Neurological presentation of spontaneous skull base defects: Retrospective study

Orlova Y.[1] (yorlova@ufl.edu), Kalamangalam G. [1], Chen S. [2], Poynter J. [2], Justice J. [2], Lobo B. [2]

[1]University of Florida, Neurology, Gainesville, FL, United States, [2]University of Florida, Otolaryngology, Gainesville, FL, United States

Background: Spontaneous defects of the skull base and acquired encephalocele can be a form of idiopathic intracranial hypertension (IIH).

Methods: We performed a retrospective chart review of patients with spontaneous skull base defects and encephalocele at our institution during 2010-2019.

Results: We identified 175 patients with encephalocele, 129 (73.7%) women. Mean age 59.6 years. Mean body mass index (BMI) 36.2.
In a pilot analysis of 43 patients (37 women, 9 men), 37 (79.1%) had craniofacial pain, 21 hearing loss (48.8%) and 20 middle ear effusion (46.5%). 9 patients (20.1%) had a history of meningitis. 34 had surgical repair of skull base defects, but only 28 had overt clinical signs of CSF leak.

Neurological presentation: Only 16 patients were seen by neurologists, 10 had headache disorder (migraine in 5, secondary headache in 4, unspecified in 2, trigeminal neuralgia in 1). IIH was diagnosed in 7, but only 2 met formal diagnostic criteria, see details in Table 1. Epilepsy diagnosed in 3 patients.
CSF opening pressure was documented in 7 patients and was normal in 4 (11-18 cm H2O) and elevated in 3 (26-47 cm H2O). Most radiological studies did not comment on imaging signs of raised intracranial pressure.

Conclusion: Only a minority of patients were adequately evaluated and meet diagnostic criteria for IIH. A further analysis of our entire cohort is in progress

Table 1

Patient No.	Sex	Age (y)	Race	BMI	CSF Opening pressure (cm H2O)	Presenting symptom	Relevant past history	Skull base defect	CSF leak	Associated radiological findings
1	F	51	White	47.5	-	Headache, clear rhinorrhea, otorrhea	Sleep apnea	R mastoid defect, thinning of L cribiform plate	Yes	Partially empty sella
2	F	78	White	34.8	-	Clear rhinorrhea,	Visual loss, papilledema, headache, meningitis, high risk for sleep apnea	R cribiform plate	Yes	Partially empty sella
3	F	44	Black	39.8	-	Seizures, chronic daily headache	Lupus	L anterior temporal bone meningoencephalocele	No	Partially empty sella, narrowing of venous sinuses, downward displacement of cerebellar tonsils
4	F	38	White	50.6	38	Headache, clear rhinorrhea	IIH with papilledema and migraine since childhood,	bilateral cribiform meningoceles	Yes	Enlarged sella, Arachnoid granulation,
5	F	37	White	39.1	-	Headache, clear rhinorrhea, papilledema	Migraine with visual aura, PCOS, moderate risk for sleep apnea	Erosive changes at the cribiform plate, dorsum sellae, posterior clinoid processes	Yes	Partially empty sella, flattening of the posterior aspect of the globe, narrowing of transverse sinuses, distention of the peri-optic subarachnoid space
6	F	31	White	60.1	25	Headache, clear rhinorrhea,	Headache, bacterial meningitis, papilledema	No, prior ethmoid CSF leak repair	Yes	Distention of perioptic subarachnoid space
7	F	61	White	50.7	-	Headache, clear rhinorrhea, otorrhea,	Sleep apnea, viral meningitis,	L ethmoid, L tegment tympani, dehiscence or near dehiscence of both superior semicircular canals.	Yes	Not documented

Reference:

1. Nelson RF, Gantz BJ, Hansen MR. The rising incidence of spontaneous cerebrospinal fluid leaks in the United States and the association with obesity and obstructive sleep apnea. *Otol Neurotol*. 2015;36(3):476-480.
2. Pérez MA, Bialer OY, Bruce BB, Newman NJ, Biousse V. Primary spontaneous cerebrospinal fluid leaks and idiopathic intracranial hypertension. *J Neuroophthalmol*. 2013;33(4):330-337.
3. Morone PJ, Sweeney AD, Carlson ML, et al. Temporal Lobe Encephaloceles: A Potentially Curable Cause of Seizures. *Otol Neurotol*. 2015;36(8):1439-1442.