Idiopathic Intracranial Hypertension- The Eyes and Beyond

Idiopathic Intracranial Hypertension (IIH), also known as primary pseudotumor cerebri is an important neurological cause of reversible visual loss. IIH was originally considered a benign syndrome of increased intracranial pressure with unremarkable imaging and cerebrospinal fluid (CSF) composition.[1] Over decades of research, it has been well understood that IIH is associated with characteristic imaging findings and has the potential of causing significant visual morbidity.[2,3] IIH has also been associated with significant extra-optic manifestations which at times may be the sole manifestation and cause of disability of this challenging entity.

Visual Morbidity in IIH

Available literature suggests that up to 96% of patients with IIH report visual abnormalities at some time during their illness. Common clinical symptoms include transient visual obscurations (TVOs) which are intermittent episodes of visual loss lasting for less than 30 seconds. Maneuvers causing a transient increase in intracranial pressure precipitate TVOs. While these may be an important cause of anxiety in patients, they do not have any significant bearing on the visual outcome.[4]

Visual field defects

Visual field deficits are the most common form of persistent visual morbidity and are often characteristic due to the involvement of optic nerve-related localized nerve bundle-like patterns. Enlargement of the physiological blind spot; constriction of the visual fields, loss of inferonasal portions of the visual field; central, paracentral, centro-caecal or arcuate scotomas and rarely altitudinal visual loss may occur. Recognizing specific patterns may provide a vital clue while analyzing the severity and response to treatment.[5,6]

Visual loss

The natural history of visual loss in IIH is highly variable. Most of the patients present with evolving deficits over weeks to months. In few studies, long-term visual morbidity due to visual loss in IIH has been equated to that in optic neuritis. However, many can even present with a delayed worsening of visual symptoms.[7]

Typically, visual acuity is affected late in the course of the disease. Significant long-term visual losses have been seen in 9-24% of patients with IIH. Progressive visual loss in poorly managed and non-compliant patients is fairly common. A small subset of patients with IIH may develop fulminant or malignant visual loss within 7 to 28 days of the onset of their symptoms. This cohort represents the patients with an acute rapid increase in intracranial pressure (ICP) causing significant loss of axoplasmic flow in the optic nerve causing loss of vision. Aggressive medical management and surgical intervention are usually required in this subgroup of patients.[8]

IIH Beyond the Optic Nerve

Apart from the involvement of the optic nerve IIH can present with the involvement of other neurological substrates. These extra-optic manifestations can be the presenting feature in IIH and are important tickers for the clinician in cases of uncertainty.

Spontaneous CSF leak

Spontaneous nontraumatic leaks of the CSF can be a presenting feature of idiopathic intracranial hypertension. Usually intermittent, it may leak into the paranasal sinuses, through the lateral recesses of the sphenoid and the ethmoid bones (rhinorrhoea), or into the temporal bone, where they may present as aural fullness and may even be misdiagnosed as serous otitis media.[9] The phenomenon merely represents the diversion of CSF secondary to increased pressure and usually requires surgical repair and adjunctive drainage procedures to prevent complications.[10]

Tinnitus

Pulsatile tinnitus has been stressed upon to occur in IIH, and both types can occur. Pulse synchronous (pulsatile) tinnitus was seen to occur in about 52% of the cases, bilateral in two-thirds and unilateral in the remaining, whereas nonpulsatile tinnitus was seen to occur in 23% of the cases in the multicentric IIHTT Trial.[11] Smaller trials report varying percentages; however, it is essential to note that the same can be present in many cases other than IIH, like dural arteriovenous fistulas, intracranial atherosclerosis, and glomus tumors, and abnormal mastoid emissary veins, etc.[12] Severity of tinnitus does not correlate with the CSF pressures. Tinnitus is usually managed with CSF drainage and diuretics.[12] Similarly, aural fullness, vertigo, and sensorineural hearing loss affecting all frequency groups can occur in the patients in varying proportions. Long-term and larger studies are awaited to study the evolving relationship between intracranial pressures and hearing loss.[13]

Cranial nerve anomalies

Extraoptic involvement of the cranial nerves has been documented in patients of IIH. Sixth nerve involvement occurs due to the downward displacement of the brainstem pressing on the sixth nerve as it crosses the petrous ridge and enters into the Dorello's canal. It has been seen in upto 12% of adults,[11] and varies widely between 9-59% in the pediatric population. The palsy resolves with the normalization of intracranial pressures.[14] Various other reports of involvement of the seventh cranial nerve and combinations of extraocular muscles are reported but are inconsistent findings.

Cognition and psychiatric anomalies

Patients of IIH have shown a significant reduction in executive functions and memory. Visuospatial functions and processing speed were also significantly affected.[15] The various quality of

life (QoL) measures (Short Form Health Survey-36, National Eye Institute Visual Function Questionnaire 25 item and 10 item questionnaires) used in the IIHTT trials have shown that the quality of life is significantly affected in patients of IIH with even minimal ocular dysfunction and is as comparable as other neurological conditions of the eye.[16]

Systemic side effects

Additionally, patients of IIH experience higher grades of fatigue and systemic side effects of drugs owing to medications like acetazolamide and topiramate.[17] Recent authors have proposed that the patients of IIH irrespective of papilloedema with symptoms of chronic fatigue syndrome may actually represent a forme fruste of the same disorder.[18]

A study of a comparative sample of IIH and healthy patients found involvement of the inferior vestibular nerve and saccular dysfunction in patients of IIH. The researchers suggested the increased intracranial pressure to affect the inner ear in a manner analogous to *hydrops*.[19] The stapedial reflex was a test that was once exhorted as a test to diagnose many neurological conditions. However, the empirical nature of the estimation methods, the low sensitivity, and specificity of these tests have limited their use in present methods. Today, many sophisticated tests have now been developed with far higher sensitivity and specificity to diagnose inner ear pathologies.

The present article attempts to discuss the role of the stapedial reflex in IIH.[20] The series has a very small number of patients with mixed results, which certainly needs to be reproduced in a much larger number to be credible. Additionally, the role of the stapedial reflex in the prediction of the intracranial pressures is not well established, as the same can occur due to many causes of the ear as well.[21] The greater message from such attempts lies in being aware of the various manifestations of IIH which can help the clinician diagnose the condition with confidence.

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