Introduction

Late onset idiopathic aqueductal stenosis (LIAS) is a pathological condition causing a triventricular hydrocephalus following an obstruction of cerebrospinal fluid (CSF) flow without a known origin. The symptomatology of this condition includes headache, gait and cognitive impairment, and urinary incontinence. Magnetic Resonance Imaging (MRI) complemented with cine phase-contrast for flow measurement has improved diagnostic capabilities. Endoscopic third ventriculostomy (ETV) is considered as the golden standard treatment for this condition. Here we present a case of a 22-year-old man with severe urinary incontinence leading to further imaging investigations and eventual diagnosis of obstructive hydrocephaly.

Case description

Age | Key events | Investigations/therapy
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19 y | First symptoms: - Hemicranial headache - Tinnitus | CT scan and MRI: mild enlargement of ventricles - Electrophysiological EEG and brainstem auditory evoked potentials (BAEP) - no abnormalities
21 y | Additional symptoms: - Progressive urinary incontinence - Nocturia - Enuresis - Cognitive impairment | Trial therapy Tamsulosine 0.4mg: no improvement - Increased psychotherapy consultations - Escitalopram - Akathisie
22 y | First Urology consultation - Recent blurred vision | Urodynamic investigations (fig 1) Brain MRI (fig 2) - Spine and cauda equina MRI: normal - Botox injections and Fesoterodine
23 y | Endoscopic third ventriculostomy | After the ventriculostomy, urinary continence completely recovered. Urodynamic testing supported these improvements (fig. 3). Likewise, the headache, tinnitus and blurred vision disappeared rapidly. His concentration improved, and his anxiety diminished. Psychiatric medication dosage was diminished.

Discussion

Late onset idiopathic aqueductal stenosis is defined as the presence of a cerebral spinal fluid flow stenosis combined with symptoms of headache, cognitive and gait impairment, and urinary incontinence (1). The CSF obstruction leads to a triventricular obstructive hydrocephalus leading to these specific symptoms. This symptomatology is similar to that of normal pressure hydrocephaly (NPH) seen in elderly patients. LIAS can thus be put in the spectrum between NPH and acute elevated intracranial pressure. Blurred vision and seizures are also common symptoms in addition to headache and other NPH symptoms (2). MRI has made diagnosing LIAS much easier by allowing visualization of the Sylvian aqueduct and the CSF flow in the canal using cine phase-contrast MRI.

The pathogenesis of this stenosis remains uncertain. The membranous ependymal-like tissue which can be found in the aqueduct suggests prior manifestation of an inflammatory process in this region. However, no basis for this inflammation has been discovered.

Normal micruntion is dependent on the autonomic reflex arc of the brainstem and the spinal cord (3). Afferent signals from the bladder reach the midbrain periaqueductal gray (PAG) which project down to the pontine micruntion center (PMC) (4). The PMC then sends suppressing efferenent signals to the spinheur motor neurons presumably through GABA and glycine interneurons (4).

Voiding function on the other hand appears to be controlled by higher brain structures including the frontal cortex. This effect appears to be inhibitory as previous animal studies have shown that lesions to the frontal cortex lead to exaggerated miction (5).

This correlates with studies demonstrating blood flow hypoperfusion in the right frontal cortex in normal pressure hydrocephalus patients whom also suffer from incontinence (6). These findings confirm that bladder (detrusor) overactivity appears to be the major cause for urinary urgency incontinence in neurological diseases (7).

Gold standard treatment for obstructive hydrocephalus is endoscopic third ventriculostomy (ETV) with good improvement of clinical symptomatology. Long term efficacy has shown to be outstanding with only a few patients receiving an implantation of a ventriculo-peritoneal shunt (8).

Conclusions

The chronic elevated pressure in the ventricles seen in LIAS and the suspected intermittent elevated pressure in NPH could possibly lead to damage to the frontal cortex, its subependymal tracts, and the PAG leading to the urinary incontinence seen in LIAS and NPH.

Elevated pressures in the bladder or neurogenic bladder patterns during urodynamic investigations for urge incontinence should be complemented with further investigations such as brain MRI with cine phase-contrast to exclude LIAS and other obstructive hydrocephaly pathologies.

Bibliography