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Case Report
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Syndrome of the Trephined: When Bone Becomes the Cure

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Abstract

Syndrome of the trephined may develop after decompressive craniectomy: It is an underestimated cause of neurologic deterioration which may appear either a few days or several years after decompressive craniectomy. We report on the case history of a 44-year-old patient who developed a syndrome of the trephined. The patient underwent a large left decompressive hemicraniectomy and evacuation of an acute subdural hematoma after traumatic brain injury. After his transfer to rehabilitation, he developed as a secondary reaction a right hemiplegia, and aphasia. The patient achieved complete recovery from his motor and speech deficits after cranioplasty. We discuss the literature and clinical implications regarding this syndrome which is a treatable complication of decompressive craniectomy. Physicians must be aware of this singularity in order to propose early cranioplasty whenever possible.

Keywords: Decompressive craniectomy; Sinking skin flap syndrome; Syndrome of the trephined, Traumatic brain injury

Introduction

Decompressive Craniectomy (DC) is used to treat intracranial hypertension when medical treatments fail to Lower Intracranial Pressure (ICP). This procedure has many known complications such as hemorrhage, external cerebral herniation, infection, seizure, Cerebrospinal Fluid (CSF) leak, wound complications, hydrocephalus and subdural hygroma [1]. Syndrome of the Trephined (ST) may also develop after DC: It is an underestimated cause of neurologic deterioration which may appear either a few days or several years after DC. Therefore, this complication concerns not only neurosurgeons and critical care specialists, but also general practitioners, neurologists and rehabilitation specialists who are increasingly involved with these patients. We report on a case history of a patient who developed a syndrome of the trephined. The patient achieved complete recovery of his motor and speech deficits after cranioplasty. We discuss the literature and clinical implications regarding this syndrome which is a treatable complication of DC.

Case Presentation

A 44-year old male with a past medical history of Sotos syndrome (cerebral gigantism) was transferred to our hospital after traumatic brain injury. Clinical examination at presentation disclosed Glasgow Coma Scale of 8 and right-side convulsions. Computed Tomography (CT) scan of the head showed a left acute subdural hematoma and contusions of the left cerebral hemisphere. The patient underwent a large left decompressive hemicraniectomy and evacuation of the subdural hematoma. A synthetic duroplasty with Neuropatch® was performed to allow brain expansion and the craniectomy was implanted in the abdominal wall fat for future cranioplasty. The patient was then admitted to the intensive care unit for hemodynamic and intracranial pressure monitoring and neurosedation. CT scan on day 2 showed disappearance of the left subdural hematoma and moderate brain edema. He was extubated on day 11 and transferred to rehabilitation on day 16. At that time, examination disclosed a slight right hemiparesis, and speech was almost normal. After his transfer to rehabilitation, the patient developed a right-side hemiplegia, and aphasia. He was referred to our Department of Neurosurgery two weeks later. Examination showed both severe motor deficit of the right side and aphasia. The patient was only able to repeat simple words. The skin flap was totally depressed. When the patient was admitted to our department for cranioplasty on day 40, he had a complete hemiplegia and could only say “yes” or “no”. His craniectomy which was inserted in the abdominal wall was re-implanted and fixed with metal wires to close the cranial defect. There was no CSF leakage during surgery. When the patient woke up postoperatively, he was able to speak almost normally with only difficulties in finding some words. Hemiplegia resolved

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immediately so that he could walk without help the day after the operation. Three months later, the neurological examination showed complete recovery of motor deficit and only mild speech problems.

Discussion

The syndrome of the trephined was first described by Grant in 1939 as a constellation of symptoms including seizure, dizziness, headache, mental depression or discomfort at the site of the defect, resolving after replacement of the bony skull [2]. He therefore concluded that cranioplasty has indications beyond the closure of the defect. Yamaura and Makino used the term of "sinking skin flap syndrome" to describe the same clinical reality, considering the concave aspect of the skin covering the skull defect after large craniectomies [3]. Several studies then reported variable clinical presentations of the syndrome, motor weakness being the most frequent symptom followed by cognitive impairment and language disturbance, impaired vigilance, headache, seizure, cranial nerve defects, vertigo and mood disorder [4,5]. These symptoms are not specific and could easily be ascribed to traumatism-induced brain damage. Even if only the reversibility of the symptoms after cranioplasty could confirm their attribution to ST, some elements may draw the attention and evoke the diagnosis: Sensitivity to orthostatic position with worsening while upright has been described, especially for headaches [6] and visual impairment [7]. The average time interval between craniectomy and onset of symptoms was 5 months but a wide range of 3 days to 7 years has been described [4]. Finally, some cases of paradoxical herniation have been described after lumbar CSF drainage or puncture [8,9], not always associated with a large skin concavity [10], thus confirming patients with DC as being in a critical period. Those clinical findings give us some clues to understanding the pathophysiological mechanisms implicated in this syndrome. By removing a large part of the skull, DC decreases ICP but also alters CSF dynamics and decreases cerebral blood flow [11]. Sinking of the skin flap suggests a negative gradient between extra and intra cranial pressure, illustrated by a lower CSF pressure measured before cranioplasty and restored after it [3]. Dujovny et al. [12] reported significant changes in the oscillatory CSF flow after cranioplasty by using dynamic phase contrast MRI. Physiologic postural ICP changes between supine and upright position were lost after DC and restored after replacement of the skull [13]. Furthermore, it has been demonstrated that cranioplasty improves cerebral blood flow on CT perfusion images [14], both on operated and contralateral sides [15]. Glucose metabolism measured by ^{18}F-fluorodeoxyglucose positron emission tomography was also impaired after DC, thus contributing to cortical dysfunction, and improved after cranioplasty [16]. A recent study on murine showed that craniectomy-related glymphatic dysfunction was associated with an astrocytic and microglial inflammatory response, which could be the substratum of the development of motor and cognitive deficits [17]. Finally, atmospheric pressure, CSF and cerebral blood flow impairments seem to be interrelated in the micropathogenic upheaval after DC, thus leading to ST. Replacement of the bony skull is crucial to alleviating ST symptoms by restoring a normal intracranial physiology. Four days were reported as the mean time interval between cranioplasty and onset of improvement, and complete resolution of symptoms as in our case may then occur [4]. Timing for cranioplasty usually depends on personal experience and local habits rather than evidence-based data. However, some studies and systematic reviews have demonstrated that early cranioplasty (within 3 months) was associated with a better functional outcome and that similar complication rates were observed when compared to delayed cranioplasty [18-20]. These observations and the spectrum of the syndrome of the trephined both argued for an early bone repair.

Conclusion

The syndrome of the trephined is a rare but an astonishing clinical worsening after DC. Physicians concerned with patients who underwent DC for acute neurological history must be aware of this singularity in order to propose early cranioplasty whenever possible.

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