7. Gastaut H, Vigouroux M, Trevisan C, Regis H. Le syndrome: "hemiconvulsion-hemiplegie-epilepsie" (syndrome HHE). Rev Neurol (Paris). 1957;97:37-52.
8. Arrese-Gispert L, Gutiérrez-Solana LG, García-Peñas JJ, Ruiz-Falcó ML. Síndrome hemiconvulsión-hemiplejia: presentación de dos casos con los hallazgos de resonancia magnética cerebral en secuencias potenciadas en difusión. Rev Neurol. 2005;41: 344-8.
9. Kawada J, Kimura H, Yoshikawa T, Ihira M, Okumura A, Mirishima T, et al. Hemiconvulsion-hemiplegia syndrome and primary human herpesvirus 7 infection. Brain Dev. 2004;26: 412-4.
10. Wakamoto H, Ohta M, Nakano N. Hypercytokinemia in hemiconvulsions-hemiplegia syndrome associated with dual infection with varicella zoster and Epstein-Barr viruses. Neuropediatrics. 2002;33:262-5.
11. Kimura M, Tasaka M, Sejima H, Takusa Y, Ichiyama T, Yamaguchi S. Hemiconvulsion-hemiplegia syndrome and elevated interleukin-6: case report. J Child Neurol. 2002;17:705-7.
12. Freeman JL, Coleman LT, Smith LJ, Shield LK. Hemiconvulsion-hemiplegia-epilepsy syndrome: characteristic early magnetic resonance imaging finding. J Child Neurol. 2002;17:10-6.
13. Morimoto T, Fukuda M, Suzuki Y, Kusu M, Kida K. Sequential changes of brain CT and MRI after febrile status epilepticus in a 6-years-old girl. Brain Dev. 2002;24:190-3.
14. Hisano T, Ohno M, Egawa T, Takano T, Shimada M. Changes in diffusion-weighted MRI after status epilepticus. Pediatr Neurol. 2000;22:327-9.
15. Scantlebury MH, David M, Carmant L. Association between factor V Leiden mutation and the hemiconvulsion, hemiplegia, and epilepsy syndrome: report of two cases. J Child Neurol. 2002;17:713-7.
16. Mondal RK, Chakravorty D, Das S. Hemiconvulsion, hemiplegia, epilepsy syndrome and inherited protein S deficiency. Indian J Pediatr. 2006;73:157-9.
17. Lee C, Born M, Salomons GS, Jakobs C, Woetfle J. Hemiconvulsion-hemiplegia-epilepsy syndrome as a presenting feature of L-2-hydroxyglutaric aciduria. J Child Neurol. 2006;21:538-40.
18. Bahi-Buisson N, Kossorotoff M, Barnerias C, Boddaert N, Burgeois M, Dulac O, et al. Atypical case of hemiconvulsion-hemiplegia-epilepsy sydrome revealing contralateral focal cortical displasia. Dev Med Child Neurol. 2005;47:830-4.
19. Aicardi J, Chevrie JJ. Consequences of status epilepticus in infants and children. Adv Neurol. 1983;34:115-25.
20. Aicardi J, Amsili J, Chevrie JJ. Acute hemiplegia in infancy and childhood. Dev Med Child Neurol. 1969;11:162-73.
21. Kataoka K, Okuno T, Mikawa M, Hojo H. Cranial computed tomographic and electroencephalographic abnormalities in children with post-hemiconvulsive hemiplegia. Eur. Neurol. 1988;28:279-84.
22. Kwan SY, Wong TT, Chang KP, Yang TF, Lee YC, Guo WY, et al. Postcallosotomy seizure outcome in hemiconvulsion-hemiatrophy-epilepsy syndrome. Zhonghua Yi Xue Za Zhi (Taipei). 2000;63:503-11.
23. Herbst F, Heckmann M, Reiss I, Hügens-Penzel M, Gortner L, Neubauer B. Hemiconvusion-hemiplegia-epilepsy syndrome (HHE). Klin Padiatr. 2002;214:126-7.

P.E. Jiménez Caballero* and S. Murcia Carretero

Servicio de Neurología, Hospital Virgen de la Salud, Toledo, Spain

*Corresponding author.
E-mail: pjimenez1010j@yahoo.es (P.E. Jiménez Caballero).

Pseudo-subarachnoid haemorrhage. A need for clinical-radiological diagnostic criteria

Pseudohemorragia subaracnoidea. Necesidad de criterios diagnósticos clínico-radiológicos

Dear Editor,

Subarachnoid haemorrhage (SAH) is a medical emergency, so its early diagnosis and treatment are vital. In cranial computerized tomography (cCT) it appears with an increased density in the basal cisterns and the subarachnoid space. Some neurological diseases have been described (hypoxic encephalopathy, hyperperfusion radical encephalopathy, extensive infarctions, viral meningoencephalitis, purulent meningitis, bilateral subdural haematoma and idiopathic intracranial hypertension)[1-5] that exceptionally present a tomographic pattern typical of SAH, without clinical or pathological haemorrhage data; this entity is known as pseudo-subarachnoid haemorrhage (pSAH).

We report a case of pSAH in the context of an advanced hypoxic-ischemic encephalopathy (HIE). The patient was a 38-year-old male smoker. He was admitted to the intensive care unit under mechanical ventilation after suffering cardiopulmonary arrest outside the hospital. He could not open his eyes or make sounds, but carried out approximation-pronation of the right arm under algesic cranial stimulation; brainstem reflexes were present and there was general spastic hypertonia. The electrocardiogram showed the classic pattern of Brugada syndrome. The initial cCT presented an early bilateral occipitotemporal hypodensity indicative of acute ischemic injury, presumably with an anoxic-haemodynamic origin, and there were no other findings (fig. 1). The electroencephalogram (EEG) showed an alpha-coma pattern. At 72h, his condition worsened, with loss of brainstem reflexes and reactivity; a new cranial CT showed diffuse cerebral oedema (DCE), hypodensity in the territory of the posterior cerebral arteries and symmetric diffuse hyperdensity in the cerebral sulci and cisterns, with an appearance of massive SAH (fig. 1). The incongruity of SAH in the clinical context led to a diagnostic reconsideration, with Hounsfield unit (HU) determination, with a value of at most 42 UH, in hyperdense sulci and cisterns; the final interpretation of the finding was pSAH related to severe diffuse cerebral oedema. A new EEG confirmed brain death.

One of the most common causes of pSAH is diffuse cerebral oedema due to HIE, which can displace the healthy brain areas into areas normally occupied by cerebrospinal fluid (CSF); neuroimaging can show the disappearance of the convexity sulci, poor differentiation between grey and white matter and generalised cerebral hypodensity. Increased intracranial pressure may also cause vascular congestion with dilation of the superficial (pial) venous structures. Consequently, the CSF (hypodense) is displaced from the subarachnoid space, where the meninges and vessels are concentrated (hyperdense). The blood-brain barrier breakdown in relation to hypoxia may contribute to an increase in CSF protein content, although this mechanism

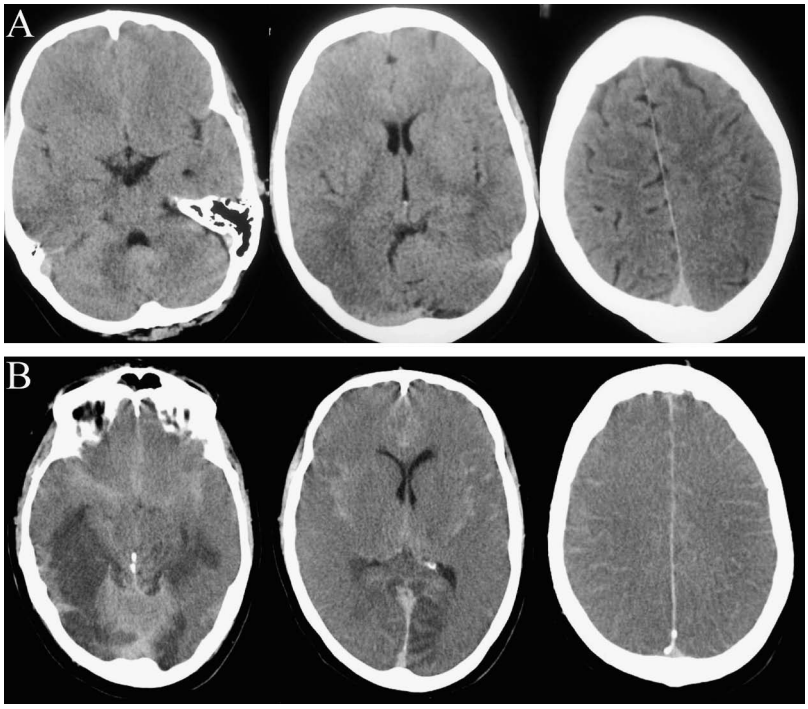

Figure 1 A: Cranial CT without contrast on the day of admission. B: Cranial CT without contrast on the third day of evolution.

is considered more relevant in cases of purulent meningitis that appear as pSAH. These events yield an increased CT density at that level which, coupled with a decreased parenchymal attenuation coefficient attributable to diffuse cerebral oedema, results in a greater density difference between the congestive zone (hyperdense) and the oedematous parenchyma (hypodense) and generates a tomographic image resembling that of SAH.[1-3] Although the literature reflects necropsy and CSF study as the only ways of confirming this suspicion,[1,2] the advent of new technologies has made digital processing of images possible; detailed measurement of the density of different visible elements, measured in HU, are now available, with specific differential values for normal and oedematous parenchyma, as well as for blood and the subarachnoid space rich in protein and congested by displaced structures.[2] Reviewing the literature, we found the following radiological data that, directly or indirectly, suggest pSAH:

- Within the indirect signs, we can count the following CT indicators of cerebral oedema: decreased relative radiographic density of white matter in patients with diffuse cerebral oedema compared to controls, thus reducing the density differential between white and grey matter in that group (both quantifiable in HU), decrease in fissural, cisternal or ventricular size or even volume reduction or non-visualisation of the venous sinuses in contrast CT.[2] The presence of these signs depends on whether there is diffuse cerebral oedema, which is common in HIE, but less prominent in other entities related to pSAH. In our case, the loss of differentiation between grey and white matter was obvious, as was a reduction in the fissure, ventricular and cisternal sizes (fig. 1). We did not quantify parenchymal densities or administer contrast.
- Within the direct signs, we highlight the possibility of measuring density in hyperdense sulci and especially in hyperdense basal cisterns. In an analysis of 7 cases of pseudo-SAH confirmed by necropsy or CSF study, the attenuation coefficient in the basal cisterns was 21 to 44 HU (mean, 29-33 HU), which was clearly lower than in all control cases consisting of patients with confirmed SAH (60-70 HU).[1] This sign would apply to all cases of pSAH, regardless of their aetiology, given that its positivity would depend on the existence of diffuse cerebral oedema. In our patient, the attenuation coefficient in the hyperdense sulci and cisterns did not exceed a maximum of 42 HU.

Given the condition of the patient, with a clearly negative prognosis after the clinical and electroencephalographic evaluation, it was decided not to perform an LP, and consent for an autopsy was not given after confirming the condition of cerebral death. Therefore there was no anatomopathological material to reliably discard SAH presence. However, considering the clinical symptoms and tomographic sequence of the patient, with appearance of diffuse cerebral oedema and a significantly lower attenuation coefficient than expected for blood in hyperdense regions, we are convinced that the diagnosis was pSAH.

Pseudo-SAH is a rare condition, but of required knowledge for every neurologist, given the prognostic and therapeutic implications that may result from it. Patients with a similar clinical and radiological situation may evolve very differently depending on the aetiology, provided there is adequate management. While most HIE-associated cases have a very poor vital prognosis,[1] there have been reports of dramatic

improvements in those attributed to meningoencephalitis or idiopathic intracranial hypertension when adequate treatment was received.[3] The elaboration of consensual clinical and radiological criteria, based on a coherent clinical context and on the aforementioned tomographic signs, would make the following possible:

1. Establishment of a firm probability diagnosis before histochemical confirmation in cases pending aetiological and pathogenic sorting, in which the study of CSF should be considered unavoidable. This would avoid erroneous early prognostics or therapeutic approaches during the interval until CSF sampling is completed (e.g., suspension of anticoagulation in a patient with a prothrombotic state and pSAH secondary to massive venous sinus thrombosis on suspicion of SAH). It could even avoid possible therapeutic omissions when the presentation severity could lead to a conservative approach supported by an erroneous presumptive diagnosis (e.g., medical management of a presumed massive SAH in a patient with a minimum score on the Glasgow Coma Scale when the true underlying cause is meningoencephalitis).
2. Avoidance of invasive diagnostics without significantly compromising the diagnosis in cases of known aetiology consistent with the disease when the prognosis is basically extremely poor.

References

1. Avrahami E, Katz R, Rabin A, Friedman V. CT diagnosis of non-traumatic subarachnoid haemorrhage in patients with brain edema. Eur J Radiol. 1998;28:222-5.
2. Given CA, Burdette JH, Elster AD, Williams DW. Pseudo-subarachnoid hemorrhage: a potential imaging pitfall associated with diffuse cerebral edema. AJNR Am J Neuroradiol. 2003;24:254-6.
3. Cucchiara B, Sinson G, Kasner SE, Chalela JA. Pseudo-Subarachnoid Hemorrhage. Report of three cases and review of the literature. Neurocrit Care. 2004;1:371-4.
4. You JS, Park S, Park YS, Chung SP. Pseudo-subarachnoid hemorrhage. Am J Emerg Med. 2008;26:521.
5. Misra V, Hoque R, Gonzalez-Toledo E, Kelley RE, Minagar A. Pseudo-subarachnoid hemorrhage in a patient with acute cerebellar infarction. Neurol Res. 2008;30:813-5.

C. de la Cruz-Cosme[a,*], G. Barbieri[a] and A. Vallejo-Baez[b]

[a] *Servicio de Neurología, Hospital Universitario Virgen de la Victoria, Málaga, Spain*
[b] *Servicio de Medicina Intensiva, Hospital Universitario Virgen de la Victoria, Málaga, Spain*

*Corresponding author.
E-mail: drcruzcosme@hotmail.com (C. de la Cruz-Cosme).