Spontaneous cerebrospinal fluid leak via foramen rotundum in a non-obese male presented as pseudo-Chiari malformation type I: a case report and literature review

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Abstract
Spontaneous cerebrospinal fluid (sCSF) leak from the skull base has been previously reported, but there are few reports of sCSF leak from the foramen rotundum due to its rare occurrence. This case report describes a 15-year-old male patient that presented with left side watery rhinorrhoea that had been present since he was 4 years of age and a history of repeated bouts of meningitis of unknown cause. A discharge sample from the nose tested positive for beta-2 transferrin. Preoperative computed tomography (CT) revealed a fistula between the cerebellopontine angle and the left sphenoid sinus. There was also a pseudo-Chiari malformation type I with ectopia of the cerebellar tonsil. Endoscopic transnasal surgery identified a leak from the foramen rotundum that was repaired using autologous material and a contralateral pedicle nasoseptal flap. At 6 months after surgery, the patient reported no recurrence of the CSF leakage. Postoperative CT imaging revealed that the cerebellar tonsil was back in the normal position, indicating that the

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preoperative Chiari malformation was possibly due to decreased CSF volume. This current case shows that a rare case of sCSF leak from the foramen rotundum can be effectively repaired using the endoscopic transnasal approach.

**Keywords**
Spontaneous cerebrospinal fluid leak, foramen rotundum, transnasal endoscopic approach, pseudo-Chiari malformation type I

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**Introduction**
Spontaneous cerebrospinal fluid (sCSF) leak is an unusual condition that involves a breach of the arachnoid and dura mater, with a defect in a pneumatized area in the skull base. It accounts for less than 4% of all CSF leakage.¹ Patients with sCSF leaks are often obese and female;² and CSF rhinorrhoea is the most prevalent initial symptom.³ This condition usually leads to fistula formation over the anterior skull base or sinonasal tract.¹ Compared with the cribriform plate and lateral sphenoid sinus, which are the most prevalent locations for anterior sCSF leaks,² leakage from the foramen rotundum is rarely reported.⁴ The present case report describes a male patient with a rare sCSF leak from the foramen rotundum that was effectively repaired using the endoscopic transnasal approach.

**Case report**
A 15-year-old male patient with body mass index of 20 kg/m² presented at the Department of Otolaryngology, University Medical Center, Ho Chi Minh City, Vietnam in December 2018 with left side watery rhinorrhoea that had been present since he was 4 years of age. His symptoms were exaggerated during the Valsalva manoeuvre or upon jugular compression. A review of his medical history showed that he had experienced 10 episodes of meningitis within the past 8 years, but all attempts failed to identify the cause. A discharge sample from the nose tested positive for beta-2 transferrin. Preoperative computed tomography (CT) revealed a fistula between the cerebellopontine angle and the left sphenoid sinus (Figures 1a & 1b). CT on sagittal sections also demonstrated a pseudo-Chiari malformation type I with ectopia of the cerebellar tonsil (Figure 1c).

The endoscopic transnasal approach was applied to repair the defect. After resecting the lateral and posterior wall of the maxillary sinus, the pterygopalatine fossa was exposed, followed by sphenopalatine artery ligation. The vidian nerve and the maxillary nerve were identified, then the foramen rotundum could be traced based on these landmarks. Clear fluid leakage was identified at the foramen rotundum site (Figure 2a). To close the defect, the temporal fascia was inserted into the space between the maxillary nerve and the foramen rotundum (Figure 2b). A piece of septal cartilage was placed over the temporal fascia and attached with tissue glue (Figure 2c). A contralateral pedicled nasoseptal flap (Hadad-Bassagasteguy flap) was then harvested and placed with complete coverage of the temporal graft and sealed with tissue glue (Figure 2d).
The patient had nasal irrigation with normal saline continuously during the recovery phase; and 6 months after the operation, the contralateral pedicled nasoseptal flap had healed and fully covered the operative region (Figures 2e & 2f). The patient remained free of symptoms and follow-up CT imaging showed that the cerebellar tonsil had returned to the normal position (Figure 3). After the surgery, no complaint of dry eyes was reported. Nevertheless, the patient felt numbness and an itchy sensation in the skin of the left lower eyelid, the prominence of the left cheek and the alar part of left nose. The left sided facial abnormal sensation went into remission on its own. The size of the paraesthesia area gradually reduced and returned to normal after 3 months.

Approval to collect and use the data from this case was provided by the Training and Scientific Research Department, University Medical Center, Ho Chi Minh City, Vietnam. Written informed consent to receive treatment and for publication of the findings were provided by the patient and his father.

**Discussion**

Spontaneous CSF leaks are reported to be more common in females, probably because of their thinner skull bases.\(^1\,^2\) In addition, there is also a relationship between sCSF leaks and obesity.\(^1\,^2\) It is likely that being overweight increases the intra-abdominal pressure and decreases the cerebral venous return, which results in elevated intracranial pressure (ICP) and further erosion of the skull base.\(^1\) While some studies reported that only 10–36% of patients undergoing repair of sCSF leak had raised ICP, it is generally accepted that ICP elevation with an underlying anatomical predisposition involving thinning of the cranial base account for the generation of sCSF leaks\(^3\,^6\) and obstructive sleep apnoea (OSA) (43%).\(^2\)

Based on the findings that patients with OSA experience transient significant spikes in ICP during episodes of sleep apnoea and the strong relationship between OSA and obesity,\(^8\,^9\) previous research demonstrated that patients with sCSF leaks were more likely to be diagnosed with OSA and had a thinning of their entire calvarium.\(^10\,^11\)
The sCSF leak in the current case, who was a non-obese young male patient without signs of raised ICP or evidence of sleep apnoea, could probably be better explained by a ‘congenital’ pathophysiology. A ‘congenital’ sCSF leak suggests that there was congenital dehiscence in the skull base. The dehiscence might also have been formed by bone remodelling or reabsorption during the patient’s life and in this

**Figure 2.** Representative intraoperative and endoscopic images of a non-obese 15-year-old male patient that presented with left side watery rhinorrhoea that had been present since he was 4 years of age and a history of repeated bouts of meningitis of unknown cause. (a) Exposure of the leakage site at the foramen rotundum (arrow) and maxillary nerve (V2). (b) Temporal fascia (f) was used as a seal to close the defect. (c) Septal cartilage (c) was placed over the temporal fascia. (d) The contralateral pedicled nasoseptal flap (PNF) was harvested to cover the temporal graft. (e) and (f) The endoscopic images at 6 months after the operation showed a fully healed pedicled nasoseptal flap without any notable defects. The colour version of this figure is available at: http://imr.sagepub.com.
case it would not suggest a congenital origin.3

Within the sphenoid sinus, sCSF leaks most often occur at the perisellar region and the lateral recess of the sphenoid sinus.3 Several factors have been reported to explain sCSF leak in the lateral recess around the foramen rotundum: (i) the extensive pneumatization of the lateral recess of the sphenoid beyond the foramen rotundum found in up to 27% of adults; (ii) the lateral cranio-pharyngeal canal (or Sternberg canal) with persistence into adulthood in 4% of patients, now being considered a misnomer for a thinned sphenoid sinus recess; (iii) and aberrant arachnoid granulations as precursors of osteodural erosion, being the most plausible responsive aetiology in this current case.3,12

Endoscopic transnasal surgery has become the preferred method for most patients with sCSF leaks suspected to be at the anterior skull base because of its comparable success rates, lower morbidity and minimal invasion compared with the transcranial approach.3,5 The choice of the graft and how to place the graft mostly relies on the surgeon’s experiences and preference. A previous study reviewed 28 patients with sCSF leaks and reported that the authors’ preferred method of graft placement was an inlay technique using autologous materials (e.g. fascia lata), along with a rigid buttress to create a watertight seal, covered with a vascularized flap when the defect was >1 cm.5

The preoperative image of a downward herniation of the cerebellar tonsil is commonly known as a condition of pseudo-Chiari malformation sign and in this current case it was an indication of possible CSF leakage.13,14 While the impending cerebellar herniation might urge clinicians to consider placing lumbar drainage to stop further herniations, this CSF leakage-related pseudo-Chiari malformation is actually due to CSF hypovolaemia and under these circumstances placing lumbar drainage after CSF leakage repair might slow down the correction of the CSF hypovolaemia. Moreover, several studies have demonstrated that the use of lumbar drains did not appear to result in improved success rates for skull base leakage repairs.2,15 Therefore, in this current case, a lumbar drain was not used postoperatively and the patient was successfully treated with the endoscopic transnasal approach using autologous material without notable recurrence of the CSF leakage.

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