on the Mini Mental State Exam (MMSE). Language and gestural/constructional praxis were spared. Specific neuropsychological test batteries evaluating executive functions, memory, and attention/concentration detected moderate impairment.

In this case, insidious neuropsychiatric symptoms developed over a 2-year course. Unlike in cases in which functional systems included in the EDSS are impaired, determining to what extent the disease may have progressed is difficult here. Although the patient's manic symptoms were florid and even required a hospital stay, we do not regard this event as a first attack. Contrast-enhanced cerebral MR in the acute phase ruled out inflammatory activity.

There is radiological evidence of dissemination in time and space: the first scan showed 3 out of the 4 Barkhof criteria, and the 6-month assessment showed 4 out of 4. In progressive forms of MS like the one described here, neurological damage tends to persist without there being any obvious inflammatory activity. Laboratory tests (oligoclonal bands in cerebrospinal fluid) also supported the diagnosis of MS. Additional complementary examinations were able to rule out other possible diagnoses. Differential diagnosis would not be completed without considering neurolupus, which often manifests with psychiatric symptoms. It was ruled out based on the patient's consistently normal autoimmune parameters and lack of systemic signs and symptoms.

The patient met the McDonald MRI criteria, and was therefore diagnosed with primary progressive MS. At present, her score on the EDSS is 3.0, reflecting moderate disability for one functional system, namely cognition.

Euphoric/manic episodes in MS have been shown to be associated with severe and progressive forms of the disease, associated cognitive impairment, frontal demyelination, cerebral atrophy, and a prolonged course of the disease. The latter description does not apply to our patient. Although euphoria and other psychiatric disorders are unusual as initial manifestations of MS, we feel that this diagnosis should be considered during initial evaluations of patients in psychiatric units.1

References


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Intravenous thrombolysis in capsulear warning syndrome: is it beneficial?12

Trombólisis intravenosa en el síndrome de alarma capsular: ¿es beneficiosa?

Capsular warning syndrome (CWS) is characterised by repeated and self-limiting episodes of motor deficit (sensory deficit is less frequent), which are caused by ischaemia limited to the internal capsule. The risk of infarction following CWS is higher than in the case of other transient ischaemic attacks (TIA).1 CWS treatment during the acute phase is controversial. The effectiveness and safety of intravenous thrombolysis (IVT) is not well-known.1,3

We present the case of a man aged 52 with a history of smoking, alcohol abuse, and obstructive sleep apnoea syndrome (OSAS), who was undergoing treatment with CPAP. He experienced 2 sudden-onset transient episodes of left-sided weakness and difficulty articulating words, each lasting less than 30 minutes. He was treated in his referral hospital, where he suffered a third identical episode before being sent to our hospital. On the way, he suffered another episode in the ambulance. Upon arrival at the hospital, he presented mild right faciobrachiocural hemi-

pa
eresy and mild dysarthria (4 on the NIHSS scale). Computed
tomography (CT) and intracranial CT angiography yielded
normal results. The patient was transferred to the stroke
unit for monitoring. After 40 minutes, the patient’s clini-
cal condition worsened; he presented left hemiplegia, an
NIHSS score of 9, and low blood pressure (minimum val-
es: 89/60 mmHg). Doctors performed an IVT (rt-PA dose:
0.9 mg per kg body weight). After bolus administration,
motor deficit showed significant improvement (NIHSS score
of 3); however, 15 minutes later (during the rt-PA infusion),
he scored 11 on the NIHSS (left hemiplegia and dysarthria)
with no subsequent improvement. The patient underwent an
emergency cranial CT that revealed a faint hypodense focus
affecting the posterior third of the right putamen (Fig. 1). As
his blood pressure remained low, intravenous fluid therapy
was administered and he was placed in the Trendelenburg
position; doctors observed no improvements.

A complementary study (electrocardiogram, echocar-
diogram, and transcranial and supra-aortic trunk neu-
ronoscopy) showed no significant changes. The routine
CT scan performed 24 hours after the IVT revealed an
ischaemic lesion affecting the posterior third of the
right putamen (Fig. 2). Eight days after admission, the
patient was discharged from hospital with left hemipare-
sis, facial paresis, and dysarthria (NIHSS = 8, modified Rankin
scale = 3).

CWS is considered a recurrent form of lacunar TIA that
is related to cerebral small vessel disease. The patho-
physiology of CWS is not well-known. The most widely
accepted mechanism has to do with haemodynamic changes
occurring in a small damaged perforating artery. However,
other proposed potential mechanisms include lipohyalinosis,
microatheromatosis, vasospasm, and cardiac or artery-to-
artery embolism.¹

Up to 42% of all CWS patients develop established lacu-
inar infarcts, which in most cases occur in the internal
capsule; however, other localisations have included the
striato-capsular area and the anterior choroidal artery.¹

Raising blood pressure by means of IV rehydration ther-
apy or drugs such as phenylephrine has been suggested as
the treatment of choice for counteracting cerebral hypoper-
fusion in the small vessels. Use of heparin or antiplatelet
drugs has not delivered conclusive results.²

IVT has been shown to be beneficial for all aetiological
subtypes of stroke.⁴,⁵ However, according to some authors,
this therapy is not as beneficial and does not affect prognosis
in cases of lacunar stroke.⁶⁷

Few articles describe the effectiveness of IVT in CWS.
The literature includes descriptions of only 5 patients, 4 of
which presented good clinical progress following IVT. Based
on these results, the authors conclude that IVT is safe and
improves prognosis in this patient subgroup.⁴,⁵

IVT did not change the natural course of CWS in our
patient. Our case seems to support the haemodynamic
hypothesis as a possible mechanism involved in this syn-
drome.

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Figure 1  Emergency cranial CT. Hypodense focus in the pos-
terior third of the right putamen.

Figure 2  Cranial CT 24 hours after IVT. Ischaemic lesion in the
posterior third of the right putamen.
Migraine triggered by laughing as a form of presentation of a Chiari Type I malformation

Cefalea desencadenada por la risa como forma de presentación de una malformación de CHIARI tipo I

Dear Editor:

Chiari malformation type I (CM1) is defined as the extrusion of the cerebellar tonsils through the foramen magnum and into the spinal canal. It can be confirmed by a radiograph showing descent of brain tissue 5 mm or more, lending a pointed appearance to the cerebellar tonsils and decreasing the subarachnoid space at the cranio-cervical junction. The clinical manifestations of CM-I are associated with direct compression of the brainstem and cervical spinal cord, or with changes in cerebrospinal fluid.

A 38-year-old man with no relevant medical history described headaches occurring throughout the preceding year. Pain was dull and moderately intense (VAS score 6) and located in the occipital region radiating to the frontal region. Headache was triggered by laughing, but not by Valsalva manoeuvres or physical exercise. Episodes lasted 10 to 30 minutes and were not accompanied by focal neurological signs. In the preceding month, pain had grown more intense (VAS 8) and was triggered not only by laughing, but also by minimal exertion and Valsalva manoeuvres such as coughing or defecating. Lateral and rotational movements of the head also caused headache episodes. The general examination was normal; a neurological examination revealed left-beating horizontal nystagmus with no other relevant findings.

Brain and cervical MRI (Fig. 1) showed that cerebellar tonsils had descended 16 mm below the foramen magnum, forming a point. The medulla had taken on an angled shape and the subarachnoid space at the cranio-cervical junction was reduced. Signs were consistent with a diagnosis of type I Chiari malformation.

The patient was initially treated with 600 mg ibuprofen every 8 hours but did not respond. Treatment was changed to 25 mg indometacin every 8 hours, still with no response. As the patient’s headaches were incapacitating, he underwent decompressive occipital surgery associated with C1 laminectomy. The headaches, vertigo, and nystagmus all resolved and have not returned in 2 years of follow-up.

Our patient’s pain met criteria for headaches caused by type 1 Chiari malformation according to the second edition of the International Headache Classification (Table 1). There have been cases in which headaches triggered by laughing, one of which was prolonged, signalled the presence of CM1. In another published case of a patient with CM1 and IV-ventricle ependymoma without hydrocephalus, headaches triggered by laughing resolved once the tumour had been removed. Although the phenomenon of headache caused by laughing is exceptional, and would rarely appear as the first manifestation of CM1, this did occur in our patient.

Laughter constitutes a Valsalva manoeuvre, and, like coughing, it raises intracranial pressure. Coughing and other activities involving Valsalva manoeuvres may cause primary cough headache. They may also trigger a headache in the

Figure 1 Sagittal, fat-suppressed cervical MR image revealing ectopia of the cerebellar tonsils 16 mm below the foramen magnum, angled medulla, and decreased subarachnoid space at the cranio-cervical junction.

Please cite this article as: Jiménez Caballero PE, Calle Escobar ML, Portilla Cuenca JC, Casado Naranjo I. Cefalea desencadenada por la risa como forma de presentación de una malformación de CHIARI tipo I. Neurología. 2013;28:446–447.