

supportive therapies to improve quality of life. The understanding of genetic variants and associated pathology may provide a revenue to disease modifying therapies.

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AN AUSTRALIAN CASE OF CASPR2 POSITIVE, MORVAN SYNDROME

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Introduction Morvan syndrome is a rare neurological disorder consisting of a combination of neuromyotonia with cognitive symptoms and dysautonomia or insomnia. It is associated with antibodies against the voltage-gated potassium channel (VGKC) associated proteins – most commonly, against CASPR2.

Case A 74-year-old male presented with six-months of worsening psychobehavioural changes, insomnia, amnesia, and neuromyotonia. At presentation, he complained of visual hallucinations and unsteadiness of gait. He subsequently developed urinary retention, labile blood pressure and atrial fibrillation. Neurologic examination revealed disorientation to time and place, dysarthria, diffuse hyperreflexia, and multifocal fasciculations.

Brain and whole cord MRI with contrast, was unremarkable. EEG showed moderate encephalopathy. CSF findings were unremarkable. Paraneoplastic antibodies and infectious and vasculitis screens were normal; malignancy was not found despite extensive investigation.

In serum, CASPR2-antibody was strongly positive, titre of 535pM, and also weakly positive for LGI1. Nerve conduction studies and electromyography showed post-CMAP after-discharges, and myokymia respectively. Overall, the study was suggestive of peripheral nerve hyper-excitability.

He was induced with intravenous immunoglobulins and pulsed with, and maintained, on steroids. He had excellent clinical response – his modified Rankin score improved from 4 to 1.

Conclusion Antibodies against the VGKC-complex proteins, most commonly CASPR-2, are associated with a wide spectrum of clinical diseases, often associated with an underlying malignancy. Morvan syndrome is one such rare and treatable manifestation. Awareness of this constellation of symptoms and consideration of prolonged clinical course are crucial in early diagnosis and prompt immunotherapy, or tumour therapy. Improvement can be pronounced, though relapse may occur.

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DOES SURGICAL CORRECTION OF REFRACTIVE ERROR ALLEVIATE HEADACHE IN PATIENTS WITH KERATOCONUS? – A RETROSPECTIVE ANALYSIS

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Objectives Refractive error has long been thought to be a cause for headache, however, historical research on this topic has major methodical limitations.

Often, patients with different headache types were examined together, making any direct conclusion between refractive error and headache difficult.

We investigate if surgical correction of refractive error with Topography Guided Photorefractive Keratectomy (TGPRK) and Collagen Cross Linking (CXL) alleviates headache in patients with keratoconus in an appropriately classified patient population, guided by the International Classification of Headache Disorders (ICHD-3).

Methods 40 patients who had keratoconus and required TGPRK and CXL met inclusion criteria.

Patients who met diagnostic criteria for headache as defined by the ICHD-3 were asked about the nature of their headaches and impact on quality of life by means of the Head Impact Score (HIT-6) questionnaire, both pre-operatively and post-operatively.

Results 24 of 40 patients reported headache pre-operatively.

Post-operatively, only 9 patients had diagnosable headaches ($p < 0.05$).

The mean number of headache days per week decreased from 4.38 ± 2.37 days/week to 0.46 ± 0.72 days/week ($p < 0.05$).

The mean duration of headache decreased from 10.8 ± 100.7 to 34.4 ± 63.5 minutes ($p < 0.05$).

The consumption of analgesia decreased from 2.42 ± 2.34 days/week to 0.56 ± 1.16 days/week ($p < 0.05$).

Post-operatively, HIT-6 scores decreased significantly. 61% of patients stopped analgesia altogether.

Conclusion Surgical correction of refractive error in patients with keratoconus can alleviate headache in a large proportion of cases, significantly improving quality of life.

It may be that the surgical treatment of keratoconus should be considered in patients as part of their headache management.

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META-ANALYSIS OF THE HEAD IMPULSE TEST (HIT) AND HEAD IMPULSE TEST, NYSTAGMUS, TEST OF SKEW (HINT) IN THE DIAGNOSIS OF STROKE AND PERIPHERAL VERTIGO

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Objectives We conducted a meta-analysis of the HIT and HINT tests to diagnose peripheral vertigo (PV) and central vertigo in the emergency department (ED).

Methods Pubmed, Google Scholar, EmBase and articles references published in English up to July 2021 were searched for keywords 'vertigo' or 'acute vestibular syndrome' or 'dizziness' and 'head impulse' and 'stroke'. Bivariate method for meta-analysis was used.

Results 11 studies HIT (8 studies, N = 417) and HINT (6 studies, N = 405). HIT and HINT were performed within 24 hours in 4 of 11 studies. Positive likelihood ratio (PLR) and negative likelihood ratio (NLR) for HIT in PV was 4.85 (95% CI 2.83 – 8.08) and 0.19 (95% CI 0.12 – 0.29, I²63.25%) respectively. The (area under the curve) AUC for HIT the diagnosis of PV and stroke was 0.90 and 0.92 respectively.

PLR and NLR for a negative HIT in stroke was 5.85 (95% CI 3.07 – 10.6) and 0.17 (95% CI 0.08 – 0.30) respectively. PLR and NLR for peripheral HINT pattern for PV was 17.3 (95% CI 8.38 – 32.1) and 0.15 (95% CI 0.07 – 0.26) respectively. PLR and NLR for central HINT pattern for stroke was 5.61 (95% CI 4.19 – 7.7) and 0.06 (95% CI 0.03 – 0.12) respectively.

Conclusion The HIT and HINT exams appear moderately good discriminators of central and peripheral vertigo. However, these results may not apply in the ED setting as most papers evaluated these tests beyond 24 hours.

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DYSTONIC STRIDOR AND LARYNGEAL MYOCLONUS IN A MEDULLOBLASTOMA SURVIVOR PRECIPITATED BY REDUCTION IN INTRACRANIAL PRESSURE

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Background Whilst stridor related to bilateral vocal cord paralysis (BVCP) is a recognised complication of increased intracranial pressure, it typically resolves with ventricular-peritoneal (VP) shunting. We present a case where tracheostomy was needed after successful VP shunting.

Case A 33-year-old-man presented with headache, slurred speech and immobility. His background included resected medulloblastoma with head-neck radiotherapy (aged 8), long-term VP shunt, and post-treatment ataxia which had progressed over 2 years with development of intermittent nocturnal stridor. He underwent emergent shunt revision for a distally blocked shunt, with resolution of hydrocephalus. Two days afterwards, he developed hiccups and worsened stridor which progressed to respiratory obstruction over 24 hours. Laryngoscopy showed tightly adducted midline vocal folds with coarse pharyngeal, palatal, and tongue myorhythmia (3 Hz). There was no response to benzodiazepines and no epileptiform activity on EEG. He had loss of gag reflex on neurological examination with normal eye movements. A cerebral and neck MRI showed resolution of hydrocephalus and post-treatment changes related to previous medulloblastoma. There has unfortunately been no improvement in vocal fold movement over 3 weeks.

Discussion Subacute stridor may be due to pathological upper motor neurone activation of the branchial motor component of the vagal nerves with this person's background brain injury.¹ Tight acute midline cord positioning and myoclonus may be unusually caused by bilateral recurrent laryngeal nerve lesions secondary to changes in intracranial pressure,^{2,3} but is not the favoured sole mechanism here. Botulinum toxin therapy could provide benefit but may compromise a future safe swallow.

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DIAGNOSTIC CHALLENGES ASSOCIATED WITH GRANULOMATOUS DISEASES: AN ILLUSTRATIVE CASE REPORT

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Case Report A 29-year-old female presented with a 4-month history of right-sided hemifacial sensory disturbance. Examination revealed reduced pain and temperature sensation in the right V1, V2 and V3 regions and a diminished right corneal reflex. Past history included intermittent herpes labialis and travel to India twice in the prior 5 years.

Enhanced brain and skull-base magnetic resonance imaging (MRI) revealed enhancement at the cisternal segment of the right trigeminal nerve and right infraorbital nerve T2-hyperintensity. Diffuse extra-axial enhancement was noted in right Meckel's cave, extending into the posterolateral cavernous sinus. Positron emission tomography (PET) imaging demonstrated markedly increased metabolism in the right Meckel's cave and metabolism in the hilar and mediastinal lymph nodes consistent with sarcoidosis. Lymph node biopsies were inconclusive but revealed a possible granuloma. Right trigeminal nerve biopsy was subsequently performed. Histopathology revealed non-caseating granulomas with few acid-fast bacilli present. Primary neuritic leprosy was diagnosed and a 12-month treatment course of rifampicin, dapsone and clofazimine was commenced.

Repeat MRI 3-months post-treatment demonstrated reduced enhancement in the right cavernous sinus. Clinically her symptoms improved with antibacterial and steroid therapy. Repeat PET imaging revealed resolution of the Meckel's cave changes but increased metabolism in thoracic and abdominal lymph nodes, raising the possibility of an alternative diagnosis (i.e. sarcoidosis) or dual pathology. Pregnancy halted further diagnostic work up.

This case highlights the complexities associated with diagnosing granulomatous disease even with access to multimodal imaging and biopsy sites, and the need to revisit the diagnosis if there are atypical features.

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THE YIELD OF EEG AND OTHER NEUROLOGICAL INVESTIGATIONS IN PATIENTS WITH NEW ONSET OF PSYCHIATRIC SYMPTOMS

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Objectives To establish the diagnostic yield of electroencephalogram (EEG) and other investigations for evaluation of limbic encephalitis in patients with new onset psychiatric symptoms.

Methods A retrospective audit was conducted of all EEGs performed between July 2020 to August 2021 for workup of new-onset psychiatric symptoms at a tertiary neurology referral centre. Data was obtained from electronic medical records including patient history, neurological examination findings, results of investigations including EEG, brain magnetic