

Background Autoimmune encephalitis is an increasingly recognised disease that presents with seizures, neuropsychiatric symptoms, dystonic movements, and autonomic dysfunction¹. As the mainstay of treatment immunosuppressive therapies are most effective when subject to early initiation and timely escalation, both of which are recognised to affect outcomes². Nevertheless approximately half of patients with NMDA-R antibody encephalitis do not respond adequately to first-line therapy, and a significant proportion (12-30%) relapse².

Cases A 19 year old lady presented with new-onset seizures and psychosis. EEG showed focal spike-and-wave discharges and MRI brain a focal area of restricted diffusion consistent with recent seizure activity. NMDA-R antibodies were present in both CSF and serum. Following early treatment with corticosteroid, plasma exchange, IVIG and rituximab the patient recovered, returning to college after 6 months.

A 50 year old gentleman presented with a two day history of myalgias and confusion. EEG showed spike-and-wave discharges and MRI brain increased T2 signal in the mesial temporal lobes. NMDA-R antibodies were present in both CSF and serum. He was treated with corticosteroid, plasma exchange, IVIG and rituximab, and continued on oral prednisone and mycophenolate. Response to treatment was poor with persistent ongoing physical and cognitive impairment at 6 months. Serial MRI showed substantial (~30%) loss of parenchymal brain volume.

Discussion These cases illustrate that timely and aggressive management of NMDA-R antibody encephalitis with favourable prognostic markers is no guarantee of recovery. Several novel clinical and immunological predictors of response to therapy have been postulated, and currently await broader validation.³

REFERENCES

1. Dalmau J, Graus F. Antibody-mediated encephalitis. *N Engl J Med* 2018 Mar 1; **378**(9):840–851.
2. Titulaer MJ, McCracken L, Gabilondo I, Armangué T, Glaser C, Iizuka T, et al. Treatment and prognostic factors for long-term outcome in patients with anti-NMDA receptor encephalitis: an observational cohort study. *Lancet Neurol* 2013; **12**(2):157–65.
3. Rüegg S, Yeh EA, Honnorat J. Forecasting outcomes in anti-NMDAR encephalitis: clearer prognostic markers needed. *Neurology* 2019 Jan 15; **92**(3):119–120.

108

THE DIAGNOSIS AND MANAGEMENT OF FIVE CASES OF SPINAL NEUROSARCOIDOSIS

¹Ariadna Fontes-Villalba, ²Dayna Griffiths, ³Natasha Gerbis, ¹Jonathan Baird-Gunning, ⁴Patrick Aouad, ⁵Suran Fernando, ¹John Parratt. ¹Neurology, Royal North Shore Hospital, Sydney, NSW, Australia; ²Neurology, Neurology Dept. Gosford Hospital, Gosford, NSW, Australia; ³Neurology, Northern Beaches Hospital, Frenchs Forest, NSW, Australia; ⁴Department of Neurology, Liverpool Hospital, Sydney, NSW, Australia; ⁵Department of Immunology, Royal North Shore Hospital, Sydney, NSW, Australia

10.1136/bmjno-2021-ANZAN.108

Objectives Neurosarcoidosis may present with longitudinally extensive transverse myelitis (LETM) posing a diagnostic challenge. We describe the clinical features, radiology and management of five patients with spinal neurosarcoidosis (SNS).

Methods We retrospectively identified five patients with a diagnosis of SNS and the clinical, radiological and pathological data were reviewed.

Results There were three females and two males who were, on average, 49 years at onset. A histopathological diagnosis of sarcoid was confirmed in two cases (lymph node and cerebral lesion) and the diagnosis was radiological in the others

(LETM with persistent enhancement in 3 patients and the trident sign in 2 patients). The average span of the spinal cord lesions was 4 vertebral bodies. Three patients had FDG-avid mediastinal lymph nodes. Cerebral disease was identified in two cases, and cardiac involvement in one. Two patients required spinal decompression surgery. All patients received intravenous and oral steroids and some had rituximab (n=2), tocilizumab (n=1), cyclophosphamide (n=1), adalimumab (n=2), and infliximab (n=4). Disease control was achieved with TNF-alpha (tumour necrosis factor-alpha) blocking in 4 cases and another responded to cyclophosphamide. The mean follow-up was 55 months.

Conclusions SNS is a cause of LETM and can be suspected by the trident sign on MRI. Persistent enhancement may be another differentiating feature. Spinal cord oedema requiring surgery may occur and patients typically respond to treatment with TNF-alpha inhibitors.

REFERENCES

1. Flanagan EP, Kaufmann TJ, Krecke KN, Aksamit AJ, Pittock SJ, Keegan BM, Gianini C, Weinschenker BM. Discriminating long myelitis of neuromyelitis optica from sarcoidosis. *Ann Neurol* 2016 Mar; **79**(3):437–47.
2. Zalewski NL, MD, Krecke KN, Weinschenker BG, MD, Aksamit AJ, Conway BL, McKeon A, Flanagan EP. Central canal enhancement and the trident sign in spinal cord sarcoidosis. *Neurology* 2016 Aug 16; **87**(7):743–4.

109

PAIR-WISE DIFFERENCES OF PENUMBRA AND CORE VOLUME ESTIMATES FROM THREE COMPUTED TOMOGRAPHY PERFUSION SOFTWARE PACKAGES ARE INFLUENCED BY SITE OF LARGE VESSEL OCCLUSION

^{1,2}Peter SW Park, ¹Robbie Chan, ¹Channa Senanayake, ¹Stanley MK Tsui, ²Alun Pope, ^{1,2}Helen M Dewey, ^{1,2}Philip MC Choi. ¹Department of Neurosciences, Box Hill Hospital, Eastern Health, Box Hill, VIC, Australia; ²Eastern Health Clinical School, Faculty of Medicine, Nursing and Health Sciences, Box Hill, VIC, Australia

10.1136/bmjno-2021-ANZAN.109

Objectives Computed tomography perfusion (CTP) data are important for hyperacute stroke decision making. Comparisons between outputs of different CTP software packages are limited. We aimed to assess the pair-wise differences in infarct and penumbra estimates produced by three CTP software packages – MIStar, RAPID, and Vitrea.

Methods Consecutive patients with suspected acute ischaemic stroke who underwent CTP between July 2020 and June 2021 at our hospital were independently reviewed by two expert readers. Pair-wise differences between software estimates of penumbra and core volumes were calculated for each patient, with analysis stratified by large vessel occlusion (LVO) status (no-LVO, proximal M2, M1 and internal carotid artery-T [ICA-T]).

Results 580 CTP studies were performed; 262 were normal, 146 technically poor, with 172 included in the final analysis. 79/172 (45.9%) had LVO; proximal M2 (n=21), M1 (n=38) and ICA-T (n=20). Overall, statistically significant pair-wise differences were seen for both penumbra and core estimates (P <0.001). The largest difference in mean core estimates were seen between Vitrea and MIStar ([mean, 95% confidence interval] no-LVO [5.8ml, 3.2–8.4]; proximal M2 [10.4ml, 3.9–17.0]; M1 [17.7ml, 8.9–26.6]; ICA-T [38.9ml, 20.2–57.7]). More comparable penumbra estimates were observed between RAPID and MIStar (no-LVO [1.79ml, -3.9–7.51]; proximal M2 [13.1ml, -0.24–26.5]; M1 [10.7ml, -5.9–27.3]; ICA-T [28.4ml, 0.78–56.0]).

Conclusion Core and penumbra volume estimates vary significantly between CTP software packages. There are minimal differences in patients with non-LVO stroke, with the greatest differences seen in patients with ICA-T occlusions.

110

BOTULINIUM TOXIN FOR A REFRACTORY HEAD TREMOR ARISING FROM CEREBELLAR METASTASES

Christopher Blair, Leon Edwards, Dennis Cordato. *Department of Neurology and Neurophysiology, Liverpool Hospital, Liverpool, NSW, Australia*

10.1136/bmjno-2021-ANZAN.110

Background Tremor is an involuntary, rhythmic, oscillatory movement of a body part that can be a clinical manifestation of a range of underlying pathologies.¹ Of those tremor subtypes for which adequate management is often elusive, head tremor is among the most debilitating. Taking ‘yes-yes’, ‘no-no’, and mixed forms, available treatments for head tremor include medication, surgery, and botulinum toxin injections.² We report a case of severe head tremor arising from focal cerebellar metastases that showed a durable response to botulinum toxin treatments in a palliative setting, despite underlying disease progression.

Case A 60 year old lady was referred to our clinic with a 3 month history of ‘no-no’ head tremor. Originally diagnosed with ER breast cancer in 2008, she underwent surgical resection but suffered a disease recurrence in 2017 when she presented with a solitary posterior fossa metastasis. This was resected and adjuvant radiotherapy was given, however in mid-2019 she developed first a left arm and then a coarse, persistent head tremor that severely limited her daily life. MRI brain revealed several new vermian and left cerebellar metastatic deposits. A combination of botulinum toxin injections to splenius capitis/sternomastoid and regular oral gabapentin effectively ameliorated her symptoms over three sessions.

Discussion Two open label studies and one RCT have shown that individualised botulinum toxin injections can be used to effectively treat essential head tremor², and we demonstrate here that such an approach may also be a useful in the management of head tremor due to rarer and more aggressive aetiologies.

REFERENCES

1. Louis ED. Diagnosis and management of tremor. *Continuum (Minneapolis)* 2016 Aug;22(4 Movement Disorders):1143–58.
2. Mittal SO, Lenka A, Jankovic J. Botulinum toxin for the treatment of tremor. *Parkinsonism Relat Disord* 2019 Jun;63:31–41.

111

DELAYED NEUROLOGICAL WORSENING IN AN IMMUNOCOMPETENT ADULT WITH *CRYPTOCOCCUS GATTII* MENINGOENCEPHALITIS

Kristen Lefever, Joel Corbett, Nabeel Sheikh, Helen Brown. *Queensland Health, Woolloongabba, QLD, Australia*

10.1136/bmjno-2021-ANZAN.111

Objective While typically considered a condition of immunocompromised patients, *Cryptococcus gattii* meningoencephalitis is increasingly observed in immunocompetent individuals, where the clinical outcomes are generally worse.^{1 2}

Methods Case report.

Results 24-year-old male presented with a three-week history of progressively worsening headache, lethargy, generalised weakness, binocular diplopia, hearing loss and unintentional weight loss. Two weeks prior, he had presented with coryzal symptoms and received outpatient treatment for community-acquired pneumonia. A lumbar puncture was performed with an opening pressure greater than 34cmH₂O, pleocytosis and positive India ink stain. *Cryptococcus gattii* was cultured at a titre of 1:2048. MRI brain demonstrated bilateral basal ganglia change and leptomeningeal enhancement consistent with Cryptococcal meningitis. Serum HIV was negative. Induction treatment with ambisome-flucytosine was initiated. Lumbar drain and subsequent VP shunt were required for management of persistent symptomatic increased intracranial pressure. After 6 weeks of therapy he was transitioned to consolidation fluconazole. Repeat CSF demonstrated improved Cryptococcal Ag titre of 1:512.

Two months into rehabilitation he suffered a seizure and rapid progressive neurological decline. EEG demonstrated a moderately severe diffuse encephalopathy. Repeat CSF cryptococcal Ag was stable. CSF limbic encephalitis and NMDA antibodies were negative. Repeat MRI brain demonstrated worsening supratentorial leptomeningeal enhancement and parenchymal vasogenic oedema, consistent with paradoxical upgrading reaction (PUR). Prednisolone 1mg/kg was initiated and the patient improved in days.

Conclusions PUR is an immune-reconstitution like event that can occur in immunocompetent patients. It represents an important cause of neurological deterioration in *Cryptococcus gattii* meningoencephalitis, requiring differentiation from relapse on consolidation therapy.

REFERENCES

1. Franco-Paredes, et al. Management of cryptococcus gattii meningoencephalitis. *Lancet Infect Dis* 2014;15(3):348–355.
2. Chen, et al. Cryptococcus gattii infections. *Clin Microbiol Rev* 2014;27(4):980–1024.

112

FAVOURABLE OUTCOME FOLLOWING EARLY TREATMENT WITH RITUXIMAB IN A PATIENT WITH PROBABLE SUSAC'S SYNDROME

Yangyang (Erin) Xiao, Elham Khalilidehkordi, Po Sheng Yang, Andrew Wong, Claire Muller. *Neurology, Royal Brisbane and Women's Hospital, Brisbane, QLD, Australia*

10.1136/bmjno-2021-ANZAN.112

Objective We report a favourable outcome following early treatment with rituximab in a patient with probable Susac's syndrome (SuS).

Background Delayed treatment of SuS leads to significant morbidity, however there is no consensus in its management.

Results A 34-year-old man presented with severe headache, subacute confusion and blurred vision developing over 4 months. The MRI brain revealed multiple supratentorial and infratentorial FLAIR/T2 hyperintense lesions in white and gray matter, including characteristic corpus callosum ‘snow ball’ lesions. The fundus fluorescein angiography (FFA) showed typical branch retinal artery occlusion, consistent with his bilateral decrease in visual fields. CSF showed high protein (3000mg/L) and pleocytosis (18X10⁶/L). Following diagnosis of probable SuS, he was treated with high-dose corticosteroids on day 3 of presentation, followed by IVIG, mycophenolate and rituximab. He had significant