improvement within 1 week. By 3 months, he returned to his neuropsychological baseline in majority of cognitive domains from moderate-severe dysfunction, with concurrent MRI demonstrating resolving white matter lesions and FFA showing less evident vasculitis. The treatment response was maintained with tapering of steroids (25mg at 12 months). He was able to return to his previous occupation as a paramedic by 1 year.

**Conclusion** SuS is a rare, immune-mediated microangiopathy in which early recognition with aggressive immunosuppression is required to achieve optimal outcome. No randomized controlled trial (RCT) exists for the management of this condition. In this report, early recognition through multidisciplinary input and aggressive immunotherapy with rituximab resulted in a favourable outcome. However, RCT evidence is needed to guide management.

**Objective** To report clinical stabilisation and improved sum- mated compound motor action potentials (CMAP) in a patient with Hereditary sensory and autonomic neuropathy type I (HSAN-1) following high dose serine therapy. Case A 52-year-old male presented in 2006 with a typical HSAN-1c phenotype and over the ensuing years had progressive distal to proximal sensory disturbance, associated lancinating pains, and mild progressive distal predominant limb weakness. A Ser384Phe mutation in the SPTLC2 gene located on chromosome 14q24 was identified in this patient in 2017. In late 2018 high dose serine therapy (11 grams TDS) was commenced, resulting in stabilisation of clinical weakness.

**Conclusions** High dose Serine replacement therapy may lead to clinical stabilisation and improved neurophysiolog- ical parameters in HSAN-1. HSAN-1, an autosomal dominant sen- soric neuropathy occurs secondary to mutations in the enzyme Serine-Palmitoyltransferase (SPT), an essential enzyme in the de-novo synthesis of sphingolipids. The administration of high dose Serine may overcome altered SPT substrate specificity in HSAN-1, which preferentially uses L-alanine and L-glycine instead of L-serine and allow the formation of typical 1-deox- ysphingolipids as opposed to atypical 1-deoxysphingolipids generally seen in this condition, with early treatment possibly preventing clinical progression.

**Objective** To present a case of painful brachial diplegia follow- ing cervical decompressive surgery. Case A 73 year-old male presented with a 6 month history tripping over his left leg, resulting in near falls. An MRI demon- strated severe spondylotic cervical canal stenosis at C4/5 with myelomalacia and he subsequently underwent a cervical decompression and fusion at this level. On post-operative day five he developed severe pain in his neck and shoulders and mild weakness of his left arm. One week later he underwent a second operation with decompression and rhizolysis at C5 to C8. On post-operative day 3 he awoke with severe pain similar to previously, followed a day later by profound weak- ness of all movements around the shoulders bilaterally, but movements around the elbows, wrists and fingers were nor- mal. While walking his arms hung limply beside him, giving him the appearance of a man in a barrel. Routine nerve con- duction studies and medium somatosensory evoked potentials were normal. Electromyography confirmed denervation within the C5 myotome bilaterally, however with selective sparing of the rhomboids bilaterally. This suggested a lesion distal to the branch to rhomboids, and a diagnosis of bilateral post-oper- ative brachial neuritis was made.

**Conclusion** Acute proximal arm weakness is an uncommon complication of cervical surgeries, referred to commonly as a post-operative C5 palsy. Investigations performed in this case however suggest that a brachial plexus lesion may be the cause of this peculiar syndrome. This syndrome may lie on the spectrum of post-surgical inflammatory neuropathies.

**Objective** Vitamin B12 is crucial for neurologic function, red blood cell production, and DNA synthesis. Deficiency can lead to a wide spectrum of haematologic and neuropsychiatric dis- orders including subacute combined degeneration of the cord. This report presents a case of a 47 y.o female who presented with subacute combined degeneration of the spinal cord with a normal active B12 level.

**Results** MRI demonstrated high T2 signal throughout the dor- sal columns of the cervical and upper thoracic cord without enhancement consistent with subacute combined degeneration of the cord. Her blood count revealed a mild macrocytic anaemia. Total vitamin B12 was <80 pmol/L and active B12 was >128 pmol/L, confirmed on repeat testing. Functional vitamin B12 deficiency was confirmed by an elevated homocysteine level of 38.6 umol/L (reference range 4.4 to 13.6 umol/L) and elevated serum methylmalonic acid of 20.75
umol/L (reference range <0.32 umol/L). Intrinsic factor and gastric parietal cell antibodies were detected.

**Conclusion** Our patient had a clinical presentation consistent with B12 deficiency with an erroneously high active B12 level. Functional assays confirmed B12 deficiency, and a serological diagnosis of pernicious anaemia was made. This case illustrates the importance of not relying on any single test to exclude B12 deficiency.

**REFERENCES**


**PARANEOPlastic PROGRESSIVE-SupRANuCLEAR PALSY LIKE BRAInSTEM SYNDROME ASSOCIATED With LUNG ADENOCARCINOMA**

1. Natasha Gerbis, 2,3Patrick Aouad, 2,3Suran Fernando, 2,1John DE Parratt, 1Northern Beaches Hospital, Frenchs Forest, NSW, Australia; 2Royal North Shore Hospital, St Leonards, NSW, Australia; 3University of Sydney, Sydney, NSW, Australia; 4Liverpool Hospital, Sydney, NSW, Australia

**Objectives** Progressive supranuclear palsy (PSP) is a neurodegenerative condition characterised by Parkinsonian features, cervical dystonia and ophthalmoparesis. Paraneoplastic PSP has previously been reported in the literature in association with several different cancer types but is very rare.1–3

**Methods** Case review.

**Results** A 74 year old Chinese man was diagnosed with Stage 1b EGFR positive lung adenocarcinoma and underwent a left upper lobectomy. Twelve months later he presented with rapidly progressive neck stiffness, reduction in motor function and gait (over 8 weeks) and recalcitrant disquilibrium.

He had hypomimia, frontalis over-activation, blepharo- spasms, blepharotremor and a supranuclear gaze palsy. There was marked axial rigidity, bradykinesia and cervical dystonia. An MRI brain and spine were unremarkable. Vestibular function tests were normal. Serum antineuronal antibodies were negative. The CSF analysis was unremarkable.

The patient responded to plasma exchange on a two to three-weekly basis with significant improvement in saccadic eye movements and Parkinsonism. However, disquilibrium remained a persistent problem despite the discovery and excision of a second EGFR wild type non-small cell lung cancer, and Rituximab was recently started. Cervical dystonia was treated partially with Botulinum toxin injections, but the patient responded poorly to L-dopa.

**Conclusions** This suspected paraneoplastic disease exhibits several features of PSP. In particular, the supranuclear palsy, Parkinsonism and dystonia are similar to the typical syndrome. However, the rapidly progressive presentation and disquilibrium are unusual and a response to plasma exchange, suggests humorally mediated neuronal pathology. In rapidly evolving PSP-like cases with cancer, investigation for immunopathology is warranted.