CASE REPORT

Sinking flap syndrome with abdominal pain: an atypical presentation

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SUMMARY

A 53-year-old man developed significant mass effect secondary to an ischaemic stroke and was treated with decompressive craniectomy. During the first few days postsurgery he developed orthostatic vertigo with nausea. After a month, with increasing mobilisation and rehabilitation, he started reporting of severe abdominal pain. No aetiology could be found despite extensive local work up and the symptoms were resistant to any symptomatic treatment. Within days postcranioplasty there was a complete resolution of all the symptoms. Conclusion: the sinking flap syndrome can cause abdominal pain and orthostatic vertigo.

BACKGROUND

The sinking flap syndrome or ‘syndrome of the trephined’ is a complication after large craniectomy. In 1939, Grant and Norcross¹ created the term ‘syndrome of the trephined’ to describe the symptoms of dizziness, fatigability, headache, depression, pain at the craniectomy site and intolerance to vibration and seizure attributed to a delayed post-craniectomy syndrome. Later, other signs and symptoms such as dysautonomic syndrome, limb paresis, akinetic mutism, a large variety of cognitive symptoms, postural headache and impaired vigilance were added to the description.^{2–7} The syndrome is usually defined by an initial improvement after decompressive surgery followed by neurological deterioration. The diagnosis is confirmed when an improvement appears after the closure of the bone flap.^{2–5}

We describe here a previously undescribed symptom of sinking flap syndrome: abdominal pain. This case opens the differential diagnosis of abdominal pain and might help to avoid misdiagnosis or unnecessary investigation/treatment in specific situations.

CASE PRESENTATION

A right-handed, previously healthy 53-year-old man was admitted to our department after an ischaemic wake-up stroke of the right middle cerebral artery due to dissection of the internal carotid artery. Initial National Institute of Health Stroke Score (NIHSS) was 19 (left hemisindrome, homonymous hemianopsia and multimodal hemineglect). Radiological ischaemic damage on initial imaging was too extensive for a late revascularisation procedure. Two days later, the symptoms worsened (sleepiness, worsening of leg paresis). A repeat head CT scan showed a significant mass effect (figure 1A). Right frontotemporoparietal

decompressive craniectomy with duroplasty was performed and a Gore-tex patch was placed. Follow-up brain CT scans showed restoration of the midline shift (figure 1B).

When the patient was mobilised the first time after craniectomy, he experienced orthostatic symptoms. As soon as the head was raised above 30° he reported of dizziness, nausea, profuse sudation and pallor. The symptoms improved within a few minutes when he was lying down. Symptomatic treatment with sulpiride and ondansetron before mobilisation was attempted with little improvement. The patient was still able to undergo partial acute rehabilitation, with limited time out of bed, and a transfer to a neurorehabilitation facility occurred 2 weeks after craniectomy. About 1 week later, he began to have symptoms of persistent nausea and continuous abdominal pain. The abdominal pain was described as epigastric, continuous, unrelated to meal, associated with abdominal distension but without any transit disorders. At this point, he presented an Methicillin sensitive *Staphylococcus aureus* (MSSA) infection of the craniectomy scar. After the transfer back to the acute care hospital, surgical revision revealed meningeal infection around the Gore-tex patch. Flucloxacillin and then sulfamethoxazole-trimethoprim and rifampicin were given and the cranioplasty was postponed. Owing to worsening abdominal pain, a thorough search for local causes was initiated. Biological tests showed results that were within normal values. Repeated abdominal CT scans, oesophagogastroduodenoscopy, abdominal, hepatic and lumbar MRI showed no abnormality. EEG examination showed no argument for an epileptic origin. The pain was resistant to treatment by domperidone, ondansetron, sulpiride, esomeprazole, scopolamine, sucralfate, ranitidine and opiates. Owing to suspected alterations of the intestinal flora, possibly responsible for intestinal cramps, a cure of oral ciprofloxacin was given without success. Another three-phase abdominal CT-angiography scan showed meteorism in the small intestine with abundant faeces in the sigmoid colon without any obstruction or dilation. We first concluded to intestinal motility disorder secondary to a parasympathetic lesion postischaemic stroke and we started treatment with prucalopride, without clinical improvement. As the pain continued to worsen, treatment with intravenous xylocaine followed by epidural analgesia was introduced, with only a transient beneficial effect. Control head MRI showed an extensive infarct in the middle cerebral artery with a sunken concave deformity at the craniectomy site without any new lesion (figure 1C).

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Figure 1 Neuroradiological findings. (A) CT scan after 48 hours showing acute ischaemic stroke in the right middle cerebral artery territory with significant mass effect. (B) CT scan postcraniectomy. (C) T2-weighted MRI showing chronic extensive infarct in the right cerebral artery and concave deformity in regard of the craniectomy site. (D) Control CT scan postcranioplasty.

OUTCOME AND FOLLOW-UP

Cranioplasty was performed 3 months after craniectomy without significant complications (figure 1D). After cranioplasty, nausea, abdominal pain and orthostatic vertigo resolved within days, allowing regular rehabilitation to be continued.

DISCUSSION

Decompressive craniectomy is often performed to reduce critically elevated intracranial pressure (ICP). Craniectomy is a life-saving procedure, but the spectrum of early and late complications in corresponding patients is increasing.⁸ Honeybul⁷ proposed the term 'neurological susceptibility to a skull defect' (NSSD) to summarise conditions such as the syndrome of the trephined, the post-traumatic syndrome, the syndrome of the sinking scalp flap and the motor trephined syndrome.

Cranioplasty does have a protective role or a cosmetic purpose, and also a functional justification. In many patients it is indeed crucial for optimal recovery and rehabilitation.⁹ Bender *et al*⁹ analysed the effect of cranioplasty delay in 147 patients and concluded that early cranioplasty is associated with improved neurological outcome.

Secondary clinical deterioration after craniectomy is a well-known entity. However, the underlying physiopathology is still controversial. Several theories have been proposed. The development of a sunken concave deformity at the operation site is

often observed, as in our patient. A hypothesis is that, in the absence of skull closure, the brain is directly exposed to atmospheric pressure which causes distortion of cerebral cortex and intracranial structures.⁷ Cerebrospinal fluid hydrodynamic changes are another proposition. In the upright position, ICP is normally negative in a closed skull. It has been shown that in patients with large cranial defects, the ICP tends to equalise with the atmospheric pressure leading to raised ICP in an upright position.^{7 5} Several authors have described a positional worsening of the symptom^{7 4 2 10} usually consisting of headaches but visual and motor symptoms, altered sensorium and dizziness have also been reported. These mechanisms might explain the orthostatic vertigo and the nausea experienced by our patient. Unfortunately, ICP pressure has not been measured in outpatient to confirm this hypothesis. Other possible explanations are disturbance in cerebral blood flow or metabolic changes that improve postcranioplasty. These changes have been reported ipsilateral to the bone defect and also in the non-operated, contralateral side, especially in the thalamus.^{11 12}

Several authors have reported the role of the ventral posterolateral nucleus (VPL) of the thalamus in visceral pain, through the dorsal columns pathway.^{13 14} This mechanism might explain the central (abdominal) pain experienced by our patient. Restoration of normal blood flow in the VPL could mediate improvement after cranioplasty. The role of the insula is

increasingly recognised in viscerosensation and also in regulation of gastrointestinal motility.¹⁵ The insular cortex was widely infarcted in our patient. However, further suffering of the insula or insula pathway induced by NSSD could also have contributed to our patient's abdominal symptoms.

Learning points

- ▶ Sinking flap syndrome can cause a large variety of neurological symptoms. This entity should always be considered in case of delayed clinical deterioration after craniectomy without clear aetiology.
- ▶ Sinking flap syndrome can cause orthostatic vertigo and abdominal pain.
- ▶ Cranioplasty is crucial for optimal recovery and rehabilitation.
- ▶ Central lesion can cause resistant abdominal visceral pain (potential implication in pain management).

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