Method The clinical and investigation findings in a series of adult patients with SMART syndrome presenting primarily with seizures were reviewed.
Results Four patients with SMART syndrome presenting with seizures were identified (mean age 51 years). Mean time from radiation therapy to SMART syndrome was 22.5 years (range 15-32 years). Indications for radiation were primary brain tumour (three patients) and haematological malignancy (one patient). Two patients had a history of seizures prior to SMART syndrome. Three patients had headaches at presentation. All patients presented with focal impaired awareness seizures with motor features. One patient had refractory non-convulsive status epilepticus requiring intravenous anaesthesia. Three patients had persistent negative motor deficits, associated with ongoing electrographic seizures with no clinical correlate, only identified on repeated EEGs or continuous EEG (cEEG). All patients failed initial anti-seizure medications (ASM), requiring a mean of five ASMs for seizure control. All patients had enhancing cortical MRI changes consistent with SMART syndrome that corresponded to the clinical deficit and ictal changes on EEG. At follow-up all patients improved but had persistent neurological deficits.
Conclusion SMART syndrome presents with seizures and less frequently status epilepticus and may be the basis for the associated clinical features and radiologic abnormalities. Judicious use of EEG and where necessary CEEG to identify non-convulsive seizures should be considered in patients with SMART syndrome presenting with prolonged neurological deficits.

099 OPTIC PERINEURITIS IN GIANT CELL ARTERITIS

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Background Optic perineuritis (OPN) is a rare orbital inflammatory disease which targets the optic nerve sheath. Although OPN is predominantly idiopathic, it can be secondary to an array of inflammatory, infective or malignant conditions, including giant cell arteritis (GCA).
Methods/Results We describe a case of a 75-year-old man with a four-week history of headaches with associated periorbital swelling, and monocular decrease visual acuity without significant constitutional or systemic symptoms. This was in the context of initially normal ESR and CRP. MRI head demonstrated bilateral OPN and GCA was subsequently confirmed based on temporal artery biopsy. He was managed with high dose prednisolone and upadacitinib.
Conclusions This case highlights the perineuritis as a rare manifestation of GCA.

100 DEVELOPING A QUALITY ASSURANCE FRAMEWORK FOR NEURO-OPHTHALMOLOGY REFERRALS USING NODE – THE NEURO-OPHTHALMOLOGY DATABASE

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Objective Quality assurance (QA) in neuro-ophthalmology (NOPH) is often lacking. The QA registry, NODE (Neuro-ophthalmology Database), was established and implemented in tertiary NOPH clinics in Australia. We developed a consensus on triage categories according to Australian standardised triage categories;P1 (consult<=30 days), P2 (consult<=30-60 days) and P3 (consult>60 days).
Methods Data on 410 patients at Alfred Hospital, Melbourne was collected with NODE. We developed a consensus on assignation of NOPH conditions to triage categories using recommendations from a panel of neuro-ophthalmologists with the modified Delphi approach. The average days from referral to triage and triage to the initial consultation were compared to the developed triage category standard.
Results Most patients presenting to the service were female (n=262, 64%), aged 21 to 30 years. Common diagnoses were Idiopathic Intracranial Hypertension, IIH (24%), Optic Neuropathy, ON (17%), Headaches, (11%) Cranial Nerve Defects, CND (9%) and Eye Movement Disorders, EOMD (9%). The mean time from referral to triage was <2 days for all the common NOPH conditions. The mean time (days, +-standard deviation) from P1 category triage to initial consult for IIH was 26 (±7), ON 27 (±11), and CND was 17 (± 5). The mean time (days) from P2 triage to initial consultant for Headaches was 27 (±12), and EOMD was (±17). The mean time (days) from P3 triage to initial consultant for Myasthenia Gravis was 30 (±10).
Conclusion We have established a consensus agreement on triage categories for neuro-ophthalmological conditions. We established a QA framework for other NOPH clinics in Australia.

101 EMBOUCHURE DYSTONIA COMPLICATING A CASE OF FOCAL CEREBRAL VASCULITIS

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Background Primary central nervous system vasculitis (PCNSV) is a rare disorder that normally develops with bilateral brain lesions. We report a case of pathologically confirmed hemispheric PCNSV with associated brain atrophy and symptoms of focal dystonia.
Method Case report.
Result A 45-year-old female presented with fluctuating apraxia, dystonia, cognitive dysfunction and numbness of the face. She had embouchure dystonia. She was taking lamotrigine for a longstanding history of well controlled partial seizures. A cerebral MRI revealed multiple left hemispheric cortical and subcortical white matter lesions. Many of the lesions enhanced. The lesions came and went over a period of three years but cortical atrophy of the fronto-parietal gyri developed. The CSF demonstrated matched oligoclonal bands. The autoimmune serology was negative. A cerebral PET demonstrated hypo-metabolism in the affected areas.A cortical lesion that was enhancing was biopsied and the histology showed several small foci of punctuate necrosis (infarcts) with lymphohistiocytic infiltration of blood vessels consistent with small vessel vasculitis. The patient was treated with prednisone and mycophenolate and improved clinically, albeit her clarinet playing