BRITISH JOURNAL OF HOSPITAL MEDICINE

CASE REPORT

Postoperative motor trephine syndrome and delayed tension pneumocephalus

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How to cite this article:
Gu TG, Wong BSS, Ho CL. Postoperative motor trephine syndrome and delayed tension pneumocephalus. Br J Hosp Med. 2020. https://doi.org/10.12968/hmed.2020.0046

Introduction

Motor trephine syndrome is an unusual condition with neurological deterioration following craniectomy. The removal of a bone flap converts the cranium from a 'closed-box' to an 'open-box' system where the intracranial contents become susceptible to differences in intracranial and atmospheric pressures. Brain herniation across intracranial boundaries results in a visible concavity of the skin flap over the scalp, hence motor trephine syndrome is also known as 'sinking skin flap syndrome'. Neurological deterioration can be exacerbated by CSF diversion, for example as a result of ventriculoperitoneal shunts. The treatment of motor trephine syndrome is cranioplasty.

Tension pneumocephalus is a potentially life-threatening neurosurgical emergency when a large amount of intracranial air causes intracranial hypertension, mass effect and neurological deterioration. Patients who have undergone cranial surgery remain at risk of the above conditions even many years later. This report highlights these unusual intracranial conditions in a young man following craniectomy and cranioplasty.

Discussion

Postoperative motor trephine syndrome

Motor trephine syndrome is a delayed, reversible complication following decompressive hemicraniectomy for traumatic brain injury, characterised by monoparesis on the contralateral

Case report

A 21-year-old-man presented to the emergency department with seizures, headache and progressive cognitive deterioration over several months. He was drowsy but arousable with no neurological deficits. There was a sunken contour of the scalp over a left craniectomy site. According to his wife, the sunken scalp deformity had worsened over the last few months. His past history was remarkable for bilateral craniectomies, evacuation of intracranial haematomas and right-sided cranioplasty 2 years before. Shortly after the last craniectomy, he underwent ventriculoperitoneal shunt insertion for hydrocephalus.

Computed tomography of the brain performed at the emergency department showed partial resorption of a right-sided cranioplasty flap and a large left frontoparietal craniectomy defect with an overlying sunken scalp flap. There was a right-ward midline shift with displacement of the left cerebral contents across the midline. This midline shift was aggravated by CSF overdrainage from a ventriculoperitoneal shunt (Figure 1). Given the findings, he was diagnosed with motor trephine syndrome. He subsequently underwent bilateral cranioplasties with titanium mesh and ligation of the ventriculoperitoneal shunt. Follow-up computed tomography of the brain showed near complete resolution of the midline shift and improvement of the ventricular size (Figure 2). Postoperative recovery was uncomplicated. His consciousness, cognitive and neurological function improved within days after surgery.

After discharge from hospital, he was well until 7 years later, when he presented again to the emergency department with seizure and worsening headaches. He had sustained a right parietal scalp laceration at the barber, which exposed the underlying titanium cranioplasty mesh. On examination, there was thinning of the soft tissue of the scalp overlying the cranioplasty mesh. Computed tomography of the brain showed a large amount of extra-axial air with a heaped-up appearance of the frontal lobes, giving a silhouette-like appearance resembling Mount Fuji. The Mount Fuji sign indicates tension pneumocephalus (Figure 3).

He subsequently underwent revision of the cranioplasty and reconstruction of the overlying scalp with a rotational flap. The postoperative recovery was uneventful with resolution of pneumocephalus and good vascularity of scalp flap.

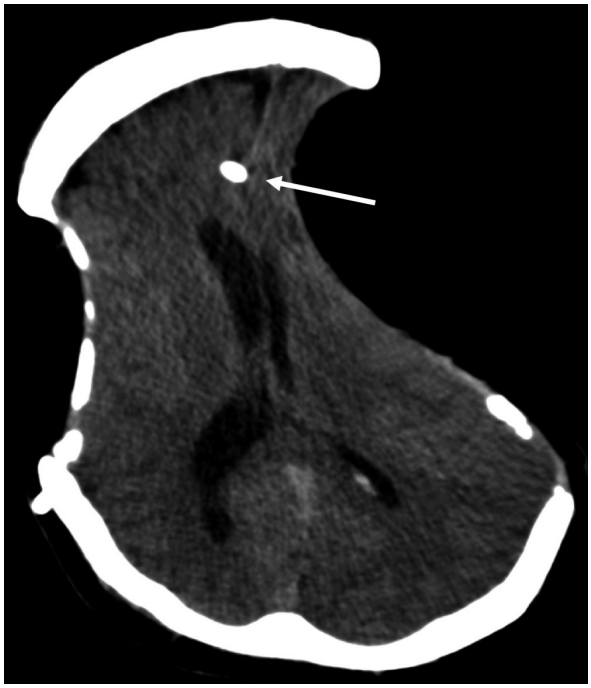

Figure 1. Computed tomography of the brain reveals partial resorption of a right frontoparietal cranioplasty flap and a large left frontoparietal craniectomy with sunken deformity of the overlying scalp. There is a right-ward midline shift with displacement of the left cerebral contents cross the midline. The midline shift is aggravated by CSF overdrainage from a ventriculoperitoneal shunt (arrow).

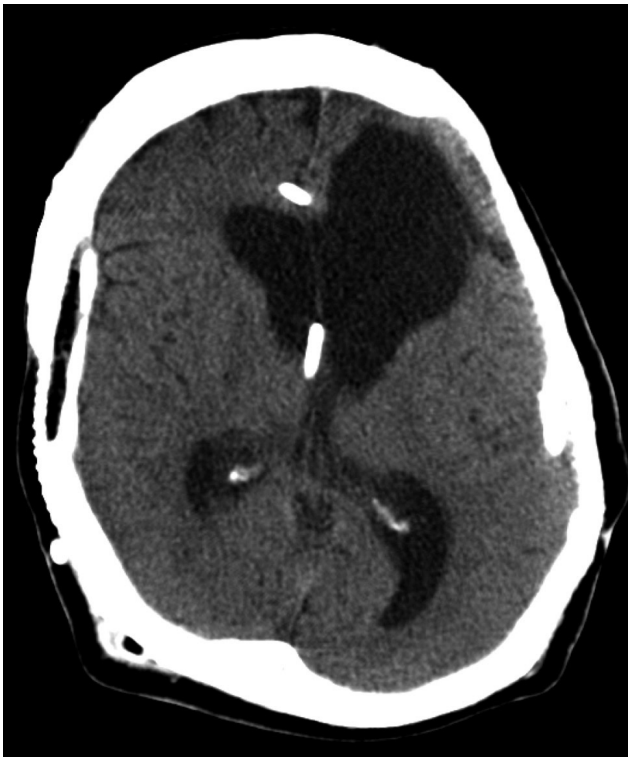

Figure 2. Computed tomography of the brain following insertion of bilateral titanium cranioplasties shows resolution of midline shift and interval improvement in ventricular size. A ligated ventriculoperitoneal shunt catheter is in-situ.

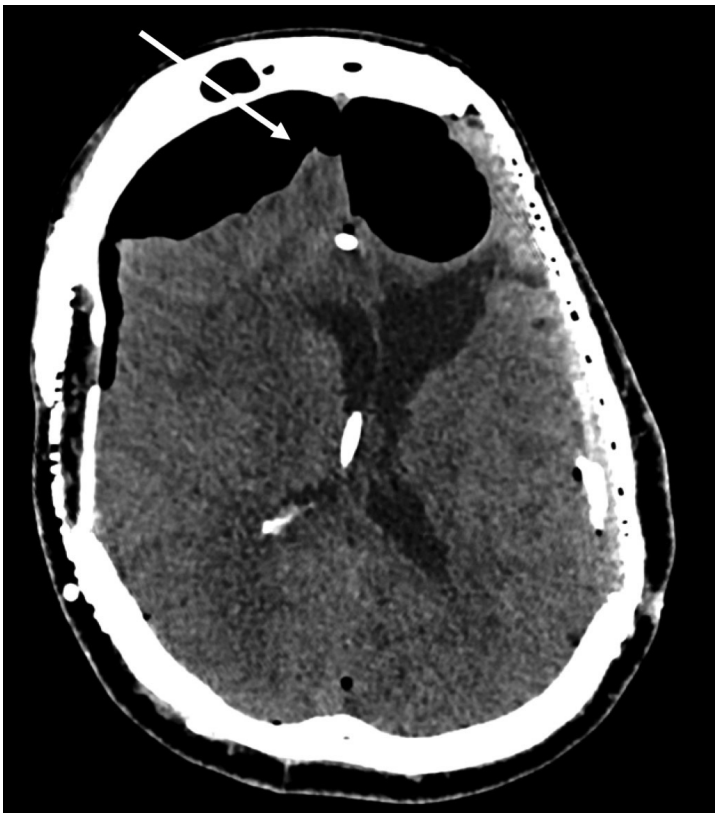

Figure 3. Computed tomography of the brain following right parietal scalp laceration shows a large amount of extra-axial air causing a heaped-up appearance of the frontal lobes – Mount Fuji sign (arrow), indicating tension pneumocephalus.

side (Stiver et al, 2008). The delayed onset of motor weakness observed in these patients may have led to the misnomer 'motor' in motor trephine syndrome even though it is not a pure motor syndrome (Stiver et al, 2008). Grant and Norcross (1939) first described motor trephine syndrome as 'syndrome of the trephined', with a cluster of symptoms including headache, dizziness, mental depression, apprehension and intolerance to vibration. Additional symptoms were subsequently recognised, including seizures, motor and sensory deficits, visual disturbances and cognitive impairment (Annan et al, 2015). The onset of symptoms may vary greatly after surgery, ranging from 3 days to 7 years, with an average of 5.1 months (Ashayeri et al, 2016).

The development of motor trephine syndrome has been attributed to a few factors. First, a cranial defect converts the intracranial cavity from a closed-box to an open-box system, allowing atmospheric pressure to act directly on the brain (Sedney et al, 2015). Next, intracranial pressure has been shown to decrease after craniectomy, and the pressure gradient between atmospheric and intracranial pressures can be further exacerbated by craniocaudal CSF flow (e.g. ventriculoperitoneal shunt) (Yamaura and Makino, 1977). Moreover, cerebral blood flow and metabolic activity in the injured brain tissues may decrease following trauma and craniectomy, which further increases the pressure gradient (Annan et al, 2015).

Owing to the non-specific presentations of motor trephine syndrome, a high index of suspicion for post-craniectomy motor trephine syndrome should be adopted in patients with the following risk factors: large craniectomy size and the presence of CSF drainage, including shunt(s) and lumbar puncture or drains (Annan et al, 2015). The treatment for motor trephine syndrome is cranioplasty, which converts the intracranial cavity back into a closed-box system. CSF pressure has been shown to normalise after cranioplasty, along with improvement in cerebral blood flow and metabolism (Yamaura and Makino, 1977). In the immediate setting, the patient should be put in a Trendelenburg position to increase CSF flow to the brain. Any shunt malfunction should also be corrected. Measures that lower intracranial pressure as per the Monro–Kellie doctrine, such as CSF drainage, shunts and hyperventilation, should be avoided (Liao and Kao, 2002). Neurological symptoms typically resolve promptly after cranioplasty, ranging from 24 hours to 2 weeks (Annan et al, 2015; Ashayeri et al, 2016).

Delayed tension pneumocephalus

The pathophysiology of pneumocephalus involves the entry of air into the intracranial cavity through a skull defect. The air is unable to escape as a result of a ball-valve effect. Small amounts of pneumocephalus are common after craniofacial surgery and can be treated conservatively. Large amounts of intracranial air cause intracranial hypertension with mass effect and neurological deterioration, hence it is called tension pneumocephalus (Jones et al, 2017).

Pneumocephalus can be acute (<72 hours) or delayed (>72 hours). Delayed tension pneumocephalus is extremely rare and must be promptly identified as it is a potentially life-threatening neurosurgical emergency. Risk factors for delayed tension pneumocephalus include craniofacial trauma, iatrogenic surgical procedures, invasive neoplasms and gas-forming infections (Markham, 1967). Hyperbaric oxygen therapy and continuous positive pressure ventilation are also causative factors (Jones et al, 2017).

Clinical presentation of delayed tension pneumocephalus is usually non-specific, and may include cranial nerve palsies, headache, seizures, giddiness or altered consciousness. Some authors have reported the presence of 'bruit hydro-aerique', a splashing sound heard during head movement (Markham, 1967). Computed tomography findings of the Mount Fuji sign (**Figure 3**) indicate tension pneumocephalus.

Postoperative pneumocephalus is expected to resolve within 3 weeks with conservative management (Reasoner et al, 1994). The current patient presented with delayed tension pneumocephalus about 7 years after cranioplasty, following laceration of the already fragile scalp soft tissue with subsequent exposure of the underlying titanium mesh. Treatment consists of surgical repair of precipitating structural defects of the scalp or skull, in this case cranioplasty revision and scalp flap reconstruction were performed. Postoperative surveillance is paramount in delayed tension pneumocephalus with particular attention to the vascularity and healing of the scalp flap.

Learning points

- Patients with motor trephine syndrome and delayed tension pneumocephalus frequently present months to years after initial surgery with non-specific symptoms.
- Treatment for postoperative motor trephine syndrome is cranioplasty which reverts the intracranial cavity from an 'open-box' to a 'closed-box' system, normalising the intracranial pressure and improving midline shift.
- Delayed tension pneumocephalus must be promptly identified as it is considered a life-threatening neurosurgical emergency. Treatment consists of surgical repair of precipitating structural defects of the scalp and skull.

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