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An exceptional cause of sudden neurological deterioration and coma☆

Una causa excepcional de deterioro neurológico repentino y coma

Dear Editor:

We are writing about an unusual case, to our knowledge not previously reported, of sudden neurological deterioration and coma.

The case is that of a 25-year old woman who had given birth 3 months before after a managed full-term pregnancy without complications. She was treated with oral iron due to postpartum anaemia but had no other relevant medical history. This patient arrived at the ER presenting with a one-month history of headaches, apathy, and bradypsychia. The headaches had worsened in the last 48 hours with incomplete response to conventional analgesic treatment. Nausea and vomiting had appeared in the last few hours. She had no fever. While in the ER she had a sudden deterioration in level of consciousness together with a tonic—clonic seizure. She was intubated. An emergent CT scan showed signs of diffuse cerebral oedema and obliterated subarachnoid cisterns. Leptomeningeal and ependymal enhancement was present (Fig. 1A and B). These findings were suggestive of cerebritis accompanied by meningeal involvement and ventriculitis.

She was admitted to the ICU and 6 hours later suddenly developed non-reactive bilateral mydriasis. A decompressive wide bifrontal craniectomy with bilateral decompression of the frontal and temporal lobes was carried out as a compassionate treatment, and a biopsy of the right frontal lobe was performed. During the intervention, we observed the brain to be congested and of a hard consistency. After the procedure an intracranial pressure sensor was placed which initially recorded pressures below 15 mmHg. After intervention mydriasis was reversed and pupillary reflexes were restored. CSF analysis showed no cytochemical abnormalities and the results from the CSF culture were negative for bacteria, viruses, and fungi. The results of blood serology tests for autoimmune diseases were also negative. A brain MRI showed supra and infratentorial diffuse involvement, especially in the frontal lobes and corpus callosum. Small areas of contrast enhancement and signs of intracranial hypertension were also observed (Fig. 1C—F).

Clinical and radiological differential diagnosis included infectious entities such as progressive multifocal leukoencephalopathy and other forms of viral encephalitis; autoimmune diseases such as acute disseminated encephalomyelitis, vasculitis or connective tissue diseases; metabolic

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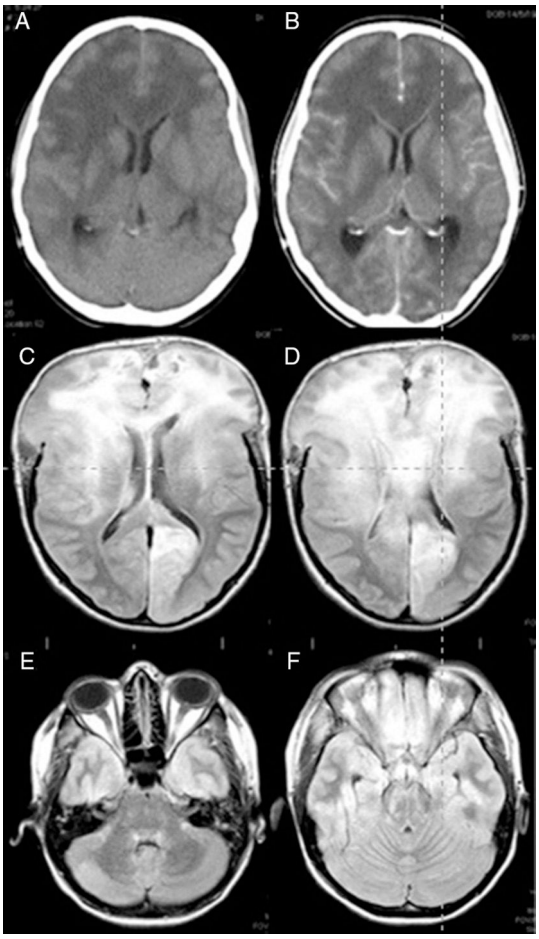

Figure 1 Brain CT (A and B) and FLAIR MRI (C—F) images. (A) Predominant presence of diffuse white matter hypodensity in both frontal lobes and involvement of the corpus callosum indicating vasogenic oedema. Convexity sulci effacement is seen. (B) Image was taken after the administration of contrast, showing no focal enhancement. (C) Signs of intracranial hypertension involving both frontal lobes and the corpus callosum are present. (D) Brain herniation persists despite extensive bilateral frontal craniectomy. (E and F) Infratentorial diffuse involvement is also present.

disorders such as certain forms of leukodystrophy; encephalopathy after radiotherapy; and such tumours as gliomatosis cerebri or primary brain lymphoma.[1,2]

Histopathological examination revealed diffuse astrocyte proliferation and low cell density with features of a low-grade glial tumour (Fig. 2). These findings added to those from the MRI were consistent with gliomatosis cerebri.

Forty eight hours after surgery the patient experienced an increase in intracranial pressure refractory to treatment. After obtaining family consent, a decision was made to limit treatment. The patient died 6 days after surgery.

Gliomatosis cerebri was first described by Nevin in 1938[1] and represents just under 1% of all astrocytomas.[3,4] It is a neoplastic disorder originating from glial cells,[3,5,6] defined by the infiltration of at least 2 lobes.[1,3,6] Despite this fact, gliomatosis cerebri typically preserves the macroscopic structure and cytoarchitecture of the CNS.[3,6]

Although this condition has been reported in children,[7] it usually occurs in patients aged between 40 and 50.[6] Its incidence appears to be slightly higher in men.[3] It is mainly located in the supratentorial level, but it often spreads to infratentorial structures.[8—10] The corpus callosum, thalamus, and basal ganglia are frequently involved.[6] In addition, expansion to the entire neuraxis has been reported.[8]

This disease is often overlooked in the early stages, so diagnosis commonly occurs in the advanced stages.[7] Common initial symptoms include the appearance of focal neurological deficits or the presence of less specific signs such as headache, nausea, vomiting, seizures, personality changes, or cognitive impairment.[5]

Neuroimaging findings of gliomatosis cerebri are characteristic but rarely specific.[9,10] This implies the need for a broad differential diagnosis. T2 and FLAIR MRI sequences usually reveal hyperintense areas with asymmetric and/or heterogeneous distribution.[9,10] The corpus callosum is usually involved and appears thickened.[9,10] Loss of differentiation between grey and white matter is also characteristic.[9,10] In type I gliomatosis cerebri there is usually no contrast enhancement, while type II commonly displays areas with contrast enhancement that correlate with anaplastic transformation.[9,10] Spectroscopy and sequences of relative cerebral blood volume can help in recognising the glial origin of this tumour.[3] These sequences can also be useful to more accurately determine areas for biopsy.[3]

Histopathological features often correspond to low-grade glial tumours,[5,6] although tumour progression showing high grade features can take place.[5,6] The most common cellular phenotype is astrocytic, but it may also present oligoastrocytic or oligodendrocytic phenotype.[5] Despite its histological appearance the clinical behaviour of this tumour correlates to at least grade III in the WHO classification.[6]

A definitive diagnosis is made with neuroimaging and histopathological features.[3,6] Nevertheless heterogeneity in histological findings may hamper the diagnosis.[3]

Because glioma are highly infiltrative and diffuse, the role of surgery is limited to biopsy for diagnostic purposes.[3,4] The mainstays of treatment are radiotherapy[11,12] and chemotherapy.[13,14] Even so, median survival time is estimated at around 14.5—18 months.[14,15] Prognostic factors are similar to those of other gliomas.[3]

A dismal clinical presentation of gliomatosis cerebri is featured in the current case. To our knowledge, intracranial hypertension syndrome with coma refractory to treatment resulting in fatality had not previously been reported as a presentation of gliomatosis cerebri.

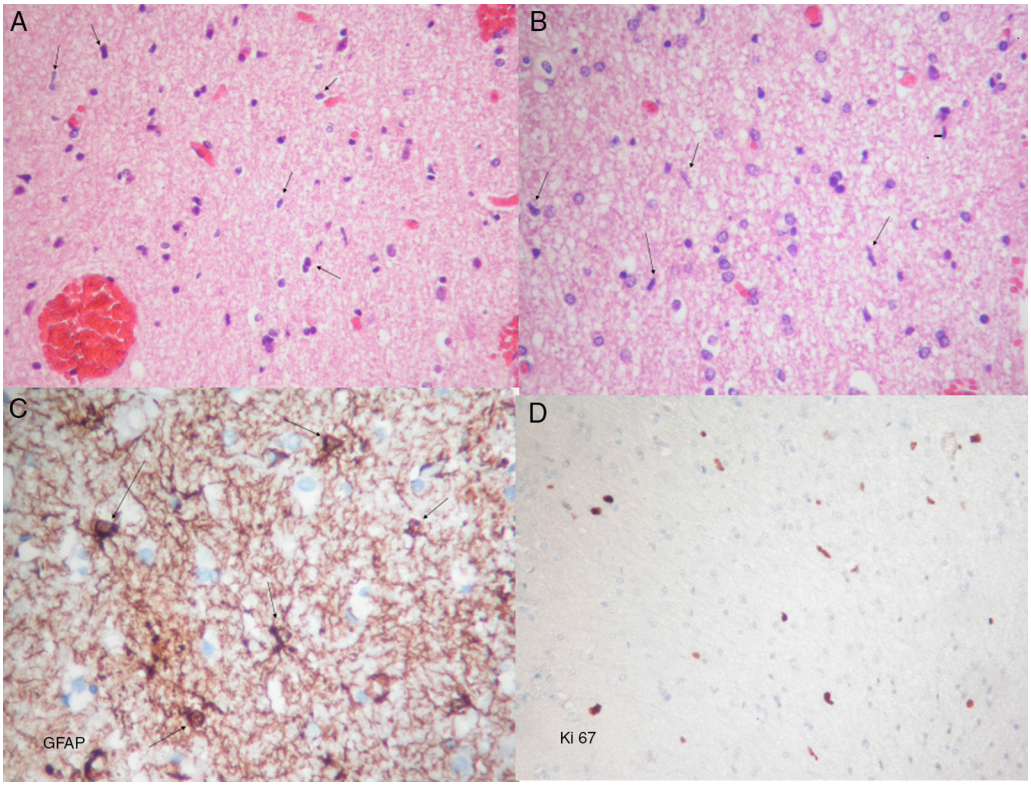

Figure 2 Photomicrographs showing frozen and permanent sections. (A) Haematoxylin—eosin stain. Proliferation of moderate cellularity characterised by the presence of naked nuclei (arrows) can be observed. (B) Haematoxylin—eosin stain. Oligodendroglial lineage cells are abundant. Glial elements composed of bare spindle-shaped moderately atypical nuclei can be seen (arrows). (C) Immunohistochemical GFAP (glial fibrillary acidic protein) staining showing atypical astrocytic elements with short irregular coarse extensions (arrows). (D) Ki67 staining showing proliferative activity around 2%. A, B, and D: original magnification 10×; C: original magnification 40×.

Conflicts of interest

The authors report no conflicts of interest concerning the materials or methods used in this study or the findings described in this paper.

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Safe thrombolysis in astrocytoma of middle cranial fossa☆

Trombólisis segura en astrocitoma de fosa craneal media

Dear Editor:

Some patients with acute ischaemic stroke occasionally present seizures at stroke onset.[1] Non-contrast brain computed tomography (CT) has some limitations in this context, and early signs of ischaemia are frequently subtle.[2] An epileptic seizure at stroke onset is considered a relative contraindication for intravenous recombinant tissue plasminogen activator (rt-PA) administration. The 2011 SEN stroke management guidelines[3] acknowledge that this should not justify ruling out thrombolytic treatment when cerebral infarction is confirmed by neuroimaging techniques. The 2013 American Heart Association/American Stroke Association guidelines deem intravenous thrombolysis appropriate given evidence that the residual deficits are secondary to ischaemia and not to a postictal event.[4]

We present the case of a 46-year old man with a history of hypercholesterolaemia and moderate alcohol consumption, who attended our emergency department due to speech disturbances and right-sided motor deficit of sudden onset. He arrived 45 minutes after symptom onset, presenting an arterial pressure of 168/92 mm Hg and a heart rate of 109 bpm. Auscultation was normal and the echocardiogram showed sinus rhythm. Neurological examination revealed global aphasia, right-sided hemiparesis of faciobrachial predominance, and right hemihypaesthesia. The National Institute of Health Stroke Scale (NIHSS) score was 15:2 in level of consciousness (LOC) questions, 2 in LOC commands, 1 in facial palsy, 4 in right motor arm, 2 in right motor leg, 1 in sensory, and 3 in language. One hour and 30 minutes after symptom onset, the patient experienced a generalised tonic-clonic seizure which resolved in 20 seconds after the intravenous administration of 10 mg diazepam. A non-contrast brain CT scan performed with a Siemens Somatom® Emotion eco (16-slice configuration) 15 minutes after the seizure yielded normal results (Fig. 1A). As we did not consider the patient's symptoms to be due to postictal changes, no change to the neurological deficits after the seizure was observed, and an advanced imaging scan could not be scheduled (time- and space-limited accessibility), we discussed the case and started treatment with intravenous alteplase, 2 hours and 15 minutes after symptom onset. Neurological improvement was observed (NIHSS 9:1 in LOC questions, 1 in LOC commands, 1 in facial palsy, 2 in right motor arm, 1 in right motor leg, 1 in sensory, and 2 in language). At 24 hours, we observed mild aphasia with no motor or sensory deficit (NIHSS 2); the contrast brain CT scan (indicated after the epileptic seizure at onset) performed at 24 hours showed a hyperdense area in the left temporal lobe (Fig. 1B). The echocardiogram and the carotid and vertebral echo-Doppler study yielded normal results. The brain MRI study (T1-, contrast T1-, T2-, T2*-, and diffusion-weighted, FLAIR sequences and apparent diffusion coefficient [ADC] maps) performed at 3 days revealed a glial tumour with cyst-like appearance in the left temporal lobe, extending to the uncus and hippocampus. It also revealed central necrosis with an infiltrative/expansive growth pattern (Fig. 1C-F). No haemorrhagic transformation or intratumoural bleeding was observed (Fig. 1C-F). Diffusion-weighted sequences and ADC maps did not show changes suggestive of cerebral ischaemia. At day 5 (with no previous corticosteroids or surgery), deficits resolved completely (NIHSS 0).

The patient was transferred to the neurosurgery department for tumour resection. Histological findings were compatible with grade III anaplastic astrocytoma.

There is limited scientific information on the use of alteplase in patients with astrocytomas mimicking stroke.[5] To our knowledge, there are only 2 published cases of patients treated with rt-PA due to suspected acute ischaemic stroke; both were finally diagnosed with glioblastoma multiforme.[6,7]

Our patient was symptomatic for more than 24 hours; however, no signs of lesions caused by cerebrovascular disease were observed. We believe that the stroke-like symptoms were caused by infiltration of the tumour into the Sylvian fissure, surrounding the left middle cerebral artery as in the case reported by García et al.[6] However, we cannot definitively rule out an ischaemia associated with the brain tumour and resolved by thrombolysis.[6]

Few cases of patients with epileptic seizures manifesting at stroke onset and safe use of rt-PA have been published,[6,8] and cases of patients with stroke-like conditions treated with thrombolysis and finally diagnosed as brain tumour are rare.[9] The Copenhagen Stroke Study suggests that an epileptic seizure at stroke onset may involve a large area of hypoperfused but potentially salvageable brain tissue.[10]

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