Paradoxical herniation due to a continuous cerebrospinal fluid drain in a previously craniectomised patient

Herniación paradójica secundaria a drenaje continuo de líquido cefalorraquídeo en un paciente previamente craniectomizado

Dear Editor:

The two large intracranial compartments divided by the tentorium normally maintain a hydrostatic pressure balance that causes the brain to float inside the skull. Sometimes this balance is lost by exposure of one of the components to atmospheric pressure[1]. In a craniectomy, a negative pressure gradient is created between the intracranial and atmospheric compartments (temporarily or permanently), which makes a cerebral herniation possible even in the absence of increased intracranial pressure. This process is perpetuated by an additional change in the spinal cord compartment, which underlies the cerebrospinal fluid (CSF) compartment; for example after a lumbar puncture or in some cases after continuous CSF lumbar drainage[2]. This description shows a severe complication that can be associated with lumbar drainage after craniectomy. We present a patient with rostrocaudal deterioration attributable to alterations in CSF dynamics that developed 12 h after placing CSF lumbar drainage in a previously craniectomised patient.

A 39-year-old man was admitted due to right temporo-parietal intracerebral haemorrhage of 160 ml with large mass effect, attributable to hypertension after renal transplantation. Neurological examination included stupor, papilloedema and dilated fixed right pupil, with left hemiparesis. He underwent a decompressive right parietal craniectomy, with evacuation of the clot, and gradual improvement was obtained. The patient was admitted to a nephrology unit 3 days after the craniectomy. High blood pressure was adequately controlled and the evolution was satisfactory. However, 15 days after the craniectomy, the patient developed low grade fever and drowsiness; the computed tomography (CT) showed a hypodense image in the surgical field. He underwent needle aspiration of the lesion and was started on broad-spectrum antimicrobial therapy. Gram staining was negative for microorganisms. The clinical condition of the patient improved partially; however, a drain of CSF through the surgical wound became clear; a continuous lumbar CSF drainage was placed in addition to medical treatment with acetazolamide. The intradural catheter was placed correctly after two attempts in lateral position and the opening pressure was reported only as "elevated". CSF cytochemical analysis showed: leukocytes, 6/μl; glucose, 38mg/dl; proteins, 35mg/dl. Twelve hours after the placement of the drain, the patient suddenly developed drowsiness that progressed to coma and non-reactive pinpoint pupils with decerebrate posture. The patient was intubated and hyperventilated, administration of mannitol was initiated and a new CT scan was obtained. He was then transferred to the intensive care unit. A new CT scan showed marked brain concavity at the site of the craniectomy with displacement of the midline structures and pneumocephalus (fig. 1). Given the concavity of brain tissue with significant displacement of the midline and distortion of the brainstem, with signs of rostrocaudal deterioration in pontine phase, an aggressive treatment with fluid administration was started; the lumbar drain was closed and the patient was placed in 0° position. The patient improved neurologically; he regained alertness and moved his right limbs spontaneously. However, he developed ventilation-related pneumonia, sepsis and systemic inflammatory response syndrome, which led to gradual deterioration and death in 15 days. In a new CT scan obtained 5 days before death, no displacement of midline structures was observed.

CSF hypovolaemia is a well-known clinical entity that often appears after lumbar puncture, continuous lumbar drainage, traumatic skull fractures and surgical procedures in which the dura mater is opened. Such CSF alterations are often expressed through mild manifestations (postural headache, vertigo and nausea). These symptoms are associated with downward traction of pain-sensitive structures and loosening of chemotactic areas; all these manifestations are benign and self-limited[3]. However, when CSF volume is reduced, it can reach a certain critical degree of hypovolaemia. This condition, which was first described in patients after craniectomy in microsurgery or aneurysm stapling, has been called "brain sag" or "sinking brain syndrome"[4]. The Monro-Kellie hypothesis defines the relationships between the 3 cranial volumes: brain tissue, blood and CSF[5]. These close relationships take place within a closed system. Under normal conditions, a CSF leakage is quickly compensated by an increase in intracranial venous volume, thus maintaining the physical support for the brain intact. When this system is "open" to atmospheric pressure, the Monro-Kellie principles do not apply and a loss in the CSF compartment leads to serious difficulties in maintaining the brain afloat. This makes possible a position dependent on cerebral herniation with subsequent neurological impairment[2] or the appearance of a focal neurological deficit[6].

Apparently, the key concept for the development of brain herniation in this context is the emergence of a pressure gradient between the cranial and spinal compartments. This may explain the neurological deterioration reported in lumbar drainages, but not in ventricular drainages, which are much more common procedures[2,7,8].

Severe hypovolaemia after craniotomy has been widely described. It produces a dramatic herniation syndrome that in most cases is reversible by placing the patient in a head-down position[9]. The literature advises that this cerebral collapse syndrome[7] should be included in the differential diagnosis of acute postoperative clinical deterioration in craniectomised patients. Specifically, in patients with aneurysmal subarachnoid haemorrhage, it has been associated with CSF hypovolaemia with general oedema in CT upon admission and with prolonged surgical time; severe hypovolaemia is therefore associated with increased morbidity and mortality[9].

However, the present case illustrates a case rarely described in clinical literature, a severe neurological deterioration after a therapeutic lumbar drainage in a previously craniectomised patient. Schwab et al.[1] reported

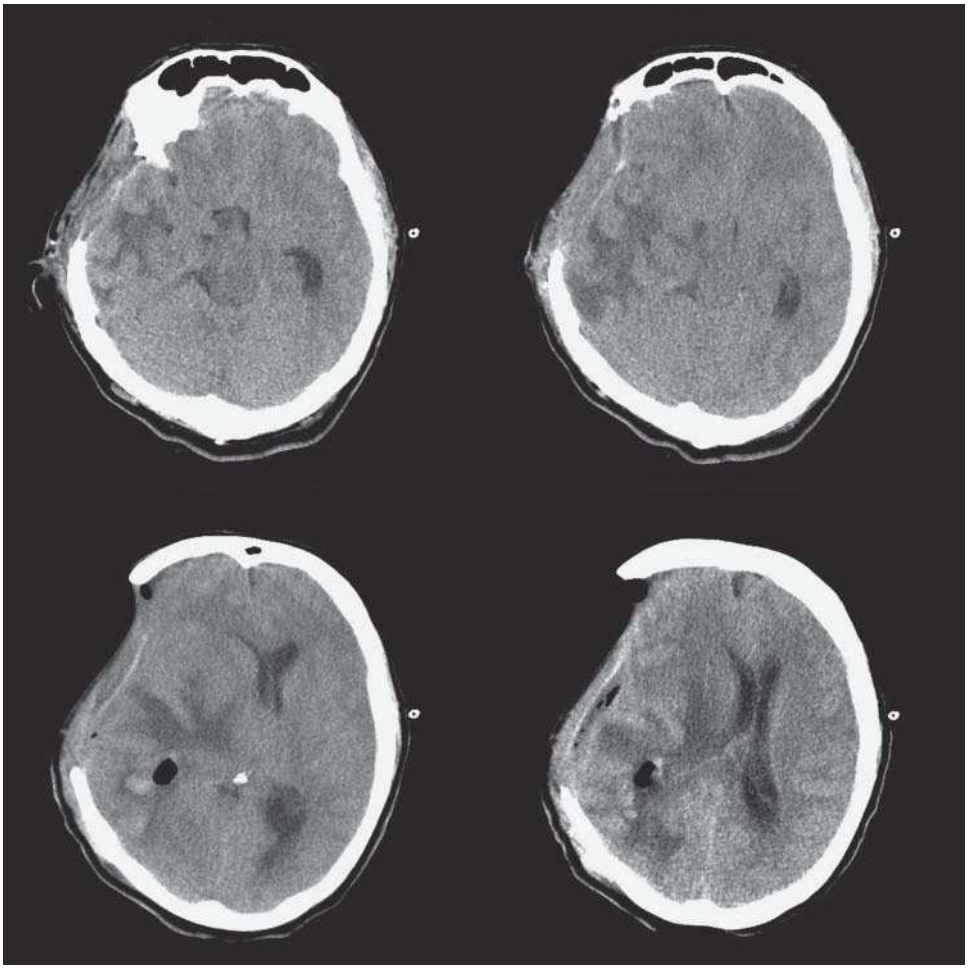

Figure 1 The computerised tomography scan shows the marked brain concavity at the site of the craniotomy with displacement of midline structures and pneumocephalus.

four cases with similar clinical and imaging characteristics and suggested the term "paradoxical herniation" to describe this syndrome. The four patients developed herniation after a lumbar puncture, but the authors postulated a hidden continuous CSF drainage with an increase of the pressure gradient between atmospheric and intracranial pressure. That caused the paradoxical herniation syndrome, which may explain the development of the syndrome in the patient described here.

According to the few descriptions available in the literature and to this case, the features that may help to recognise a paradoxical herniation are: *a*) clinical: temporal association with a CSF drainage procedure and marked concavity at the craniectomy site, and *b*) imaging: subfalcine, uncal or transtentorial herniation and presence of pneumocephalus.

Since the indications for decompressive craniectomy are increasing, and they are often used by neurosurgeons and neurologists, it is of great importance to be aware of and recognise the clinical characteristics of paradoxical herniation and perhaps include large cranial defects among the relative contraindications for lumbar puncture and continuous lumbar drainage. Given the absolute necessity of such procedures, the lumbar puncture should be performed positioning the head downwards and obtaining the minimum possible amount of CSF. The immediate measures in case of damage include therapy with fluids and the flat (0°) or head-down position (Trendelenburg). The role of an early "emergency" cranioplasty is still controversial, particularly in patients with rapid deterioration. Finally, it is obvious that the current understanding of CSF dynamics between intracranial compartments is limited and there is also a great need to develop experimental models that take into account parameters such as the anatomy of the subarachnoid cisterns and arachnoid or Pacchioni granulations, CSF viscosity, meningeal membrane elasticity and the presence of bridging veins. The available models are too simple[10] and cannot explain pathological processes such as the one reviewed in this case.

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