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Case Report

Unusual presentation in syndrome of trephined – A unique case observation

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ABSTRACT

Background: Syndrome of trephined (SoT) is a well-recognized complication of decompressive craniectomy (DC). The understanding of SoT has improved more recently with the increasing utilization of DC for severely raised intracranial pressure. It usually presents after a period of weeks or months with a cluster of adverse neurological symptoms, most commonly with worsening of motor strength.

Case Description: An elderly gentleman with traumatic brain injury underwent DC. He later developed a sinking flap and unexplained agitation which responded to cranioplasty by returning to a state of calm. His cognitive function further improved over a period of 6 months. This is an unusual observation reported in this case.

Conclusion: Timely recognition of the cognitive complications of craniectomy that may respond to early cranioplasty promises to decrease the length of hospital stay and enhance rehabilitation in such patients.

Keywords: Cognitive dysfunction, Cranioplasty, Decompressive craniectomy, Sinking flap syndrome, Syndrome of trephined

INTRODUCTION

Syndrome of trephined (SoT) is a delayed sequel of decompressive craniectomy (DC), or any other neurosurgical procedure involving removal of a part of the cranium, leaving the brain without adequate bony coverage, and presents with a symptomatic deterioration of clinical features that are independent of the location and improve after cranioplasty.[2] A number of clinical presentations have been reported in the literature including motor dysfunction, altered sensorium, headache, psychosomatic symptoms, seizures, tremors, cranial nerve deficits, and varying severity of cognitive or language-related problems.[2,13]

Appreciation of SoT has increased over time and its reported frequency has increased from an uncommon entity to up to 26% of decompressive craniectomies.[17] Motor symptoms develop in 57% of the patients diagnosed with SoT.[2] A purely cognitive SoT is rare and as motor deficits predominate, cognitive dysfunction is often overlooked. Carota *et al*. reported the first case of sinking skin flap syndrome with pure "dysexecutive" cognitive deterioration.[4]

Cognition includes language, memory, perception, recognition, reasoning, problem-solving, conceptualization, learning, and many other overlapping processes that define our knowledge

and judgment.[14] Agitation is a common manifestation of a number of cognitive and neuropsychiatric disorders.[7] It is widely studied in the context of psychiatric disorders although it is seen in neurological and medical conditions as well. According to a recent consensus by the International Psychogeriatric Association, agitation is defined as a state occurring in patients with cognitive impairment, exhibiting behavior consistent with emotional distress along with excessive motor activity, and verbal or physical aggression that causes excess disability not solely due to another disorder.[7]

Here, we present a patient who developed severe agitation 5 weeks after DC and markedly improved immediately after cranioplasty.

CASE

A 69-year-old hypertensive male was transferred to us with traumatic brain injury after sustaining a ground-level fall. On admission, his Glasgow Coma Scale was 12/15 (E4V2M6). CT scan depicted a left temporal hematoma with extensive acute subdural hematoma, diffuse edema and mass effect, and midline shift of 6.6 mm [Figure 1]. He was admitted to the special care unit for observation where his GCS dropped to 9/15. He then underwent left-sided emergent DC and was discharged 4 weeks later. At the time of discharge, he was awake, alert, and intermittently following simple commands without gross motor or sensory deficits. His speech was slurred and intermittently, he had mildly agitated behavior (Richmond Agitation Sedation Scale or RASS + 1) with a Modified Rankin Scale of 4.[15]

One week later, the patient returned to the hospital with progressively worsening agitation without a decline in consciousness. CT scan showed interval development of the right chronic subdural hematoma with interval resolution of the previous acute hematoma. The hematoma was evacuated through burr-hole craniotomy and drain placement. The procedure was uneventful but after the surgery, his agitation increased (RASS + 3) and cognitive dysfunction did not improve. Mini-Mental Score could not be assessed as his comprehension was not intact. At this point, patient was extensively evaluated for other causes of agitation such as electrolytes disturbances, endocrine abnormalities, infection, sepsis, drug intoxication, or alcohol withdrawal, all workup was within normal limits.

On serial CT scans, the right subdural collections decreased and his craniotomy skin flap started to sink. Suspecting "syndrome of trephined" and in view of his persistently unexplained behavioral dysfunction, we planned for early cranioplasty which was performed 6 weeks after the primary procedure [Figure 2]. Within 48 h of cranioplasty, his agitation started to improve. On the 4th day, his RASS was + 1.

At 6-month follow-up, the patient was found able to walk unattended and only required minimal assistance with his activities of daily living (ADL) (MRS 3). His comprehension had also improved considerably and he was able to follow two-step commands with consistency.

DISCUSSION

DC can be a life-saving procedure for certain neurosurgical emergencies such as traumatic brain injury and acute vascular occlusions. A recently published meta-analysis of RCTs for DC in traumatic brain injury found a mortality benefit over the best medical treatment.[10] Similar mortality benefit related to supratentorial as well as infratentorial brain infarcts following DC is available in the literature but the role

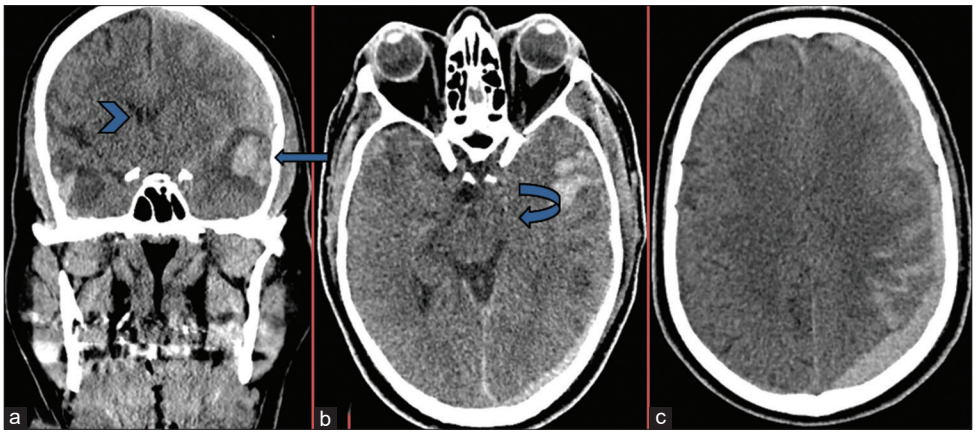

Figure 1: (a-c) Coronal and axial images from unenhanced computed tomography scan brain reveal a large left temporal hemorrhagic contusion (blue arrow) with associated surrounding edema, resulting in mass affect and midline shift to right (Arrow head). There is accompanying effacement of the left-sided perimesencephalic basal cisterns raising the possibility of impending uncal herniation (curved). Extensive left acute subdural hematoma and subarachnoid hemorrhage.

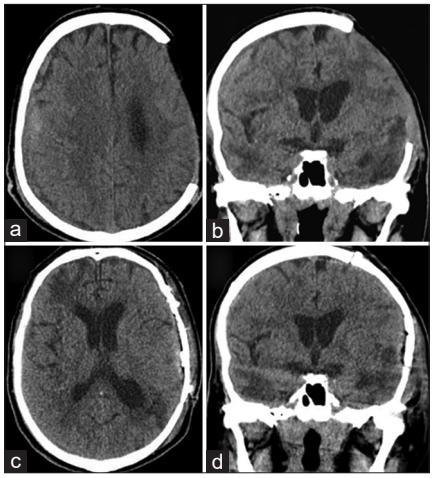

Figure 2: Axial and coronal images from unenhanced computed tomography scan brain. (a and b) showing a craniectomy defect. (c and d) are the images taken 1 week after cranioplasty.

of this surgery in improving functional outcome remains debatable.[3,16] A number of early and delayed complications have been described in association with DC. SoT is a constellation of late onset neurological deficits that improve with cranioplasty.[11] Most of the published reports describe the motor symptoms of SoT, with or without cognitive impairment. Few case reports mention the recovery of cognitive dysfunction.[6,12] However, to the best of our knowledge, isolated agitation as the presenting feature of SoT and responding with marked improvement to cranioplasty has not previously been published.

Di Rienzo *et al*. described SoT as a sunken scalp flap irrespective of posture with radiological evidence of depressed parenchyma and a new onset physical or cognitive deterioration.[8] The motor deficits associated with the sunken flap are more readily identified as compared to new onset cognitive abnormalities in a severe traumatic brain injury or stroke survivor.[2] A case reports have been published which describe a significant improvement in ADL, instrumental ADL, functional independence measure (FIM) index scores, the executive interview (EXIT 25), and neurobehavioral Cognitive Status Examination (Cognistat).[1,6] One prospective case series reported a 16% of improvement on the FIM scale within 7 days of cranioplasty.[12] However, the majority of these patients had motor deficits as the foremost symptom.

Data regarding the quantitative use of neuropsychological or cognition assessment tools before and after cranioplasty are not established in the literature so the extent of improvement is not clearly defined.[1] Di Stefano *et al.* reported a series of four cases, all with significant improvement in cognition and motor deficits.[9] Due to the heterogeneity of the cognition assessment tools in these patients, statistical tests could not be applied. However, improvement was reported in the "number of failed cognitive tests" and this was significantly reduced after cranioplasty.[9]

Cranioplasty results in an improvement in 3–53% of cognitive symptoms as reported in different case series.[5,18] However, across the literature on SoT, cognition is discussed rather vaguely and as a broad entity with infrequent use of cognitive measurement scales. This vague use of "cognition" hinders our understanding of the cognitive spectrum of the disease symptomatology. In the case reported here, mini-mental state examination and other cognitive scales could not be utilized as the patient's comprehension was impaired along with expressive dysphasia. Thus, despite having no motor deficit, this patient's communication barrier made him a difficult candidate for rehabilitation. As his alertness improved, his agitation worsened. Assessment for subclinical seizures, hyponatremia, or metabolic disorders was noncontributory. Neurological and psychiatric evaluation along with the increasing use of antipsychotic drugs did not seem to help his agitation. The very impressive recovery observed promptly after the cranioplasty strongly suggested that the sinking of the flap was the sole reason for the delayed onset of agitation in our patient. The timely repair of his cranial defect returned our patient to a calm state of cognition and this allowed early initiation of rehabilitation. There was also a reduction in his requirements for antipsychotic medications and a reduction in the costs of hospital stay.

CONCLUSION

With the wider use of cranial decompression in emergency neurosurgical practice, SoT is now being encountered more frequently. Combative and agitated behavior without a deteriorating level of alertness can be a rare manifestation of SoT and if recognized may respond well to early cranioplasty.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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