Idiopathic intracranial hypertension case series

Bsteh et al. included patients from the Vienna-Idiopathic-Intracranial-Hypertension (VIIH) database with Idiopathic intracranial hypertension according to Friedman criteria and cranial MRI performed at diagnosis. The presence of empty sella (ES), perioptic subarachnoid space distension (POSD) with or without optic nerve tortuosity (ONT), posterior globe flattening (PGF) and transverse sinus stenosis (TSS) was assessed and multivariable regression models regarding visual outcome (persistent visual impairment/visual worsening) and headache outcome (headache improvement/freedom of headache) were fitted.

They included 84 IIH patients (88.1% female, mean age 33.5 years, median body mass index 33.7). At baseline, visual impairment was present in 70.2% and headache in 84.5% (54.8% chronic). Persistent visual impairment occurred in 58.3%, visual worsening in 13.1%, headache improvement was achieved in 83.8%, and freedom of headache in 26.2%. At least one MRI feature was found in 78.6% and 60.0% had \geq 3 features with POSD most frequent (64.3%) followed by TSS (60.0%), ONT (46.4%), ES (44.0%), and PGF (23.8%). In multivariable models, there was no association of any single MRI feature or their number with visual impairment, visual worsening, headache improvement, or freedom. Visual impairment at baseline predicted persistent visual impairment (odds ratio 6.3, p<0.001), but not visual worsening. Chronic headache at baseline was significantly associated with a lower likelihood of headache freedom (odds ratio 0.48, p=0.013), but not with headache improvement.

MRI features of IIH are neither prognostic of visual nor headache outcome 1).

A retrospective observational study of patients diagnosed with IIH over a 12-year period at a single centre was completed via database review. Demographic data, symptoms at baseline and last visit, treatments undertaken, and duration of follow-up were included. Visual outcomes, including visual acuity, colour vision, 30-2 Humphrey automated perimetry data, and retinal nerve fibre layer thickness (RNFL), were collected at baseline and last visit.

Results: IIH was diagnosed in 132 patients (90.9 % female) with a median of 2.8 years (range: 0-9.1) follow-up. Mean BMI was 35.9 \pm 7.9 kg/m2. Symptoms at presentation were headache (87.6 %), pulsatile tinnitus (27.2 %) and transient visual obscurations (27.2 %). First-line management was acetazolamide in 86.4 %, with 34.2 % of these patients ceasing treatment because of adverse events. Visual field measures and RNFL at last follow-up improved when compared to baseline (median MD: -1.99 dB (IQR -3.6 to -0.9) to -0.85 (-2.1 to 0.0) (p < 0.001), median RNFL: 132 μ m (IQR 116 - 183) to 103 (92 - 113) (p < 0.001)). Some patients (6.1 %) required surgery for more severe IIH.

Conclusions: Long-term symptomatic and visual prognosis in IIH patients is excellent. However, a subset of patients with more severe disease require surgical intervention. Adverse events of treatment lead to high medication discontinuation rates ²⁾.

A total of 51 patients with IIH were included. Patient demographics, symptoms, imaging data, ophthalmological and clinical findings were collected.

At the time of diagnosis the mean age was 32.5years (SD 10.7), the body mass index was 37.1 kg/m² (SD 7.4), and the opening pressure 29.1 mmHg (SD 6.2). A total of 88.2% of patients were female and 45.1% were diagnosed with a psychiatric co-morbidity prior to IIH diagnosis. The mean follow-up time was 4.4 years (SD 5.4). The overall treatment outcome was significantly poorer on a group of patients with psychiatric diagnosis when compared to individuals without such history (p = 0.001), but there were no differences in the resolution of papilledema (p = 0.405). Patients with IIH and psychiatric disorders had more often empty sella on their imaging at diagnosis when compared to patients without psychiatric co-morbidity (p = 0.044).

Psychiatric disorders are highly prevalent in patients with IIH and associate with worse subjective outcomes. These findings advocate for monitoring the mental health of patients with IIH and warrant further multidisciplinary research to understand the potentially underlying psychosocial and neuroendocrinological mechanisms ³⁾.

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Sankey et al. performed a retrospective review of 31 patients (169 total emergency department visits) who presented to the emergency department for IIH-related symptoms between 2003 and 2015. Demographics, comorbidities, symptoms, IIH diagnosis and treatment history, ophthalmological examination, diagnostic lumbar puncture (LP), imaging findings, and data regarding admission and management decisions were collected. Multivariable general linear models regression analysis was performed to assess the predictive factors associated with admission and shunt revision. RESULTS Thirty-one adult patients with a history of shunt placement for IIH visited the emergency department a total of 169 times for IIH-related symptoms, with a median of 3 visits (interquartile range 2-7 visits) per patient. Five patients had more than 10 emergency department visits. Baseline factors associated with admission included male sex (OR 10.47, 95% CI 2.13-51.56; p = 0.004) and performance of an LP (OR 3.10, 95% CI 1.31-7.31; p = 0.01). Contrastingly, older age at presentation (OR 0.94, 95% CI 0.90-0.99; p = 0.01), and a greater number of prior emergency department visits (OR 0.94, 95% CI 0.89-0.99; p = 0.02) were slightly protective against admission. The presence of papilledema (OR 11.62, 95% CI 3.20-42.16; p < 0.001), Caucasian race (OR 40.53, 95% CI 2.49-660.09 p = 0.009), and systemic hypertension (OR 7.73, 95% CI 1.11-53.62; p = 0.03) were independent risk factors for shunt revision. In addition, a greater number of prior emergency department visits (OR 0.86, 95% CI 0.77-0.96; p = 0.009) and older age at presentation (OR 0.93, 95% CI 0.87-0.99; p = 0.02) were slightly protective against shunt revision, while there was suggestive evidence that presence of a programmable shunt (OR 0.23, 95% CI 0.05-1.14; p = 0.07) was a protective factor against shunt revision. Of note, location of the proximal catheter in the ventricle or lumbar subarachnoid space was not significantly associated with admission or shunt revision in the multivariable analyses.

The decision to admit a shunt-treated patient from the emergency department for symptoms related to IIH is challenging. Knowledge of factors associated with the need for admission and/or shunt revision is required. In this study, factors such as male sex, younger age at presentation, lower number of prior emergency department visits, and performance of a diagnostic LP were independent predictors of admission. In addition, papilledema was strongly predictive of the need for shunt revision, highlighting the importance of an ophthalmological examination for shunt-treated adults with IIH who present to the emergency department ⁴⁾.

Levitt et al., reviewed all cerebral venograms with manometry performed for headache between January 2008 and May 2015. Patient demographics, headache etiology, intracranial pressure (ICP) measurements, and radiographic and manometric results were recorded. CVPG was defined as a difference ≥ 8 mm Hg by venographic manometry. RESULTS One hundred sixty-four venograms were performed in 155 patients. There were no procedural complications. Ninety-six procedures (58.5%) were for patients with IIH. The overall incidence of CVPG was 25.6% (42 of 164 procedures): 35.4% (34 of 96 procedures) in IIH patients and 11.8% (8 of 68 procedures) in non-IIH patients. Sixty procedures (36.6%) were performed in patients with preexisting shunts. Seventy-seven patients (49.7%) had procedures preceded by an ICP measurement within 4 weeks of venography, and in 66 (85.7%) of these patients, the ICP had been found to be elevated. CVPG was seen in 8.3% (n = 5) of the procedures in the 60 patients with a preexisting shunt and in 0% (n = 0) of the 11 procedures in the 77 patients with normal ICP (p < 0.001 for both). Noninvasive imaging (MR venography, CT venography) was assessed prior to venography in 112 (68.3%) of 164 cases, and dural venous sinus abnormalities were demonstrated in 73 (65.2%) of these cases; there was a trend toward CVPG (p = 0.07). Multivariate analysis demonstrated an increased likelihood of CVPG in patients with IIH (OR 4.97, 95% CI 1.71-14.47) and a decreased likelihood in patients with a preexisting shunt (OR 0.09, 95% CI 0.02-0.44).

CVPG is uncommon in IIH patients, rare in those with preexisting shunts, and absent in those with normal ICP ⁵⁾.

1)

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