

Introduction

- There is increasing evidence that idiopathic intracranial hypertension (IIH) is a cause of spontaneous skull base CSF leaks.
- A spontaneous CSF leak can make diagnosing underlying IIH challenging since the leak acts like a shunt, reducing intracranial pressure (ICP) and preventing development of typical symptoms.
- We aimed to compare the use of ophthalmic exam for papilledema against lumbar puncture (LP) in the assessment of IIH in patients after repair of a spontaneous CSF leak to evaluate how often elevated ICP is not being diagnosed and treated after surgery.

Methods

- A retrospective review was performed on all patients presenting to a tertiary Neurotology practice from 2008 through 2022 with a diagnosis of spontaneous encephalocele with CSF otorrhea.
- Inclusion criteria were as follows: age ≥18 years, diagnosis of spontaneous temporal bone encephalocele repaired by the senior authors, and post operative evaluation for IIH with either LP or funduscopic exam. Patient demographics, post operative lumbar puncture opening pressure if available, and funduscopic eye exam results if available were collected.
- Intracranial hypertension was defined as an opening pressure greater than 25 cm H₂O.
- The post operative protocol for evaluation of possible IIH at this practice changed from LP to funduscopic exam over the collection timeframe of this study due to a meeting with the Neurology and Ophthalmology departments, during which they recommended only funduscopic exam post operatively.

Table 1: IIH diagnostic criteria	
1.	Required for diagnosis of pseudotumor cerebri syndrome
	A. Papilledema
	B. Normal neurological examination except cranial nerve abnormalities
	C. Neuroimaging: normal brain parenchyma without evidence of hydrocephalus, mass or structural lesion
	D. Normal CSF composition
	E. Elevated lumbar puncture opening (>250 mm H ₂ O in adults)
2.	Diagnosis of pseudotumor cerebri syndrome without papilledema
	A. If B-E are present with unilateral or bilateral abducens nerve palsy
	B. In the absence of papilledema or sixth nerve palsy the diagnosis can be suggested if B-E above are satisfied and at least 3 of the following are satisfied.
	i. Empty sella
	ii. Flattening of posterior aspect of the globe
	iii. Distension of the perioptic subarachnoid space with or without a tortuous optic nerve
	iv. Transverse venous sinus stenosis

Table 1: Proposed diagnostic criteria of IIH

*A diagnosis of IIH is definite in patients that fulfill criteria A-E in section 1 and considered if criteria A-D are met but the CSF pressure is lower than specified

Results

- 34 patients underwent surgical repair of a lateral skull base encephalocele with cerebrospinal fluid (CSF) leak and then were evaluated for possible idiopathic intracranial hypertension (IIH) with either funduscopic eye exam or lumbar puncture (LP) with opening pressure. 14 patients underwent a post operative lumbar puncture and 20 underwent funduscopic eye exam after surgical encephalocele repair of spontaneous CSF otorrhea. 2 underwent both exams. The majority of patients were female (88%).
- The mean patient age was 59.3 (range, 39-82). All patients were overweight or obese with a mean BMI of 38.5 k/m² (range, 25.1-53.4 kg/m²). 14 patients with lumbar puncture, 64% had elevated opening pressure with the mean opening pressure of 24.5 cm H₂O (range, 10-40 cm H₂O). 20 patients who underwent funduscopic eye exam, 2 (10%) were found to have papilledema. 2 patients with a negative funduscopic eye exam but an elevated opening pressure on lumbar puncture.

Table 2: Patient Demographics						
	Total		Lumbar Puncture		Funduscopic Eye Exam	
Patients (n)	34		14		20	
Age mean	59.3		56.8		61.7	
Age range	39-82		39-71		44-82	
Male	4	11.8%	2	12.5%	2	9.5%
Female	30	88.2%	12	87.5%	18	90.5%
BMI mean	38.5		38.7		38.2	
BMI range	25.1-53.4		25.1-50.6		25.1-53.4	
Elevated Opening Pressure > 20 cm H ₂ O (n, %, mean)			9	64.3%	24.5	cm H ₂ O
Elevated Opening Pressure > 25 cm H ₂ O (n, %, mean)			8	57.1%	28.8	cm H ₂ O
Papilledema (n, %)					2	10%

Table 2: Combined data with demographic means for patients that were evaluated with LP and funduscopic exams following encephalocele repair.

Discussion

- Current diagnostic criteria for IIH includes several criteria including papilledema, abducens palsy, and elevated CSF opening pressure. However, there is no standardized method of post operative diagnosis or treatment of IIH after surgical repair of a spontaneous CSF leak.
- There is a significant difference in the percentage of patients identified to have elevated ICP based on funduscopic exam versus LP.
- Demographics between the two groups were similar, mostly overweight or obese middle-aged females.
- It is unclear whether prolonged or continued elevated intracranial hypertension increases a patient's risk of recurrent or new CSF leak. One patient in this study developed a new leak on the contralateral side repaired just prior to submission of this study. She had a negative funduscopic exam after her initial surgery and was not treated for IIH prior to her second leak.
- Though we did not study recurrence, treating patients medically to lower their intracranial pressure would plausibly lower their risk of recurrence, as well as other comorbidities. We thus raise the question of whether these patients are being underdiagnosed and, therefore, under treated.

Conclusion

- We demonstrate a discrepancy between rates of elevated ICP on LP and papilledema on funduscopic exam in patients with prior surgery for spontaneous CSF leak.
- Further studies are warranted to evaluate possible long-term manifestations of IIH, including possible sequential or recurrent CSF leaks.

References

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