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N. Morollón*, P. Guerrero, J. Duarte

Sección Neurología, Complejo Asistencial de Segovia, Segovia, Spain

*Corresponding author.
E-mail address: noemimorollon@gmail.com (N. Morollón).
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Spontaneous acute epidural haematoma of the cervical spine with an atypical onset resembling ictal symptom☆

Hematoma epidural agudo cervical espontáneo de inicio atípico simulando cuadro ictal

Dear Editor,

Spontaneous acute epidural haematoma (SAEH), described by Jackson in 1869, is an infrequent condition with an estimated annual incidence of one case per million population.[1] It accounts for less than 1% of all spinal space-occupying lesions, with peak incidence between the sixth and seventh decades of life.[1—3] SAEH may cause rapid, irreversible neurological impairment and is associated with a mortality rate of 6% to 8%.[3] These lesions are preferentially located in the cervicothoracic region, mainly in the posterior portion of the spinal canal.[1] Cases where no underlying cause or predisposing factor for bleeding (blood dyscrasia, anticoagulant or antiplatelet treatment, tumours, pregnancy, or trauma) can be identified are considered spontaneous; this occurs in 40% to 50% of patients.[2] Most authors suggest a haemorrhagic venous aetiopathogenic mechanism, due to the rupture of valveless epidural venous plexuses in response to a sudden pressure increase at the thoracic or abdominal level.[4] However, other authors suggest bleeding is arterial in origin,[5] secondary to the rupture of epidural radicular arteries, based on the rapid clinical deterioration typically associated with SAEH.[6]

Our patient is a 79-year-old man with no relevant history who presented symptoms of intense headache and neck pain and sudden-onset left hemiparesis of 3 hours' progression. In view of these symptoms, we activated the code stroke protocol and the patient was transferred to his reference hospital. Motor examination revealed paresis of the left arm (3/5) and left leg paralysis associated with contralateral hypaesthesia without sensory extinction, left extensor plantar reflex, and right flexor plantar reflex. An emergency brain CT scan did not show signs of intra- or extra-axial bleeding or acute brain ischaemia. Suspecting spinal involvement, we performed an emergency cervical CT scan, which showed a hyperdense epidural cervical collection located in the left posterolateral region from C3 to C7 (Fig. 1). After being diagnosed with cervical SAEH, the patient underwent an emergency C3 to C7 posterior cervical laminectomy with microsurgery; the epidural cervical collection was completely evacuated. No underlying vascular anomalies at the C3 to C7 level were identified during the procedure; the anatomical pathology study ruled out the presence of such anomalies in the surgical specimen. A cervical MRI scan performed after the surgery (Fig. 2) revealed the presence of typical postsurgical changes, absence of signs of cervical myelopathy, and small remnants of the epidural collection. The postsurgical study ruled out the presence of any clotting disorders which may have triggered the symptoms. After surgery, the patient displayed good clinical progress, recovering mobility of the left side of the body, with no residual motor or sensory deficits after completing rehabilitation.

SAEH clinically manifests as acute, local, lancinating pain accompanied by motor, sensory, and/or autonomic neurological deficits, normally symmetrical, developing over a period of minutes or days.[7] Unilateral symptoms are infrequent and may lead to misdiagnosis, and diagnostic and therapeutic delay. In these cases, a differential diagnosis of cerebrovascular stroke is essential,[8] given the catastrophic effect of treatment with fibrinolytic agents. Acute neck pain is probably the only baseline clinical finding that may lead us to suspect spinal involvement. The distinctive feature of the case described is that the patient's condition was initially interpreted as a supratentorial ischaemic stroke.

In MRI studies, SAEH in the acute or hyperacute stage appears isointense on T1-weighted sequences and hyperintense on T2-weighted sequences.[9] Although MRI is the diagnostic method of choice, spine CT is a widely available

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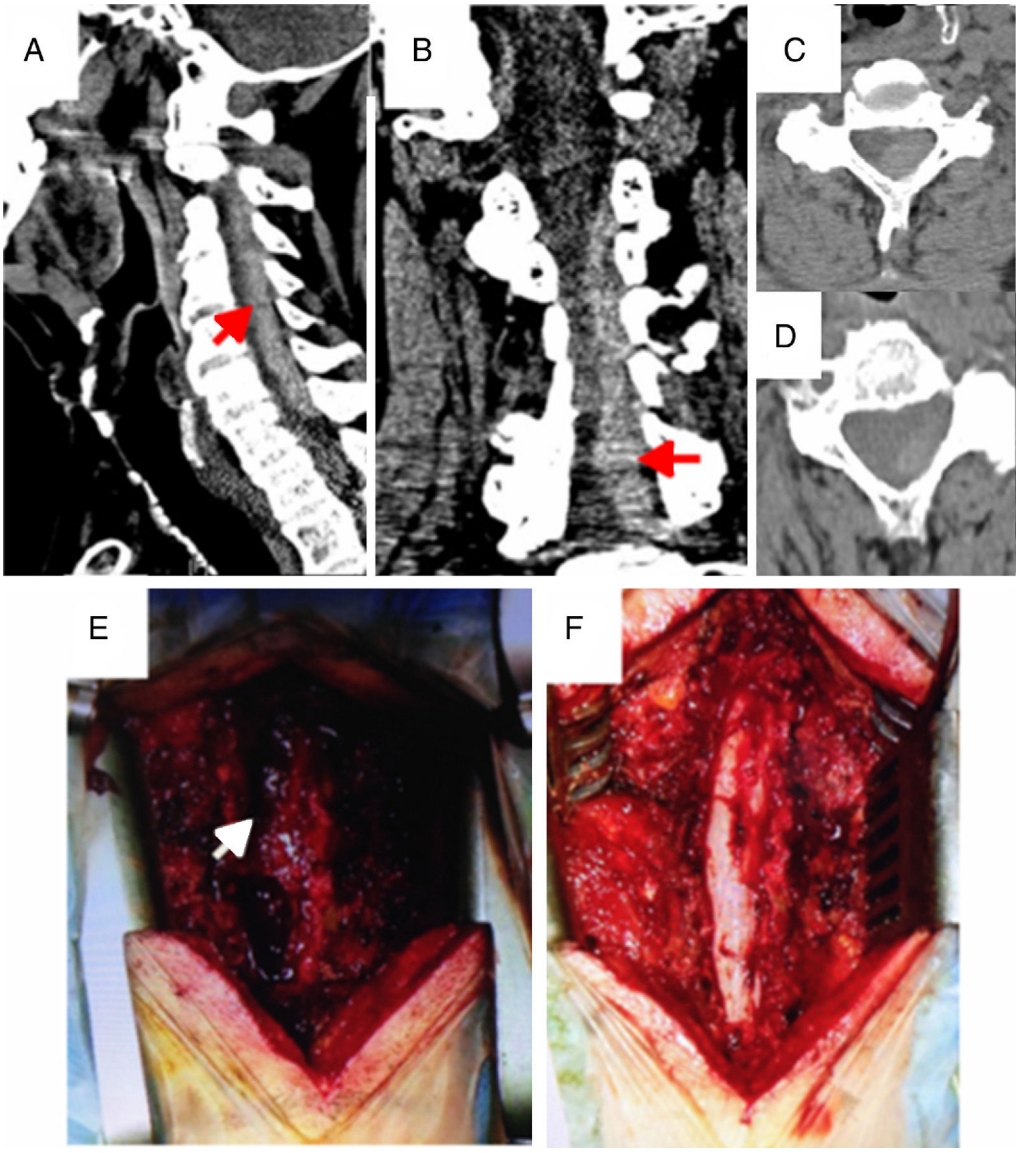

Figure 1 Presurgical images from the emergency cervical CT scan. Sagittal (A), coronal (B), and axial (C and D) slices revealing the presence of an extensive hyperdense collection at the left posterolateral epidural level C3-C7 (arrows), compatible with SAEH. This collection causes a pronounced reduction of the spinal canal and displacement of the spinal cord to the right. Images after C3-C7 cervical laminectomy showing the previously described acute cervical epidural collection at the left posterolateral level (E). Image during surgery after complete evacuation of SAEH (F).

test which is fast to perform and suitable for unstable patients under invasive monitoring, and can show the location and extension of the haemorrhage. Further studies such as MR-angiography may provide relevant information on underlying lesions.[10] Spinal MR-angiography is not routinely indicated; it would only be justified in those cases in which spinal MRI and/or CT findings give rise to strong suspicion of an underlying vascular lesion responsible for SAEH.[11]

SAEH is considered a surgical emergency and most published series recommend early surgical treatment, since surgical decompression in the first 12 to 24 hours after symptom onset is associated with better functional outcomes.[2] Decompressive laminectomy and complete evacuation of the epidural collection is currently considered the surgical treatment of choice, achieving the best results.[12] Conservative symptomatic treatment may be indicated in cases of incidental diagnosis, patients with

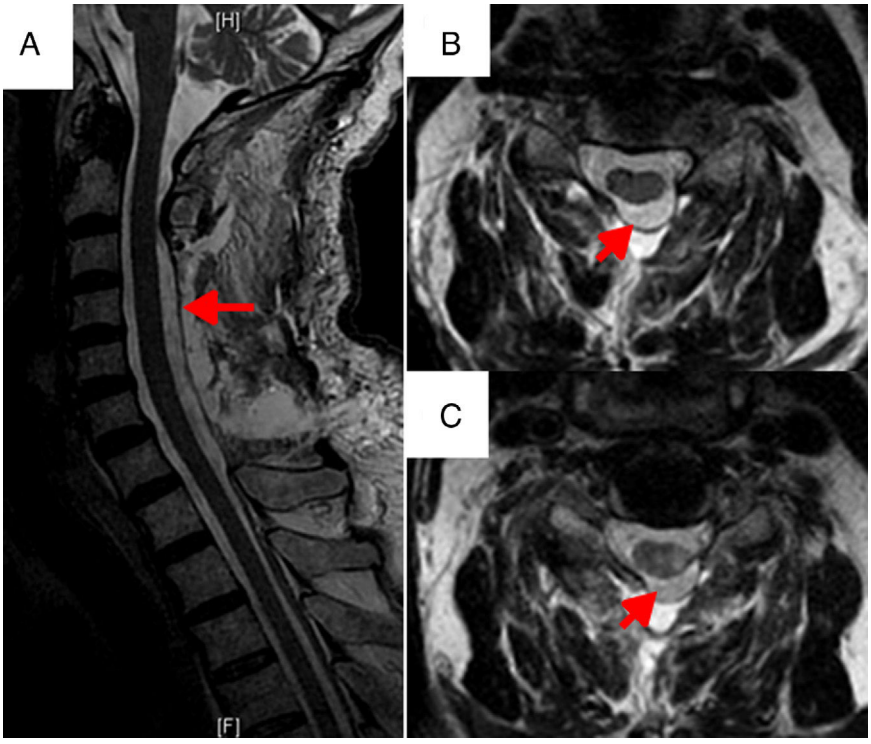

Figure 2 Cervical MRI scan performed 24 hours after surgery. Sagittal (A) and axial (B and C) slices showing adequate decompression of the spinal canal at vertebrae C3-C7 (arrows), with no sign of new blood collections or myelopathy at that level.

minimal or no clinical signs of spinal cord compression, in patients with severe comorbidities and poor prognosis of short-term survival, or in patients with severe clotting disorders resulting in high surgical risk.[2] The baseline degree of spinal involvement, as well as the time between symptom onset and surgical decompression, may be the most important prognostic factors for functional recovery.[13]

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Conflicts of interest

The authors have no conflicts of interest to declare.

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A. Arévalo*, M. Navas, P. Pulido, R. García de Sola

Servicio de Neurocirugía, Hospital Universitario de la Princesa, Madrid, Spain

* Corresponding author.
E-mail address: praimale@hotmail.com (A. Arévalo).

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Aphasia in a patient with acute hepatic encephalopathy associated with multifocal cortical brain lesions☆

Afasia en paciente con encefalopatía hepática aguda asociada a lesiones corticales cerebrales multifocales

Dear Editor,

Acute hepatic encephalopathy (AHE) is typically characterised by a wide range of neuropsychiatric manifestations including behavioural, cognitive, or mood changes associated with asterixis and different degrees of alteration to the level of consciousness, which may progress to stupor or coma in some cases.[1] This situation may be due to acute liver failure, cirrhosis, portal hypertension, or a transjugular intrahepatic portosystemic shunt.[2,3] Brain magnetic resonance imaging (MRI) of patients with AHE may reveal signal alterations in different brain areas, caused by vasogenic and cytotoxic oedema related to the toxic effect of ammonia in the brain.[4,5] These alterations may disappear after resolution of the encephalopathy,[5] or may progress to cortical laminar necrosis in cases with poor outcomes.[6] Presentations with focal manifestations are very infrequent in AHE, and may lead to diagnostic errors.

Clinical case. The patient was a 50-year-old man who was diagnosed in 2002 with portal vein thrombosis secondary to protein S deficiency, which was treated with a transjugular intrahepatic portosystemic shunt (TIPS) and oral anticoagulation. He was admitted to another hospital in May 2015 due to a sudden loss of consciousness which progressively and spontaneously improved over the following hours. The patient was transferred to our hospital to rule out the possibility of cerebrovascular accident, since he presented difficulties speaking after recovering a normal level of consciousness. During the neurological examination at admission, the patient was awake and presented mild asterixis in both hands; the most striking finding, however, was the presence of mixed aphasia characterised by hypofluent speech with presence of frequent phonological paraphasias in spontaneous language and difficulty understanding complex commands. Word repetition was preserved, although he showed difficulties repeating simple sentences. The neurological examination yielded otherwise normal results. The blood test revealed chronic anaemia, altered liver profile (ASAT 58 IU/L, total bilirubin 3.3 mg/dL) and moderately elevated levels of ammonia (91 μmol/L). The peritoneal fluid analysis and a brain computed tomography showed no abnormalities. However, the brain MRI showed altered high intensity signal on the fast fluid-attenuated inversion recovery (FLAIR), T2-weighted, and diffusion-weighted imaging (DWI) sequences in the left frontal parasagittal, insular, temporal, and cingulate cortices, as well as the right insular cortex, with no abnormal diffusion restriction in the apparent diffusion coefficient (ADC) map (Fig. 1A). The MR-angiography showed permeability of the intra- and extracranial vessels. An EEG study revealed presence of a delta rhythm with bi- and triphasic waves suggestive of toxic-metabolic dysfunction, with no associated epileptiform activity. We finally diagnosed the patient with acute aphasia as a manifestation of AHE associated with multifocal cortical brain lesions, predominantly involving the left insular and temporal cortices, and in the context of a TIPS. The patient received cathartic therapy, with aphasia progressively improving and resolving several weeks later. An additional MRI scan performed 2 months later showed that the cortical lesions had completely disappeared (Fig. 1B).

Discussion. Few cases have been described of patients with TIPS who, in the context of AHE, developed focal neurological signs associated with presence of cortical lesions in neuroimaging.[7–9] Two of the patients described in the literature presented sudden loss of consciousness with posterior aphasia, with brain MRI scans showing cortical lesions with a similar location to that described in our patient.

Correlation of the level of ammonia with the type of manifestation and the clinical severity of encephalopathy is a controversial issue.[10] In our case, as in that reported by Babington et al.[7], blood ammonia levels increased only moderately. This suggests that the pathophysiology of these pseudo-ictal discharges may involve factors other than ammonia, such as inflammatory mechanisms or other potential toxins,[1,11] the individual susceptibility of the brain to the toxic effect of ammonia, or the rate of increase of ammonia levels in the brain, favoured by TIPS.

Diagnosis of AHE in case of acute focal symptoms in patients with liver disease or TIPS is important since it may have prognostic consequences. It is well known that

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