LETTER TO THE EDITOR

To The Editor

Rare Neurologic Deterioration to GCS 3 in a Patient with the Syndrome of the Trephined

Keywords: Craniectomy, Syndrome of the Trephined, Sunken Skin Flap Syndrome

The Syndrome of the Trephined (ST) is a rare complication of a craniectomy. Over time, the definition has evolved. Common symptomatology includes headaches, sensorimotor impairment, cognitive disturbances, and disorders of vigilance.[1] Additional associated symptoms include visual changes, hemineglect, tinnitus, insomnia, irritability, epilepsy, difficulty with concentration, hemiparesis, and hemiplegia.[1–3] Of particular relevance is the postural exacerbation of symptoms upon standing and valsalva maneuvers.[3] Fostad et al. suggest that for a diagnosis of ST to be made, the reversibility of symptoms with cranioplasty must be present. Prognosis post-cranioplasty has been reported to be optimal with most patients having full and relatively prompt recovery.[1]

Here we present, to the best of our knowledge, previously unreported GCS of 3 in a patient diagnosed with the Syndrome of the Trephined. While previous studies have commented on reports of altered level of consciousness (LOC), none have explicitly reported a GCS of 3. This conclusion is based on a PubMed search for articles with the key words 'GCS 3' and 'sunken skull flap' or 'syndrome of the trephined'.

This 48-year-old male patient was transferred to our hospital from another tertiary center with a declining LOC and a GCS of 3. Two months prior he had sustained a seizure accompanied by an acute, traumatic subdural hematoma and left temporal contusion treated with a hemicraniectomy and evacuation of the hematoma. During this hospital stay, he continued to have recurrent seizures. Fifty-one days after the initial seizure, he was transferred to another hospital where his LOC deteriorated. CT imaging showed effacement of the normal sulci and gyri pattern of the left cerebral hemisphere with a 15-mm midline shift (Figure 1) resulting in the decision to transfer to our hospital. On arrival to The Ottawa Hospital, he had a GCS of 3, with brainstem reflexes intact, and required intubation. The source of his decreasing LOC was unclear; a complete metabolic workup was found to be unremarkable, and an EEG demonstrated no seizures or seizure-like activity and therefore was determined not to be post-ictal status. The decision was made to proceed with a left-sided artificial cranioplasty with a titanium mesh implant. The scalp was sunken significantly at the level of the decompressive craniectomy. Once the scalp was dissected off the dura, it was noted that the brain started to come out toward the opening leading to the belief that the midline shift had improved. Pre- and post-operative diagnoses were consistent with ST. The patient was extubated within 48 hours post-operatively, for respiratory optimization, and recovered from a GCS of 3 to a GCS of 12 (Figure 2).

In hospital, he had right-sided weakness which was not in agreement with post-operative CT findings. MRI imaging revealed rightward mass effect on brain parenchyma without evidence of acute infarct and stable left temporal encephalomalacia. The patient's baseline GCS improved to 14, oriented only to person, unable to provide descriptions of baseline function or identify common objects. His thoughts were disorganized and tangential. The Physiotherapy service consultation reported independent mobility without gait aid. Occupational Therapy service consultation reported prior to admission the patient, who was previously independent, required cueing for basic self-care tasks. Neurology consultations were provided regarding seizure activity. Twenty-one days after admission to The Ottawa Hospital, the patient was transferred to an Acquired Brain Injury rehab for 42 days. At the time of discharge, the patient had a GCS of 15, tangential speech, and ⅘ strength in the upper extremity and was ambulating independently. Neuropsychiatric testing revealed low attention and executive function, processing speed, and poor memory encoding.

Two months post-diagnosis of ST showed progressive improvement of post-operative changes with no adverse interval change (Figure 2). At the latest follow-up, the patient had returned to baseline and close to his normal level of functioning.

In addition to the aforementioned clinical presentation, the presence of clinical or radiologic evidence of a depressed skin flap has also been reported.[1,3] The most common imaging findings include a sunken skin flap, paradoxical herniation (away from site of craniectomy), and midline shift.[3] A study comparing patients with post-craniectomy ST and those without a diagnosis of ST (with or without the sunken skin flap sign) reported that the size of the third ventricle and CSF volume may be beneficial in diagnosis.[3] The same study reported 92.3% of patients presenting with sulcal effacement on the affected side in the ST group and 78.95% in the control group.[3] This study demonstrated that midline shift and paradoxical herniation were not specific for ST and the presence of a sunken skin flap sign is not necessary for diagnosis.[3]

The reported significance of age is varied in the literature with Sedney et al. reporting a non-statistically significant preponderance toward older patients.[4] Vasung et al. did not find any significance in age distribution of patients with ST, although the group also alludes to conflicting results in the literature. A range of time to presentation has been reported in the literature with the longest reported being 138 weeks. The average time to onset has been reported to be between 1 month and 1 year.[1,3,4]

While the true pathophysiology of ST is yet to be elicited, some hypotheses have been succinctly summarized by Annan et al. to include atmospheric pressure causing depression of the skin flap which further causes brain compression.[1,5,6] Another explored hypothesis includes changes in CSF dynamics resulting in low CSF volumes particularly when standing.[1,7] Cerebral blood flow and metabolism changes have also been theorized to contribute to the pathogenesis of ST.[1,8] It has been proposed that the atmospheric pressure in combination with and causing redistribution of CSF due to postural changes results in the presentation of symptoms.[3,5] CSF-related pathogenesis was supported by this study which demonstrated decreased CSF volumes in patients with ST.[3]

In conclusion, we have presented the previously unreported and exceedingly rare case of the Syndrome of the Trephined in a patient with a GCS of 3 at presentation post-hemicraniectomy. The patient underwent a cranioplasty and has recovered well with almost no deficits.

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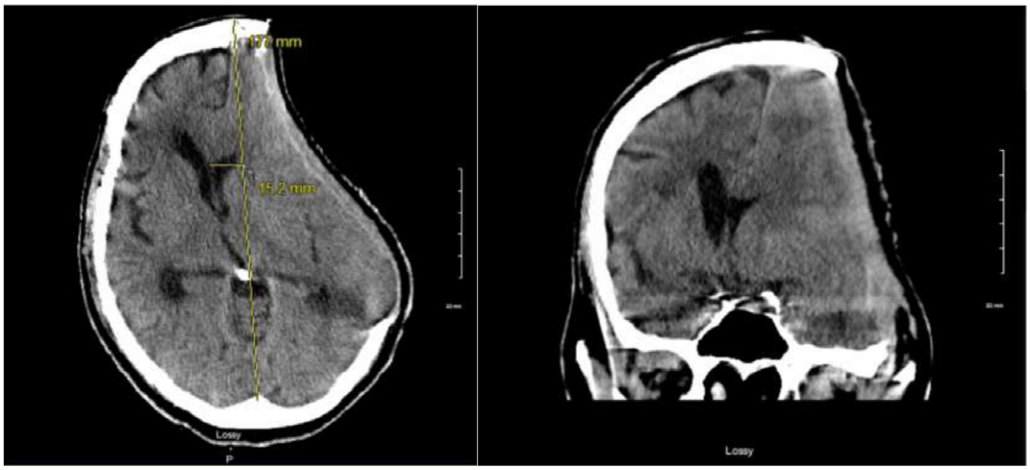

Figure 1: *CT non-contrast axial (left) and coronal (right) views of sunken craniectomy site with associated midline shift.*

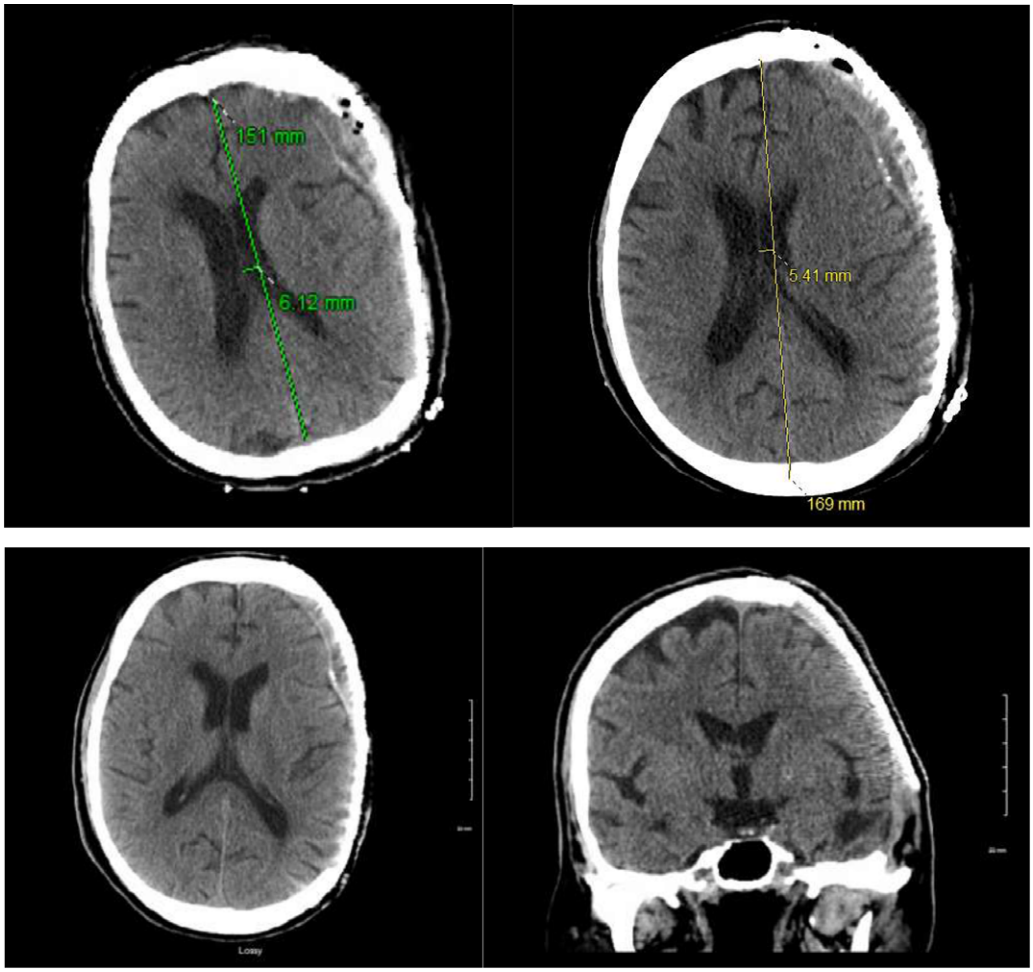

Figure 2: *Top: CT non-contrast axial 6 hours (left) and 48 hours (right) post-operatively from an artificial cranioplasty demonstrating resolution of midline shift and sunken skin flap. Bottom: CT non-contrast axial (left) and coronal (right) views 8 weeks post-operatively.*

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Disclosures

The authors have no conflicts of interest to disclose.

Statement of Authorship

AM was involved in conceptualization, literature review, and drafting and editing of the manuscript.

AA was involved in conceptualization, literature review, and drafting and editing of the manuscript.

SM was involved in conceptualization and drafting and editing of the manuscript.

Anahita Malvea
Faculty of Medicine, University of Ottawa, Ottawa, ON, Canada

Ahmed Alnemari
Division of Neurosurgery, Department of Surgery, Faculty of Medicine, University of Ottawa, Ottawa, ON, Canada

Safraz Mohammed
Division of Neurosurgery, Department of Surgery, Faculty of Medicine, University of Ottawa, Ottawa, ON, Canada

Correspondence to: Dr. Safraz Mohammed, Division of Neurosurgery, Civic Campus, The Ottawa Hospital/L'Hôpital d'Ottawa, C2214-1053 Carling Avenue, Ottawa, Ontario, K1Y 4E9, Canada. Email: smohammed@toh.ca

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