ORIGINAL ARTICLE

Cerebrospinal Fluid Rhinorrhea- An Overview

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Abstract

Background:*The commonest cause of cerebrospinal fluid (CSF) rhinorrhea is trauma, especially craniofacial trauma: iatrogenic or accidental.*

This paper presents the clinical features, investigative techniques (most of which are not yet in common use in our environment) and finally, the current treatment modalities of this clinical entity.

Methodology: *A review of literature was done using Medline search, relevant journals and text on the topic were reviewed the treatment modalities for cerebrospinal Fluid Rhinorrhea were highlighted.*

Result: *Road traffic accident is the leading cause with the involvement of motorcycles on the increase. As the motorcycle has been allowed to become an important part of public transportation in our society and laws on the use of protective helmets not strictly adhered to, the incidence of craniofacial injuries and invariably that of CSF rhinorrhea is on the increase.*

Conclusion: *other etiological factors of CSF rhinorrhea should be noted as a small percentage is attributable to non-trauma.*

Therefore, the presence of a clear rhinorrhea following craniofacial trauma for example should raise the suspicion of a CSF fistula.

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Introduction

Cerebrospinal fluid (CSF) rhinorrhea is defined as CSF leakage into the nasal cavity.

This occurs as a result of disruption in the arachnoid and dura mater with an osseous defect in the floor of the anterior cranial fossa coupled with a CSF pressure gradient either continuously or intermittently greater than the tensile strength of the disrupted tissues [1].

First reported in the 17th century, with the first repair utilizing a bi-frontal craniotomy and placement of a fascia lata graft reported by Dandy in the 20th century [2].

The most significant etiological factor is trauma, iatrogenic or accidental especially affecting the craniofacial region and the presence of a clear rhinorrhea following this should draw suspicion to a CSF rhinorrhea [3].

With the motorcycle becoming an important part of public transportation in our environment where riders mostly do not use protective helmets, the incidence of head injuries and invariably that of CSF rhinorrhea has increased [4].

It has also been reported that the incidence of CSF rhinorrhea has increased with the advent of endoscopic sinus and anterior skull base surgeries [5].

It carries the risk of meningitis when ignored, therefore early detection and adequate treatment is essential [6].

In our environment, a recent study found an increase of 3.3% with a male

preponderance [7]. However, 25% was found in patients in Europe [8].

Etiology/Classification

The classification of CSF rhinorrhea is based on its etiology therefore it is classed under 2 main subdivisions as follows;

1. Traumatic a) Accidental which may be,

   i) Surgical and

   ii) Non-surgical

   b) Iatrogenic

2. Non-traumatic (spontaneous) of which there are 2 types,

   a) High pressure and

   b) Normal pressure.

Trauma accounts for about 90% of CSF leaks [9] with 2% to 9% occurring in head injured patients. This incidence

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increases to 25% in those complicated by fractures of the paranasal sinuses [10,11].

CSF leaks may follow such surgical procedures as trans-sphenoidal hypophysectomy and endoscopic sinus surgery.

Traumatic non-surgical leaks are commonly as a result of road traffic accidents and falls from heights.

The onset of traumatic CSF rhinorrhea is variable, being classified as immediate i.e. occurring within 48 hours or delayed. Of the delayed cases, 95% are known to present within 3 months of injury [9]. Most CSF leaks from non-surgical trauma present immediately and 50% of iatrogenic leaks present within the first week.

Non-traumatic high pressure CSF rhinorrhea are rare and they result from pituitary tumors, empty sella syndrome (ESS), hydrocephalus (obstructive or communicating)[12], more recently in acoustic neuroma [13] and benign intracranial hypertension (BIH) [14].

Normal pressure non-traumatic CSF leaks occur in congenital defects in the floor of the anterior cranial fossa, focal atrophy of the meninges or in osteomyelitic erosions of the floor of the anterior cranial fossa [15].

Cerebrospinal fluid leakage from the middle cranial fossa into the nose may result from the persistence of the craniopharyngeal canals [16].

Pathophysiology

About 20mls of CSF is produced per hour by the choroid plexus. It circulates from the ventricles through the foramina of Monro (interventricular foramina) into the third ventricle, aqueduct of Sylvius and the fourth ventricle and out through the foramina of Luschka and Magendie to the subarachnoid space and resorbed by the arachnoid villi.

The total CSF volume is 140mls and its pressure ranges from 40mm of water in infants to 140mm of water in adults.

CSF pressure is maintained by the relative balance between CSF secretion and resorption.

The CSF resorption rate plays a pivotal role in determining CSF pressure [17].

Immediate CSF leakage follows either a dural tear, a bony defect or a fracture. Delayed

Traumatic leakage may possible be caused by a previously intact dural layer that has slowly herniated through a bony defect, finally tearing the dura and causing a CSF leak. According to another theory, the tear and bony defect are present form the time of original injury, but the leak presents only after the dissolution of a masking hematoma [18].

Dura of the anterior cranial base is subject to wide variations in CSF pressure because of several factors including normal arterial and respiratory fluctuations. The other stresses on the dura include Vasalva-like actions that occur during nose blowing. This stress can lead to dural tear in areas of bony floor abnormalities 18. Increased intracranial pressure is not necessary for non-traumatic CSF leaks to occur.

Theories for primary non-traumatic CSF leaks include focal atrophy, rupture of arachnoid projections that accompany the fibers of he olfactory nerve and persistence of an embryonic olfactory lumen [18].

Clinical presentation

Symptoms

The most prominent symptom is a clear nasal discharge which may be provoked or increased by physical work or a change in posture.

Some patients may complain of persistent salty taste in their mouths and others may complain of a persistent headache [1].

A history of headache and visual disturbances indicates increased intracranial pressure.

An antecedent history of trauma or surgery helps in making a diagnosis. Often, a misdiagnosis of allergiv and vasomotor rhinitis is made especially in delayed CSF leakages [19].

Signs

A thorough otologic, rhinologic, head and neck and neurological examination is required which may reveal a n encephalocele or meningocele.

Often, asking the patient to perform a vasalva manouver or compression of both jugular veins can provoke drainage of CSF. This is the *Queckenstedt-Stokey* test [20].

A positive reservoir sign which is the gush of CSF collected in the paranasal sinuses on hange in head position is diagnostic.

In head injured patents, mixture of blood and CSF may make the diagnosis difficult.

CSF separates from blood when placed on filter paper producing clinical detectable ring sign, double ring sign or halo sign. The presence of this ring is not exclusive to CSF and can lead to false-positive results [21].

In contrast to unilateral rhinorrhea, the presence of bilateral rhinorhea gives no clue of the laterality of the defect. However, even in this situation, exceptions have been known to occur. Paradoxical CSF rhinorrhea occurs when midline structures that act as barriers (crista galli, vomer) are dislocated. This allows CSF to flow to the opposite side and presents at the contralateral naris [22].

Investigations

This is to confirm the diagnosis and to localize the site of CSF leak.

The investigations carried out are grouped into laboratory investigations and imaging Studies.

Laboratory investigations

Cerebrospinal fluid is usually clear with a specific gravity of 1.004-1.008 but in contrast to pure nasal secretions it contains glucose which can be tested using the glucose oxidase paper. This test is however unreliable giving false-positive results [23]. The appropriate test to carry out is the measurement of the glucose content of the CSF undertaken carefully to ensure that the result is consistent with concurrently drawn lumbar CSF [24].

However, all positive results should be confirmed with the more reliable immunoelectrophoretic identification of beta2-transferrin (a protein present in perilymph, serum of newborns, vitreous humor, patients with liver disease and CSF but highly specific for human CSF). This is a non-invasive assay with high sensitivity andspecificity [25].

Imaging studies

Plain radiographs are seldom helpful in localizing the site of CSF leakage but can demonstrate a fracture of the skull, an air-fluid level in the sinus or an aerocele in the cranial vault. The presence of a pneumocephalus on plain radiograph is an almost pathognomonic sign of a large tear in the dura [2].

A high resolution computed tomographic (CT) scan is very useful in delineating the

fracture site that underlies a traumatic leak or it may reveal an underlying tumor or hydrocephalus i.e. providing information on the brain parenchyma in the region of the leak. Other features that may be seen are an air-fluid level in the sphenoid sinus and a pneumocephalus [26]. A deviated crista galli is a radiological sign supporting a congenital bony dehiscence as the cause for a CSF rhinorrhea [22].

CT cisternography may identify the site of CSF leak but if it does not, a modified technique called digital subtraction cisternography has been developed to do so [27].

The diagnostic yield of CT scan is further improved by metrizamide CT cisternography (MCTC) which is injecting metrizamide (a water soluble non-ionic tri-iodinated contrast material). Another agent used is iohexol (omnipaque) [28]. These agents are injected intrathecally. They give the precise location of CSF leakage in most patients with active leaks [29]. Injecting these agents carries low morbidity rate but reports of nausea, headaches and acute organic psychosyndromes have been given.

In patients with slow leaks who are poor candidates for MCTC, a repeat study can be done after increasing the intracranial pressure (ICP) by the vasalva maneuver, coughing or by the intrathecal infusion of isotonic sodium chloride (normal saline) solution.

Magnetic resonance imaging (MRI) is not recommended in the evaluation of CSF rhinorrhea because it demonstrates bony defects poorly. However, a heavy T2-weighted image can reveal a brisk leak [30].

Radioactive isotopes like radioactive iodine I 131, radioactive iodinated serum albumin (RISA), ytterbium Yb 169 diethylenetriamine pentaacetic acid (DPTA), indium In 111

DPTA, technetium Tc99m human serum albumin and technetium Tc99m pertechnetate can be introduced into the CSF via lumbar or suboccipital puncture with the distribution of these agents being determined by single photon emission computed tomography (SPECT) or scintiphotography [31]. Another option is the introduction of nasal pledgets which can be analysed for the presence of the tracer.

The drawbacks of using tracers are as follows;

1. They cannot precisely identify the CSF leakage site.

2. Patient positioning can cause distal pledgets to take up isotope incorrectly

3. Readings of radioactivity should be quite high to determine a true CSF leak. Borderline results are unreliable and false-positive results are seen in 33% of patients.

4. Isotope is absorbed into the circulatory system and can contaminate extracranial tissues.

Overpressure radionuclide cisternography (ORNC) is also helpful in confirming the diagnosis of a CSF rhinorrhea [32].

Treatment

Majority of traumatic CSF leaks heal spontaneously but some require surgical repair [36]. Until the leak ceases, the patient is at risk of developing pneumococcal meningitis and brain abscess [6].

Conservative treatment

This involves bed rest for 1 to 2 weeks with the patient nursed in a head-up position. Coughing, sneezing, nose blowing, lifting heavy weights should be avoided. Stool softeners can be used to reduce the strain and increased intracranial pressure associated with bowel movements.
The use of prophylactic antibiotics is controversial even though no prospective controlled study resolves this issue. The impression is that prophylactic antibiotics gives room for the development of resistant organisms. However, in high risk individuals, their use is recommended to reduce the risk of meningitis. Leech and Paterson considered that surgical repair should be done if CSF rhinorrhea persists longer than 7 days as the protection afforded by long time antibiotic prophylaxis diminished after that period [37].

Surgical treatment

This is indicated in the following conditions;

1. Failure of conservative treatment.
2. Patients with large high-volume CSF leaks.
3. Non-traumatic leaks.
4. Prolonged leaks regardless of etiology.
5. Recurrent leaks.
6. Patients with open wounds that are connected to the dural defect.
7. Closed heads injuries with intracranial complications and CSF leaks caused by and detected at intracranial or nasal surgeries.

Two surgical procedures are used in the treatment of CSF rhinorrhea. They are the intracranial and extracranial approaches. While the neurosurgeons prefer the intracranial approach, otolaryngologists prefer the extracranial [38].

Intracranial approach

This has been the standard mode of repair until recently. Advantages of this method of repair include the ability to inspect the adjacent cerebral cortex, direct visualization of the dural defect and better ability to seal the leak in the presence of increased ICP. Even when pre-operative localization of a leak fails, this approach has been done with successful outcome. It entails a frontal anterior fossa craniotomy and different techniques have been used for repair and they include free or pedicled periosteal or dural flaps, muscle plugs, mobilized portions of the falx cerebri, fascial grafts and flaps with fibrin glue [39]. The complications of this approach are increased morbidity, anosmia, hematoma, cognitive dysfunction, seizures, brain edema, hemorrhage. Failure rates of up to 27% are recorded [40].

Extracranial approach

Primary surgical repair of CSF rhinorrhea is usually extracranial unless intracranial exploration is necessary to repair other damages [41]. This method of repair can be accomplished by an external route or by the use of endoscopes.

External approach

Repair of CSF leaks using external routes can be by the following methods;

a. Anterior osteoplastic flap.
b. Bicoronal or eyebrow incision.
c. External ethmoidectomy.
d. Sphenoidotomy which could be transethmoidal or transseptal.
e. Tranantral route.

The graft materials for sealing the leak is the same as those mentioned above. The graft is placed superior to the bony skull base inferior to the dural tear. In all the techniques, lumbar CSF drainage is advisable for a few days post-operatively to maintain a constant low pressure on the closure. The disadvantages of the external approach include the inability to treat concomitant intracranial abnormalities, difficulty with frontal and sphenoidal repair and the relative ineffectiveness of repairing high pressure leaks from below [42].

Endoscopic technique

This is usually successful in 90% of cases following a first attempt at repair [43].

It has more advantages than the external approach in that it gives a better field of vision with enhanced illumination and magnified angle visualization. Other advantages include the ability to clean the mucosa off the adjacent bone without increasing the size of the defect and accurate positioning of the graft material [44]. The use of endoscopic telescopes

in a transseptal transsphenoidal approach to localize sphenoid CSF leaks has also been reported [45].

The success rate for extracranial repair of CSF leaks ranges from 86% to 100% [42].

Management of underlying intracranial hypertension improves the outcome of endoscopic repair of spontaneous CSF leaks with success rates reaching 95% [4]

The future

The ability to precisely localize sites of CSF leakage and hence an increase in the success rates for repair will be further enhanced with the advent of computer assisted image guided surgical techniques.

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