

Isolated spontaneous cerebrospinal fluid rhinorrhoea as a rare presentation of idiopathic intracranial hypertension: Case reports with comprehensive review of literature

Priti Soin, Umer M Afzaal¹, Pranav Sharma¹, Puneet S Kochar¹

Department of Pathology and Laboratory Medicine, Weil Cornell College of Medicine, New York, ¹Department of Radiology, Yale New Haven Health Bridgeport Hospital, Bridgeport, CT, USA

Correspondence: Dr. Puneet S Kochar, 267 Grant Street, Bridgeport, CT, USA. E-mail: drkochar.puneet@gmail.com

Abstract

Isolated cerebrospinal fluid (CSF) rhinorrhoea as a sole presenting symptom of idiopathic intracranial hypertension (IIH) is extremely rare. IIH typically presents with headache, pulsatile tinnitus, dizziness, nausea, vomiting, and visual disturbance. We report two cases which presented with acute onset spontaneous CSF rhinorrhoea without any other symptom. In addition, we discuss in detail imaging features of IIH with review of its literature.

Key words: Cerebrospinal fluid rhinorrhoea; idiopathic intracranial hypertension; idiopathic intracranial hypertension; spontaneous

Introduction

Idiopathic intracranial hypertension (IIH) is a syndrome of headache characterized by raised intracranial pressure (or cerebrospinal fluid [CSF] pressure) in the absence of an intracranial mass or hydrocephalus. Clinical exam is almost always normal except for occasional cranial nerve VI palsy and papilledema.^[1] Papilledema is the hallmark of IIH. Papilledema can be bilateral, asymmetrical, or even unilateral; though it can be absent in some cases. To the best of our knowledge, there are very few cases in literature in which patient presented with CSF rhinorrhoea as an isolated symptom. In this case series, we describe two cases which presented solely with spontaneous rhinorrhoea.

Cases

Case 1

A 38-year-old female presented with acute onset of spontaneous CSF rhinorrhoea, which started a month ago. The patient disregarded the symptom and related it to rhinosinusitis. However, she again had episodes of intermittent nasal discharge on left side. She denied history of headache, vomiting, blurring, visual disturbance, or head injury. The ophthalmoscopic examination revealed papilledema [Figure 1] with normal visual acuity and field charting. The laboratory analysis of nasal discharge

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Cite this article as: Soin P, Afzaal UM, Sharma P, Kochar PS. Isolated spontaneous cerebrospinal fluid rhinorrhoea as a rare presentation of idiopathic intracranial hypertension: Case reports with comprehensive review of literature. Indian J Radiol Imaging 2018;28:406-11.

Access this article online

Quick Response Code:



Website:
www.ijri.org

DOI:
10.4103/ijri.IJRI_228_18

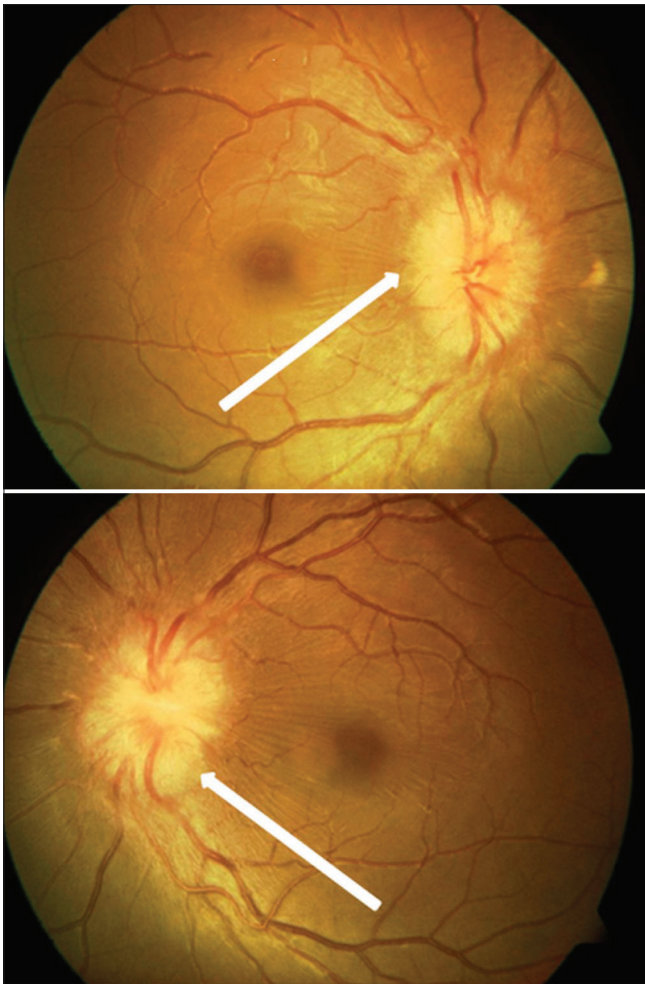


Figure 1: Ophthalmoscopic exam in case 1 demonstrated bilateral optic disc edema. (white arrows)

tested positive for beta-2 transferrin. The magnetic resonance imaging (MRI) cisternography was requested to demonstrate site of leakage. The patient was advised to get MRI on the day of active nasal discharge. The MRI was acquired in prone position with a dry cotton pledget placed in the left nasal opening. A tiny focus of CSF leakage was identified in the left cribriform plate [Figure 2A; zoomed up D], which was seen continuing posteroinferiorly into the nasal cavity [Figure 2B]. The dry cotton pledget got soaked with CSF and appeared hyperintense on T2 weighted images [Figure 2C]. In addition, MRI demonstrated perioptic CSF distension [Figure 3A], vertical tortuosity of the optic nerves [Figure 3B and C], empty sella [Figure 4A], and very prominent CSF distended bilateral Meckel's cave [Figure 4B]. No intracranial mass was noted. Moreover, MR venography showed bilateral distal transverse sinus stenosis [Figure 5]. Imaging diagnosis of IIH was suggested.

Lumbar puncture was performed 2 weeks later at the time of absence of CSF rhinorrhoea, which demonstrated increased opening pressure of 30 cm of H₂O (normal 10–18 cm of H₂O) with normal CSF composition on laboratory analysis. This

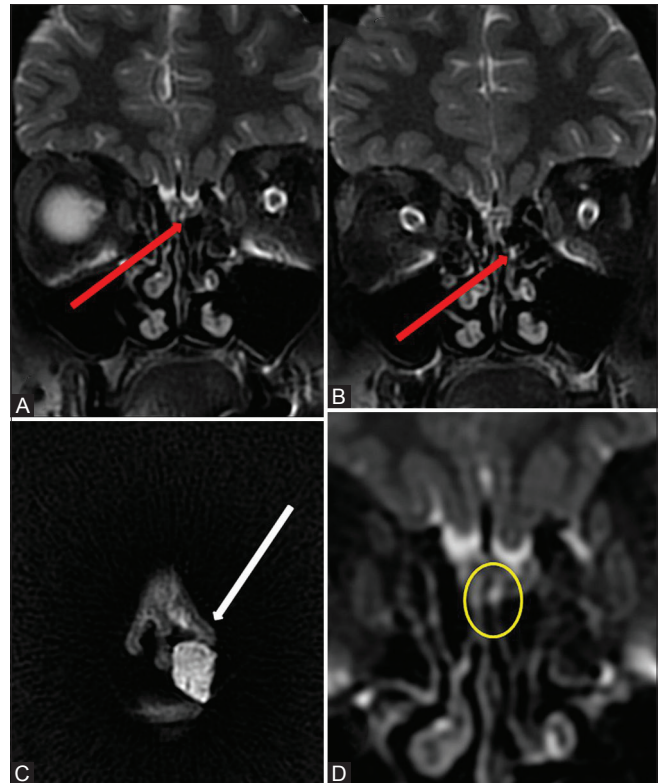


Figure 2 (A-D): Coronal T2 fat saturated images in case 1 demonstrate a small site of leakage in the left cribriform plate (red arrow; image A) with a small track of fluid extending posteriorly and inferiorly (red arrow; image B). A dry cotton pledget was placed at the left nasal opening which got soaked with fluid, appearing T2 hyperintense (image C; white arrow). Image D is a zoomed image demonstrating site of leakage

confirmed imaging diagnosis of IIH. The patient was offered computerized tomography (CT) scan, which she denied. The patient was treated medically and was offered endoscopic surgical repair. Eventually, patient was lost to follow-up.

Case 2

A 55-year-old female presented with sudden onset of copious spontaneous CSF rhinorrhoea, which started 3 weeks ago noted on the right side. She denied history of headache, vomiting, blurring, visual disturbance, or head injury. The ophthalmoscopic examination revealed normal fundus exam, visual acuity, and field charting. The laboratory analysis of nasal discharge tested positive for CSF. The MRI cisternography was requested. The MRI was acquired in prone position. A moderate sized focus of CSF leakage was identified in the right cribriform plate [Figure 6A], which was seen continuing posteroinferiorly into moderate amount of CSF in the right ethmoid air cells [Figure 6B]. The MRI also demonstrated a ballooned empty sella [Figure 7A], perioptic CSF distension [Figure 7B], and vertical tortuosity of the optic nerves [Figure 7C-D]. No intracranial mass was noted. Imaging diagnosis of IIH was suggested. CT scan showed a corresponding defect in the right cribriform plate [Figure 8]. The patient was referred for endoscopic/neurosurgical surgical repair. Surgery confirmed the site of leakage noted on MRI/CT and was repaired with fascia lata patch.

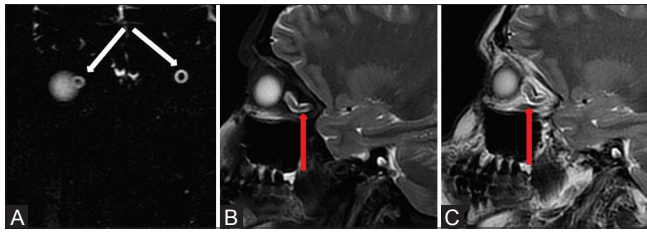


Figure 3 (A-C): Coronal T2 weighted images in case 1 through the orbits demonstrates CSF distension of the bilateral perioptic subarachnoid space (white arrows; image A). Oblique sagittal T2 weighted images are showing vertical tortuosity of bilateral optic nerves (red arrows; images B and C)

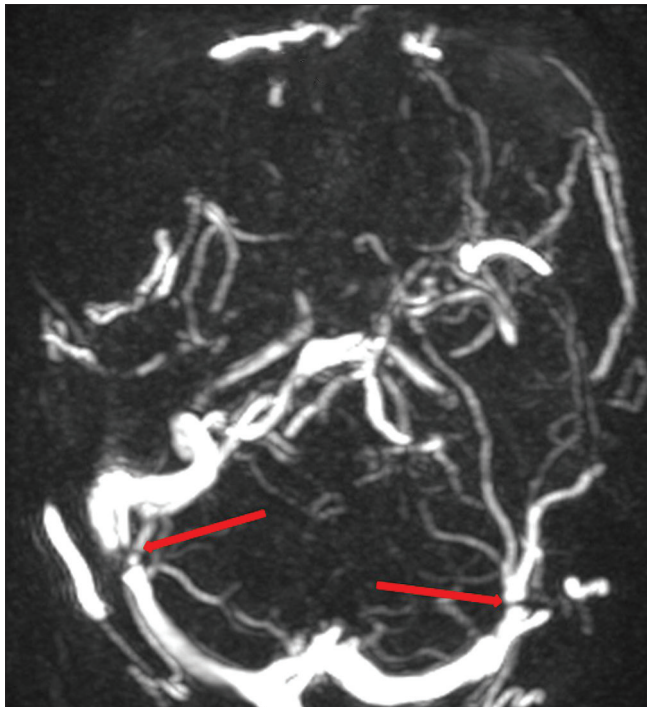


Figure 5: MR venogram axial Maximum Intensity Projection (MIP) image from case 1 demonstrates severe stenosis of bilateral distal transverse sinuses with non opacification of left sigmoid sinus and jugular bulb (red arrows)

Discussion

IIH is not an uncommon clinical condition, primarily affects young obese women with estimated incidence in this population of 19 per 100,000. This is 20-fold higher than in normal-weight individuals.^[2-4] The typical presentation is headache, pulsatile tinnitus, dizziness, nausea, vomiting, visual disturbance, papilledema, and elevated CSF pressures, with symptoms and signs often not always present together and varying with time.^[5] CSF rhinorrhoea is a rare complication of benign intracranial hypertension with cribriform plate being the most frequent site of CSF fistula formation, followed by the sella turcica.^[6]

To the best of our knowledge, there are very few cases in the literature with IIH patients solely presenting as CSF

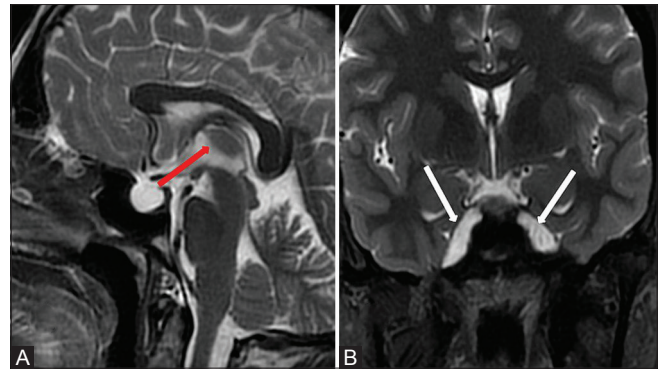


Figure 4 (A and B): Sagittal and coronal T2 weighted images in case 1 demonstrating empty sella (image A; red arrow) and prominent CSF distended bilateral Meckel's cave (image B; white arrows)

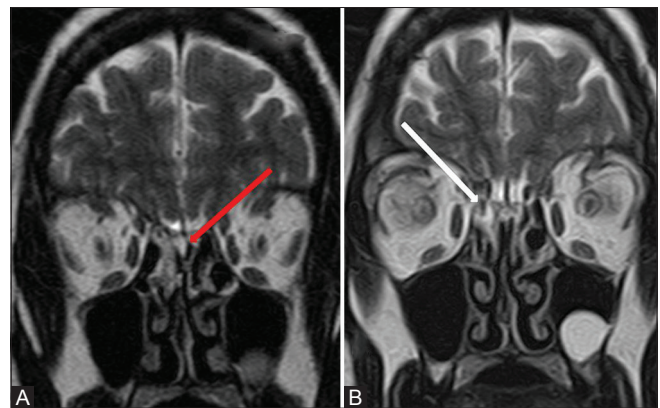


Figure 6 (A and B): Coronal T2 weighted images in case 2 demonstrate a moderate sized leakage of CSF at the level of right anterior cribriform plate (red arrow; image A). Moderate amount of CSF is seen accumulating in the right ethmoid air cells (white arrow; image B)

rhinorrhoea. Saifudheen *et al.* (2010) described one case confirmed on MRI and increased CSF opening pressure.^[7] Rosenfeld *et al.* (2013) described 4 cases of spontaneous CSF leakage, with one patient having CSF rhinorrhoea as a presenting symptom.^[8] Perez *et al.* (2013) described one case with isolated CSF rhinorrhoea.^[9] Both our cases presenting with sudden onset isolated spontaneous CSF rhinorrhoea.

Epidemiology

The incidence of IIH is 1–2 in 100,000 population but increases to 19 in 100000 young obese women of reproductive age.^[2-4] There is a female predilection over men ranging from 3:1 to 15:1 in the literature.^[2,3,10] IIH may also occur in children, but association with obesity, and female sex is less common in prepubertal children.^[11]

Pathophysiology

Although recognized as a clinical entity for more than 100 years now, the pathogenesis of IIH remains unclear. Several mechanisms have been proposed, such as parenchymal edema, increased cerebral blood volume, excessive CSF production, venous outflow obstruction, compromised CSF resorption, and contribution of

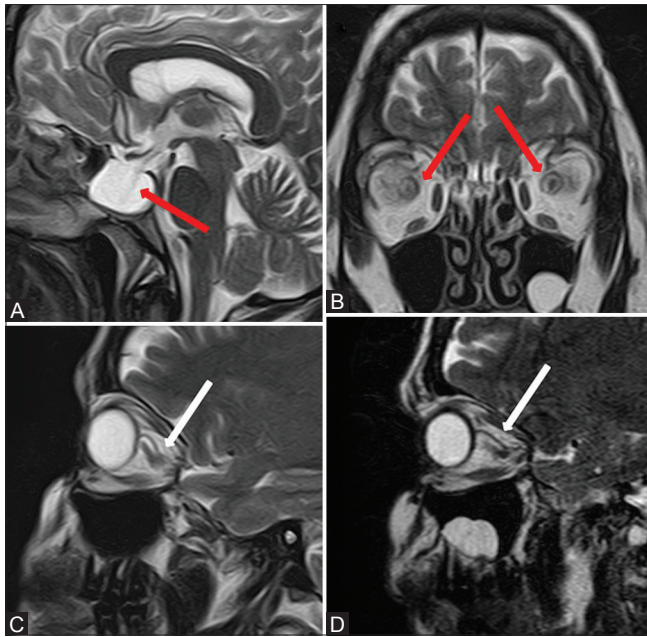


Figure 7 (A-D): Sagittal T2 weighted image in case 2 demonstrates ballooned empty sella (red arrow; image A). Coronal T2 image shows CSF distended bilateral perioptic subarachnoid space (red arrows; image B). Oblique sagittal T2 weighted images show vertical tortuosity of bilateral optic nerves (white arrows; images C and D)

inflammatory factors.^[12,13] Despite numerous theories, there is no consensus about the exact cause of IIH.

Says and Joynt reported microscopic evidence of intracellular and extracellular edema,^[14] which was later supported by MRI.^[15] Dandy first hypothesized increase in either cerebral blood volume or amount of CSF in IIH.^[16] An excessive CSF production and abnormally increased outflow resistance to the CSF has also been proposed.^[10,12,17]

The most important risk factors of IIH are obesity and female gender with majority of men with IIH are also obese.^[18] There is a positive correlation between body mass index and the risk of IIH.^[19] Successfully treated IIH with increased weight was found to be associated with the recurrence of this disease.^[20] The severity of obesity correlated with more advanced clinical symptomatology, which significantly reduced even with a small weight reduction.^[21,22]

Despite a stronger risk factor, it remains undetermined how obesity causes IIH. Few suggestions include increased intra-abdominal pressure probably leading to reduced cerebral venous drainage and chronic inflammation associated with obesity causing pro-thrombotic and pro-inflammatory state abnormal metabolism of vitamin A.^[10,13,23-25]

Sex hormones have also been attributed to IIH because of the preferential occurrence among post-pubertal, pre-menopausal women with no gender predilection before puberty.^[18,26] IIH has been linked to use of oral contraceptive

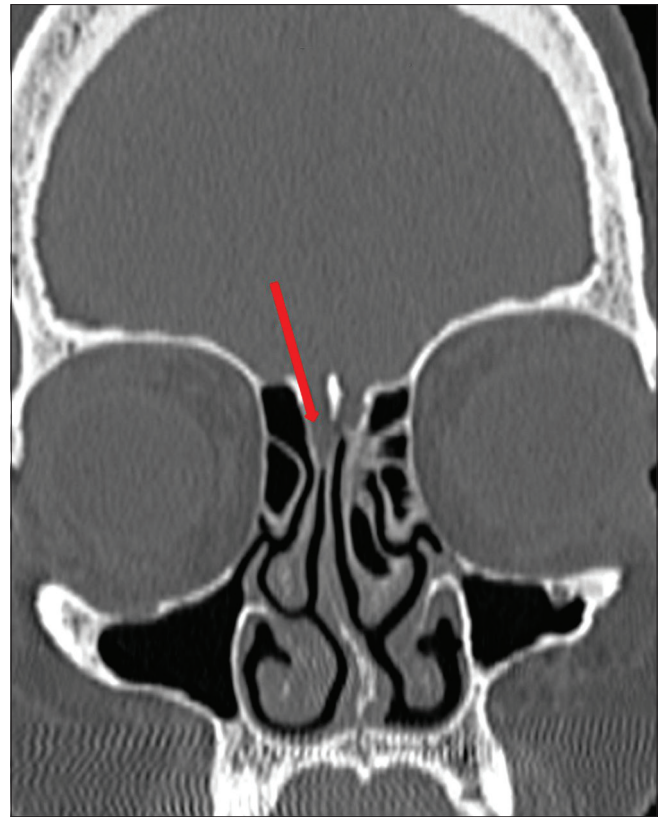


Figure 8: Coronal high resolution CT scan in case 2 demonstrates a defect in right cribriform plate (red arrow)

pills and polycystic ovarian syndrome (PCOS). There is a high prevalence of PCOS in women referred for IIH ranging from 39-57%.^[27,28]

Recent study by Skau *et al.* evaluated the natriuretic peptide system as a possible cause of altered intracranial pressure autoregulation in patients with IIH.^[29] Several drugs such as retinoids and steroids have been shown to induce Aquaporin (AQP) 1 expression suggesting the causative role of AQP1 in the pathogenesis of drug-induced IIH.^[30,31]

Recently, there have been reports that some patients with IIH have bilateral transverse sinus stenosis (BTSS).^[32] This led to a debate on whether such a stenosis is the cause or effect of IIH.^[33] Venous sinus stenting was, therefore, used in some patients with conflicting results.^[33-37] Another important observation was that one-third of the patients with BTSS had normal CSF pressure, suggesting that BTSS is only one of the contributing factors causing IIH.^[38]

Clinical presentation

Majority of the times IIH presents as headache, papilledema, visual symptoms and signs, without any lateralizing findings in the neurological examination, and normal CSF findings.^[39] The headache in IIH should be progressive with at least one of these characteristics; daily occurrence, diffuse and/or constant (non-pulsating), and aggravated by

coughing or straining.^[40] Infrequently, IIH may present in the absence of papilledema.^[41]

The diagnostic criteria of IIH were first defined by Dandy in 1937^[16] and were later modified as “Modified Dandy Criteria” by Smith in 1985.^[42] The current modified Dandy criteria used for the diagnosis are (i) signs and symptoms of increased intracranial pressure; (ii) no other neurological abnormalities or impaired level of consciousness; (iii) elevated CSF opening pressure with normal CSF composition; (iv) a neuroimaging study that shows no etiology for increased intracranial pressure; and (v) no other cause for intracranial hypertension found.^[43]

Imaging

MRI or contrast-enhanced CT are the imaging that can be performed for typical IIH patients.^[44]

Friedman *et al.* proposed updated criteria for the diagnosis which required:^[45]

1. Papilledema, normal neurologic examination except for intracranial nerve abnormalities
2. Neuroimaging findings including normal brain parenchyma
3. Normal CSF composition and elevated lumbar puncture opening
4. In the absence of papilledema, the diagnosis would be suggested with at least 3 of the following neuroimaging features were present additionally:
 - A) Empty sella
 - B) Flattening of the posterior aspect of the globe
 - C) Distention of the perioptic subarachnoid space with or without a tortuous optic nerve.
 - D) Transverse venous sinus stenosis.

Additional MRI findings that have been seen in IIH are flattening of posterior sclera and intraocular protrusion of the prelaminar optic nerve [Figure 9A and B].^[46]

Both high-resolution noncontrast CT and MR cisternography have been utilized for the diagnosis of a CSF leak. The sensitivity, specificity, and accuracy of CT cisternography were 92%, 100%, and 93%, respectively compared to 87%, 100%, and 89%, respectively for MR cisternography. In addition, CT and MR cisternography complement each other, with CT demonstrating the bone defect and MR demonstrating CSF leak leading to combined sensitivity, specificity, and accuracy improving to 95%, 100%, and 96%, respectively.^[47]

Conclusion

Spontaneous CSF rhinorrhoea is a known complication of IIH, however, sole presenting symptom in IIH is extremely rare. IIH typically presents with headache, pulsatile tinnitus,

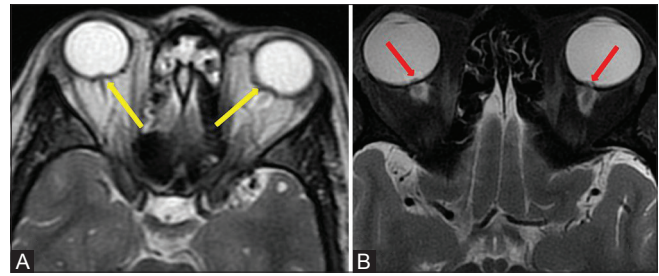


Figure 9 (A and B): Axial MRI images demonstrate flattening of posterior sclera in known patients with IIH (image A; yellow arrows) and intraocular protrusion of the prelaminar optic nerves (image B, red arrows)

dizziness, nausea, vomiting, and visual disturbance. In the absence of trauma, MR cisternography along with MRI brain should be offered to the patients to identify the site of leakage as well as changes of IIH. These include empty sella, distended perioptic subarachnoid space, vertical tortuosity of the optic nerves, flattening of posterior sclera, and intraocular protrusion of the prelaminar optic nerve. In addition, MR venography may be performed to exclude bilateral venous sinus stenosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Soler D, Cox T, Bullock P, Calver DM, Robinson RO. Diagnosis and management of benign intracranial hypertension. *Arch Dis Child* 1998;78:89-94.
2. Durcan FJ, Corbett JJ, Wall M. The incidence of pseudotumor cerebri. Population studies in Iowa and Louisiana. *Arch Neurol* 1988;45:875-7.
3. Radhakrishnan K, Ahlskog JE, Cross SA, Kurland LT, O'Fallon WM. Idiopathic intracranial hypertension (pseudotumor cerebri). Descriptive epidemiology in Rochester, Minn, 1976 to 1990. *Arch Neurol* 50:78-80.
4. Jensen RH, Radojicic A, Yri H. The diagnosis and management of idiopathic intracranial hypertension and the associated headache. *Ther Adv Neurol Disord* 2016;9:317-26.
5. Shaw GY, Million SK. Benign intracranial hypertension: A diagnostic dilemma. *Case Rep Otolaryngol* 2012;2012:814696.
6. Clark D, Bullock P, Hui T, Firth J. Benign intracranial hypertension: A cause of CSF rhinorrhoea. *J Neurol Neurosurg Psychiatry* 1994;57:847-9.
7. Saifudheen K, Gafoor A, Arun G, Abdurahiman P, Jose J.

- Idiopathic intracranial hypertension presenting as CSF rhinorrhea. *Ann Indian Acad Neurol* 2010;13:72-3.
8. Rosenfeld E, Dotan G, Kimchi T, Kesler A. Spontaneous cerebrospinal fluid otorrhea and rhinorrhea in idiopathic intracranial hypertension patients. *J Neuroophthalmol* 2013;33:113-6.
 9. Pérez MA, Bialer OY, Bruce BB. Primary spontaneous cerebrospinal fluid leaks and idiopathic intracranial hypertension. *J Neuroophthalmol* 2013;33:327-34.
 10. Baykan B, Ekizoglu E, Altioğka Uzun G. An update on the pathophysiology of idiopathic intracranial hypertension alias pseudotumor cerebri. *Agri*. 2015;27:63-72.
 11. Phillips PH, Repka MX, Lambert SR. Pseudotumor cerebri in children. *J AAPOS* 1998;2:33-8.
 12. Skau M, Brennum J, Gjerris F, Jensen R. What is new about idiopathic intracranial hypertension? An updated review of mechanism and treatment. *Cephalgia* 2006;26:384-99.
 13. Sinclair AJ, Ball AK, Burdon MA, Clarke CE, Stewart PM, Curnow SJ, *et al.* Exploring the pathogenesis of IIH: An inflammatory perspective. *J Neuroimmunol* 2008;201-2:212-20.
 14. Sahs AJ, Joynt RJ. Brain swelling of unknown cause. *Neurology* 1956;6:791-803.
 15. Moser FG, Hilal SK, Abrams G, Bello JA, Schipper H, Silver AJ. MR imaging of pseudotumor cerebri. *AJR Am J Roentgenol* 1988;150:903-9.
 16. Dandy WE. Intracranial pressure without brain tumor: Diagnosis and treatment. *Ann Surg* 1937;106:492-513.
 17. Malm J, Kristensen B, Markgren P, Ekstedt J. CSF hydrodynamics in idiopathic intracranial hypertension: Along-term study. *Neurology* 1992;42:851-8.
 18. Bruce BB, Kedar S, Van Stavern GP, Monaghan D, Acierno MD, Braswell RA, *et al.* Idiopathic intracranial hypertension in men. *Neurology* 2009;72:304-9.
 19. Daniels AB, Liu GT, Volpe NJ, Galetta SL, Moster ML, Newman NJ, *et al.* Profiles of obesity, weight gain, and quality of life in idiopathic intracranial hypertension (pseudotumor cerebri). *Am J Ophthalmol* 2007;143:635-41.
 20. Ko MW, Chang SC, Ridha MA, Ney JJ, Ali TF, Friedman DI, *et al.* Weight gain and recurrence in idiopathic intracranial hypertension: A case-control study. *Neurology* 2011;76:1564-7.
 21. Szewka AJ, Bruce BB, Newman NJ, Biousse V. Idiopathic intracranial hypertension: Relation between obesity and visual outcomes. *J Neuroophthalmol* 2013;33:4-8.
 22. Johnson LN, Krohel GB, Madsen RW, March GA Jr. The role of weight loss and acetazolamide in the treatment of idiopathic intracranial hypertension (pseudotumor cerebri). *Ophthalmology* 1998;105:2313-7.
 23. Sugerman HJ, DeMaria EJ, Felton WL 3rd, Nakatsuka M, Sismanis A. Increased intra-abdominal pressure and cardiac filling pressures in obesity-associated pseudotumor cerebri. *Neurology* 1997;49:507-11.
 24. Tabassi A, Salmasi AH, Jalali M. Serum and CSF vitamin A concentrations in idiopathic intracranial hypertension. *Neurology* 2005;64:1893-6.
 25. Warner JE, Larson AJ, Bhosale P, Digre KB, Henley C, Alder SC, *et al.* Retinol-binding protein and retinol analysis in cerebrospinal fluid and serum of patients with and without idiopathic intracranial hypertension. *J Neuroophthalmol* 2007;27:258-62.
 26. Cinciripini GS, Donahue S, Borchert MS. Idiopathic intracranial hypertension in prepubertal pediatric patients: Characteristics, treatment, and outcome. *Am J Ophthalmol* 1999;127:178-82.
 27. Glueck CJ, Aregawi D, Goldenberg N, Golnik KC, Sieve L, Wang P. Idiopathic intracranial hypertension, polycystic-ovary syndrome, and thrombophilia. *J Lab Clin Med* 2005;145:72-82.
 28. Glueck CJ, Iyengar S, Goldenberg N, Smith LS, Wang P. Idiopathic intracranial hypertension: Associations with coagulation disorders and polycystic-ovary syndrome. *J Lab Clin Med* 2003;142:35-45.
 29. Skau M, Goetze JP, Rehfeld JF, Jensen R. Natriuretic propeptides in idiopathic intracranial hypertension. *Regul Pept* 2010;164:71-7.
 30. Johansson PA, Dziegielewska KM, Ek CJ, Habgood MD, Møllgård K, Potter A, *et al.* Aquaporin-1 in the choroid plexuses of developing mammalian brain. *Cell Tissue Res* 2005;322:353-64.
 31. Umenishi F, Schrier RW. Induction of human aquaporin-1 gene by retinoic acid in human erythroleukemia HEL cells. *Biochem Biophys Res Commun* 2002;293:913-7.
 32. Riggeal BD, Bruce BB, Saindane AM, Ridha MA, Kelly LP, Newman NJ, *et al.* Clinical course of idiopathic intracranial hypertension with transverse sinus stenosis. *Neurology* 2013;80:289-95.
 33. McGeeny BE, Friedman DI. Pseudotumor cerebri pathophysiology. *Headache* 2014;54:445-58.
 34. Stevens SA, Previte M, Lakin WD, Thakore NJ, Penar PL, Hamschin B. Idiopathic intracranial hypertension and transverse sinus stenosis: A modelling study. *Math Med Biol* 2007;24:85-109.
 35. Puffer RC, Mustafa W, Lanzino G. Venous sinus stenting for idiopathic intracranial hypertension: A review of the literature. *J Neurointerv Surg* 2013;5:483-6.
 36. Kumpe DA, Bennett JL, Seinfeld J, Pelak VS, Chawla A, Tierney M. Dural sinus stent placement for idiopathic intracranial hypertension. *J Neurosurg* 2012;116:538-48.
 37. Rohr A, Dörner L, Stingele R, Buhl R, Alfke K, Jansen O. Reversibility of venous sinus obstruction in idiopathic intracranial hypertension. *AJNR Am J Neuroradiol* 2007;28:656-9.
 38. Bono F, Messina D, Giliberto C, Cristiano D, Broussard G, Fera F, *et al.* Bilateral transverse sinus stenosis predicts IIH without papilloedema in patients with migraine. *Neurology* 2006;67:419-23.
 39. Wall M, George D. Idiopathic intracranial hypertension. A prospective study of 50 patients. *Brain* 1991;114:155-80.
 40. Headache Classification Subcommittee of the International Headache Society. The International classification of headache disorders: 2nd Edition. *Cephalgia* 2004;24(Suppl 1):9-160.
 41. Bono F, Quattrone A. Idiopathic intracranial hypertension without papilloedema in headache sufferers. *Cephalgia* 2009;29:594.
 42. Smith JL. Whence pseudotumor cerebri? *J Clin Neuroophthalmol* 1985;5:55-6.
 43. Wall M. Idiopathic intracranial hypertension. *Neurol Clin* 2010;28:593-617.
 44. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology* 2002;59:1492-5.
 45. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology* 2013;81:1159-65.
 46. Suzuki H, Takahashi J, Kobayashi K, Nagasawa K, Tashima K, Kohno Y. MR imaging of idiopathic intracranial hypertension. *Am J Neuroradiol* 2001;22:196-9.
 47. Shetty PG, Shroff MM, Sahani DV, Kirtane MV. Evaluation of high-resolution CT and MR cisternography in the diagnosis of cerebrospinal fluid fistula. *AJNR Am J Neuroradiol* 1998;19:633-9.