Spontaneous Recurrent CSF Rhinorrhoea: A Rare Case and Review of Literature

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Abstract Cerebrospinal fluid (CSF) rhinorrhoea is the leakage of CSF through the communication between the subarachnoid space and the nasal cavity. Surgical repair is indicated in all cases of spontaneous leaks, recurrent leaks, leaks that do not stop after conservative management and cases with history of meningitis. We present a rare case of spontaneous (delayed onset post traumatic) CSF rhinorrhoea with multiple defect sites, which was treated with an endonasal endoscopic repair. The patient was asymptomatic for 9 years after surgery, and then presented with a spontaneous left frontal recess CSF leak, which was closed using endonasal approach.

Keywords Recurrent cerebrospinal fluid rhinorrhoea · β -2-Transferrin · CT cisternography · Endonasal endoscopic repair · Multiple defect CSF leak · High pressure CSF leaks · Meningitis

Introduction

Cerebrospinal fluid (CSF) rhinorrhoea is the leakage of CSF through the communication between the subarachnoid space and the nasal cavity. CSF rhinorrhoea can be congenital or acquired. It can be acquired by trauma (iatrogenic or non-iatrogenic), tumour/neoplastic, or

spontaneously. Spontaneous fistula can be further subdivided as primary cases, where no underlying cause can be found, or they can be secondary to intracranial pathology (normal or high intracranial pressure). High pressure leaks include tumours, benign intracranial hypertension and hydrocephalus. Normal pressure leaks are caused by tumours, congenital defects, infection, arachnoid granulations, meningoencephaloceles and idiopathic conditions.

Usually, CSF rhinorrhoea is secondary to head trauma with an associated skull base fracture. Most of the traumatic leak stops with conservative management. Only 3–4 % of leaks are spontaneous. The majority occurs in adults in the fourth decade of life, with females outnumbering males (2:1). Diagnosis is made by clinical, biochemical and radiological examination. Detection of β -2-transferrin in watery nasal discharge is diagnostic of CSF. CT cisternography is the gold standard for diagnosis of CSF rhinorrhoea. Intrathecal sodium fluorescein can also be used to localize the site of the defect intra-operatively. Alternatively, plain CT with MR cisternography can be also performed.

CSF rhinorrhoea causes a potential risk of ascending infection, which mandates surgical repair. Surgical repair is indicated in all cases of spontaneous leaks, recurrent leaks, leaks that do not stop after conservative management and cases with history of meningitis or with a history of CSF rhinorrhoea.

Case Report

A 46 year male patient presented with complains of watery discharge from right nostril since 2 months. He presented with a history of minor head trauma 15 years back. ENT examination and general examination were within normal

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limits. CT cisternography showed bony defects along with active CSF leak in the right cribriform plate (Fig. 1), left anterior fovea (Fig. 2) and right lateral sphenoid (Fig. 3). There was a doubtful leak on left cribriform plate as well.

An endonasal endoscopic repair of CSF rhinorrhoea under general anaesthesia was planned. A lumbar drain was inserted prior to surgery. The nose was packed with cotton patties soaked in saline adrenaline (1:1000) solution. The surgery commenced with a right uncinectomy, maxillary ostium widening, and anterior and posterior ethmoidectomy. The sphenoid sinus ostium was visualised and widened by removing the anterior sphenoid wall. The postero-medial wall of the maxillary sinus was removed to gain access to the lateral wall of the sphenoid sinus. The sphenopalatine artery was identified and cauterized. A small encephalocele with CSF leak was seen in the infero-lateral wall of the sphenoid sinus with a 45° endoscope (Fig. 4). The encephalocele was cauterized. Fascia lata and fat was harvested from the thigh. The defect was closed by fascia lata, tissue glue and fat. A small meningocele with CSF leak was identified in the right cribriform defect (Fig. 5) and repaired using fascia lata and tissue glue. The mucosa of middle turbinate and adjacent septum was made raw and bolgerisation was done. There was a defect on the left cribriform plate as well, in communication with the right side. This was closed similarly using fascia lata and tissue glue.

Next, a left uncinectomy, maxillary ostium widening, and anterior and posterior ethmoidectomy was performed. The frontal recess was cleared and the ostium seen. A large bony defect with an encephalocele was seen in the anterior fovea (Fig. 6), which was cauterized and the defect was closed by nasal septal cartilage (Fig. 7), fascia lata, tissue glue and fat. Haemostasis was achieved and nose was packed for five days. Avoidance of sneezing or coughing, the use of stool softeners, elevation of the head, and bed-rest was given in the post operative period. Patient was on prophylactic antibiotics for 7 days. The lumbar drain was

Fig. 1 Coronal CT scan with cisternography showing defect in right cribriform plate

Fig. 2 Coronal CT scan with cisternography showing defect in left anterior fovea (red arrow)

Fig. 3 Coronal CT scan with cisternography showing defect in infero-lateral wall of right sphenoid sinus

Fig. 4 Operative photo showing encephalocele with CSF leak in infero-lateral wall of right sphenoid sinus, seen by 45° endoscope

removed on 5th post-operative day. Patient was followed weekly, for 6 weeks for endoscopic nasal cleaning. At 1 year follow up both the nasal cavities healed well with few synechiae, and no breathing difficulties. The patient was asymptomatic for nine years after surgery, and then presented with a spontaneous left frontal recess CSF leak, which was closed using fascia lata, tissue glue and fat via an endonasal approach.

Fig. 5 Operative photo showing encephalocele with CSF leak in right cribriform plate, seen by 45 endoscope

Fig. 6 Operative photo showing encephalocele in left anterior fovea, seen by 45 endoscope

Fig. 7 Operative photo showing left anterior fovea defect closed with septal cartilage

Discussion

The most frequent cause of CSF rhinorrhoea is iatrogenic or traumatic. The cribriform niche adjacent to the middle turbinate anteriorly, superiorly and lateral walls of the sphenoid are the most common sites for congenital defects. Head trauma, blunt and penetrating facial injuries are responsible for 90 % of all cases of traumatic CSF leaks. It could be immediate (within 48 h) or delayed. In traumatic cases, the leak is usually at the cribriform plate and fovea ethmoidalis. Immediate CSF rhinorrhea is due to a bony defect or fracture in conjunction with a dural tear. Delayed CSF leak usually presents within 3 months after the injury and is due to an intact dural layer slowly herniating through the bony defect, finally tearing it and causing CSF leak. Occasionally these leak occurs only after the masking hematoma dissolves.

The cribriform area is particularly vulnerable to development of spontaneous leaks because of the presence of maldevelopments with extension of the subarachnoid space through the foramina of the cribriform plate [1]. Spontaneous leaks usually manifests in adulthood and accounts for 4 % of CSF leaks [2, 3]. The dura of the anterior cranial base is subject to wide variations in CSF pressure due to normal arterial and respiratory fluctuations and Valsalva like manoeuvres during nose blowing or straining. It may be secondary to elevated intracranial pressure (ICP) due to idiopathic intracranial hypertension (IIH) or obstructive sleep apnea. Elevated ICP results in bone remodeling and thinning due to the pressure exerted and hence a defect is formed. This further leads to formation of a meningocele or an encephalocele, depending on the size of the defect. Other nontraumatic causes of spontaneous CSF rhinorrhea include focal atrophy, rupture of arachnoid projections that accompany the fibers of the olfactory nerve, and persistence of an embryonic olfactory lumen.

Iatrogenic causes include neurosurgical and otolaryngologic surgeries including functional endoscopic sinus surgery. The anterior skull base is the most frequent site for iatrogenic defects. In endoscopic sinus surgery, the lateral lamella, cribriform plate and posterior ethmoid roof are sometimes accidentally breached. In neurosurgical procedures, CSF leaks may occur less commonly after transsphenoidal pituitary surgery. Most of the CSF leaks are unilateral, and only few bilateral and multiple CSF leaks have been reported [4, 5].

CSF rhinorrhoea is diagnosed by clinical, biochemical and radiological examination. β -2 transferrin is a protein present in CSF, perilymph and aqueous humor [6]. β -2

transferrin provides an accurate, non invasive method to establish the diagnosis of an active CSF leak but does not provide information on the location of the leak. However, testing nasal secretions for β -2 transferrin is a specific and sensitive technique with low chances of false positive and false negative [7].

High resolution Computed Tomography (HRCT) scan gives good bone details regarding fracture or defect and indirect evidence of CSF fistula. HRCT can accurately localize fistula from 22 to 100 %, with greatest accuracy in anterior cranial fossa [3, 8]. A Computed Tomographic cisternography (CTC) is more helpful in defining the exact site of leak and the extend of bone defects in patients with CSF rhinorrhoea [2]. CT cisternograms are performed after injection of contrast material, usually iohexol and iopamidol, as they have low incidence of side effects as compared to older compound like metrezamide [9]. CTC is more helpful with leaks in the sphenoid or frontal area, as these sinuses act as reservoirs in the case of persistent but not intermittent leaks. The main drawbacks of CTC scans are, the bony defect may not correspond to the site of dural tear and their high radiation to the lens. It is invasive and time consuming, and requires a lumbar puncture and intrathecal injection of contrast. It also requires the patient to have an active leak at the time of the investigation. The side effects include headaches, seizure and infection. Intrathecal fluorescein endoscopy can be used to locate the site of CSF leaks intraoperatively. A few reports of reversible complication when using intrathecal fluorescein are mentioned in the literature [10–12], though most of the studies show that fluorescein can be safely used [13–15].

The role of MRI in the evaluation of patients with CSF fistula results from its superior demonstration of CSF and its continuity through dural fistula with the paranasal sinuses or the middle ear [16]. A fast spin echo T2 weighted sequence is also useful if a soft tissue mass is present in the sinus, to differentiate between mucosal oedema or tissue herniation such as meningocele or meningoencephalocele. It is not necessary in the setting of trauma as it does not image bone very well. It is non invasive and has no radiation risk.

An increased intracranial pressure leads to headache and visual disturbances. Also, associated symptoms can assist in localizing the leak, e.g. anosmia indicates an injury in the olfactory area and anterior fossa. Optic nerve deficits suggests a lesion in the region of tuberculum sellae, sphenoid sinus, or posterior ethmoid cells. Patients with recurrent meningitis, especially pneumococcal meningitis, should be evaluated for a defect that exposes the intracranial space to the upper airway, regardless of the presence or absence of CSF rhinorrhea.

An untreated CSF leak poses risk of ascending infection resulting in meningitis in 9–41 % of untreated fistulas and recurrent pyogenic meningitis [3]. Endonasal endoscopic approach has a success rate of over 90 % with lower

morbidity as compared to intracranial techniques. Over the years, many types of graft materials have been used to repair these anatomical defects. They can mainly be divided into pedicled grafts, and free grafts which may be simple mucosal grafts or composite grafts. These grafts may be laid in a single or multilayer fashion. Tissue adhesives such as tissue or fibrin glue have also been used to maintain the position of these grafts with or without degradable packing substance such as absorbable gelatin sponge. The aim is to avoid displacement of the grafts during removal of nasal packing post-operatively.

Surgical techniques for the closure of defects can be employed by underlay or onlay techniques or by obliterating the affected sinus. Once the defect is identified, the surrounding mucosa should be debrided before the laying of grafting material because sinus mucosa contains mucus producing glands which can lift the graft from the recipient bed [17]. When operating in the sphenoid sinus, if obliteration of the sinus is desired, all of its mucosa is stripped. This is to prevent the formation of mucocele by any mucosal remnants. Fat obliteration is preferred because it maintains its size and consistency as opposed to muscle that shrinks to a fibrous reticulum [18].

The role of a lumbar drain is still debatable and its use is a matter of surgical preference. It reduces the intracranial pressure at the skull base and prevents displacement of the graft [19]. Post operative lumbar drain may be necessary in high pressure leaks or recurrent leaks. Many argue that a lumbar drain just lengthens the hospital stay and increases the risk of ascending meningitis [20].

Endoscopic repair is less suited for defects in the frontal sinuses with prominent lateral extension and for defects greater than 1.5 cm in diameter involving the skull base. Multiple bilateral traumatic defects are generally repaired trans-cranially. Inaccessibility to the skull base limits the use of an endoscopic approach, especially on the posterior wall of the frontal sinus or due to the extensive size of the frontal defect. Hegazy et al. [21] did a meta analysis of previous studies and concluded that endoscopic technique has a success rate of 90 % after the first attempt, and a 97 per cent overall success rate after the second attempt. The risk of major complications from a endonasal endoscopic approach for CSF leak repair is less than one percent [18]. These include meningitis, subdural haematoma, intracranial abscess and pneumocephalus [21].

Conclusion

Very few cases of bilateral, multiple and with a long recurrence interval of CSF rhinorrhea have been reported in the literature especially without a history of recent trauma. Our case report is a rare presentation of delayed

traumatic or spontaneous CSF rhinorrhea. The aetiology for this may be attributed to risk factors like middle age, obesity, and hypertension, which might have lead to increased ICP. Surgical repair of the defect is preferred to prevent recurrent meningitis. The endonasal endoscopic approach with its short hospital stay, minimal morbidity, preservation of olfaction and the possibility of revision by the same route, along with its limited disadvantages, is now regarded by most, as the preferred mode of treatment of CSF rhinorrhoea. Long-term success following endoscopic repair is very good and is determined by the etiology of the leak, with higher failure rates among patients with increased intracranial pressure and other risk factors.

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Compliance with Ethical Standards

Conflict of interest All authors declare that they have no conflict of interest. This material has never been published and is not currently under evaluation in any other peer reviewed publication.

Ethical Approval This case report does not contain any studies with human participants or animals performed by any of the authors. All procedures performed involving human participant 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 the patient included in the study.

References

- Milford CA (1997) In: Mackay IS, Bull TR (eds) Scott Brown's otolaryngology. Rhinology 4: 14/1–4/12/12
- Colquhoun IR (1993) CT cisternography in the investigation of cerebrospinal fluid rhinorrhea. Clin Radiol 47:403–408
- Eljamel MS, Fay PM (1990) Acute traumatic CSF fistula: the risk of intracranial infection. Br J Neurosurg 4:381–385
- Ramsden JD, Rogan C, Bates G (2000) Bilateral cerebrospinal fluid rhinorrhoea. J Laryngol Otol 114:137–138
- Shetty P, Shroff M, Kirtane M, Karmarkar S (1997) CSF otorhinorrhea in patients with defects through the lamina cribrosa of the internal auditory canal. AJNR 18:478–481
- Schlosser RJ, Blger WE (2004) Nasal cerebrospinal fluid leaks; critical review and surgical consideration. Laryngoscope 114:255–265
- Skedros DG, Cass SP, Hirsch BE, Kelly RH (1993) Sources of error in use of β -2 transferrin analysis for diagnosing perilymphatic and cerebral spinal fluid leaks. Otolaryngol Head Neck Surg 109:861–864
- Llyod MN, Kimber PM, Burrows EH (1994) Post traumatic CSF rhinorrhoea: modern high definition computed tomography is all that is required for the effective demonstration of the site of leakage. Clin Radiol 49:100–103
- Manelfe C, Guirand B, Tremoulet M (1997) Diagnosis of CSF rhinorrhea by computerized cisternography using metrizamide. Lancet 2:1073
- Mahaley MS Jr, Odom GL (1996) Complication following intrathecal injection of fluorescein. J Neurosurg 25(3):298–299
- Wallace JD, Weintraub MI, Mattson RH, Rosnagle R (1972) Status epilepticus as a complications of intrathecal fluorescein. J Neurosurg 36:659–660
- Moseley JJ, Carton CA, Stern WE (1978) Spectrum of complications in the use of intrathecal injection of fluorescein. J Neurosurg 48(5):765–767
- Roberto E, Guimaraes S et al (2002) Localization of skull based cerebrospinal fluid leaks by using intrathecal fluorescein in a hypodense solution during the surgery. Rev Braz Otorrinolaringol 68(6):788–792
- Lund VJ, Savy L, Llyod G et al (2000) Optimum imaging and diagnosis of cerebrospinal fluid rhinorrhea. J Laryngol Otol 114:988–992
- Lund VJ (2002) Endoscopic management of cerebrospinal fluid leaks. Am J Rhinol 16:17–23
- McConachie NS, Joseph BV, Eljamel MS, Stafford Johnson BD (1995) Magnetic resonance cisternography of CSF otorhinorrhea. Int J Neuroradiol 1:177–181
- Schlosser RJ, Bolger WE (2002) Traumatic cerebrospinal fluid rhinorrhoea. Otolaryngol Head Neck Surg 13:302–308
- Wormald P, Mc Donogh M (1997) Bath-plug technique for the endoscopic management of cerebrospinal fluid leaks. J Laryngol Otol 3:1042–1046
- Casiano RR, Jassir D (1999) Endoscopic cerebrospinal fluid rhinorrhoea repair: is a lumbar drain necessary ? Otolaryngol Head Neck Surg 121:745–750
- Briggs RJS, Wormald PJ (2004) Endoscopic transnasal intradural repair of anterior skull base cerebrospinal fluid fistulae. J Clin Neurosci 11:597–599
- Hegazy HM, Carrau RL, Snyderman CH, Kassam A, Zweig J (2000) Transnasal endoscopic repair of cerebrospinal fluid rhinorrhoea: a meta analysis. Laryngoscope 110:1166–1172