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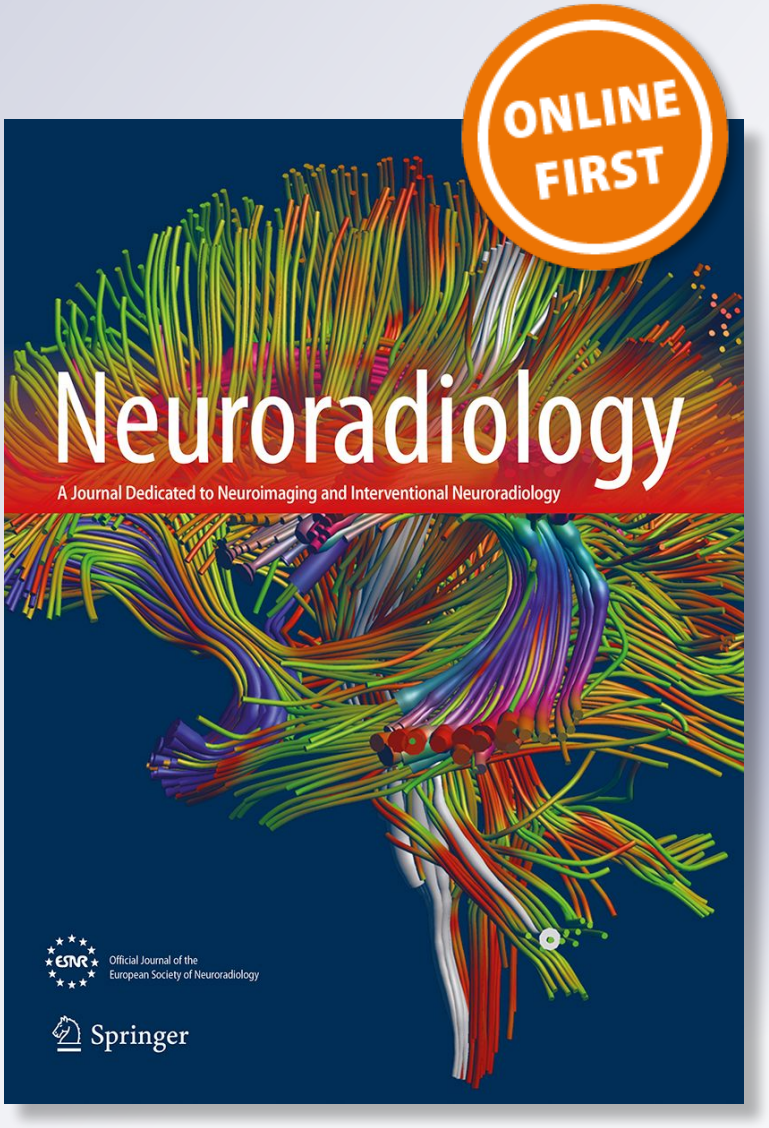
Neuroradiology
A Journal Dedicated to Neuroimaging and Interventional Neuroradiology

ISSN 0028-3940

Neuroradiology
DOI 10.1007/s00234-019-02251-8



Springer

Neuroradiology
https://doi.org/10.1007/s00234-019-02251-8

SHORT REPORT

Dural venous sinus stenting as a stand-alone treatment for spontaneous skull base CSF leak secondary to venous pseudotumor cerebri syndrome

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Received: 3 April 2019 / Accepted: 21 June 2019

Abstract

Most spontaneous CSF leaks (SCSFL) are associated with an underlying pseudotumor cerebri syndrome (PTCS). Treatment generally includes surgical leak repair and PTCS correction, as untreated PTCS carries a risk of recurrence. We describe a 72-year-old woman with rhinorrhea, aural fullness, and posterior nasal drip. CT and MRI showed signs of CSF hypovolemia and PTCS, as well as bilateral transverse sinus stenoses. CT and MRI cisternography documented CSF leaks through the right cribriform plate and the posterior aspect of the petrous bone. Opening CSF pressure was 6 cm H_2O. Dural venous sinus stenting (DVSS) was performed after failed conservative treatment. Rhinorrhea resolved 3 days after stenting, aural fullness 1 month later. After 6 months, signs of CSF hypovolemia had disappeared on MRI and the stents were patent. After 9 months, the patient had a transient, spontaneously resolving episode of rhinorrhea. She has been symptom-free for the remaining 39 months of follow-up.

Keywords Spontaneous CSF leak · Pseudotumor cerebri · Dural venous sinus stenosis · Dural venous sinus stent

Abbreviations

AG	Arachnoid granulation
CSF	Cerebrospinal fluid
DVSS	Dural venous sinus stenting
PTCS	Pseudotumor cerebri
SCSFL	Spontaneous CSF leak

Electronic supplementary material The online version of this article (https://doi.org/10.1007/s00234-019-02251-8) contains supplementary material, which is available to authorized users.

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Introduction

Spontaneous CSF leaks (SCSFL) occur secondarily to one or multiple skull base osteodural defects caused by increased CSF pressure. SCSFL represent a variant of the pseudotumor cerebri syndrome (PTCS) [1, 2]. Factors such as hyperpneumatization and skull base thinning from chronically elevated CSF pressure may favor their formation and account for PTCS manifesting with SCSFL rather than with classical findings of PTCS [1, 2]. The leak likely provides a CSF diversion pathway that "protects" the patient from developing complications of increased CSF pressure [3]. CSF pressure elevation following fistula repair [1] accounts for the recurrence after successful surgical management [1, 4].

SCSFL may present with signs of CSF hypovolemia secondary to rhinorrhea/otorrhea or with ascending meningitis. Management of SCSFL includes conservative treatment, surgical repair of the leak site, and adjunctive measures to treat PTCS such as CSF diversion to reduce the risk of SCSFL recurrence [5].

We present a case of SCSFL secondary to PTCS caused by dural venous sinus outflow obstruction treated with dural

Published online: 27 July 2019

venous sinus stenting (DVSS) with the hypothesis that reversal of PTCS would lead to SCSFL cessation.

Case report

Informed consent was obtained prior to all procedures and imaging examinations including off-label uses of a carotid stent and intrathecal Gadolinium injection.

A 72-year-old woman with no past medical, surgical, or trauma history consulted for spontaneous high flow rhinorrhea, right aural fullness, and postnasal drip. General, ENT, and neurological examination were normal. BMI was normal (< 25 kg/m^2). β-2 transferrin was detected in the nasal fluid confirming CSF.

Brain MRI revealed pachymeningeal thickening and enhancement. In addition, there was a markedly enlarged "empty sella," bilateral petrous apex meningoceles, enlarged CSF spaces around the oculomotor nerves, middle cranial fossa arachnoid pits, slight optic nerve tortuosity, and bilateral transverse sinus and right sigmoid sinus stenoses caused by arachnoid granulations (AG) (Fig. 1, Figure 3 in Supplementary material). CT-cisternography (intrathecal injection of Iodixanol - Visipaque 270, GE Healthcare) revealed contrast material in the right nasal fossa (Fig. 1) and possibly in the right petrous air cells. Opening CSF pressure was 6 cm H_2O. Eight days later, rhinorrhea and post-nasal drip were resolved. SCSFL secondary to PTCS due to venous outflow obstruction was diagnosed. Given the spontaneous resolution, no treatment was proposed.

SCSFL with identical symptoms recurred 4 months later. CT and MRI cisternography were performed (intrathecal injection of Iodixanol and 0.7 ml of Gadopentetate dimeglumine (Magnograf, Bayer)). CSF leaks were found in the same locations (Figure 3 in Supplementary material). Therapeutic options after 4 weeks of failed conservative treatment included surgical fistula repair and / or correction of the underlying PTCS. In the absence of encephaloceles, surgical repair was not thought to be necessary provided that PTCS was corrected. Dural venous sinus stenting was preferred over CSF shunting as it offered the advantage of treating the cause of PTCS. Catheter DVS pressure measurements revealed non-significant pressure gradients of 2–3 mmHg across the stenosis, judged not relevant in the setting of CSF hypovolemia. After multidisciplinary discussion, the patient was prepared with 100 mg of acetylsalicylic acid and 75 mg of clopidogrel QD 7 days prior to stenting for which no resistance was found (continued for 6 months). Two 6 × 40 mm self-expandable stents (Precise Pro RX, Cordis) were telescoped in the right transverse and sigmoid sinuses across the stenosis (Fig. 1). Rhinorrhea and postnasal drip ceased on day 3 after stenting, with aural fullness disappearing by week 4.

Post-contrast MRI and CT performed at 6 months showed patent stents without stenosis and regression of CSF hypovolemia signs (Fig. 2, Figure 3 in Supplementary material). At 9 months, the patient presented a few episodes of isolated possible rhinorrhea that resolved spontaneously after a few days; MRI was unremarkable. Outside this isolated episode, the patient has been symptom-free without recurrence of rhinorrhea throughout the 39 months of follow-up period.

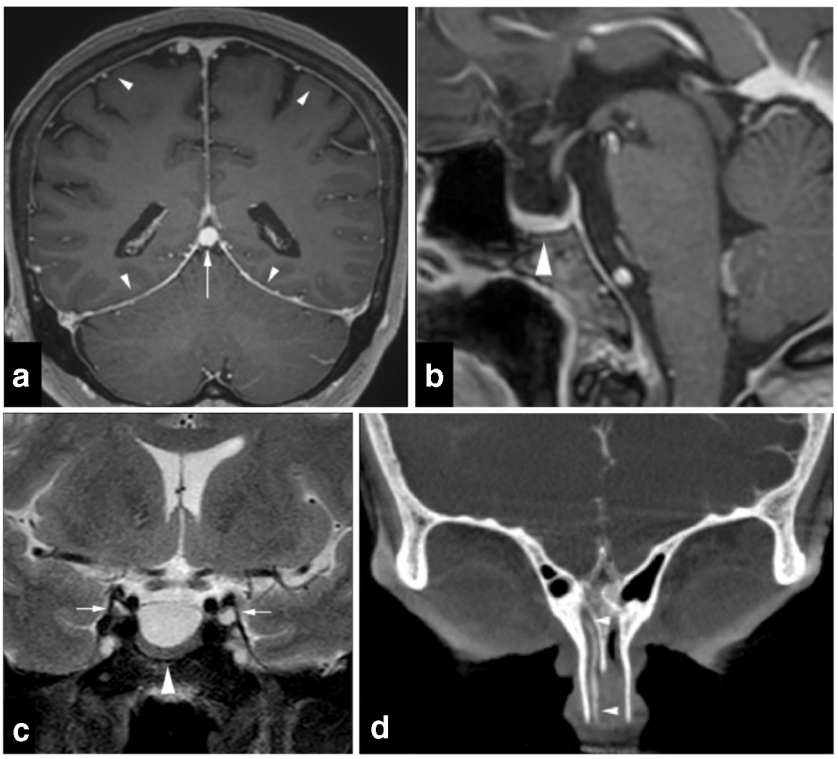

Fig. 1 Initial MRI study. **a** and **b** Gadolinium-3DT1 MPRAGE coronal and sagittal plane reconstructions. **c** Fat sat T2 coronal plane. **d** Dyna-CT-cisternography, coronal oblique plane reconstruction. Images show signs of CSF hypovolemia in **a**: pachymeningeal thickening and enhancement (small arrowheads) and dilated straight sinus (arrow). These signs co-exist with signs of pseudotumor cerebri: enlarged "empty sella" (large arrowhead, **b** and **c**) and bilaterally dilated oculomotor nerve sheath around its course through the lateral wall of the laterosellar space (arrows, **c**). Dyna-CT cisternography after intrathecal contrast material injection demonstrating CSF leak into the right nasal cavity (arrowheads, **d**)

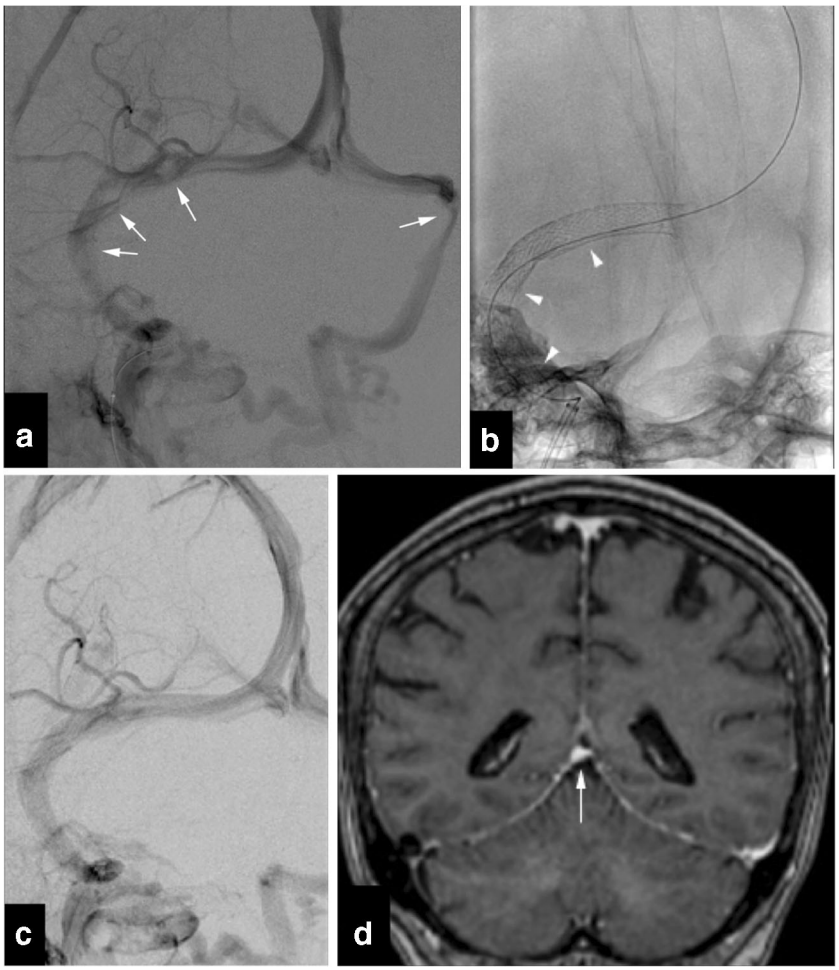

Fig. 2 **a**–**c** DSA, left anterior oblique projections. **d** Gadolinium-3DT1 MPRAGE coronal plane reconstruction. **a** Venous phase of a right common carotid artery injection showing bilateral transverse sinus and right sigmoid sinus stenosis (arrows). **b** Two overlapping stents (arrowheads) are deployed in the distal two-thirds of the transverse sinus and sigmoid sinus on the right. **c** Venous phase of a right common carotid artery injection showing the right transverse and sigmoid sinus stenosis being relieved after stenting. **d** MRI 6 months after stenting showing resolution of abnormal pachymeningeal thickening and enhancement and normalization in size of the vein of Galen (arrow—compared with pre-stenting MRI; Fig. 1a)

Discussion

The current literature suggests that fistula repair in SCSFL without treating the underlying PTCS carries a high risk of recurrence [1, 6]. Treatment strategies for SCSFL include surgical repair and management of the underlying PTCS with weight loss, acetazolamide, and CSF diversion. Surgical repair generally precedes PTCS treatment, often because PTCS is overlooked until signs appear following surgery. Though DVSS is increasingly used for treating PTCS [7], it is exceptionally employed in cases associated with SCSFL after surgical repair of the leak [8]. Recently, Iyer et al. [9] reported two additional cases of surgically treated SCSFL where DVSS was subsequently performed to treat an underlying PTCS that became clinically apparent following surgery. DVSS for PTCS is a safe and effective treatment [7]. The hemodynamic effect of DVSS on the venous circulation is immediate, allowing for rapid lowering of CSF pressure [10]. We found no prior instance of SCSFL succesfully treated exclusively by measures aimed at correcting the underlying PTCS.

In our case, the patient presented with recurrent SCSFL from two remote fistulous sites, PTCS, and venous outflow obstruction caused by AG. Pressure measurements across the stenoses revealed non-significant pressure gradients of 2–3 mmHg. However, CSF hypovolemia could have accounted for low-pressure gradients. Indeed, CSF hypovolemia, following the Monro-Kellie doctrine, leads to compensatory dural venous dilatation that could have countered the intraluminal stenosis and reduce the pressure gradient. Arguments in favor of DVSS included (i) failed conservative management; (ii) challenging repair of coexisting supratentorial and infratentorial CSF leaks; (iii) the need to treat the underlying PTCS; (iv) potential complications of CSF diversion, including frequent shunt revisions; (v) risk of decreased CSF volume following derivation leading to symptomatic CSF hypovolemia; (vi) the fact that DVSS addresses the underlying etiology of PTCS; (vii) and the hypothesis that PTCS correction would normalize the intracranial pressure and resolve the CSF leaks. In our patient, SCSFL resolved 3 days after DVSS and has persisted throughout the 39-month follow-up period, with the exception of one possible episode of rhinorrhea spontaneously resolving after a few days. Imaging at 6 and 9 months showed patent stents, with no residual or recurrent signs of CSF hypovolemia.

The present case suggests that SCSFL with PTCS can be successfully managed by DVSS. Each case must be discussed by a multidisciplinary team of interventional neuroradiologists, ENT surgeons, and neurosurgeons. Surgery may be warranted in cases of large osteodural defects with encephalocele in conjunction with DVSS. In those cases, DVSS should follow surgical repair as antiplatelet therapy or anticoagulation is needed after DVSS.

Funding No funding was received for this study.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in the studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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