



Spontaneous cerebrospinal fluid (CSF) otorrhoea in Western Australia – an emerging entity?

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Background: Spontaneous cerebrospinal fluid (CSF) otorrhea appears to be an emerging clinical entity. The aetiology, characteristics and treatment outcomes of these group of patients are poorly described so we aimed to further investigate this growing cohort.

Methods: A retrospective review across the three largest tertiary hospitals in Western Australia (WA) over a 5-year period was conducted. Twenty-eight patients with non-iatrogenic, and non-traumatic spontaneous CSF otorrhea were included. Baseline demographics, presenting symptoms, past medical history, clinical examination, investigation (biochemical and radiological), management and outcomes were analysed. The study was approved by the South Metropolitan Health Services Ethics Committee (approval number RGS0000001008).

Results: The majority of patients were female (61%), obese (mean BMI =29.5 kg/m²) and presented with a unilateral middle ear effusion and conductive hearing loss. Beta-trace protein sampling of the fluid was positive for CSF in 26 of the 28 patients. The most frequent site of leak was at the tegmen mastoideum and middle cranial fossa repair was the most commonly performed procedure with excellent outcomes. Only one patient had confirmed benign intracranial hypertension and two patients presented emergently to hospital with meningitis thought to be due to the CSF leak.

Conclusions: Spontaneous CSF otorrhea should be suspected in unilateral middle ear effusion of unknown cause, especially occurring in at-risk individuals including those whom are obese and of female gender. Clinical or radiological diagnosis alone appears inadequate in diagnosing a spontaneous CSF leak unless clinicians have a high index of suspicion. This emerging entity does not appear to be solely a consequence of raised intracranial pressure and further studies to elucidate the cause are required.

Keywords: Cerebrospinal fluid (CSF); otology; skull base; otorrhoea; diagnosis; treatment; outcomes

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Introduction

Cerebrospinal fluid (CSF) is produced by the choroid plexus of the ventricles in the brain, the ependyma and through capillary ultrafiltrate. It has numerous roles including

lubrication, buoyancy and regulation of brain function through nutrient transport (1). Given its importance in brain function, protection and the small volume produced, leakage of CSF is extremely serious. Potential sequelae of a

leak can include intracranial hypotension, brain herniation and susceptibility to life-threatening intracranial infections such as meningitis (2). Although there is an estimated 10–20% risk of meningitis in patients with CSF leaks through the nose, the risk of meningitis due to CSF otorrhoea is unclear and the literature is limited. Furthermore, the cumulative risk of meningitis in these patients with chronic CSF leaks is unknown (3–6)

The causes of CSF otorrhoea vary and subsequent management is largely dependent on the aetiology however commonly it occurs as a result of trauma—either related to base of skull fractures or following surgery (7). CSF otorrhoea or rhinorrhoea does not infrequently (20%) complicate temporal bone fractures and the risk of a persisting leak post endoscopic resection of skull base tumours is up to 15% (8–13). Diagnosis of CSF leaks is often made through the use of biochemical markers in conjunction with radiological imaging modalities. Beta-trace protein (BTP) and beta-2 transferrin are commonly used biochemical markers for the detection of CSF. Both markers have high diagnostic accuracy and are non-invasive tests however BTP testing allows for more rapid detection and is less labour-intensive as compared to beta-2 transferrin analysis (14–16). An important consideration in the interpretation of BTP is the concentration variation seen in patients with particular disease states of which clinicians should be aware of. Lower BTP levels are observed in the CSF in patients with bacterial meningitis for example which is of relevance to this topic as compared to elevated BTP levels in patients with chronic kidney disease (15).

There appears to be an increasing number of patients presenting with spontaneous CSF otorrhoea. In fact, surgical repair of spontaneous CSF leaks in the USA have nearly doubled in the past ten years (17). These leaks are occurring in the absence of an obvious trigger and the aetiology, pathophysiology and optimal management is yet to be fully described. The commonest theory is of patients with benign intracranial hypertension (BIH) resulting in elevated CSF pressure that exerts force on weak skull base sites (18,19). Consequently, there is thinning and eventual dehiscence at this anatomical site with subsequent leakage (18,20–24). Obesity and obstructive sleep apnoea (OSA) have recently been observed as potential risk factors for spontaneous CSF leaks for similar mechanisms (17,25–27). These findings have commonly been observed in obese female patients (18,25,28). Arachnoid granulations embedded in the skull base bones and causing erosion and defects have also been described as potential culprits for

spontaneous CSF leaks (29,30). The literature however is largely limited to small case series and predominately focuses on spontaneous CSF rhinorrhoea rather than CSF otorrhoea or CSF within the middle ear (ME) space. Given the poorly understood pathophysiology of the disease itself, the diagnosis and management of these patients is challenging which is cause for concern given the risk of serious morbidity and mortality.

This study aims to identify the key features on history and examination in patients presenting with non-iatrogenic, non-traumatic spontaneous CSF otorrhoea in order to further characterise this apparently emerging population. Additionally, it will aim to describe the significance of radiological imaging and biochemical markers in the diagnosis of spontaneous CSF otorrhoea.

Methods

A retrospective cohort study was conducted across the three largest tertiary hospitals in Western Australia. Patients identified with spontaneous CSF within the ME (with and without otorrhea) and subsequently treated by two otology and skull base surgeons (J Kuthubutheen, S Rodrigues) with neurosurgical assistance (A Bala) were included. Twenty-eight patients over a 5-year period (August 2013 to August 2018) made up the final cohort of the study.

The population demographics of age, gender and body mass index (BMI) were recorded. The presence of symptoms at the time of first presentation to the specialist surgeon was recorded and included the presence of hearing loss, otorrhea, vertigo, tinnitus, otalgia, headache and aural fullness. A history of meningitis prior to the first presentation, as documented by hospital admission was also documented. Relevant past medical history, including previous ear surgery, smoking status, alcohol consumption and vaccination status, where available, was also recorded. A documented history of OSA and BIH were also assessed. Patients who were suspected of having OSA had to have had a formal polysomnogram and a diagnosis made by a sleep or respiratory physician. A diagnosis of BIH was confirmed on the documented presence of raised opening CSF pressures above 20 cmH₂O on lumbar puncture.

Clinical examination findings on first presentation to the specialist surgeon was recorded and included otoscopic examination as well as the audiological assessment which included both pure tone audiometry (PTA) and tympanometry. A history of ventilation tube (VT) placement and tympanocentesis was recorded. Results of the

Table 1 Patient characteristics (total N=28)

Characteristics	Value
Gender, n [%]	
Male	11 [39]
Female	17 [61]
Mean age at diagnosis [range] (years)	66 [29–89]
Mean BMI whole cohort (kg/m ²)	29.5
Male	29.2
Female	29.8
Presenting symptoms, able to have >1, n [%]	
Unilateral hearing loss	22 [79]
Bilateral hearing loss	1 [4]
Meningitis	2 [7]
Tinnitus	14 [50]
Aural fullness	9 [32]
Otagia	7 [25]
Otorrhoea	7 [25]
Headache	5 [18]
Imbalance/vertigo	4 [14]
Side of CSF leak, n [%]	
Right	16 [57]
Left	10 [36]
Bilateral	2 [7]
Mean preoperative air conduction threshold (dB) (range) (n=26)	44.8 (10–86.6)
Smoking status	
Smoker	5 [18]
Ex-smoker	4 [14]
Non-smoker	19 [68]

BMI, body mass index; CSF, cerebrospinal fluid.

subsequent biochemical analysis of ME fluid sampled and sent for BTP was recorded. A positive result was considered >1.3 mg/L as per the hospital laboratory cut-off value. Where available, imaging results from both high-resolution temporal bones computed-tomography (HRCT) scans and magnetic resonance imaging (MRI) scans of the temporal lobe and middle cranial fossa were recorded. Unfortunately, due to the variety of imaging protocols used by the external

radiological providers, several HRCT scans (n=4) were sub-optimal with non-dedicated temporal bone algorithms (non-fine cut images) and motion artefact. All MRI scans utilised T2-weighted sequences. Surgical procedures and management outcomes together with post-operative follow-up and surveillance were examined.

The study was approved by the South Metropolitan Health Services Ethics Committee (approval number RGS0000001008). A waiver of consent was granted for the study in line with the NHMRC National Statement on Ethical Conduct in Human Research [2007]. Statistical analysis was performed using SPSS Version 24. For all analysis we considered an alpha value of 0.05 as being statistically significant.

Results

Twenty-nine patients were originally identified with spontaneous CSF otorrhoea however one was excluded due to the confounding presence of a recurrent cholesteatoma. The remaining 28 patients were included forming the final cohort (see *Table 1*). There was a female predominance (n=17 or 61% of the cohort) and the mean age at diagnosis was 66 years (with a range from 29 to 89 years). The mean BMI of the cohort was 29.5 kg/m². One patient passed away soon after CSF leakage was diagnosed due to non-related comorbidities.

Unilateral hearing loss was the most frequent presenting complaint identified in a large majority of patients (79%). Tinnitus (50%), aural fullness (32%) and otorrhoea (25%) were the next most common symptoms although most patients presented with a combination of symptoms rather than a single symptom. Two patients (7%) who were both females presented emergently to hospital with meningitis as the main trigger for ongoing investigation. Both had positive cultures for streptococcal pneumonia on blood cultures and in the CSF. The first patient was subsequently found to have multiple defects in the tegmen mastoideum and as well as the posterior cranial fossa bony plate for which she underwent a subtotal petrosectomy and blind sac closure. The second patient was found to have bilateral tegmen tympani dehiscences on CT with a positive CSF BTP sample from her left ear and is awaiting middle fossa craniotomy (MFC) and repair.

Otological examination demonstrated the presence of middle ear effusions (MEE) on the affected side (86%), in keeping with the presenting complaint of unilateral hearing loss. Hearing assessment was available for 26 out of 28 of

Table 2 Investigations and management

Investigations and management	Value
Mean beta-trace protein (range) (mg/L) (n=26)	13.8 (2.24–57.7)
Vaccination education provided (yes), n [%]	17 [61]
Radiological imaging findings (CT and MRI), able to have >1, n [%]	
Opacified middle ear/mastoid	25 [89]
Partial dehiscence	14 [50]
Total dehiscence	9 [32]
Normal	1 [4]
Management and approach (n=28), n [%]	
Middle fossa craniotomy	13 [46]
Transmastoid	1 [4]
Combined middle fossa craniotomy + transmastoid	2 [7]
Conservative surveillance	4 [14]
Awaiting surgery	8 [29]

the included patients. Type B tympanograms were the most common finding on tympanometry (n=17) as expected. Forty-two percent of patients presented with mixed hearing loss and 38% with purely conductive hearing loss (CHL) also in keeping with the clinical presentation. Mean PTA was 44.8 (range, 10–86.6). In more than half of the patients, a right sided leak was present (n=16) with only two patients presenting with bilateral leaks. No patients presented with CSF rhinorrhoea.

Eleven patients had a history of ischemic heart disease, hypertension or hypercholesterolaemia. Four suffered from hypothyroidism and five had sustained some degree of significant head trauma at varying time frames prior to their CSF leak diagnosis. Two patients had confirmed OSA on polysomnography but these were the only two patients to have undergone sleep study assessment in the cohort. Lumbar puncture for the assessment of opening pressures was also only performed in two patients. One had confirmed BIH as evidenced by an opening lumbar CSF pressure of 25 cmH₂O. The other patient's lumbar pressure was within normal limits however she had suspected radiological signs of BIH including optic nerve sheath hydrops and an empty sella turcica.

Five patients (18%) received VTs for their MEEs. In three of these patients, persistent otorrhoea prompted sampling of the fluid resulting in positive BTP results.

The remaining two patients had VT insertions specifically to assess the fluid for CSF. Twelve patients underwent tympanocentesis without prior VT insertion and fluid analysis for BTP due to clinical suspicion and/or concerning evidence of a CSF leak on prior radiological investigation.

BTP biochemical analysis was performed in 26 of the 28 patients (see *Table 2*). The mean BTP was 13.8 mg/L (range, 2.24–57.7 mg/L) (standard deviation =14.7 mg/L). In one of the patients whose MEE was not sampled, imaging confirmed a 5 mm × 5 mm right tegmen tympani dehiscence with an encephalocele in the epitympanum and he underwent a combined MFC and transmastoid repair. In the other patient, imaging demonstrated a right tegmen tympani dehiscence and an opacified ME, however, the risks of surgery were too great for this patient, so they were conservatively managed with regular surveillance.

All patients underwent radiological investigation in the form of a HRCT with or without an MRI scan (*Table 2*). Twenty-three patients (82%) had both imaging modalities performed. The remaining few had unimodality imaging either in the form of a HRCT or MRI alone. Twenty-five (89%) patients had imaging demonstrating opacified ME's. Of the patients without this finding (n=3), all had undergone either tympanocentesis or VT insertion of a clinical unilateral MEE (demonstrating CSF through elevated BTP levels) prior to radiological investigation (which no longer reported any middle ear fluid).

Partial or total skull base dehiscences were identified in 82% of patients and were defined according to radiological findings. Partial dehiscence was defined as the radiological localised thinning of the skull base but still with the presence of a bony tegmen. Total dehiscence was defined as the radiological absence of bony tegmen at the skull base. Of concern, 23% of CT reports were initially reported as showing no obvious skull base dehiscences but after subsequent MRI and/or positive BTP results, a second review of these scans confirmed the presence of skull base dehiscences. Multiple defects in the affected side were frequently described (n=26) and the tegmen mastoideum was the most common specific site of dehiscence.

Surgical repair was the most common method of management in this cohort of patients with 16 patients undergoing surgical repair (57%) (*Table 2*). Four patients were conservatively managed with regular observation by their own preference on the understanding of the risk of meningitis as a life-threatening complication for failing to repair the leak. At the time of writing the manuscript, a further eight patients were awaiting surgical repair, bringing

a total of 86% of patients who would have received surgery. The most common surgical approach used was the MFC (n=13) in keeping with the commonest site of leak. One patient had a transmastoid repair in view of both middle and posterior cranial fossa dehiscence's and two patients had a combined MFC and transmastoid repair due to a concurrent encephalocele in one patient and concurrent pre-sigmoid arachnoid granulations in another patient. Repair techniques utilised multi-layered closures with varying combinations of temporalis fascia, hydroxyapatite cement and muscle or fat plugs. Multiple tegmen defects were found intra-operatively in 12 cases and single defects present in three cases. Two patients had obvious arachnoid granulations. Unfortunately, the precise locations of all tegmen defects were not recorded relative to the skull base or landmarks, but the most common sites were the tegmen tympani and tegmen mastoideum.

Of the patients that underwent surgery, follow-up ranged from 1 to 26 months, with a mean of 8 months. Otherwise at the time of writing the manuscript, follow-up for the whole cohort (including those awaiting surgery or having conservative surveillance) ranged from 1 to 64 months, with a mean of 19 months.

Surgical repair was effective with only two short-term recurrences of CSF otorrhoea (12.5%). One recurrence was within 2 weeks of the initial MFC with fluid within the middle ear confirmed with a positive BTP level. Subsequent surveillance CT imaging demonstrated satisfactory repair of the tegmen dehiscence's and over a 3-month period, the fluid resolved spontaneously without further surgical intervention required. The other recurrence occurred 6 months post-operatively and was hypothesised as a sequela of a plane flight and the patient having difficulty performing the Valsalva manoeuvre, possibly resulting in pressures forcing CSF through the repair site. This was also managed conservatively.

One patient developed an infected subdural haematoma post craniotomy that required a craniectomy however there was no ongoing CSF leak post operatively. And one patient developed a small haematoma at the surgical site that was managed conservatively. Post-operative adverse events were otherwise limited to pain around the surgical site (n=3). Post-operative hearing assessment was only available in 11 patients in the cohort. Pure tone audiometry was improved in eight patients and unchanged in three. Subjective symptomatic improvement occurred in ten patients, worsened in one (this patient had recurrent CSF otorrhoea), and remained similar in three.

Discussion

Non-iatrogenic, non-traumatic spontaneous CSF otorrhoea can result in life-threatening complications such as meningitis. Establishing an accurate and prompt diagnosis for these patients is difficult given the chronicity and subtleness of presenting symptoms that often masquerade as common otologic conditions such as a unilateral hearing loss or serous MEE. Unfortunately, the underlying aetiology and pathophysiology of spontaneous CSF otorrhoea is poorly understood therefore making it difficult for clinicians to apply evidence-based principals to the diagnosis and management of these patients.

The pathophysiology of spontaneous CSF otorrhoea has been recently investigated in a number of studies and it appears to be multifactorial. Raised intracranial pressure (ICP) almost certainly plays a role through increased hydrostatic forces on weak skull base sites and subsequent leakage (18,19,31-33). Although acknowledged as an important factor, Allen *et al.* explained that it is not the sole factor in the development of spontaneous CSF leaks with only a third of his cohort having elevated ICPs on lumbar puncture (19). One patient in our cohort was confirmed as having BIH however only two patients underwent lumbar puncture for assessment of ICPs therefore limiting the ability to draw conclusions regarding BIH in our study. There is the potential also that patients with CSF leaks may not exhibit the classical clinical signs, symptoms or radiological findings of ICP because they are in fact actively leaking CSF (24). Additionally, there is likely a spectrum of patients with intermittent elevations in ICPs but who do not meet the criteria for a formal BIH diagnosis. Therefore, a preoperative lumbar puncture to measure raised ICPs may in fact not be an accurate reflection of the likely underlying cause for this condition.

Another factor that appears to be contributory with regards to the pathophysiology of spontaneous CSF otorrhoea is obesity (17,19,25-28,34). Our study demonstrated 81% of the cohort classified as overweight (BMI >25 kg/m²) and almost half (42%) obese (BMI >30 kg/m²) in keeping with this hypothesis. The likely mechanism has been proposed to be due to elevated intra-abdominal and intravenous pressures resulting in raised intracranial hypertension and subsequent increased hydrostatic forces on the skull base (33,35,36). Obesity is furthermore associated with OSA which has been identified as a common co-morbid condition in those presenting with spontaneous CSF otorrhoea. Nelson *et al.* described higher than average rates of OSA

occurring in patients with spontaneous CSF otorrhoea (37). A significant correlation between the two was demonstrated through a prospective study of 18 patients that presented with spontaneous leaks and underwent subsequent polysomnography to confirm OSA (37).

Nelson *et al.* further investigated the association between calvarial skull base thinning and OSA given the importance of skull base thickness in middle cranial fossa CSF leaks (27). PSG confirmed the presence of moderate-severe OSA in 92 patients with imaging identifying thinner calvaria and skull bases in this cohort compared to those without OSA. A dehiscent tegmen mastoideum was noted as occurring more frequently in the moderate-severe OSA group (27). The mechanism of progressive skull base thinning may be due to OSA however further studies are required to determine the incidence of OSA in patients with spontaneous CSF leaks such as in our cohort.

Interestingly, there were no patients in our cohort that presented with CSF rhinorrhoea in addition to otorrhoea. We can only hypothesise that leaks via the nose may be of too smaller volume or too intermittent for detection by the patient. In addition, it is possible that the leak predominately affects the mastoid rather than the middle ear thereby creating a reservoir of fluid. We also did not provoke manoeuvres to detect CSF rhinorrhoea given this would not have changed the diagnosis or management arguably. However, this is certainly an important point that we could examine for in future cases.

Almost all of the patients included in this study presented with hearing loss and examination findings consistent of a MEE. The typical management of a unilateral MEE has been to exclude a post nasal space obstructing lesion and in the absence of spontaneous recovery, insertion of a VT of which was evident in our cohort. This has the potential to worsen the disease process and increase the patient's risk of meningitis if spontaneous CSF otorrhoea is not suspected. It highlights the importance of considering the various aetiologies of MEEs and especially in at-risk individuals (obese, females) identified through this study.

Surgical treatment is known to be effective and demonstrated in our cohort with a low incidence of adverse events and recurrent CSF otorrhoea (7%) (38-40). Nelson *et al.* revealed similar recurrence rates of spontaneous CSF otorrhoea (6.7%) to our study in his cohort of 60 patients that underwent MFC repair (40). Multiple approach and repair techniques have been described including MFC, transmastoid, and combined MFC/transmastoid approaches, as well as single and multi-layered defect repair techniques.

An MFC approach appears advantageous due to increased exposure of the middle fossa which is especially important in the setting of multiple defects and/or large cephaloceles (32,40,41). Additionally, rates of recurrent CSF leaks have been demonstrated as higher in those undergoing transmastoid approaches (38,42). Conversely, avoidance of trauma or retraction to intracranial tissue is beneficial in transmastoid surgery. Multi-layered closures are favourable over single-layered techniques with lower rates of recurrence and failure (43). Both patients in our cohort that developed recurrent CSF leaks initially underwent MFC repairs with multi-layered closures (temporalis fascia and hydroxyapatite cement). Interestingly, multiple tegmen tympani and mastoideum defects in addition to several arachnoid granulations in one of these patients were noted intra-operatively which may be reflective of more extensive disease in these individuals and difficulty in repair. Prospective assessment on approach and repair techniques would allow for further evaluation of success of surgical intervention in these patients however based on our study and the evidence to date, an MFC approach with multi-layered closure techniques are extremely effective.

Our study was limited by the retrospective nature of study which inherently introduces bias. However, the sample size albeit small is in fact comparable if not larger than most other retrospective cohort studies assessing spontaneous CSF otorrhoea. Whilst the minority of our patients had symptoms and signs of BIH, only two patients had lumbar CSF pressures measured to completely exclude this and not all patients had documented examinations of the optic disc to exclude papilloedema. In addition, not all patients had polysomnograms to exclude OSA. Future studies will require examination of these risk factors more closely. The variety of imaging protocols utilised by external radiological providers may have impacted the detail and/or content of the scans and this was the case in four patients whereby HRCTs were sub-optimal.

Our study's strength is that it included analyses of audiological findings and biochemical results which have been poorly described in the literature thus far. We have also demonstrated that gender and obesity appear to be important risk factors in disease development, factors which are consistent with what has been reported in the literature although the incidence of this condition in the wider community is yet unknown and needs to be studied. Prompt biochemical analysis of middle ear fluid for CSF in conjunction with multi-modality imaging techniques is crucial for accurate diagnosis given the frequently obscure

symptoms and signs these patients often present with. Future studies may need to examine alternative methods for diagnosing CSF to ensure accuracy and consistence of the results obtained since the need for surgical intervention is predicated on this result.

Conclusions

This multicentre study is the first to our knowledge in Australia assessing patients with spontaneous CSF otorrhoea with comprehensive evaluation of diagnosis, investigations, management and follow-up. Of concern is that clinical or radiological diagnosis alone appears inadequate in diagnosing a spontaneous CSF leak unless clinicians have a high index of suspicion. We suggest an approach adopting both clinical and radiological investigation in addition to analysis of middle ear fluid for BTP to confirm CSF. Unilateral MEE and conductive hearing loss are common presenting symptoms and so health professionals should be aware of the increasing incidence of this condition to ensure early and accurate diagnosis and referral for management thereby reducing the risk of life-threatening intracranial infections such as meningitis.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at <http://dx.doi.org/10.21037/ajo.2019.01.07>). JK serves as an unpaid editorial board member of *Australian Journal of Otolaryngology*. The other authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the South Metropolitan Health Services Ethics Committee (approval number RGS0000001008). Informed consent was waived due to the retrospective nature of the study.

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