

Spontaneous cerebrospinal fluid rhinorrhea as a presenting symptom of cerebellopontine schwannoma

Sir,

Cerebrospinal fluid (CSF) leak (cerebrospinal fluid fistula) usually occurs when there is an abnormal communication between subarachnoid space and either the sinuses or the ear (from the nose, CSF rhinorrhea or the ear, CSF otorrhea).^[1-4] Spontaneous CSF rhinorrhea is rare as a primary presenting symptom and a long-standing spontaneous CSF rhinorrhea may indicate the possibility of a concurrent intracranial pathology.^[1,5,6] A 50-year-old gentleman presented with watery intermittent discharge from the nose off and on since the last one year and was accentuated by coughing and bending forwards, was more profuse when arising from bed and changing positions particularly tilting the head to right side. He had one episode of fever with chills and rigors, headache 4 months back, features suggestive of meningitis. There was no history of focal weakness, seizures. The patient also reported mild headache before nasal discharge that used to relieve after the nasal discharge. His general and systemic examination was normal. Fundus examination was normal, and there were no motor, sensory, or cerebellar signs, and no signs of meningitis. His magnetic resonance (MR) imaging of the brain showed a left cerebellopontine angle tumor with erosion of the petrous bone, middle ear effusion, and hyperintensity in the left side of the sphenoid sinus [Figure 1]. There was no evidence of empty sella or hydrocephalus on imaging. The patient underwent left retromastoid suboccipital craniectomy and near total decompression of the tumor. The CSF rhinorrhea ceased completely after the surgery.

Most commonly CSF rhinorrhea occurs following head trauma or as a postoperative complication following intracranial surgery. Spontaneous CSF rhinorrhea is an uncommon complication (3–4%) and can be a presenting feature of several benign and malignant intracranial conditions including tumors.^[1,3-17] Spontaneous rhinorrhea is an extremely rare presentation of acoustic neuroma^[4,8,18,19] and as seen in the present case tumors it produces CSF rhinorrhea by destructive lesions along the skull base.^[3,5,12,14] Probably it may be due the erosion of the temporal bone by the tumor growth producing communication between the air cell and subarachnoid space, a mechanism similar to as described by Kaufman *et al.*^[20] that pneumatization of the middle fossa floor a lateral extension of the sphenoid sinus into the greater wing of the sphenoid.

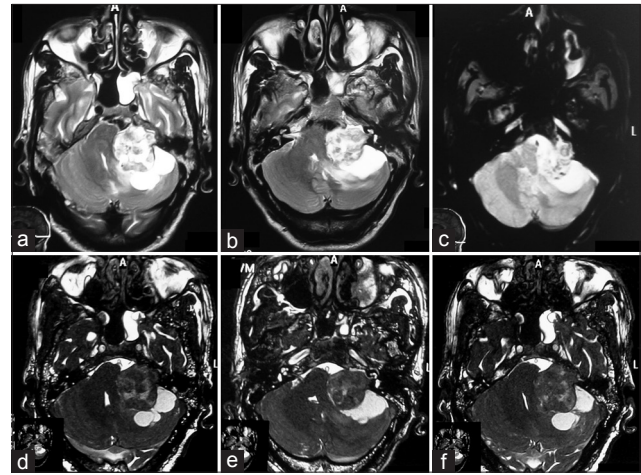


Figure 1: MRI T2W axial images showing left cerebellopontine angle tumor with mass effect and midline shift

The majority of spontaneous cases occur in adult females in their fourth decade of life and as spontaneous CSF leak is very uncommon, the initial insidious onset can often be mistaken for a feature of rhinitis that requires a high index of suspicion for diagnosis including a detailed history, endoscopic examination, and appropriate investigations.^[16] Clear nasal discharge that is positive for glucose has been used historically to diagnose CSF rhinorrhea but is not diagnostic of CSF leak, to overcome this b-2 transferrin has become a diagnostic test for CSF fistula with a sensitivity nearing 100% and specificity of 95%.^[16,21] Precise preoperative localization of CSF leak is essential for surgical repair.^[1,16] High resolution CT scans have been advocated as the noninvasive radiological investigation of choice.^[10,16,17] MRI is helpful to investigate the soft tissue and possible site of leak as it is noninvasive and has no radiation risk; however, bony defects can be better visualized in CT scans.^[16] The management of spontaneous high-pressure CSF rhinorrhea includes the removal of the primary pathology (i.e. tumors)^[11] or diversion of CSF (i.e. hydrocephalus)^[11] and/or surgical closure of the defect.^[14,16,22] This can be achieved *via* a frontal craniotomy with the success rate of between 60% and 80%^[16,22] or can be performed by a less invasive transnasal endoscopic approach enabling the surgeon to achieve good visualization of the site of the leak^[16,23] having a success rate of between 76% and 95%.^[23] In summary, spontaneous CSF rhinorrhea may be a manifestation of underlying brain tumors where it act as a natural vent or protective mechanism preventing rise in intracranial pressure. Physicians should be careful and

should have high degree of suspicion for the diagnoses of such cases.^[1,16]

Amit Agrawal, Lakshmi Narayan Garg¹, Amit Mittal², Sunil Sampley³, Ravish Aneja³

Department of Neurosurgery, Narayana Medical College Hospital, Chinthareddypalem, Nellore, Andhra Pradesh, ¹Departments of ENT, ²Radiology, and ³Surgery, MM Institute of Medical Sciences and Research, Mullana, Ambala, Haryana, India.
E-mail: dramitagrawal@gmail.com

REFERENCES

- Muzumdar D, Nadkarni T, Goel A. Spontaneous cerebrospinal fluid rhinorrhea as a presenting symptom of aqueductal stenosis--case report. *Neurol Med Chir (Tokyo)* 2003;43:626-9.
- Abuabara A. Cerebrospinal fluid rhinorrhoea: Diagnosis and management. *Med Oral Patol Oral Cir Bucal* 2007;12:E397-400.
- Ommaya AK. Cerebrospinal fluid rhinorrhea. *Neurology* 1964;14:106-13.
- Ommaya AK, Di Chiro G, Baldwin M, Pennybacker JB. Non-traumatic cerebrospinal fluid rhinorrhoea. *J Neurol Neurosurg Psychiatry* 1968;31:214-25.
- Hanel RA, Prevedello DM, Correa A, Antoniuk A, Araujo JC. Cerebrospinal fluid fistula as the presenting manifestation of pituitary adenoma: Case report with a 4-year follow-up. *Arq Neuropsiquiatr* 2001;59:263-5.
- Kane PJ, Mendelow AD, Keogh AJ, Symon L. Cerebrospinal fluid rhinorrhoea associated with colloid cyst. *Br J Neurosurg* 1991;5:317-20.
- Morley TP, Hetherington RF. Traumatic cerebrospinal fluid rhinorrhea and otorrhea, pneumocephalus, and meningitis. *Surg Gynecol Obstet* 1957;104:88-98.
- Rovit RL, Schechter MM, Nelson K. Spontaneous "high-pressure cerebrospinal rhinorrhea" due to lesions obstructing flow of cerebrospinal fluid. *J Neurosurg* 1969;30:406-12.
- Vaquero J, Cabezudo JM, Salazar AR, Brasa J. Symptomatic intrasphenoidal meningoencephalocele after removal of a parasagittal meningioma. *Acta Neurochir (Wien)* 1981;57:61-5.
- Hsieh CT, Liu MY, Su WF, Ju DT. Spontaneous cerebrospinal fluid rhinorrhea: A rare initial presentation of clival chordoma. *Neurol India* 2009;57:513-4.
- Cole IE, Keene M. Cerebrospinal fluid rhinorrhoea in pituitary tumours. *J R Soc Med* 1980;73:244-54.
- Norsa L. Cerebrospinal rhinorrhea with pituitary tumors. *Neurology* 1953;3:864-8.
- O'Connell JE. primary spontaneous cerebrospinal fluid rhinorrhoea. *J Neurol Neurosurg Psychiatry* 1964;27:241-6.
- Clark D, Bullock P, Hui T, Firth J. Benign intracranial hypertension: A cause of CSF rhinorrhoea. *J Neurol Neurosurg Psychiatry* 1994;57:847-9.
- Bhatjwale M, Goel A, Muzumdar D. Spontaneous cerebrospinal fluid (CSF) rhinorrhoea in spongiform dysplasia of the cranium: An unusual presentation of neurofibromatosis. *Br J Neurosurg* 1998;12:592-3.
- Gendeh BS, Wormald PJ, Forer M, Goh BS, Misiran K. Endoscopic repair of spontaneous cerebro-spinal fluid rhinorrhoea: A report of 3 cases. *Med J Malaysia* 2002;57:503-8.
- Fatemi N, Dusick JR, Gorgulho AA, Mattozo CA, Moftakhar P, De Salles AA, *et al.* Endonasal microscopic removal of clival chordomas. *Surg Neurol* 2008;69:331-8.
- Bhayani R, Bafna P, Goel A. Cerebrospinal fluid rhinorrhoea: Unusual presentation of acoustic neurinoma. *Br J Neurosurg* 1993;7:705-7.
- Cushing H. Further concerning the acoustic neuromas. *Laryngoscope* 1921;31:209-28.
- Kaufman B, Nulsen F, Weiss M, Brodkey J, White R, Sykora G. Acquired spontaneous, nontraumatic normal-pressure cerebrospinal fluid fistulas originating from the middle fossa. *Radiology* 1977;122:379-87.
- Reisinger PW, Hochstrasser K. The diagnosis of CSF fistulae on the basis of detection of beta 2-transferrin by polyacrylamide gel electrophoresis and immunoblotting. *J Clin Chem Clin Biochem* 1989;27:169-72.
- Hughes RG, Jones NS, Robertson JJ. The endoscopic treatment of cerebrospinal fluid rhinorrhoea: The Nottingham experience. *J Laryngol Otol* 1997;111:125-8.
- Mattox DE, Kennedy DW. Endoscopic management of cerebrospinal fluid leaks and cephaloceles. *Laryngoscope* 1990;100:857-62.

Access this article online

Quick Response Code:



Website:

www.ijns.in

DOI:

10.4103/2277-9167.102292