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Brain sagging syndrome, a potentially reversible cause of subacute ataxia and dementia. A case report

Brain sagging syndrome, una causa potencialmente reversible de ataxia y demencia. Reporte de un caso

Dear Editor:

Spontaneous hypotension is an infrequent, potentially fatal condition, usually related to a Cerebrospinal Fluid (CSF) leak producing intracranial hypotension (ICH), leading to a downward displacement of the brain known as ''brain sag''.[1]

Brain sagging syndrome (BSS) has recently been described[2,3] as clinically indistinguishable from behavioral variant frontotemporal dementia (Bv-FTD) but with neuroimaging findings suggestive of ICH,[3] and a potentially reversible course.

We report a 53-year-old male, without relevant medical history, who developed progressive bilateral tinnitus and orthostatic headache for six months, and later on: recurrent falls, behavioral changes (irritability, disinhibition, being socially inappropriate), delusional ideation and insomnia, progressively worsening over two months, requiring hospital admission.

On examination, relevant findings were cerebellar syndrome (four-limb dysmetria, gait ataxia with retropulsion, Video-A), kinetic tremor and severe cognitive decline (evaluated with Trail Making Test, Frontal Assessment Battery, Boston Naming Test, Complex Figure of Rey, etc.), consisting in dysexecutive frontal syndrome, severe behavioral impairment with apathy and impulsivity. Blood test resulted normal. CSF study showed low opening pressure (4 cm H_2O), without other alterations. Cranial-CT scan showed bilateral subdural hygromas. During hospital admission clinical condition worsens, presenting urinary incontinence, inability to walk, and severe behavioral impairment (quetiapine 50 mg is started with partial response), being dependent for all activities.

Given the rapidly progressive dementia and ataxia, we proposed the following differential diagnosis: Vascular etiology was ruled out given the progressive course and absence of focal symptoms. The patient did not fulfill 2018 diagnostic criteria for Creutzfeldt—Jakob disease given the absence of cardinal symptoms or radiological findings. Other neurodegenerative diseases were unlikely given the aggressive course and lack of typical radiological findings. Negative serologies (HIV, Whipple) and lack of fever ruled out infectious cause. Autoimmune etiology was also ruled out given the normal blood analysis (including onconeural antibodies) and absence of systemic manifestations.

Brain MRI showed descent of the cerebellar tonsils, transtentorial herniation, distortion of the brainstem structures and descent of the splenium of the corpus callosum (Fig. 1), findings suggestive of ICH. Spinal MRI demonstrated descent of the spinal cord, cisternography showed CSF leak point at D5 level.

Final diagnosis was rapidly progressive dementia (suggestive of Bv-FTD) and cerebellar ataxia, probably secondary to ICH, an entity described as BSS.

We treated ICH with an epidural blood patch, first lumbar (with transient improvement of gait), and later on at dorsal level, achieving progressive significant improvement, both radiologically (resolution of brain sagging, Fig. 1) and clinically, with resolution of ataxia (improvement of SARA scale from a score of 16—0) and recovery of cognitive-behavioral symptoms. Clinical response was maintained six months after discharge (Video-A).

The classic symptoms of ICH include orthostatic headache, tinnitus[4] and, in severe cases, coma and death.[1] In 2002,[5] Hong et al. reported a patient with ICH symp-

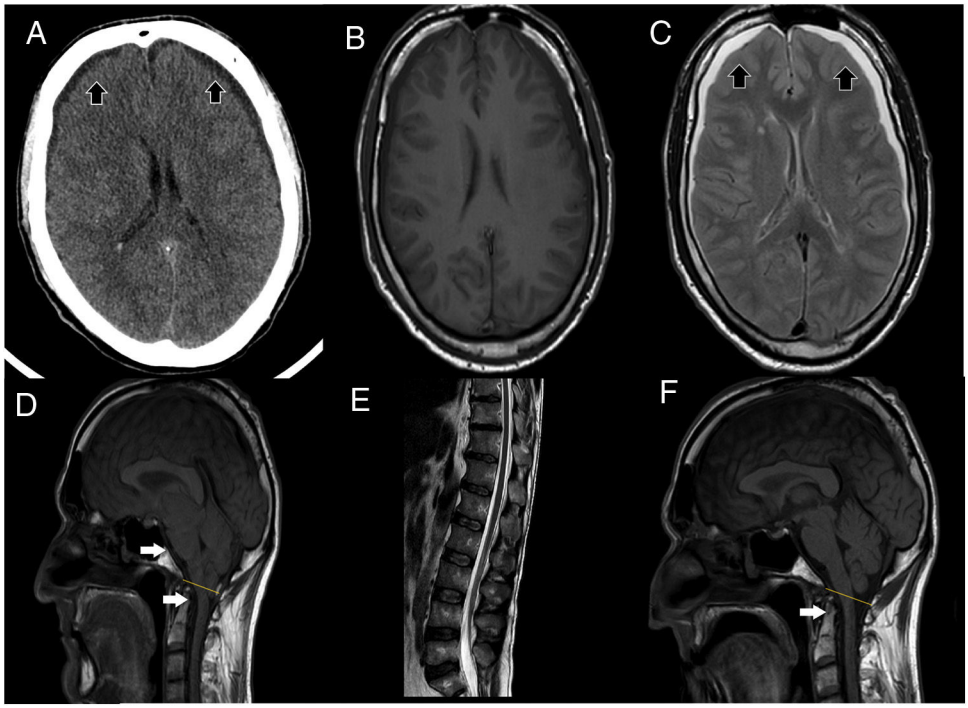

Figure 1 (A) Cranial CT scan shows bilateral subdural collections (as the black arrows point), without associated mass-effect, suggestive of subdural hygromas. (B, C) It stands out collapsed lateral ventricles, also bilateral subdural collections, hypointense in T1 (B), hyperintense in T2 (C) sequences (as the black arrows point), without enhancement after contrast administration, supporting the suspicion of subdural hygromas. (D) T1 weight sagittal sequence showed severe downward displacement of cerebellar tonsils (as the white arrows point) below the McRae line (represented by the yellow line), distortion of the brainstem structures and descent of the splenium of the corpus callosum. (E) Spinal MRI showing inferior and ventral displacement of the medullary cord. (F) Resolution of downward displacement of brainstem (as the white arrows point) and cerebellar structures (cerebellar tonsils are placed above the McRae line, represented by the yellow line). Resolution of the rest of the signs suggestive of brain sagging.

toms and cognitive deterioration suggestive of Bv-FTD, with neuroimaging signs of ICH, without frontotemporal atrophy. In 2011, Wicklund et al.[3] described 8 cases with ICH, similar cognitive impairment and other neurological symptoms such as tremor, chorea, dysphagia and gait disturbance, proposing the term BSS to describe this entity. Since then, different cases[1–3] have been reported, confirming the probable link between ICH and BSS. The few pathological studies in BSS diagnosed patients showed absence of typical Bv-FTD pathological findings.[3] The descent of the brain structures would exert traction on the frontal—temporal cortex, brain stem, cerebellum and their networks,[3] leading to dysfunction and the symptoms described. Why some ICH patients develop BSS remains unexplained, BSS could be an extreme presentation of ICH.

BSS affects predominantly middle-aged males (median 53 year).[3] Clinical features (usually with an insidious onset)[1] includes dementia (present in almost all cases, Bv-FTD profile),[4] cerebellar syndrome (66—75% of cases),[3,4] movement disorders[3,4] (tremor,[1] chorea[1,3]), sleep disorders,[3,4] urinary incontinence,[1] ICH classic symptoms (orthostatic headache, tinnitus)[4] and death.[1]

Most authors recommend prioritizing ICH treatment, either with a blood patch (multiple times if needed) or surgery of the CSF leak.[4] The most extensive series reported good functional recovery in treated patients (up to 72%).[4] Different symptomatic treatments (donepezil, quetiapine) have been tried,[1,3,6] with diverse results. More studies are needed to draw more weighty conclusions in this regard.

Our case highlights the importance of suspecting BSS in patients presenting subacute ataxia and dementia, especially with another accompanying neurological symptoms (like movement and sleep disorders). In the case described, it stands out that repetitive epidural blood patches were needed, with a suboptimal response the first time. Neuroimaging studies should be reviewed in detail looking for characteristic BSS findings, as it is a rare but potentially reversible entity, with a fatal course in the absence of treatment.[2]

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.nrl.2022.04.002.

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Position is important: retrograde air embolism after central venous catheter removal

La posición importa: embolismo aéreo retrógrado tras retirada de catéter venoso central

Dear Editor:

Retrograde venous air embolism (RVAE) occurs when central venous pressure (CVP) is lower than atmospheric pressure, as is the case with deep inhalation, vertical positions above 45°, and hypovolaemia. The pressure gradient favours the entry of air into venous circulation, travelling to the right ventricle and pulmonary artery, and potentially even leading to an obstructive shock and right ventricular dysfunction.[1,2] Some studies show that the air may retrogradely ascend to the cerebral venous circulation when the patient is in a vertical position, due to the lower specific weight of air in comparison with blood. This phenomenon will depend on the size of the bubble, the diameter of the vein, and the patient's cardiac output.[3,4] Causes of RVAE include trauma, vascular surgery, diving, barotrauma due to mechanical ventilation, and insertion and extraction of central venous catheters. Incidence is difficult to determine, ranging from 1.6% to 55.3%; it is an underestimated entity due to the difficulty of establishing a diagnosis, which requires presence of a known risk factor, compatible clinical signs, no right-to-left shunting in the echocardiography, and imaging studies showing the presence of air in the intravascular space. The most frequent neurological complications are altered level of consciousness, coma, stroke, and seizures.[5] Patients may also present haemodynamic and respiratory alterations including dyspnoea, tachypnoea, chest pain, arterial hypotension, low cardiac output, and even obstructive shock and cardiorespiratory arrest. Electrocardiographic alterations include sinus tachycardia, right ventricular overload signs, non-specific changes in the ST segment/T-wave, and elevated markers of myocardial damage. Definitive diagnosis is established by head CT scan revealing air bubbles in the cerebral intravascular space and parenchyma, sometimes accompanied by diffuse cerebral oedema. In addition to symptomatic treatment with volume therapy, treatment for RVAE includes vasoactive amines, antiepileptics, oxygen therapy with high FiO_2, and placing the patient in the left lateral decubitus position (Durant manoeuvre) or the Trendelenburg position. Hyperbaric oxygen therapy may be considered in severe cases.[6]

Patient 1

Our first patient was a 77-year-old man who was admitted due to perforated sigmoid diverticulitis. The patient had a central venous catheter in the right jugular vein, which was removed with the patient in a seated position. Immediately after removal, he presented arterial hypotension and decreased level of consciousness with spontaneous opening of the eyes, fixed gaze and inability to follow commands, and pain with left hemiparesis. A head CT scan revealed air bubbles in the cavernous sinuses and basal cisterns but no other alterations (Fig. 1A). An electrocardiography (ECG) study showed ST-segment elevation in precordial leads and negative T-wave in leads V5, V6, I, and aVL (Fig. 1B), with elevated markers of myocardial damage. He was transferred to the intensive care unit (ICU) a few hours late due to a generalised tonic-clonic seizure; we ruled out toxic, metabolic, and infectious aetiology. A transthoracic echocardiography study showed no atrial septal defect. We started treatment with fluid replacement, oxygen therapy using a high-flow mask, and antiepileptics, which led to favourable progres-

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