Abstract

We present our experience in managing spontaneous CSF rhinorrhoea which could be devastating if untreated. This is a retrospective study of all the cases of spontaneous CSF leaks treated by the first author between 1991 and 2016 in a tertiary referral centre.

A total of 450 patients underwent endoscopic repair of CSF fistula, with an overall success rate of 98.8%. 14 patients required revision, of which nine patients underwent second surgery successfully. The remaining opted to get treated elsewhere. Apart from three patients who had minor post operative early meningitis, which were treated promptly, no serious complications were encountered. Patient follow up ranged from 6 months to 25 years.

The principles of diagnosis of CSF rhinorrhoea, surgical management and results are discussed. Precise identification of the bony defect, meticulous preparation of the graft bed, careful elevation of dura, placement of cartilage in the extra-dural pocket and judicious use of just enough tissues and adequate graft support are key factors in a successful repair.

ABBREVIATIONS:

CSF: Cerebrospinal Fluid; CT: Computed Tomography; SE: Sphenoidethmoid

INTRODUCTION

CSF rhinorrhoea was first described by Miller in 1826 [1]. The first surgical intervention for repair of cerebrospinal fluid leak was carried out by Dandy in 1926 [2] by using an intracranial approach through a bifrontal craniotomy. This condition was exclusively treated by neurosurgeons, transcranially or through external transtentorial approaches, until endoscopes were introduced for nasal and sinus diseases. Wigand in 1981[3] and Stankiewicz in 1987[4] respectively, were the first to report on endoscopic repair which improved success rate to more than 90%. Thus, through the decades, trans-cranial approaches have been gradually replaced by endonasal endoscopic surgery, greatly reducing the number of complications like intracerebral hemorrhage, retraction related brain edema and recurrence rates.

Since our first repair which was a pure accident in 1991, we have done endoscopic repair of over 450 cases of spontaneous CSF rhinorrhoea with skull base defects in the last 25 years. In this article we present our diagnostic protocol, evolution of surgical technique, pearls, perils and results.

MATERIALS AND METHODS

A total of 450 patients (290 females, 160 males with a mean age 45 years) were clinically diagnosed with CSF rhinorrhoea. We have confined our study to the cases of spontaneous leaks. The duration of symptoms ranged from 3 months to 2 years. All patients underwent thorough clinical examination. The two important tests we rely on for the diagnosis of a CSF leak are (i) the CSF drip test (Figure 1) and (ii) high resolution computed tomography (CT). With 1 mm slices we are able to detect the bony defect in the vast majority of the cases. Sometimes a 3 mm thickness routine CT of paranasal sinuses reveals the defect (Figure 2).
Patients are taken up for surgery under general anesthesia, with systemic antibiotic cover. In all our cases, an endoscopic sphen-o-fronto-maxillo-ethmoidectomy is done as the initial step. The middle turbinate is sacrificed if the defect is in the cribriform plate or lateral lamella. In all cases, invariably, a glistening white arachnoid cyst, pseudo meningocele, or meningocele was seen at the site of defect. The arachnoid cyst or pseudo cyst may present some distance away from the skull base. It is followed up to the bony defect. The margin of the defect is clearly defined and graft bed is prepared by removing mucosa and soft tissues in the vicinity. Bare bone, all around the defect is an ideal graft bed. Diathermy may be used to burn the excess soft tissue or reduce an encephalocele. The size of the bony defect ranged from 3 to 20 mm in its maximum diameter, with a mean of 10 mm. In defects larger than 4 mm, we created an extra-dural pocket on the cranial side, to receive an inlay cartilage graft. The cartilage graft should be 2-3 mm larger than the bony defect in at least two opposing directions (Figure 3). The insertion of the cartilage through the bony defect into the extra-dural pocket is done carefully. A piece of temporalis fascia is used on the nasal side to cover the bare bone around the defect and the cartilage graft. In defects smaller than 4 mm, we used only an onlay graft. Merocel® is used to support the grafts from below after placing an intervening layer of plastic sheet. We used the bath plug technique only in 5 cases and sphenoid sinus obliteration with fat in 14 cases.

Patients were made ambulant from the first post operative day. They were prescribed post operative antibiotics. The CSF leak continues for a few days after the repair, before it stops. A patient was discharged after 72 hours and packs removed after 1 week.

RESULTS

Complete overall stoppage of rhinorrhoea was observed in 98.8% of our cases. In 14 patients, a recurrence was noted, of which, 9 were successfully repaired at the second attempt and the rest were lost to follow up. There were no procedure related complications. 2 patients had meningitis post surgery, due to sinus infection, which was treated with intravenous antibiotics. The largest defect encountered was 2 x 1.5 cm in the fovea ethmoidalis, which received a septal cartilage graft in the extra-dural pocket. The longest patient follow up period was 25 years and the shortest was six months. The median follow up period was 18 months.

DISCUSSION

Abnormal communication between the subarachnoid space and nasal cavity is called cerebrospinal fluid fistula. When it occurs without any trauma it is termed as non-traumatic spontaneous leak. Non-traumatic spontaneous CSF rhinorrhoea forms a distinct group of patients presenting with CSF rhinorrhoea. In the cribriform plate area the preformed perforations which transmit the olfactory nerve fibres predispose to CSF leak. Studying the anatomy of this area one finds that the dura, the toughest layer of the meninges, passes through these perforations, only to continue as the peristeum on the nasal side (Figure 4). The pia and arachnoid continue as the perineurium of the nerves, which is the weak layer. With the dura constantly pulsating and compounded by raised intracranial pressure the pia and arachnoid prolapse by the side of the nerves, making the normal perforations in the cribriform plate into a pathological defect. The prolapsed pia and arachnoid form a cyst filled with CSF on the...
nasal side which ruptures to produce CSF leak [5]. In a few cases we have noted pulsating arachnoid cyst on the nasal septum some distance away from the skull base, and also on the vertical lamella of the middle turbinate and both of them pulsating; on dissecting the arachnoid cysts, they led to a common defect in the cribiform plate indicating that pia-arachnoid is the weak spot in the cribiform plate anatomy and the cysts have followed the path of the olfactory fibers which were forced to run over the septum and middle turbinate. Since the CSF lacks the cells for self repair, the fistula continues to leak. For this reason, we are of the view that "Spontaneous CSF rhinorrhoea, does not close spontaneously" and it has to be closed surgically.

Non-traumatic spontaneous CSF leaks from the fovea ethmoidalis, sphenoid sinus and frontal sinus are developmental in nature and need to be surgically closed too. As observed by D S Deenadayal et al [6] we also noted obese females in the 5th and 6th decades are the commonest group to suffer from spontaneous CSF rhinorrhoea. Badia et al. [7], also suggested a relationship between obese females and increased risk of developing a primary spontaneous CSF rhinorrhoea.

As for the diagnostic protocol, we keep it simple. The CSF drip test is the most important test for us. Crystal clear, watery fluid coming out one nostril, is unlikely to be anything other than CSF. By demonstrating the CSF drip, the opposite side is eliminated from exploration. If enough CSF can be collected, we send it for biochemical studies. Nandagopal et al.[8], stated Beta-2 transferin as a valuable and sensitive means of confirming the diagnosis of CSF leaks. Since β₂-transferin is not widely available in India, except in one centre, we have not done that in any of our cases. In sporadic leakers, CSF drip may not be positive in a single visit. Patients may be asked to come again at a time when it is leaking. Alternatively, patients may be provided a container to collect the watery fluid whenever it comes.

Imaging studies are paramount in the workup of presumed CSF leak [9]. Numerous investigations like cisternography and intrathecal fluorescein have been advocated for diagnosis in literature [10]. Being a referral centre, patients already carry films of imaging studies done elsewhere. If not, we order high resolution 1mm slices of CT Scan of the skull base which readily reveal the bony defect in vast majority of the cases, a fact corroborated by Lloyd et al [11]. Sometimes, a 3 mm thickness routine CT Scan of the paranasal sinuses reveal the bony defect and we do not order any other imaging study at all (Figure 1).

We look for the arachnoid cyst or pseudo meningocele on the table. White, glistening cysts of varying sizes may be seen by shifting the middle turbinate laterally in cribiform plate defects. Wherever there is prolapse of the meninges, they appear typically white, quite different from the nasal mucosa. Large meningoceles and encephaloceles are useless part of the brain and they are to be sacrificed. Sometimes pulsating arachnoid cysts could be seen on the septum, some distance away from the skull base. They are typical arachnoid cysts following the path of olfactory fibres. The arachnoid cysts, pseudo-meningocele, meningoceles and encephaloceles give the surgeon a rough indication of the bony defect in the vicinity. The prolapse of the meninges and arachnoid cysts resulting from lateral lamella defects and fovea ethmoidalis cannot be readily visualized unless an ethmoidectomy is done.

We do an endoscopic sphenofronto-maxillo-ethmoidectomy as a first step in all cases for a wide exposure of the skull base. We adhere to strict mucosa preservation techniques. Small arachnoid cysts are reduced by diathermy. Larger meningoceles and encephaloceles are removed and sent for histopathology. All soft tissues including the mucosa is removed in the region of the bony defect. Whenever there is prolapse of brain tissue into the nasal cavity it is separated and returned to the cranium. Bare bone all around the bony defect is an excellent graft bed. In small defects less than 4mm in diameter, a simple onlay connective tissue such as temporalis fascia or fascia lata is adequate. The graft is trimmed to fit the size of the graft bed and supported from below by Merocel®. Often we find the bone in the vicinity is extremely thin and while trying to delineate edges of the bony defect we end up making the bony defect bigger. It does not matter and it is better if one removes all the fragile bone until thick strong bone is encountered, which provide a stable bed for the grafts (Table 1).

In larger defects, larger than 4mm in diameter, we believe in reconstructing the defect layer by layer. An extradural pocket is created on the cranial side using sharp and blunt instruments. By staying close to the bone on the cranial side the dura can be elevated to create an extradural pocket. A piece of cartilage 2-3mm larger than the bony defect is pushed into the extra dural pocket, until it clicks into place. The cartilage graft is held in place by the weight of the intracranial structures and the pulsations stop immediately. The tragal cartilage is our preferred graft, because it is harvested with the perichondrium intact. The flexibility and the tightly adherent perichondrium on either side does not allow the cartilage to break. Manipulation of this cartilage is easier than the septal cartilage and the layer of perichondrium goes to repair the dural defect. With the insertion of tragal cartilage, duraplasty and skeletal reconstruction are achieved in one shot. An appropriately sized temporalis fascia graft is used to cover the exposed bone and cartilage graft on the nasal side, which is supported from below by Merocel®. We avoid unnecessary stripping of the nasal mucosa; just enough tissues are used, not more not less. The tremendous ability of the nasal mucosa for healing is taken advantage of and we have never felt the need for a pedicled vascularised flap.

We do not believe a vascularised flap improves the success rate as stated by others [12]. There is no substitute for a

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<td>SITE</td>
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<td>Cribriform plate – SE Recess</td>
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meticulous preparation of the graft bed. A vascularised flap on a poorly prepared graft bed can lead to failure. Besides, large vascularised flap [13] distorts the anatomy and physiology of the nasal cavity, and is fraught with healing problems. We are of the view; it is an overkill to mobilize a large mucosal flap about 7 cm x 7 cm, completely denuding the nasal septum and nasal floor to cover a defect of less than 10 mm. Also, more area needs to be denuded to seat the flap in the recipient site. The advocates of large pedicled flap [14] may argue that the donor area heals without any problem, with a little nurturing. The same logic can be applied to the healing of repair site without any pedicled flap. This is supported by the fact that we have excellent success rate in one of the largest series where no pedicled flap is used at all. We are of the view that large pedicled flaps may be reserved for very large skull base defects with large areas of exposed bone [15]. The largest defect we closed is 2 cm x 1.8 cm and we did not use a pedicled flap. Our results are comparable or even better than some of the studies which used vascularised flaps [16]. We have used the bath plug technique and sphenoid obliteration with fat only in a few suitable cases (Table 2).

Chin CJ et al used Dura Seal in five patients to enhance graft strength and form a watertight seal [17]. Nishihira et al., [18] suggested that fibrin glue, by its adhesive sealing properties, enhances the results for the treatment of cerebrospinal fluid rhinorrhoea. Though many surgeons world over use tissue glue routinely [19], we have not used any and we have never felt the need for it. Fibrin present in the blood is a very potent adhesive and all we do is create an atmosphere, conducive for the body's natural healing to take place. Patients are made ambulant from the 2nd post operative day onwards and we have noticed some amount of CSF leak for a few days before the graft gets adherent. Allen et al [20] advocated the use of lumbar drain which was further supported by studies by Ackerman et al [21] who propogated the use of lumbar drain after CSF leak repair as they are of the opinion, that it helps to minimize post-operative leak. We have not used any CSF diversion techniques, though we prescribe tablet Furosemide 250mg twice a day for one month post-operatively.

CONCLUSIONS

Demonstration of the CSF drip test is a very important test for the diagnosis of CSF rhinorrhoea. Crystal clear, watery fluid coming out of one nostril is unlikely to be anything other than CSF. It is the bony defect that we want to visualize and high resolution 1mm thickness CT scan seems to do just that. Sometimes a 3mm coronal CT scan of the paranasal sinuses is enough to visualize the bony defect. Demonstration of CSF drip combined with characteristic CT findings on the same side points to the site of leak. Since the CSF lacks the cells for self repair, non-traumatic CSF rhinorrhea does not close by itself and we postulate the dictum “spontaneous CSF rhinorrhoea does not close spontaneously”. They have to be closed surgically. The technique of placing an extradural cartilage inlay with just enough fascias to cover the raw area on the nasal side for defects larger than 4 mm is standardized. For smaller defects, just a fascia onlay is all that is required.

We have neither used large pedicled vascularised grafts, which leave most of the nasal cavity denuded, nor commercially available tissue glue. There is no statistical evidence to suggest that employment of these two agents improve the results. We are of the view there is no substitute for precise location of the bony defect, meticulous preparation of the graft bed, excision or return of the meningeal tissue to the cranium, and use of just enough tissues for layer by layer reconstruction.

REFERENCES


