CASE REPORT

Sinking skin flap syndrome in glioblastoma

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SUMMARY

Sinking skin flap syndrome (SSFS) is a rare neurological complication in patients with traumatic haemorrhage, stroke or cerebral oedema who undergo decompressive craniectomy to relieve increased intracranial pressure. Hallmark of SSFS is the sinking of the scalp to a plane lower than the edges of the skull defect in the setting of neurological deterioration. Our objective is to report that SSFS can present after small craniotomy without cerebral cortex compression and to share our diagnostic/therapeutic approach. A 62-year-old woman with a glioblastoma developed SSFS after a small craniectomy and tumour resection without cerebral cortex compression but a decrease in the surgical cavity volume. Brain MRI showed decreased size of the surgical cavity. Interestingly, the patient also developed posterior reversible encephalopathy syndrome (PRES).

This case highlights an atypical presentation of SSFS and the possible association with PRES. It also illustrates how an early cranioplasty can successfully reverse SSFS.

BACKGROUND

The sinking skin flap syndrome (SSFS) is a rare neurological complication in patients with traumatic haemorrhage, massive stroke or severe cerebral oedema who undergo decompressive craniectomy to relieve increased intracranial pressure (ICP). In these patients, an initial short-lasting clinical improvement is followed by progressive neurological deterioration. The hallmark of SSFS is the concaving or sinking of the scalp to a plane lower than the edges of the skull defect. To date, there are few case reports in the literature. Correction of the sunken scalp improves the neurological deterioration.^{1 2} In all cases, SSFS developed after decompressive craniectomy and the sunken skin compressed the underlying cerebral cortex. We describe a patient with a glioblastoma (GBM) who developed SSFS after a small craniectomy and tumour resection without cerebral cortex compression but a decrease in the surgical cavity volume.

CASE PRESENTATION

A 62-year-old woman with prior history of colon cancer treated with colectomy and chemotherapy with no evidence of recurrent disease, who presented with confusion and gait imbalance. MRI of the brain showed a right temporal mass that corresponded to a GBM. Postoperatively, she was treated with concurrent chemoradiation with temozolomide. Unfortunately, there was tumour progression soon after completion of chemoradiation, and the patient underwent a second craniotomy with gross

total resection of the enhancing component of the lesion. One month later, she developed dehiscence of the operative wound, cerebrospinal fluid (CSF) leak and meningitis by a multiresistant strain of *Pseudomonas aeruginosa*, requiring removal of the bone flap. The meningitis was successfully treated, but she developed gait difficulty, urinary incontinence and progressive cognitive decline. Repeat imaging confirmed a communicating hydrocephalus for which she underwent placement of a left frontal ventriculoperitoneal (VP) shunt. She was transferred to the inpatient rehabilitation service, improving remarkably. On discharge she was able to walk and perform most activities of self-care.

Three weeks later, the patient presented with confusion and generalised tonic-clonic seizures. The patient was hypertensive (153/95 mm Hg), and laboratory studies revealed hyponatraemia (132 mEq/L), and azotaemia (31 mg/dL).

INVESTIGATIONS

A brain MRI showed the interval development of bilateral, asymmetric T2/fluid-attenuated inversion recovery hyperintense signals in the occipital, parietal, temporal and frontal lobes, suggesting an atypical presentation of posterior reversible encephalopathy syndrome (PRES), but with no evidence of tumour progression (figure 1A, B). Several EEG showed mild to moderate diffuse slowing, with no electrographic seizures. CSF analysis did not reveal any evidence of infection or malignancy. Skin examination shows surgical wound without any concavity nor signs of infection (figure 1C). Hypertension was controlled and his cognition improved. Two weeks later, her neurological examination revealed worsening of left hemiparesis, a receptive aphasia in the setting of abulia and severe impairment of cognitive function. Repeat MRI showed improvement of PRES (figure 1D) but decreased size of the surgical cavity (figure 1E). Physical examination revealed the sinking of the temporal scalp flap over the craniotomy defect (figure 1F).

TREATMENT

She underwent a right frontal temporal cranioplasty with titanium mesh and revision of left VP shunt.

OUTCOME AND FOLLOW-UP

Over the first 72 h, she gradually improved, manifest as increased level of alertness and awareness, allowing active participation in physical therapy.

DISCUSSION

SSFS can manifest in patients after undergoing an extensive decompressive craniectomy with

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Figure 1 Radiological findings before developing sinking skin flap syndrome (SSFS). (A) Brain MRI with and without contrast: axial T2 sequence showing extensive subcortical vasogenic oedema involving the temporal, parietal and occipital lobes bilaterally. (B) Coronal T1 postcontrast sequence showing surgical cavity measuring 50.4 and 36.9 mm without evidence of tumour recurrence or infection. (C) Surgical wound without any concavity or signs of infection. (D–F) Radiological findings at SSFS onset. (D) Brain MRI with and without contrast: axial T2 sequence showing improvement of the subcortical vasogenic oedema previously described. (E) Coronal T1 postcontrast sequence showing decreased size of the surgical cavity measuring 45.6 and 34.8 mm. (F) Evident concavity of the scalp over the skull defect.

symptoms such as headache, decreased level of alertness, seizures, vertigo, dysaesthesias or paresis.³ It may present with dysautonomia as well.⁴ These manifestations are potentially reversible without any sequelae. Yamaura *et al* reviewed 300 cases of decompressive craniectomy. Four out of 39 patients who developed sinking skin flap/flat type syndrome underwent cranioplasty with significant improvement in their neurological symptoms. Review of all the reported cases revealed that the symptoms usually occurred anywhere from the second day up to 18 months after the inciting procedure.⁵ Our patient developed the symptoms about 6 months after the second craniotomy.

There have been many theories postulated to explain the pathogenesis of SSFS, such as direct compression of the underlying cortex by atmospheric pressure, alternation in the cerebral blood flow (CBF) and alteration in the CSF flow dynamics.⁶ The Monro-Kellie doctrine states that cranial compartment is incompressible and the volume inside the cranium is fixed. Loss of volume (brain tissue/blood/CSF) in a fixed space such as the cranium creates a vacuum effect; the loss of skull bone results in a gradient between the atmospheric pressure and the ICP. This results in the inward displacement of the skin flap over the skull defect.^{2–6} The normal upper limit of the ICP is 20–25 mm Hg in a closed skull and the normal atmospheric pressure is 760 mm Hg. With a large skull defect, the ICP tends to equalise with the atmospheric pressure gradually which results in an increased ICP. In a closed space, the pressure and volume are inversely related. In order to accommodate the increase in ICP, the blood, CSF and brain parenchyma have to be displaced. Some reports in the literature have demonstrated abnormal blood flow in the area of the skull defect and also development of CSF hygromas indicating altered CBF/CSF dynamics.^{2–3}

Patients who after craniotomy have lost a large volume of CSF or who have undergone procedures to divert CSF, such as VP shunt or EVD placement, are at higher risk to develop SSFS. In some cases, the upright position has been also associated with worsening of symptoms.¹ The Trendelenburg position restores the CSF volume and has helped with reversal of symptoms in those patients.

In our patient, despite sinking of the skin to a level under the edges of the bone flap, we did not observe compression of the brain because she had a temporal lobectomy. We observed compression or a decrease in the CSF filling the surgical cavity, which we believed was related with drainage of CSF as expected with any VP shunt placed to treat hydrocephalus. The unique feature of SSFS without evidence of radiographic compression of the brain parenchyma, to our knowledge, has not been reported in the literature. In addition, this is the first report of an association between SSFS and PRES, which could be secondary to abnormal CBF/CSF dynamics. There are no reports showing PRES as a risk factor for development of SSFS or vice versa. Loss of brain parenchyma, early CSF leak and VP shunt placement for hydrocephalus could have contributed to the development of SSFS in our patient.

Kemmling *et al* used MR perfusion evaluation immediately before and 4 weeks after cranioplasty in a patient with SSFS. They compared the CBF, cerebral blood volume and mean transit time of both right and left hemispheres. The brain perfusion was lower on the right when compared with the left. The perfusion maps revealed a focal deficit particularly in the compressed central cortical region beneath the skin flap. The blood perfusion improved in the right hemisphere after the cranioplasty. This improvement in the brain perfusion correlated with clinical improvement.³ There have been a few case series that

have demonstrated improvement of CBF and brain metabolism by using transcranial doppler and FDG-PET scan, respectively, after undergoing a cranioplasty.⁷ These studies emphasise that the mechanism of SSFS is associated with alterations in the CBF.

The most accepted hypothesis of the cause of PRES is the failure of cerebral vascular autoregulation typically due to hypertension that results in brain hyperperfusion, cerebral vasoconstriction, capillary bed injury and subsequent brain vasogenic oedema.⁸ Cerebral autoregulation occurs when the mean arterial pressure ranges between 60 and 120 mm Hg, but it may be disrupted with sudden changes in blood pressure or severe hypertension above the mentioned limit. Both PRES and SSFS appear to be caused by derangement in the CBF and CBF/CSF dynamics, respectively. We hypothesise that the relationship between both clinical entities is the failure of autoregulation. The autoregulatory mechanism fails when the cerebral perfusion pressure and the blood flow to the cortical region beneath the skin flap

decrease, resulting in local vasodilation and increase in systemic blood pressure. There is a resultant increased pressure gradient over the skin flap and the autoregulatory mechanism would fail to increase the blood flow to the region just beneath the skin flap, while the remainder of the brain would become hyperperfused and prone to developing PRES. Additionally, this process would affect mainly the posterior circulation, a circuit that has a relative lack of sympathetic innervation.⁹ The goal behind restoring the neurological function is to restore the normal ICP, CBF and CSF flow. This can be achieved by decompression of the sunken scalp with cranioplasty in patients with large skull defects or to increase the ICP in patients with intracranial hypotension.

Contributors CK-M and SS were involved in study concept and design. CET and IWT-L were involved in revision of the manuscript. MRG was involved in study supervision.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Early recognition of the symptoms and physical manifestations of sinking skin flap syndrome (SSFS) is important as the process and neurological symptoms can be reversible.
- ▶ This condition is often overlooked because although there are many patients who undergo craniectomy there are only few reported cases of SSFS.
- ▶ SSFS is more uncommon after a small craniotomy and tumour resection without evidence of cerebral cortex compression.
- ▶ Early cranioplasty might be a better choice of therapy in patients with large skull defects.
- ▶ The concordance of SSFS with PRES reported here suggests that the development of SSFS may increase the risk to develop PRES; however, confirmation of this will require additional evaluation.

REFERENCES

- 1 Han PY, Kim JH, Kang HI, *et al.* "Syndrome of the sinking skin-flap" secondary to the ventriculoperitoneal shunt after craniectomy. *J Korean Neurosurg Soc* 2008;43:51–3.
- 2 Bhat AR, Kirmani AR, Nizami F, *et al.* "Sunken brain and scalp flap" syndrome following decompressive "extra-craniectomy". *Indian J Neurotrauma* 2011;8:105–8.
- 3 Kemmling A, Duning T, Lemcke L, *et al.* Case report of MR perfusion imaging in sinking skin flap syndrome: growing evidence for hemodynamic impairment. *BMC Neurol* 2010;10:80.
- 4 Romero FR, Zanini MA, Ducati LG, *et al.* Sinking skin flap syndrome with delayed dysautonomic syndrome—an atypical presentation. *Int J Surg Case Rep* 2013;4:1007–9.
- 5 Yamaura A, Sato M, Meguro K, *et al.* Cranioplasty following decompressive craniectomy—analysis of 300 cases. *No Shinkei Geka* 1977;5:345–53.
- 6 Joseph V, Reilly P. Syndrome of the trephined. *J Neurosurg* 2009;111:650–2.
- 7 Winkler PA, Stummer W, Linke R, *et al.* Influence of cranioplasty on postural blood flow regulation, cerebrovascular reserve capacity, and cerebral glucose metabolism. *J Neurosurg* 2000;93:53–61.
- 8 Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: controversies surrounding pathophysiology of vasogenic edema. *AJNR Am J Neuroradiol* 2008;29:1043–9.
- 9 Le EM, Loghin ME. Posterior reversible encephalopathy syndrome: a neurologic phenomenon in cancer patients. *Curr Oncol Rep* 2014;16:383.

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