Egyptian Journal of Radiology and Nuclear Medicine

CASE REPORT

Open Access

Spontaneous cerebrospinal fluid fistula secondary to hyper-pneumatized paranasal sinuses and skull base: two case reports

Shweta Raviraj Poojary, Divya Vishwanatha Kini*, T. R. Kapilamoorthy, Kavitha B. Chittaragi and Balasubramanian Gurumurthy

Abstract

Background Spontaneous cerebrospinal fluid (CSF) fistulas occur due to various reasons other than well-identified causes such as trauma, neoplasia or infection. Various contributory factors are attributed to formation of spontaneous CSF leaks such as idiopathic intracranial hypertension leading to prominent arachnoid granulations. Further, presence of hyper-pneumatized paranasal sinuses or the skull base weakens the bone and predisposes to development of spontaneous defects and further fistulas. This case report highlights two cases of spontaneous CSF leaks associated with hyper-pneumatized petrous bone and sphenoid sinus.

Case presentation A 26-year-old female patient with history of right rhinorrhea with imaging evidence of bilateral hyper-pneumatized petrous bones and a bony defect in the right petrous bone on computed tomography (CT). Subsequent CT cisternography demonstrated CSF leak extending into the right pneumatized petrous apex cells, Eustachian tube, middle ear cavity, aditus, antrum and mastoid air cells. Pooling of contrast in the right nasal cavity and ethmoid cells was also seen. A 49-year-old female patient with history of right rhinorrhea with features of hyper-pneumatization of sphenoid bone involving right greater wing of sphenoid bone and bilateral pterygoid process with a bony defect in the right greater wing of sphenoid was demonstrated on CT. Corroborative magnetic resonance imaging (MRI) brain Constructive interference in steady state (CISS) sequence revealed a meningoencephalocele. Additionally, a suspicious focal dehiscence was observed in the right cribriform plate CSF pockets herniating into right ethmoid sinus.

Conclusion Hyper-pneumatized petrous bone and paranasal sinuses can be attributed as a risk factor for formation of spontaneous CSF leaks.

Keywords Cerebrospinal fluid, Hyper-pneumatization, Petrous bone, Sphenoid sinus, Fistula

Background

Cerebrospinal fluid (CSF) fistulas are classified based on their etiology into congenital and acquired fistulas. The acquired fistulas are further classified as nontraumatic, traumatic and spontaneous [1, 2].

Spontaneous CSF leaks may arise from defects in the skull base, in relation to the Tegmen tympani, Tegmen mastoideum, sigmoid sinus and posterior semicircular canal. Leaks in these locations cause indirect CSF rhinorrhoea implying a communication of the subarachnoid space with the middle ear cavity. A constellation of extensive pneumatization, arachnoid pits and empty sella plays a role in the pathogenesis of sphenoid sinus fistulae [3] apart from congenital skull base defects. Arachnoid granulations in relation to the temporal bone are another

*Correspondence:
Divya Vishwanatha Kini
drkinidivya@gmail.com
Department of Radiology, JSS Hospital, Ramanuja Road, Agrahara, Mysore 570004, Karnataka, India

contributing factor for spontaneous CSF otorrhea and rhinorrhoea [1]. Detection of these defects is best done with computed tomography (CT) and CT Cisternography with an additional role of MRI in assessment of associated parenchymal herniation with utmost accuracy. In this study, we wish to highlight the existence of spontaneous CSF leak as an entity associated with hyper-pneumatized petrous bone and sphenoid sinus.

Case presentation

Case 1

A 26-year-old female patient presented with history of right sided rhinorrhoea for 1 month. There was no history suggestive of meningitis, trauma, visual blurring. Patient was conscious, oriented. No meningeal signs were seen. Fundoscopy was normal. Clinically patient appeared of appropriate built and nourishment for age and gender. Blood work-up revealed no significant abnormality. No previous similar complaints were noted in the past.

Plain CT study showed hyper-pneumatization of the petrous bones with associated soft tissue thickening within (Fig. 1). In addition, a bony defect of ~ 2 mm with adjacent bone scalloping involving the anterolateral aspect of petrous part of right temporal bone (Fig. 1) was demonstrated. CT cisternography showed contrast opacified CSF extending into the pneumatized petrous apex cells, Eustachian tube, middle ear cavity, aditus, antrum, mastoid air cells (Fig. 2). However, no contrast opacification of cochlea and vestibular apparatus was observed. Pooling of contrast in the right nasal cavity and ethmoidal air cells was also seen (Fig. 2). Corresponding MRI brain revealed features of right mastoiditis extending to involve the pneumatized petrous bone (Fig. 3). Diagnosis of right petrous defect with paradoxical CSF rhinorrhea was hence made.

Follow up: Patient underwent right temporal craniotomy and CSF leak repair. Intra-operatively, the petrous bone was thin with cobble stoning and few areas of defect over the petrous ridge. A defect of 1×1 cm was seen toward the lateral aspect close to the anterior petrous ridge. A temporal muscular pedicle flap was used as a covering fascia. Patient tolerated the procedure well and was discharged with no CSF leak. Patient is currently asymptomatic.

Case 2

A 49-year-old female patient presented with history of right sided watery nasal discharge over the last 15 days. There was no history suggestive of meningitis, trauma or visual blurring. Patient had no other comorbidities with no similar prior complaints. Clinically patient appeared of appropriate built and nourishment for age and gender. Blood work-up revealed no significant abnormality.

On HRCT of the temporal bone, hyper-pneumatization of sphenoid bone involving right greater wing of sphenoid bone and bilateral pterygoid processes was noted (Fig. 4). Also, bony defect measuring 1.4 mm noted in the right greater wing of sphenoid bone (lateral to the foramen rotundum) (Fig. 5). Corroborative MRI brain CISS sequence revealed predominant downward herniation of meninges and adjacent minimal brain parenchyma through the above-mentioned defect into the hyper-pneumatized right greater wing of sphenoid bone and right pterygoid process (Fig. 6).

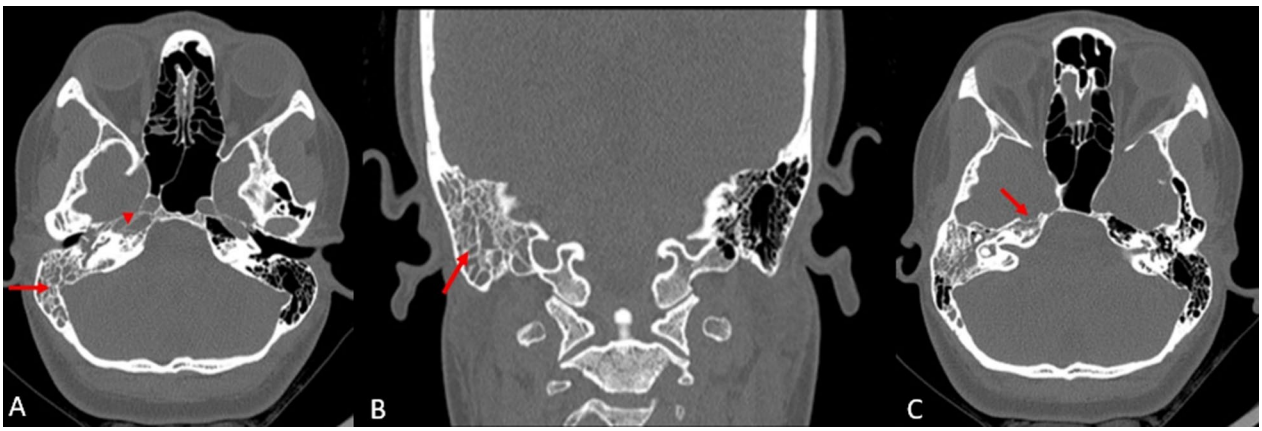

Fig. 1 A 26-year-old female presented with history of right sided rhinorrhoea. **A** Plain CT PNS and temporal bone axial section showing bilateral hyper-pneumatized petrous bones with overcrowding of the right mastoid air cells (red arrow), opacification of the hyper-pneumatized petrous bone (red arrow head) and mild right ethmoid sinusitis. **B** Plain CT temporal bone coronal section demonstrating the overcrowding of the right mastoid air cells by mucosal thickening (red arrow). **C** Plain CT PNS and temporal bone axial section demonstrating the scalloping along the antero-medial aspect of the right petrous bone (red arrow)

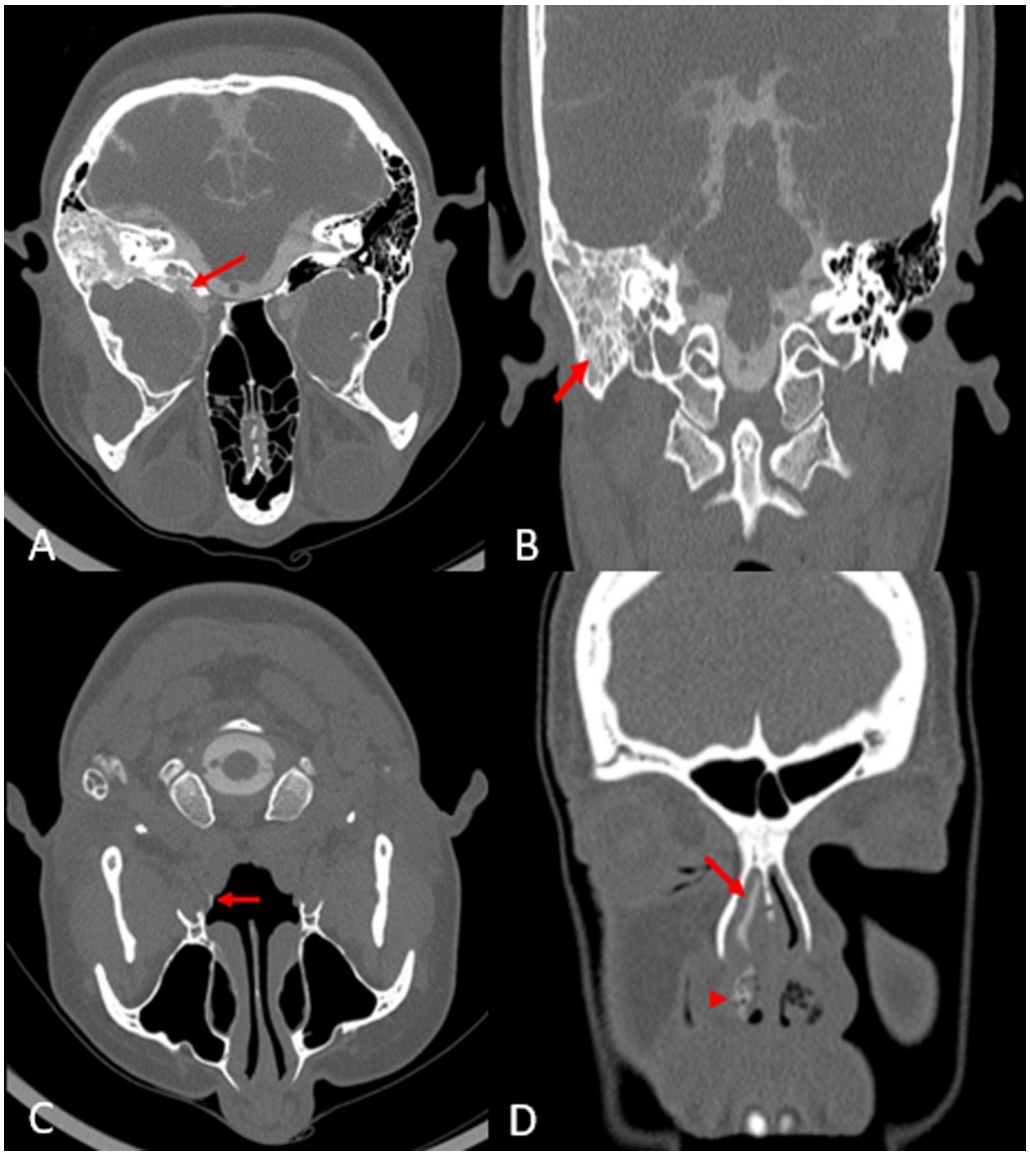

Fig. 2 **A** CT cisternography performed in prone position depicting the scalloping along the antero-medial aspect of the right temporal bone (red arrow) with contrast leak through this into the adjacent hyper-pneumatized petrous bone. **B** CT cisternography coronal section demonstrating the increased density corresponding to the contrast leak into the right mastoid air cells (red arrow) and middle ear and **C** CT cisternography axial section demonstrating contrast leak at the nasopharyngeal opening of the right Eustachian tube (red arrow). **D** CT cisternography coronal section demonstrating the contrast leak into the right nares (red arrow) with the nasal pledgets showing contrast soakage (red arrowhead)

Follow up: Patient was lost to follow-up and hence information regarding further management could not be provided.

Discussion

The development of skull base defects is caused by the interaction of CSF pulsation with the bone. The anatomical predisposition differs between the anterior, middle and posterior cranial fossa. Site of CSF leaks in temporal bone in various studies are summarized in Table 1 according to the age of the patients [4–8].

The novelty in the first case lies in the fact that none of these cases presented with a defect in the anterior ridge of the petrous bone. Unlike the thin tegmen tympani and tegmen mastoideum, the anterior ridge of the petrous bone is thicker and does not have any congenital dehiscences. Due to the presence of petrous apex hyper-pneumatization, the otherwise thicker anterior ridge could have given way in response to the raised

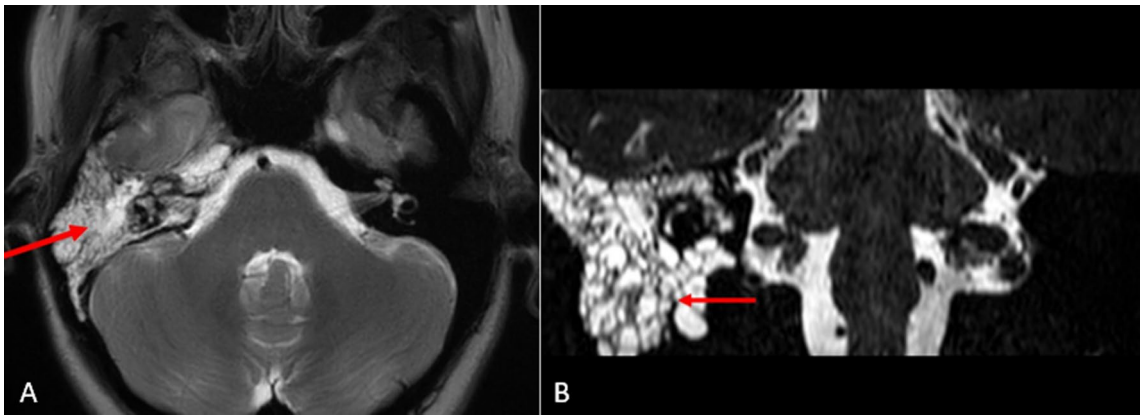

Fig. 3 **A** T2W axial section showing T2 hyperintensity within the right mastoid air cells (red arrow) and middle ear cavity. It is further seen extending to involve the hyper-pneumatized right petrous body. **B** MRI CISS coronal section showing the T2 hyperintensity within the right mastoid air cells (red arrow)

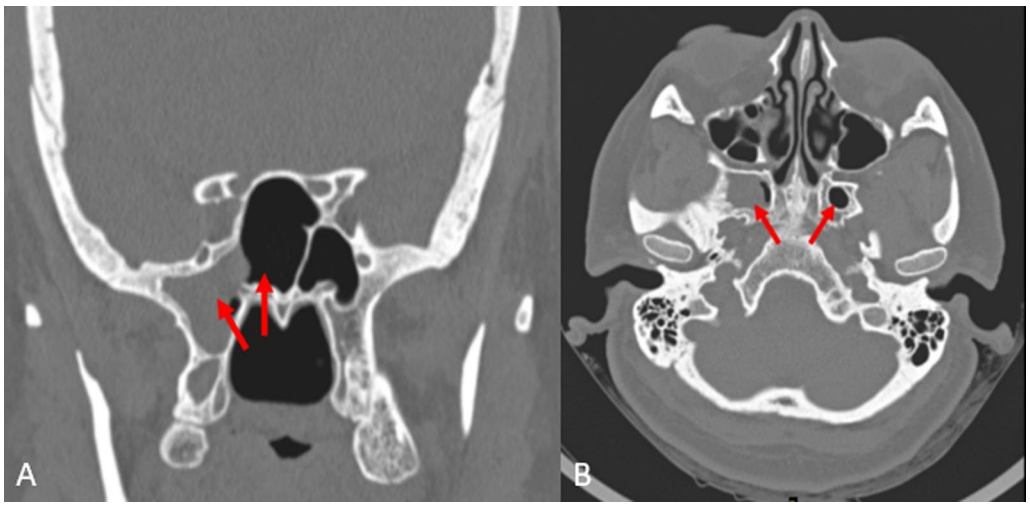

Fig. 4 A 49-year-old female presented with history of right sided watery nasal discharge. **A** Plain CT PNS coronal section showing the hyper-pneumatized right sphenoid bone (vertical red arrow) with mucosal thickening within (oblique red arrow) and **B** axial section showing the hyper-pneumatized bilateral pterygoid bones (red arrows) with opacification on the right side by mucosal thickening

intracranial pressure. The scalloping of the anterior ridge of petrous bone indicated a chronic process in the background of raised CSF pressure. Other factors for spontaneous CSF leak such as arachnoid granulations defects and idiopathic intracranial hypertension were ruled out in this patient [1]. Multiple defects were detected in the right petrous bone in this case. Difference between the previous cases and this case was that despite having a temporal bone defect, the patient presented with rhinorrhea instead of otorrhea. This indicated that the tympanic membrane was intact. The cause of rhinorrhea could be attributed to paradoxical leak into the nasopharynx via the eustachian tube and further into the ethmoid sinus and nasal cavity. It is very important to highlight here the identification of the middle fossa defect, which if left unrepaired may result in persistent leak. Also emphasizes the role of radiologists in accurate localization of site of leak which decides the appropriate surgical strategy [1, 4–8].

The second case highlighted the association of CSF rhinorrhea with presence of meningoencephaloceles and hyper-pneumatized paranasal sinuses. Literature has thrown light on the likely pathogenesis of development of meningoencephaloceles secondary to CSF fistulas. The presence of raised intracranial pressure has been strongly associated with the presence of spontaneous CSF fistulas-they are seen to promote formation of prominent arachnoid granulations [1, 2, 9]. Owing to

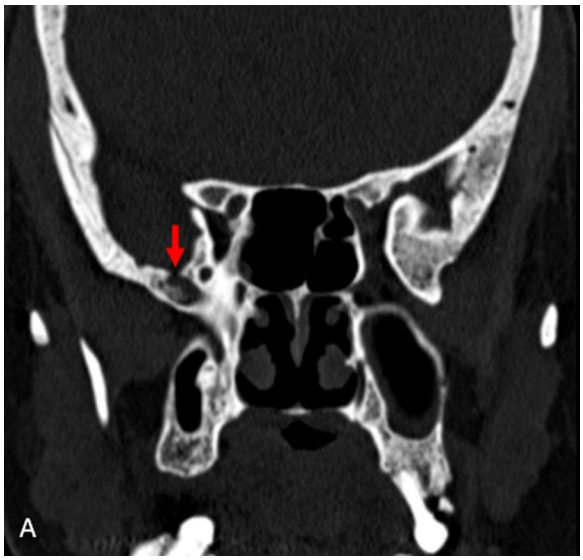

Fig. 5 CT cisternography coronal section in bone window demonstrating the bony defect in the right greater wing of sphenoid bone (red arrow) and resultant contrast opacification (red arrow)

the altered CSF dynamics within these, there is a path of reduced resistance created through which the meninges and cerebral parenchyma tend to easily herniate resulting in creation of a spontaneous CSF fistula [1]. However, the current accepted theory is that of a multifactorial process involving the combination of elevated intracranial pressure and anatomical deficiencies (viz. hyper-pneumatized paranasal sinuses) [10, 11] aiding in the development of meningoencephaloceles and spontaneous CSF fistulas [1].

High resolution computed tomography (HRCT) of the paranasal sinuses as well as the temporal bones is a baseline imaging modality which helps identify skull base defects irrespective of the presence of CSF leaks [2, 11]. However, the detection of dural defects and differentiation of mucus secretions from actual CSF is not possible. To aid in this, use of additional CT Cisternography and MR cisternography is beneficial. CT Cisternography utilizes intra-thecal injection of iodinated contrast in order to opacify the CSF and thus, it accurately demonstrates the site of leak with visualization of active leak through the defect. MR Cisternography (MRC) utilizes the property of high signal intensity of CSF on heavily T2 weighted images in the subarachnoid spaces with the added advantage of its excellent soft tissue resolution thereby helping detect the presence of an associated dural defect with super-added meningoencephaloceles [2, 10, 11]. However, bony details maybe sub-optimally evaluated on MRI and hence interpretation of the imaging findings must be performed along with CT. Additional modalities that have been proven to aid in the diagnosis are contrast enhanced MRC and Radionuclide cisternography [10]. At our institute, we follow a protocol

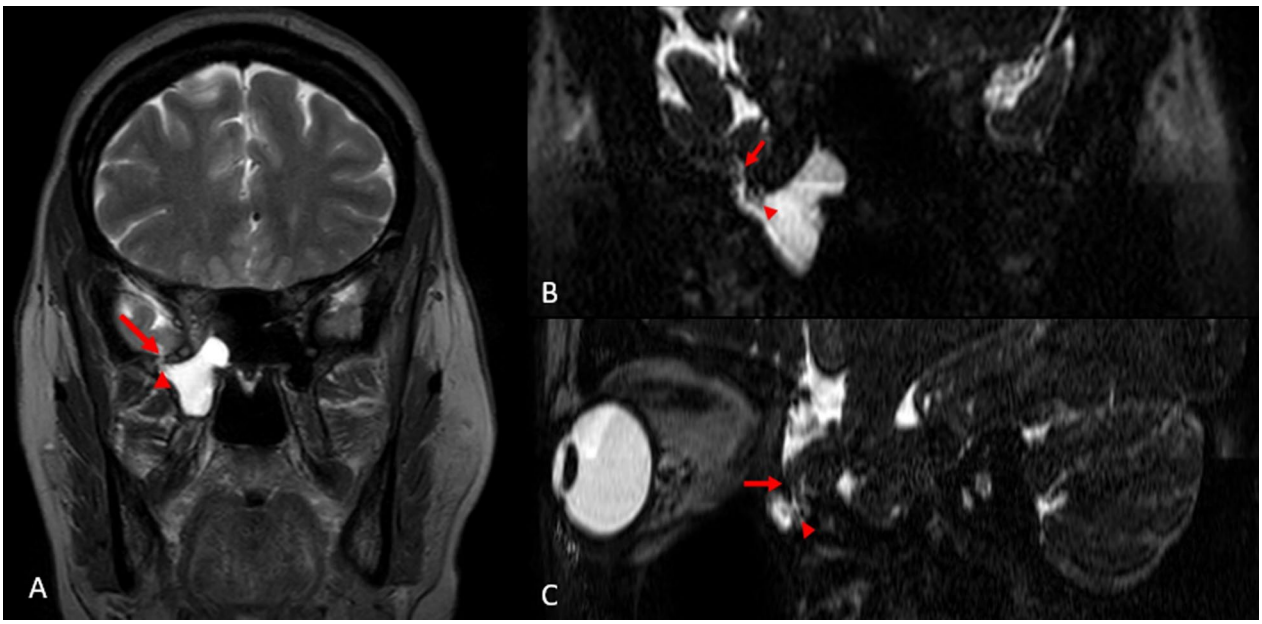

Fig. 6 **A** T2W image coronal section showing the defect in the right greater wing of sphenoid bone (red arrow) with predominant downward herniation of meninges and adjacent minimal brain parenchyma (red arrowhead). MRI brain CISS sequence **B** Coronal and **C** sagittal sections showing the defect in the right greater wing of sphenoid bone (red arrow) with downward herniation of meninges and adjacent minimal brain parenchyma (red arrowhead)

Table 1 Site of CSF leaks in temporal bone in various studies—table adapted and modified from study by Nadaraja et al. [8]

Study author	Year	Age	Gender	Side affected	Defect site
Schuknecht et al. [4]	1982	53	Female	Left	Between superior petrosal and sigmoid venous sinus
Luessen et al. [5]	2004	64	Male	Right	Posterior fossa plate
Luessen et al. [5]	2004	57	Female	Left	Trautmann area
Rao et al. [6]	2005	75	Female	Right	Medial to Sigmoid sinus
Lee et al. [7]	2008	41	Male	Left	Bilateral defects, left greater than right
Nadaraja et al. [8]	2011	68	Female	Right	Superior to the endolymphatic sac
Nadaraja et al. [8]	2011	37	Female	Right	Large defect from sigmoid sinus to posterior semicircular canal
Moreira et al. [9]	2021	42	Male	Right	Multiple small tegmen tympani and mastoid defects

consisting of HRCT of the paranasal sinuses and temporal bones followed by MR brain with MR Cisternography (CISS sequences—heavily T2 weighted images). This is further followed by a confirmatory CT Cisternography to demonstrate site of active leak with use of dynamic maneuvers facilitating the leak.

As demonstrated in these cases, in the presence of normal intracranial pressure, hyper-pneumatization is a significant risk factor for the development of spontaneous CSF fistulas. Such correlation has not been documented in previous studies. Hence, we tend to propose a novel hypothesis of hyper-pneumatization predisposing to formation of spontaneous CSF leaks. As a part of routine reporting protocol, documentation of presence of hyper-pneumatization is essential to estimate the risk of development of such fistulas.

Limitation

The second case was lost to follow-up and thus a confirmation regarding the radiological diagnosis by surgical correlation could not be made.

Conclusions

The recognition of spontaneous CSF leaks is vital and is to be made with accurate detection of the site, etiology and severity of the same as poorly managed cases result in deleterious complications. HRCT of the involved region with further CT cisternography and corroborative MRI is an essential and beneficial imaging protocol. Careful scrutiny of the skull base to look for hyper-pneumatized sinuses and anatomical dehiscences should be performed in a meticulous manner.

Abbreviations

CSF Cerebrospinal fluid
CT Computed tomography
MRI Magnetic resonance imaging
CISS Constructive interference in steady state

Acknowledgements

We sincerely thank our esteemed institution JSS Academy of Higher Education and Research, Mysore, for encouraging and providing support in all our endeavors. We also take this opportunity to express my gratitude to all the faculty, post graduates, technical and supporting staff of Department of Radiology.

Author contributions

Dr. SRP conceptualized and designed the study, analyzed and interpreted the data, prepared the draft manuscript and supervised the project. Dr. DVK conceptualized and designed the study, analyzed and interpreted the data and prepared the draft manuscript. Dr. TRK conceptualized and designed the study, analyzed and interpreted the data and supervised the project. Dr. KBC and Dr. BG supervised the project and analyzed and interpreted the data. All the above authors have read and approved the manuscript.

Funding

No funding was obtained for this study.

Availability of data and materials

The datasets generated and/or analyzed during the current study are not publicly available due to privacy of the study participant.

Declarations

Ethics approval and consent to participate

Non-identifiable images of the patient have been used. Careful attention has been provided to prevent patient identifiable information being revealed in the images.

Consent for publication

Written informed consent has been obtained from the patient.

Competing interests

The authors declare no competing interests.

Received: 2 September 2022 Accepted: 30 December 2022
Published online: 09 January 2023

References

1. Alonso RC, de la Peña MJ, Caicoya AG, Rodriguez MR, Moreno EA, de Vega Fernandez VM (2013) Spontaneous skull base meningoencephaloceles and cerebrospinal fluid fistulas. Radiographics 33(2):553–570. https://doi.org/10.1148/rg.332125028. (**PMID: 23479713**)
2. Lloyd KM, DelGaudio JM, Hudgins PA (2008) Imaging of skull base cerebrospinal fluid leaks in adults. Radiology 248(3):725–736. https://doi.org/10.1148/radiol.2483070362. (**PMID: 18710972**)

3. Shetty PG, Shroff MM, Fatterpekar GM, Sahani DV, Kirtane MV (2000) A retrospective analysis of spontaneous sphenoid sinus fistula: MR and CT findings. AJNR Am J Neuroradiol 21(2):337–42
4. Schuknecht HF, Zaytoun GM, Moon CN Jr (1982) Adult-onset fluid in the tympanomastoid compartment. Diagnosis Manag Arch Otolaryngol 108(12):759–765. https://doi.org/10.1001/archotol.1982.00790600003002. (**PMID: 6890798**)
5. Welge-Luessen A, Probst R (2004) Spontaneous cerebrospinal fluid otorrhea in the posterior fossa as a rare cause of adult bacterial meningitis. Otolaryngol Head Neck Surg 130(3):375–377. https://doi.org/10.1016/j.otohns.2003.07.001. (**PMID: 15054385**)
6. Rao AK, Merenda DM, Wetmore SJ (2005) Diagnosis and management of spontaneous cerebrospinal fluid otorrhea. Otol Neurotol 26(6):1171–1175. https://doi.org/10.1097/01.mao.0000179526.17285.cc. (**PMID: 16272936**)
7. Lee MH, Kim HJ, Lee IH, Kim ST, Jeon P, Kim KH (2008) Prevalence and appearance of the posterior wall defects of the temporal bone caused by presumed arachnoid granulations and their clinical significance: CT findings. AJNR Am J Neuroradiol 29(9):1704–1707. https://doi.org/10.3174/ajnr.PMID:18617585;PMCID:PMC8118775
8. Nadaraja GS, Monfared A, Jackler RK (2012) Spontaneous cerebrospinal fluid leak through the posterior aspect of the petrous bone. J Neurol Surg B Skull Base 73(1):71–75. https://doi.org/10.1055/s-0032-1304560.PMID:23372998;PMCID:PMC3424026
9. Moreira BCB, Azevedo AF, Castro MCM (2021) Spontaneous temporal cerebrospinal fluid leak. J Otolaryngol ENT Res 13(2):22–27. https://doi.org/10.15406/joentr.2021.13.00485
10. Hiremath SB, Gautam AA, Sasindran V, Therakathu J, Benjamin G (2018) Cerebrospinal fluid rhinorrhea and otorrhea: a multimodality imaging approach. Diagn Interv Imaging 100(1):3–15. https://doi.org/10.1016/j.diii.2018.05.003. (**Epub 2018 Jun 15 PMID: 29910174**)
11. Hiremath SB, Gautam AA, Sheeja K, Benjamin G (2018) Assessment of variations in sphenoid sinus pneumatization in Indian population: a multidetector computed tomography study. Indian J Radiol Imaging 28(3):273–279. https://doi.org/10.4103/ijri.IJRI_70_18

Publisher's Note