Japanese encephalitis (JE) typically presents as fever, headache and mental state change due to meningoencephalitis, but more rarely can also cause a polio-like illness without encephalitis. We report a case of an Australian expatriate who presented with acute flaccid paralysis due to JE Virus (JEV), in which the atypical presentation contributed to a significant delay to diagnosis.

**Initial Presentation**

A 57-year-old Caucasian man living northern rural Thailand became unwell in mid-May 2013. After a week of lethargy, he developed mild diarrhoea, fevers, headache, diplopia, vomiting and progressive dyspnoea. He was admitted to hospital 3 days later after he collapsed at a train station.

Over the next 72hrs, our patient deteriorated dramatically. His illness evolved to progressive asymmetric flaccid weakness of all 4 limbs, left eyelid ptosis, disconjugate eye movements and dysphagia, acute respiratory failure requiring intubation and referral to a tertiary centre in Bangkok. He was empirically treated with meropenem, moxifloxacin and doxycycline. Up until intubation he had no meningism and maintained normal consciousness.

His past medical history included recurrent fungal sinuitis, spontaneous pneumothorax and ureteric calculi. He had no close zoonotic contacts but may have sustained multiple insect bites prior to his illness. He had not sought any vaccination advice prior to emigrating to Thailand and has since lived there for 3 years with his female partner. In particular he had never been vaccinated against JEV nor heard of the disease.

Salient abnormalities on his initial pathology panel were mild neutrophil, haemoparaesthesia, modest thrombocytopaenia and CRP 245mg/L. Septic screen was unremarkable. Serological investigations in Thailand had also excluded HIV, dengue, leptospirosis, scrub and murine typhus and melioidosis.

Nerve conduction studies in Thailand suggested Guillain-Barré syndrome. From day 14 he received Flebogamma for 3 days as well as ongoing broad-spectrum antibiotic therapy. By day 17 his fevers resolved with rapid improvement in tidal volumes post IVIG, his eye movements also normalised and bulbar function and limb weakness improved to Grade 2-3 power.

**Repatriation to Australia**

On day 25 our patient was repatriated to Westmead ICU. Profound weakness remained and was worse proximally. Additional investigations include mildly elevated CK 382 U/L, unremarkable heavy metal screen and autoimmune screens, as were syphilis, mycoplasma, legionella and hepatitis serology. His HTLV-1 serology was positive but his neurological deficits were atypical for HTLV-associated myelopathy or tropical spastic paraparesis and thought to be a false positive following IVIG.

CSF examination on day 26 showed 23 leucocytes (100% mononuclear), no erythrocytes, high protein 1.32g/L, glucose 4.5mmol/L (62% of peripheral glucose). CSF cryptococcal Ag, PCRs for enterovirus, HSV, CMV, EBV and TB were also not detected.

Multiple imaging abnormalities on MRI brain and spine were noted:

- probable cavernomas in the right temporal lobe, right pons and pontomedullary junction
- possible right carotid dissection, likely related to central line insertion
- T1-T3 intramedullary spinal cord haemorrhage, possible underlying cavernoma
- nonspecific mild enhancement of cauda equina nerve roots
- diffuse enhancement of the ector spinae muscles

Abnormal EEG with generalised slowing, consistent with diffuse cerebral dysfunction. Repeated nerve conduction and EMG suggested an upper-motor neuron-like dysfunction in lower limb muscles and excluded motor neuropathy and neuromuscular junction disorders.

**Definitive Diagnosis & Outcome**

Confirmatory diagnosis of JE was confirmed on CSF serology (ELISA) which was strongly positive for JE IgM. Peripheral blood JE IgM was also strongly positive at a titre of 1:320, IgG 1:160 and neutralising titre of 1:2560.

Our patient was managed with supportive treatment and required prolonged admission in high dependency for nearly 3 months. He remained dependent on non-invasive ventilation and bedbound due to grade 2-4 tetraplegia. He was discharged to nursing home care and passed away 5 months after initial illness onset.

**JEV causing flaccid paralysis**

Flaccid paralysis without encephalitis is a very rare presentation of JEV. There are 13 such cases published in the English literature to date, and Solomon at al showed that over a 12-month period at a referral hospital in Vietnam, 55% (12/22) paediatric presentations with flaccid paralysis were due to acute JEV infection. Features in these patients include:

- NCS & EMG studies indicated damage to the anterior horn cells
- At 12-month follow-up, patients with paralysis due to JEV had greater disability & more muscle atrophy than JEV-negative causes

Studies in imaging characteristics in JE suggest the thalamus is the most common site of involvement, with low density lesions on CT and high signal intensities on T2 weighted images in addition to haemorrhagic changes.

**Conclusion**

JE can very rarely present with a polio-like illness. To our knowledge this is the first report in Australia of flaccid paralysis due to JEV.

Particularly in the context of recent poliovirus outbreaks abroad, JE should be recognised as an uncommon but important differential diagnosis of acute flaccid paralysis.

There is a need for greater awareness of risk factors for JE amongst GPs and travellers as it is potentially fatal but vaccine-preventable disease.