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Original Research Article

Endoscopic cerebrospinal fluid leak repair combined with thecoperitoneal shunting for spontaneous cerebrospinal fluid rhinorrhea: MERF experience

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ABSTRACT

Background: Spontaneous cerebrospinal fluid (CSF) rhinorrhoea is encountered as a diagnosis of exclusion in clinical practice once all other aetiologies have been meticulously ruled out and can mostly present from preformed pathways due to benign intracranial hypertension (BIH) over time. Such leaks need to be clearly located for planning appropriate repair along with adequate control of CSF pressure to avoid recurrence. Thus, the aim of our study was to review the efficacy and outcomes of a combined endoscopic multilayered repair along with simultaneous theco-peritoneal shunting (TPS) for patients with spontaneous CSF rhinorrhoea in the background of BIH.
Methods: Retrospective observational study analyzing the clinical presentations, surgical techniques and the immediate / long term outcomes of this combined approach.
Results: The 46 patients who had endoscopic CSF repair with TPS done between 2008-2019 were reviewed. The mean opening CSF pressure in these cases were >25 cm H_2O. The commonest site of leak was from cribriform plate followed by fovea ethmoidalis. Multilayered closure was done with autologous grafts along with TPS simultaneously. The 92% were successful, 6% had shunt problems needing revision shunts and 2% also needed revision endoscopic repair.
Conclusions: Overall outcomes shows that the combined approach was safe and effective in arresting the CSF leak in 98% patients. Therefore, the rationale for a combined approach for the management of spontaneous CSF leaks in the background of BIH stands proven based on our Institutional experience.

Keywords: Spontaneous CSF rhinorrhea, Endoscopic repair, BIH, Intracranial pressure, TPS

INTRODUCTION

Spontaneous CSF rhinorrhoea was first described by Miller.[1] Non-traumatic/spontaneous or primary CSF fistula accounts for 3-4% of all CSF rhinorrhea and can be further classified into high pressure and normal pressure leaks.[2] Due to the serious potential complications of CSF rhinorrhea like meningitis, encephalitis and pneumocephalus, prompt management with repair of all CSF rhinorrhea cases should be attempted. CSF leaks are often challenging to diagnose, since clinical features maybe subtle and may be present for years before manifesting with morbidity. Spontaneous CSF leaks are known to have the highest rate of recurrence, mostly due to raised intracranial pressure (ICP) post-surgery.[3]

As per protocol a diagnostic lumbar drainage and neuro-ophthalmic examination is recommended to rule out BIH prior to surgery in all cases, although the diagnostic features do not become apparent until the leak is actually sealed. There have been situations experienced previously in our institute, wherein BIH manifested following repair leading to failure of seal at leak site and further leaks at a different site. This ironical situation

triggered interest in the study whereby a combination of endoscopic leak repair performed along with simultaneous CSF diversion was anticipated to be safe and efficacious providing optimal wound healing time while avoiding post-operative risk of raised ICP.

Chaaban et al on the basis of their 5-year prospective study on 46 patients with spontaneous CSF leaks, concluded that successful treatment of elevated ICP in combination with endoscopic repair can provide high success rates (93% primary and 100% secondary) approaching that of other etiologies.[4] Previous to this current study period, an institutional audit showed about 25-30% recurrences among the spontaneous CSF rhinorrhea patients which had been managed only by endoscopic repair. This situation had triggered interest in the experimentation of a novel combination of leak repair performed along with simultaneous CSF diversion to bring down ICP. The combination proved safe and efficacious providing optimal wound healing time while avoiding post-op raised ICP. The reflection of long-term outcomes from this large cohort over 10 years has been discussed in this study.

The main objective of this study is to highlight the surgical technique and to determine the efficacy and immediate/long term outcomes of this combined single-stage approach.

METHODS

This is a retrospective observational study conducted in our tertiary care centre. This study included a total of 46 consecutive patients who underwent endoscopic repair for CSF rhinorrhea along with TP shunting as a combined procedure over a period of 10 years from 2008 was performed, with a follow-up period of 18 months for each patient.

Patients diagnosed as spontaneous CSF rhinorrhea who underwent the combined approach were included in the study. The exclusion criteria were post- traumatic/ surgical CSF rhinorrhea, revision cases and patients undergoing surgical techniques other than the combined endoscopic CSF leak repair with TPS shunting.

Details pertaining to the various clinical presentations, surgical techniques and hospitalization details were collected from our medical records department after obtaining the institutional ethical clearance and patient's consent. The diagnosis was confirmed based on the aetiology, diagnostic nasal endoscopy, CSF analysis by Tau protein / beta 2 transferrin assay, HRCT/ T2-MRI and CT/MR cisternography. The management protocol followed is depicted in Figure 1. All the patients underwent endoscopic CSF leak repair (multi-layered technique of defect closure) performed by otorhinolaryngologists with simultaneous TP shunt placement by the neurosurgeons. Postoperatively, patients were reassessed for the development of elevated ICP and TP shunt functioning. Patients were followed up on regular basis. Outcomes were statistically analyzed using t test with significance at $p<0.05$.

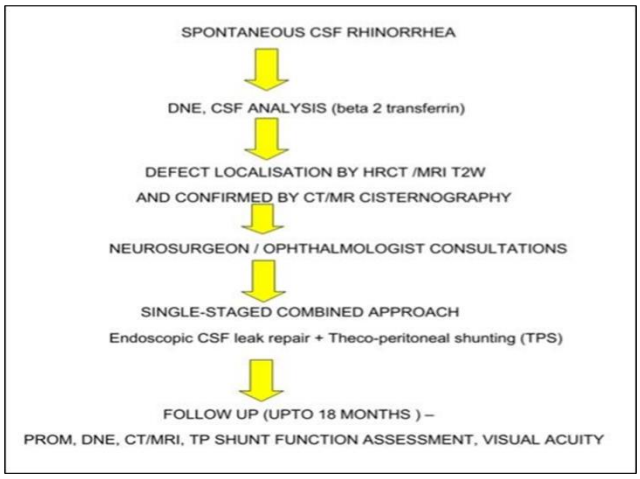

Figure 1: Institutional protocol for management of spontaneous CSF rhinorrhea.

DNE-Diagnostic nasal endoscopy, PROM-patient related outcome measure.

RESULTS

The most common age group among our patients was above 50 years (58.6%). The male: female ratio of 1:6 (6 males, 40 females). In our cohort, the maximum number of patients (56.5%) were just about overweight, followed by 32.6% patients who were obese (BMI >29.9). Patients demographical and clinical data are detailed in Table 1. Among 46 patients, the most common major presenting major symptoms encountered by patients were clear rhinorrhea (98%). 33 patients (71.7%) had presented with headache, 7 patients (15.2%) had anosmia and 4 patients had reduced visual acuity at the time of presentation.

Table 1: Summary of the clinical and operative data.

Variables	Data
Age (Years)	>50 (58.6%)
Gender	6 males, 40 females
BMI (kg/m^2)	26-29.9=56.6%
	>29.9=32.6%
Pre-operative clinical presentations	Headache=33 (71.7%)
	Anosmia=7 (15.2%)
	Visual acuity (<20/20)=4 (8.6%)
	Opening CSF pressure range (31±6.4 cm H_2O)=39 patients
	Meningocele=3 patients
Operative findings	Site of defect, CP=21, FE=8, FR=5, LrS=6, CP+FE=4, CP+LrS=2
	Size of defect, 5-7 mm= 84.4%
TPS related complications	Low pressure headache=4
	TP shunt migration=1
	TP shunt obstruction=1

BMI-body mass index, CP-cribriform plate, FE-Fovea ethmoidalis, FR- Frontal recess, lrS-Lateral recess of sphenoid.

All patients underwent lumbar puncture and opening pressure was checked pre-operatively. CSF analysis was found to be normal in all of them. Opening CSF pressure range is generally about 21-33 cm of H_2O. Patients were grouped based on the opening pressure into the following classes-7 patients had opening pressure between 20-25 cm of H_2O, 39 had opening pressure greater than 25. CSF leak was seen commonly through the Cribriform plate region in 21 patients (45.7%) followed by Fovea ethmoidalis in 8 patients (17.3%), through frontal sinus/recess (5 patients, 10.8%), lateral recess of sphenoid in 6 patients (13%) and 6 out of 46 had a combination of these sites. Also 2 patients presented with bilateral leaks which were sealed simultaneously.

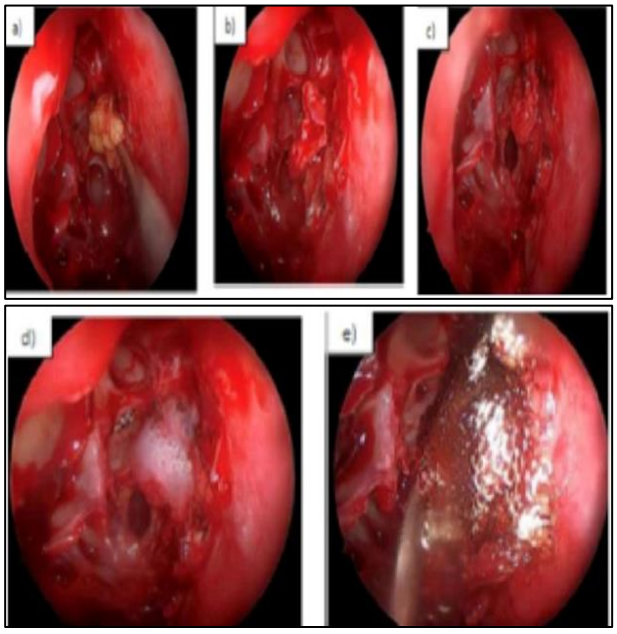

Figure 2 (A-E): Endoscopic images of steps of layered closure of skull-base defect. Steps-fat, abdominal fascia, tissue glue and surgical.

The leak size was determined by high resolution CT and measurement of the defect with curettes in the surgical field. In 84.4% patients it was 5-10 mm (most of them being 5-7 mm in size), while 6.5% had 11-15 mm size of leak and 8.7% patients had a leak size <5 mm. Depending on the size and location of cranial defect, layered closure of the CSF leak site (Figure 2) was widely used by underlay/overlay or combined technique. In our cohort, graft materials used for repair of the skull base defect was fat with fascia lata (50%), septal flap (23.9%), middle turbinate flap (19.6%) as most of the defects were less than 8 mm. For larger defects more than 10mm the naso-septal (Haddad) flap was used (6.5%).

Meningoencephalocele was noted in 2 patients with an opening CSF pressure of 36-40 cm of H_2O and the other patient had an opening CSF pressure of 31-35 cm of H2O. Bipolar cautery laser or radiofrequency energy may be used to fulgurate a concomitant meningoencephalocele. In the immediate follow up period, 40 patients recovered uneventfully, while 6 patients (13%) developed TP shunt related problems. Of these 6 patients, 4 of them developed low-pressure headache which was transient and was managed conservatively. One patient (2.1%) developed recurrent CSF leak from the same site within 1 month of surgery due to TP shunt obstruction for which combined revision surgery was done. One more patient (2.1%) also presented with signs of raised ICP due to TP shunt migration within 1 month of surgery and so, repositioning of the shunt was done and this was sufficient to bring down the ICP. Hence the overall success rates of this novel combined procedure were noted to be 95.7% with failure rate being 4.3%, which was in keeping with the results across world literature for spontaneous leaks.

During the follow-up period, our series showed a significant (p=0.001) improvement in headache at the 1st week post- surgery follow-up itself. This continued throughout the period of follow-up with no recurrence. Pre-operative visual assessment in our series showed 4 patients (8.6%) with below normal visual acuity (less than 20/20 Rosenbaum chart). Post-surgery all the patients showed improvement in visual acuity from the first follow-up onwards. There was progressive visual acuity improvement during all follow-up and we had a complete resolution of impaired visual acuity in all the patients. None of these patients had papilledema in the long term follow up.

DISCUSSION

Most of the spontaneous leaks are considered due to elevated ICP (which may be subclinical and undiagnosed), leading to disruption of the arachnoid and dura with pressure induced erosion of the bone leading to fistula formation.[5] Mirza et al in their 10 years retro-prospective analysis, observed that 46% of their patients (13 of 29 patients) with spontaneous CSF leaks had evidence of raised ICP /BIH and also 6 of those 13 patients with raised ICP (46%) had a recurrence of leaks.[6] Normal pressure CSF leaks have also showed recurrences if not managed thoroughly. If the patient was actively leaking CSF at the time of lumbar puncture, the opening pressure may be normal, because the leak has decompressed the ICP; thus, the increased pressure may only be apparent after a site of leakage has been surgically repaired. This has been studied well and published by Schlosser and colleagues.[7] Some patients may have symptoms of increased ICP even when the leak is active but most often, patients develop symptoms and signs of intracranial hypertension only after the CSF leak has been repaired, likely because the leak acts as a route of spontaneous CSF diversion.[8] Numerous studies have documented raised ICP in the post-operative period.[9,10]

In our cohort, the maximum number of patients were females (86.9%) and within the age-group of 50-60 years (78.3%) which was consistent with literature.[2,4] In our cohort, the maximum number of patients (56.5%) were

just about overweight, followed by 32.6% patients who were obese (BMI >29.9), which is in contrast to literature stating spontaneous CSF rhinorrhea to be more common among the obese.[10,11]

Literature frequently quotes the cribriform plate and the lateral recess of the sphenoid sinus, as the most common locations of spontaneous CSF.[10,12] In our study, the most common site of spontaneous leak was the cribriform plate (45.7%). Meningoencephalocele was noted in 3 out of 46 patients, of which 2 patients had defect in the sphenoid sinus. Meningoencephaloceles are reported to be more common among sphenoid sinus leaks due to congenital dehiscences in the lateral sphenoid roof that occur as a result of persistence of the lateral craniopharyngeal canal (Sternberg canal). The pulsatile CSF pressure results in erosion of bone and protrusion of brain and/or meninges forming meningocele and/or encephalocele. Therein pulsatile pressure finally causes rupture of arachnoid without having support of underlying bone leading to formation of fistula.[13,14] In our study meningoencephalocele was found to be associated with high CSF opening pressure (p=0.022). This is consistent with the study by Omer et al which showed meningoceles are significantly more common in patients with increased ICP s than in the controls and can be considered as an additional imaging sign for BIH.[15]

TPS is a better option for long term CSF diversion with lesser morbidity. Once in place this Theco-peritoneal shunt is used to drain the excess CSF from the brain via the subarachnoid space and transport it into the sterile peritoneal cavity, where it is eventually absorbed. The TPS thus decreases the ICP in the post-operative period and provides a dry seal for the repaired fistula in the anterior cranial base. This technique is well established in literature and has helped to reduce the recurrence rates both in traumatic and in spontaneous CSF fistulas. A small number of our patients developed shunt related problems. Four patients presented with persistent low-pressure headache on upright posture in the post-operative period, which was transient in nature and weaned off over time. They were managed conservatively with appropriate bed rest and analgesics. One of the patients developed TPS obstruction leading recurrence of CSF rhinorrhea from the same site, for which a revision combined approach surgery was done. One other patient presented at 1month follow-up with shunt failure due to migration of the thecal end of the catheter, for which repositioning of TPS was done, as the patient had developed symptoms of raised ICP. Shunt migration towards peritoneal cavity has been reported in literature and can be due to the intestinal peristaltic movement, which created a pulling effect on the shunt tube. Valveless shunt systems in all our patients, but we did not have any incidence of over drainage complications such as Arnold Chiari malformation as suggested in literature.[16,17] It has been suggested that placement of short length of thecal end catheter could result in over drainage complications.[18] Hence, intrathecal catheter length as 10cm in all the cases and this found to be a desirable length to reduce the low-pressure symptoms. Over drainage complications could be reduced by using programmable shunts.[19]

Only one of our patients had developed TP shunt block in the post-operative period which was diagnosed by TP shuntography. The intrathecal administration of in-111 DTPA (diethylene-triamine-pentaacetic acid) and sequential images of the abdomen and of the head can also be used to assess shunt patency.[20] The risk of infection of the TP shunt is known to be very low at less than 3%, in comparison to lumbar drains.[19] There were no TP shunt / wound infection in our series. Currently with an experience of over 10 years using this combined approach, being performed by our well-trained skull base team of ENT and neurosurgeons who work in synchrony, this novel approach has become our institutional protocol for managing spontaneous leaks. The overall long-term results which we achieved with this combined approach has helped us to propose this technique as a safe and effective option for the surgical management of CSF leaks especially in the background of suspicious BIH.

Clinical significance

There is currently no perfect one-stop surgical treatment for spontaneous CSF rhinorhea, since there is a background of raised ICP playing a part in their surgical failures. Though endoscopic CSF repair is the gold-standard primary treatment, it may not suffice if the opening CSF pressure is high which suggests a suspicion of BIH. In such cases it is prudent to combine the leak repair along with CSF diversion. The decision regarding CSF diversion techniques in a particular patient must be individualized and should be decided by a multidisciplinary approach, based on available literature and surgeon's experience. Our novel combined approach has stood the test of time proving to be safe and effective in the long term and we recommend this protocol to achieve the most optimal outcomes when managing this challenging entity.

CONCLUSION

The results of this study allow us to make a number of observations regarding the efficacy of TPS for spontaneous CSF rhinorrhea. A functioning TPS is an effective treatment in alleviating the signs and symptoms of idiopathic intracranial hypertension. The recurrence rate of spontaneous CSF rhinorrhea is negligible in patients with this combined endoscopic repair with TPS, which proves the safety and efficacy of this single-stage procedure. A substantial percentage of patients who undergo placement of a TPS for spontaneous CSF rhinorrhea do not require a revision of the shunt.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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