

Spontaneous Cerebrospinal Fluid Rhinorrhea as the Presenting Symptom of Idiopathic Intracranial Hypertension: A Case Series

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Abstract- Although rare, Cerebrospinal Fluid (CSF) Leakage can result in deadly complications such as meningitis and brain abscess. Previously, primary spontaneous CSF leakage was referred to leakages without any detectable causes. However, it has been found recently that it may be related to abnormal increased intracranial pressure (ICP). Here, we reported demographic, clinical, and therapeutic features in addition to the outcomes of five patients with Idiopathic Intracranial Hypertension (IIH) presented with spontaneous CSF leakage as the initial symptom. Four of our patients were female. The mean age was 38 years old. Rhinorrhea was the first manifestation of the CSF leakage in our patients. Ethmoidal cells were the most common site of leakage. The mean opening pressures (OP) was 31.3 cmH₂O. The computed tomography (CT) scan and magnetic resonance imaging (MRI) of the brain was normal in all patients except one patient showing fullness in left ethmoidal cells. In all of the patients, cerebral CT cisternography was diagnostic to detect the site of leakage. CSF leak in two patients resolved with medical therapy but CSF diversion procedure was mandatory in other three patients. CSF leakage resolved in all of them. CSF leakage can be the first and only presenting symptom of abnormal increased ICP. The key point in patient treatment is controlling the elevated ICP, even though some patients may need to CSF diversion procedure eventually.

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Introduction

Cerebrospinal fluid leakage is a rare entity but it can result in awful complication such as meningitis and brain abscess (1). The cause of CSF leak is disruption in Dura and defect in bone accompanying to intracranial pressure greater than defect strength (2). The term primary spontaneous CSF leak referred to CSF leak due to undetectable causes, but investigators recently found that it is caused by elevated intracranial pressure (3,4). It also can be a rare complication of idiopathic intracranial hypertension (IIH) (1). Here we present five cases of IIH presenting with CSF rhinorrhea as the initial symptom.

Case Report

Case 1

A 34-year-old woman presented with spontaneous CSF rhinorrhea without any other complaint. Her body

mass index (BMI) and the OP was normal. Brain MRI revealed no remarkable abnormality and brain CT cisternography showed CSF leakage from ethmoidal cells. The patient underwent surgical repair procedure. The patient represented with headache and diplopia one month after surgery. Fundoscopy showed papilledema. Lumbar puncture (LP) revealed normal CSF composition but an OP of 255 mmH₂O. She has gone under medical treatment for IIH (Acetazolamide) by which CSF leakage has stopped and ICP stabilized in the normal range.

Case 2

A 43-year-old woman presented with CSF rhinorrhea in left nostril from 2 months before admission. She was overweight. The brain CT scan and MRI showed no remarkable abnormality. The brain CT Cisternography showed CSF leakage from right ethmoidal cells. LP showed an OP of 250 mmH₂O with normal CSF composition. After medical therapy failure, the patient

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Spontaneous cerebrospinal fluid rhinorrhea

underwent a lumboperitoneal shunt insertion surgery, and CSF leak was stopped.

Case 3

A 46-year-old man with the complaint of CSF leakage of his nose was admitted to our institute. Fundoscopy and OP were normal with no other abnormal findings in physical examination. BMI was in normal range. Brain imaging showed no abnormal finding while the CT cisternography revealed CSF fistula from ethmoidal cells. The defect was repaired surgically, but recurrent intermittent leakage continued. Fundoscopy examination repeated and showed papilledema. LP revealed an OP of 330 mmH₂O and normal CSF composition. Since the conservative treatment was failed, the patient went under lumboperitoneal shunt insertion by which the symptoms improved and CSF leakage stopped.

Case 4

A 25-year-old woman presented with CSF rhinorrhea with no other accompanying symptom like headache, nausea, vomiting or fever. She had a history of head trauma and left frontal lobe brain contusion from three years earlier. The brain CT scan showed porencephaly in left frontal lobe secondary to previous trauma. LP demonstrated normal OP and CSF composition. The CT cisternography demonstrated CSF leakage from frontal sinus. Because of the history of trauma, transcranial repair of frontal sinus defect was done. The patient represented recurrent leakage about one month later. LP revealed an OP of 430 mmH₂O with normal CSF composition. The patient underwent a lumboperitoneal shunt, with resolution of CSF leak. During follow-up, she was asymptomatic and had no CSF leak.

Case 5

A 42-year-old woman presented with CSF rhinorrhea from left nostril since 4 months ago, was admitted to our neurosurgical department. She had a history of endoscopic paranasal sinus surgery because of sinonasal problem. Brain CT scan, MRI, and CT cisternography showed an encephalocele through bone defect associated with CSF leakage in left ethmoidal cells. In order to repair the defect, she underwent bifrontal craniotomy. One year later, she was still complaining of recurrent CSF leakage when the LP revealed an OP of 300 mmH₂O with normal CSF composition. Medical treatment administered and CSF leakage stopped with successful control of ICP.

Discussion

The idiopathic intracranial hypertension (IIH) also named pseudotumor cerebrii (PTC) is the disorder of increased intracranial pressure (ICP) without ventriculomegaly or mass lesion, and with normal cerebrospinal fluid (CSF) composition (5). It is a rare incidence that PTC presents initially with CSF leakage (1). In the cases of active leakage, other symptoms of raised ICP, may not present or may occur after repair of skull base defect (6). CSF diversion should be made in order to treat the IIH patients with CSF leak that are refractory to conservative treatment (7).

In our study, four patients of five were female. The mean age was 38-year-old. None of them had history of raised ICP or its suggestive symptoms. Two patients had history of previous head trauma and endonasal surgery.

The primary presenting symptom in all of them was CSF rhinorrhea. Although, this result is similar to some previous reports (8-10) but the literature usually reported CSF leak in the course of the IIH, not as the first presenting symptom (7,11,12). In four patients, ethmoidal cells were the site of the leak as other studies reported that the cribriform plate is the most common site for leakage (8). As mentioned earlier, IIH is most prevalence in obese middle age female and in our series four of my patient was female, and three of them had the BMI higher than normal (8,10,11,12). In our series, patients with IIH presented only CSF leak, no other symptoms. All of the patient revealed rhinorrhea. The OP was high in all patients. The CT scans and MRIs showed no abnormality causes ICP elevation and CT cisternography detected site of leakage. CSF leak in two patients resolved with medical treatment (Acetazolamide). One patient with CSF leak had a history of previous trauma, but the latest diagnosis for her was IIH, not traumatic fistula. She did not respond to the surgical transcranial repair of frontal sinus defect, then CSF diversion procedure successfully resolved leak.

In all our patients, the surgery failed because of raised ICP, but lowering of ICP by medical and surgical treatment, cut out CSF leakage. The surgical procedure of choice was lumboperitoneal shunt, and no complication was found related to the procedure.

Tables 1-3 compare some of the most important studies regarding CSF leak.

CSF leakage can be the first and only presenting symptom of increased ICP. The key point in patient treatment is controlling the elevated ICP, even though some patients may need to CSF diversion procedure eventually.

Table 1. Demographic, clinical and therapeutic features of patients in this survey

Case	Sex	Age	BMI	Primary Symptom	Another symptom	Site of defect	CTscan	MRI	CT cisternography	OP (cmH ₂ O)	Treatment	Outcome
1	F	34	Normal ¹	Rhinorrhea	None	Ethmoidal cells	NL	NL	Showed leakage	25.5	1-TC repair of the defect 2-Medical therapy	Stop leakage
2	F	43	Overweight	Rhinorrhea	Blurred vision	Ethmoidal cells	NL	NL	Showed leakage	25	1- Medical therapy 2-LumboPritoneal shunt	Stop leakage
3	M	46	Normal	Rhinorrhea	None	Ethmoidal cells	NL	NL	Showed leakage	33	1-TC repair of the defect 2-Medical therapy 3- LumboPritoneal shunt	Stop leakage
4	F	25	Overweight	Rhinorrhea	None	Frontal sinus	NL ²	NL ²	Showed leakage	43	1-TC repair of the defect 2-Medical therapy 3- LumboPritoneal shunt	Stop leakage
5	F	42	Obese	Rhinorrhea	None	Ethmoidal cells	Ethmoidal cells fullness	Ethmoidal cells fullness	Showed leakage	30	1-TC repair of the defect 2-Medical therapy	Stop leakage

1-Normal (20<BMI<25), overweight (25<BMI<30), obese (30<BMI)

2-Brain CT scan and MRI showed left frontal lobe porencephaly due to previous head trauma

Table 2. Previous reports of IIH-induced CSF leak and our study

Study	Number	Sex	Age	Leak	Leak as initial symptom	OP	Treatment	Outcome
(Rosenfeld, Dotan et al. 2013) ⁷	1	F	48	Rhinorrhea	No	28	EN repair+ medical therapy	Resolved
	2	F	35	Rhinorrhea	No	NR	EN repair+ medical therapy	Resolved
	3	F	42	Rhinorrhea	No	27	LumboPritoneal shunt	Resolved
	4	F	44	Otorrhea	No	37	LumboPritoneal shunt	Resolved
(Choh 2006) ¹¹	1	F	Elderly	Rhinorrhea	No	NR	Repair of defect+ medical therapy	Resolved
	1	F	33	Rhinorrhea	No	30	LumboPritoneal shunt	Resolved
(Clark, Bullock et al. 1994) ¹²	2	F	44	Rhinorrhea	No	34	LumboPritoneal shunt	Intermittent leak
	3	F	42	Rhinorrhea	No	34	Repair of defect+ multiple LP	Resolved
	4	F	34	Rhinorrhea	No	30	LumboPritoneal shunt+ fistula repair	Resolved
	1	M	40	Rhinorrhea	Yes	↑	Repair of defect	Resolved
(Deenadayal, Vidyasagar et al. 2013) ⁸	2	F	39	Rhinorrhea	Yes	↑	Repair of defect	Resolved
	3	F	40	Rhinorrhea+Cough	Yes	↑	Repair of defect	Resolved
	4	F	40	Rhinorrhea+Headache	Yes	↑	Repair of defect	Resolved
	5	M	28	Rhinorrhea	Yes	↑	Repair of defect	Resolved
	6	F	54	Rhinorrhea+Sneezing	Yes	↑	Repair of defect	Resolved
	7	F	58	Rhinorrhea	Yes	↑	Repair of defect	Resolved
	1	F	49	Rhinorrhea	Yes	37	Repair of defect+LumboPritoneal shunt	Resolved
(Perez, Bialer et al. 2013) ¹⁰	2	F	32	Rhinorrhea+Headache	No	42	Repair of defect+CSF diversion procedure	Resolved
	3	M	43	Headache+Rhinorrhea	No	33.5	VP shunt+dural repair	Resolved
	1	F	34	Rhinorrhea+Headache	Yes	36	Repair of defect+medical therapy	Resolved
(Matsubara, Akutsu et al. 2014) ⁹ Our study	1	F	34	Rhinorrhea	Yes	25.5	TC repair of defect+medical therapy	Resolved
	2	F	43	Rhinorrhea	Yes	25	LumboPritoneal shunt	Resolved
	3	M	46	Rhinorrhea	Yes	33	TC repair of defect+medical therapy+ LumboPritoneal shunt	Resolved
	4	F	25	Rhinorrhea	Yes	43	Medical therapy+TC repair of defect+ LumboPritoneal shunt	Resolved
	5	F	42	Rhinorrhea	Yes	30	TC repair of defect+medical therapy	Resolved

EN: Endoscopic Nasal/ LP: Lumbar Puncture/ TC: Transcranial

Table 3. Previous reports of IHH-induced CSF leak and our study

Study	Year	No of cases	Age (average) years	BMI (average) kg/m ²	F/M	The most common site of the leak	CSF leak as the initial symptom	OP(average) Cm H ₂ O
(Woodworth, Prince <i>et al.</i> 2008) ¹³	2008	55	61	36.2	43/12	Lateral Sphenoid sinus	81% cases	27
(Chaaban, Illing <i>et al.</i> 2014) ¹	2014	46	51.2	35.6	32/14	Frontal sinus	98% cases	32.3
(Yang, Wang <i>et al.</i> 2011) ⁴	2011	21	53	31.2	18/3	Ethmoid sinus(13/21)	14% cases	25.5
(Deenadayal, Vidyasagar <i>et al.</i> 2013) ⁸	2013	7	42.7	NR*(81.2 kg)	5/2	Anterior cribriform plate	100%cases	NR(raised)
Our study (Rosenfeld, Dotan <i>et al.</i> 2013) ⁷	2015	5	38	#	4/1	Ethmoidal cells	100%cases	31.3
(Clark, Bullock <i>et al.</i> 1994) ¹²	2013	4	42.2	34.7	4/0	Cribriform plate	None	30.6(in 3 patients)
(Perez, Bialer <i>et al.</i> 2013) ¹⁰	1994	4	38.2	32	4/0	NR	None	NR
(Choh 2006) ¹¹	2013	3	41.3	NR	2/1	Cribriform plate	2 from 3 patients	37.5
(Matsubara, Akutsu <i>et al.</i> 2014) ⁹	2006	1	NR	NR	1	Cribriform plate	none	NR
(Menku, Tucer <i>et al.</i> 2004) ¹⁴	2014	1	34	34.1	1	Olfactory cleft	1	36
	2004	1	42	NR	1	Sphenoid sinus	1	NR

*NR: Not Reported

One patient was obese, two patients were overweight and two patients had normal weight, respective

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