Letter to the editor

Syndrome of the trephined, a case report

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1. Introduction

Syndrome of the trephined also called “sinking skin flap syndrome” is a rare and late complication of the craniectomy. It appears in the weeks or months (3 months in average) after the surgery and is characterized by a neurological deterioration, not explained by other etiologies. This syndrome also associates various symptoms such as headaches, motor impairments, cognitive disorders, alertness disorders and also in some cases autonomic dysfunction. This clinical picture can be aggravated by orthostatism and dehydration. A characteristic neurological improvement of the patient’s neurological state after having replaced the skull bone flap will validate the diagnosis [1–5].

2. Observation

A 76-year-old male came to the emergency room presenting with intense holocranial headaches (pain VAS 9/10) for the past week, with no notion of recent trauma. A brain CT-scan performed as an emergency procedure (Fig. 1A) evidenced a bilateral subdural hematoma with a mass effect (subfalcine and left temporal cerebral herniation).

In the patient medical history, we noted an infarct in the right middle cerebral artery, 10 years earlier during surgical management for descending thoracic aorta replacement, which left him with an aphasia characterized mostly by speech disorders.

Two days later, a first drainage was performed by our neurosurgical team via a right fronto-parieto-temporal bone flap approach.

Postoperative care is characterized by a deterioration of the neurological state (sleepiness) in immediate postoperative care, evidenced by a CT-scan (Fig. 1B) which unveiled the recurrence of the subdural hematoma.

The next day a second drainage was performed, via the same surgical approach, with transfer to the intensive care unit, where the patient was monitored for 9 days.

After 45 days, the patient was transferred to our neurological rehabilitation center. Upon admission, the clinical evaluation showed major functional deficits with quadriplegiasis globally scored at 4/5 on the Medical Research Council (MRC) Scale, static gait disorders, and swallowing disorders requiring the placement of a gastrostomy tube (G-tube). Furthermore, the patient’s state of consciousness was preserved with a good understanding of simple orders.

Two and a half months after his admission to the emergency room, he presented with fever and frontal swelling on the right side. A CT-scan performed the same day showed a cerebral empyema right below the surgical site (right fronto-parieto-temporal bone flap). The bone flap was removed and replaced simultaneously during the drainage procedures for the bilateral subdural hematoma. An immediate neurosurgical management of the empyema was performed by removing the bone flap and cement, as well as draining and profusely rinsing the infected area.

A long term IV antibiotic therapy was implemented by the infectious disease team (6 weeks) following the positive result of the cerebrospinal fluid to a multisensitive Staphylococcus aureus.

Following this multidisciplinary care in our neurological rehabilitation center, the patient showed an improvement of his static disorders and quadriplegiasis (4/5 on the MRC). G-tube feeding was continued due to the severe swallowing disorders.

Two months after removing the skull bone flap, the patient presented with a sudden deterioration (< 24 hours) of his neurological state characterized by a state of sleepiness, aggravation of the left hemispheres globally evaluated at 2/5 on the MRC (previously at 4/5) in a left-handed patient and major balance disorders. Blood tests and infectious results were reassuring. An emergency CT-scan showed a retraction of the cerebral parenchyma with loss of right hemispheric convexity. An NMRI (Fig. 2) performed three days later did not evidence any acute phenomenon that could explain the patient’s symptoms. The NMRI showed the loss of right hemispheric convexity with retraction of the skin flap (Fig. 2A), compression of the right superior frontal sulcus and compression of the right hemisphere between the skin flap and the falx cerebri (Fig. 2B).

Based on the clinical and imaging data, the neurosurgical and infectious disease teams agreed to replace the bone flap.

Right from the next day after surgery, the patient showed a clear neurological improvement, improved left hemiparesis and balanced disorders; he recovered the neurological status he had prior to his clinical deterioration.

An NMRI evaluation (Fig. 3), performed two months after the bone flap surgery, showed the reappearance of the right hemispheric convexity and sulci, thus underlining the disappearance of the mechanical constraint on the cerebral parenchyma.

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3. Discussion

Decompressive craniectomy is usually performed in the surgical management of intracranial hypertension secondary to diverse causes such as traumatic brain injury, ischemic stroke, subarachnoid hemorrhage or infection. Even though this surgery is technically simple to perform, it is not without risks. There are several complications to this procedure such as: mortality, hernia, subdural effusion, infection, hydrocephalus, epileptic seizures and syndrome of the trephined [6].

The syndrome of the trephined was described for the first time by Grant and Norcross in 1939, who evidenced a series of symptoms such as headaches, epileptic seizures, mood swings and behavioral disorders in patients with a large bone flap. In a similar manner, Yamaura and Makino in 1977 described in patients a sudden improvement of neurological deficits after cranioplasty and invented the term “syndrome of the sinking scalp flap” [5–7].

The syndrome of the trephined or its synonym the “syndrome of the sinking scalp flap” describes a neurological deficit due to a cortical dysfunction caused by brain herniation under the bone flap. On a clinical level, this syndrome most often associates motor, cognitive or alertness disorders, headaches and in rare cases dysautonomia. Several researchers have tried to explain the complex pathophysiology of this phenomenon and several factors were identified as being related to this syndrome. Some authors [2,3,5–7,9] promote the theory that atmospheric pressure would be directly transmitted to the brain structures in the absence of the bone flap. It would be responsible for the internal displacement of

Fig. 1. A. Initial status showing a “mixed” bilateral subdural hematoma with fresh blood (hyperintense signal) and older blood (hypointense signal). The hematoma is thicker on the right side because of the parenchymal “emptiness” caused by the sequelae of a right-side ischemia in the sylvian fissure (stroke that occurred in 2006). There is median diencephalic herniation (not illustrated on the picture). B. Status after the first surgical procedure showing the right hemispheric craniectomy and the effective drainage of the peripheral layers of a partitioned hematoma where the deep layers are quite important (arrow). C. Status after the second surgical procedure showing an effective drainage of the entire right hematoma.

Fig. 2. NMRI evaluation after retraction of the skin flap: T2WI slides. A. Cross-sectional view via the vertex showing the retraction of the skin flap and the compression of the right superior frontal sulcus with disappearance of the hyperintense signal of the CSF (arrows). B. Coronal view showing the compression of the right hemispheric parenchyma between the skin flap on the outside and the falk cerebri on the inside (between the arrows). Let us note that no compressive suffering was visualized in diffusion-weighted imaging at the level of the compressed parenchyma (not illustrated).
the scalp as well as dysfunction of the cerebral blood flow and/or cerebrospinal fluid (CSF) locally at the craniectomy site.

Patients with small-surface craniectomy are usually older with a greater infarct volume thus more sensitive to atmospheric pressure. The hypothesis being that major cerebral atrophy after a stroke would contribute to reduce brain volume, and consequently intracranial pressure. Furthermore, a smaller-size bone flap could lead to secondary lesions because of an inadequate hemispheric decompression in regards to the width of the hemispheric lesion [10]. More recently, other authors [5,8,9] proposed the idea that the negative pressure gradient between atmospheric pressure and intracranial pressure is aggravated by the decreased pressure of the CSF related to its hypovolemia. Thus, drainage of CSF in patients with hydrocephalus or meningitis would imply an increase in negative pressure gradient through the craniectomy site and would promote brain distortion. Prolonged dehydration and an upright position could precipitate this phenomenon. Finally, authors using dynamic CT-scan and xenon-enhanced CT-scan to quantify this phenomenon [7–9] were able to highlight increased cerebral blood flow after cranioplasty not only on the side of the craniotomy site but also on the contralateral side of the bone flap.

The rate of patients affected by this syndrome is difficult to determine since many of them are in the neurological rehabilitation phase following major brain lesions. The onset of the syndrome is usually delayed in the weeks or months (three months in average) following the craniectomy [7].

The main treatment for the syndrome of the trephined consists of replacing the bone flap or doing a cranioplasty if necessary, as soon as possible. Its objective is to restore the pressure exerted by the depressed craniectomy site. If there is no immediate neurological clinical improvement, in spite of bone flap replacement or cranioplasty, the diagnosis should be reassessed [1–7].

The prevention of various complications related to craniectomy, in the context of the syndrome of the trephined, would also consist in a quick replacement of the bone flap or a cranioplasty, ideally in the first 2 to 3 months. In this framework, an early neurosurgical consultation could be proposed to our patients. Other authors [DECIMAL study] also promoted the relevance of initially performing a wide bone flap, which presents a lesser risk of developing the syndrome of the trephined [10–12].

4. Conclusion

The syndrome of the trephined is a rare and delayed complication of craniectomy. It should be considered in patients with a bone flap, especially in the context of infections or hydrocephalus, presenting with neurological deterioration unexplained by other causes. The diagnosis will be validated by the quick improvement of the patients’ neurological state after bone flap replacement or cranioplasty.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


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