CASE REPORT

Longitudinal Extensive Transverse Myelitis as a Neurological Sequelae post-Sea Urchin Stings: A Case Report

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ABSTRACT

Puncture injury from sea-urchin stings may lead to a local and systemic inflammatory reaction. We are reporting a case of longitudinal extensive transverse myelitis (LETM), which occurred ten days post-sea-urchin stings, where the patient presented with bilateral lower limb weakness. MRI showed multilevel segment spinal cord T2-weighted hyperintensity. Prompt intravenous methylprednisolone was administered, and the patient had a full recovery. To date, there is no case report of LETM associated with sea-urchin stings. Possible mechanism due to delayed immunological hypersensitivity to sea-urchin venom. This case demonstrates the potential serious neurological sequelae that may be associated with post-sea-urchin sting and the importance of prompt recognition and management in aiding recovery.

Keywords: Acute transverse myelitis, Neuromyelitis Optica spectrum disorder, Sea-urchin, Delayed hypersensitivity

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INTRODUCTION

Sea Urchins are marine spiny invertebrate Echinoderms with a globular body, which dwell on the sea bed, rocky shores and coral reefs. There are roughly 80 species of sea urchins known to be toxic to humans (1) and 12 species indigenous to Malaysia, including the poisonous Toxopneustes pileolus mainly found in peninsula Malaysia.

Puncture injuries by sea urchin spine are known to cause local and systemic inflammatory reactions. Some patients may have spines fragments retained in their bodies for weeks, which may lead to delayed complications and hypersensitivity attributed to foreign body reactions and infections (1).

We are reporting a case of Longitudinal Extensive Transverse Myelitis (LETM), which occurred ten days after a sea urchin spine puncture over both feet with retained spines.

CASE REPORT

A 36-year-old man with underlying well-controlled Type 2 Diabetes Mellitus, accidentally stepped on a sea urchin and had multiple spines puncturing the soles of both feet while swimming along the beach of Langkawi Island, Malaysia. As part of folk's remedy, he was told not to remove the spines and had one spine remained in the sole of his left foot for the next one week while other smaller spines were removed. He did not seek immediate medical attention.

One week later, he developed fever with myalgia and arthralgia. On the third day of fever, he developed bilateral lower limb weakness, which progressed until he was unable to walk on day four of illness. It was associated with numbness of bilateral lower limb and urinary retention. There was no history of back trauma before this presentation.

Upon presentation to the emergency department, his Glasgow coma scale (GCS) was full, afebrile, and hemodynamically stable. Neurological examinations of lower limbs revealed pyramidal weakness with power over bilateral knee more in the flexors reduced to 2/5 and bilateral ankle more in the dorsiflexion reduced to 3/5. Both knee and ankle jerks were present with negative plantar reflex. Muscle tones were hypotonia. There was evidence of sensory level up to L1 bilaterally. Proprioception was intact. Neurological examinations of upper limbs were normal. Cranial nerve examinations were normal, with no evidence of optic neuritis (ON). Local examination on the soles of both feet reveals multiple healed puncture wounds with one of the wounds was tattooed with some pigmented stain. No foreign body or sea-urchin spine noted on both soles. Other systems were unremarkable.

The white cell count was not raised at 8.3x 103/uL (no predominant eosinophilia), with mildly elevated his C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), which valued at 19.3 mg/L and 22 mm/hr, respectively. Other blood investigations, e.g., B12, folate level, and thyroid function test was normal.

An urgent gadolinium contrasted magnetic resonance image (MRI) of the whole spine revealed long multilevel segment spinal cord T2 weighted hyperintensity from T1 to T12 with mild cord expansion, which suggestive of LETM (Fig. 1 & 2). However, conus medullaris was spared, and no contrast enhancement (Figure 3). On the cross-sectional view, the hyperintensity occupies two-third of the cord (centrally), affecting both halves of the cord. Visualised MRI brain was normal. Subsequently, lumbar puncture reveals a clear cerebrospinal fluid (CSF), normal glucose value, no pleocytosis with high protein value of 1809 mg/dL. CSF and blood culture did not grow any organism. Serum atypical bacterial serology screening was negative. Acid-fast bacilli smear and Mycobacterium tuberculosis culture were negative. CSF Aquaporin 4 and the oligoclonal band were negative. MOG-Abs (myelin oligodendrocyte glycoprotein antibodies) were unavailable as the test need to be sent to a private lab in our case.

He was started on intravenous methylprednisolone 1g once daily for five days on day 2 of admission, followed by a tapering dose of oral prednisolone for 14 days. No antibiotic was commenced. He regained his bilateral lower limb muscle power and sensation gradually after glucocorticoid therapy commenced. Upon completing IV methylprednisolone on day five, he was already walking with aid, able to pass urine without difficulty and normal sensation. The patient was discharged walking. The patient was followed up one month after admission revealed fully recovered neurological deficit with subsequent follow up at four months showed no new neurological deficit or relapsing course.

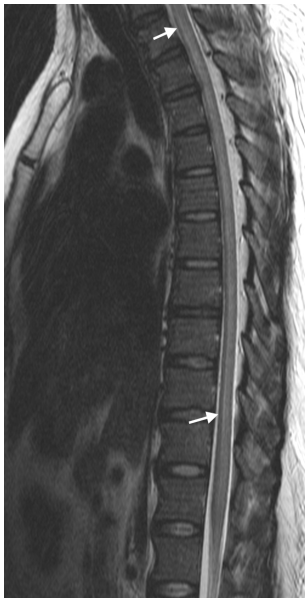

Figure 1: Sagittal T2-weighted MRI image showing long segment of abnormal high signal intensity of the cord from the thoracic to lumbar level as shown by arrows

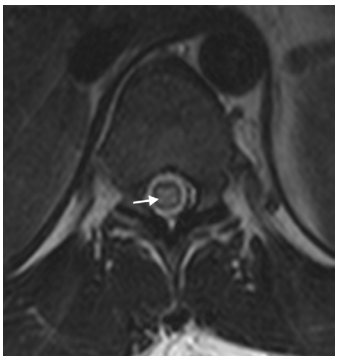

Figure 2: Axial T2-weighted MRI image showing central high signal intensity within the spinal cord as shown by arrow

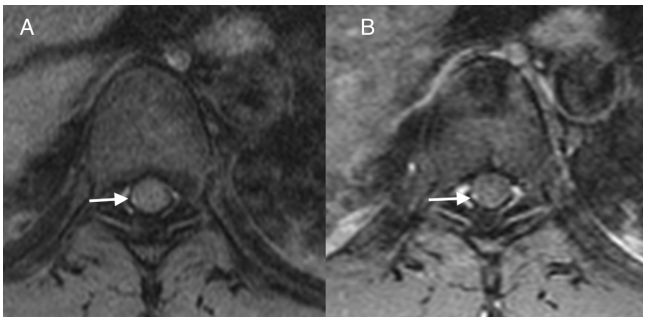

Figure 3: Axial T1-weighted MRI, pre- contrast (3A) and post contrast (3B) image showing no enhancement of the spinal cord. The spinal cord is as shown by arrows

DISCUSSION

Our patient presented with bilateral lower limb weakness with normal reflexes and sensory level evidence suggesting a cord lesion. MRI showed long segment T2 weighted hyperintensities, which main differentials may include demyelination, ischaemic, infective, and inflammatory lesions of the spinal cord. Diagnosis of LETM was made, as evidenced by the long multilevel segment spinal cord lesion supported by the clinical presentation.

LETM is an immune-mediated inflammatory disease of spinal cord diagnosed by having contiguous lesions extending over three or more vertebral segments on spinal MRI with variable contrast enhancement and located centrally within the spinal cord (2). The clinical presentation is usually dramatic with patients having acute or subacute paraparesis or tetraparesis depending on the location of lesion on the spinal cord with sensory disturbances and alteration of gait, bladder, bowel and sexual dysfunction (2). In our case, we postulated the pathophysiology of immune-mediated LETM secondary to sea-urchin stings. However, other differential diagnoses need to be ruled out in particular neuromyelitis optica spectrum disease (NMOSD), multiple sclerosis (MS), and infectious origin (2). NMO is a rare chronic inflammatory central nervous system

that requires the presence of either ON or acute myelitis, brain imaging not meeting the diagnosis of MS, and the presence of antibodies against aquaporin-4 (AQ4-Abs) (2, 3). Recently, the presence of serum MOG-Abs can be assigned as NMOSD (3). In NMOSD, most common presentations were more predominantly ON, where it might occur up to 54% and 64% in AQ4-Abs and MOG-Abs, respectively (3). In our case, ON was absent together negative AQ4-Abs in CSF analysis. However, although we did not send MOG-Abs, myelitis in MOG-Abs related NMOSD present only in 18 to 33% of the case, with ON usually being the predominant presentation unlike in our case (3).

MS, on the other hand, is a demyelinating autoimmune disease, where acute myelitis is common as an initial presentation (2). Conversely, spinal cord lesion in MS usually patchy, extend over one or two vertebrae and localized to the lateral or dorsal aspect of the spinal cord (2). Furthermore, LETM in MS is unusual and usually will have lesions in the MRI brain (2). Our patient presents with LETM, predominantly central cord lesion, normal MRI brain with a negative oligoclonal band to suggest MS. An infectious cause may include viral, bacterial, parasitic infections, or mycobacterium tuberculosis (2). Bacterial and mycobacterium tuberculosis infection was less likely in this patient given sterile CSF picture, no growth on blood and CSF culture coupled with mildly elevated CRP and ESR. Besides that, other infectious causes include herpes viruses (herpes simplex, varicella-zoster, Epstein-Barr virus, and cytomegalovirus), human-deficiency virus, human T-cell lymphotropic virus type 1, Borellia burgdogeferi, Treponema Pallidum, and schistosomiasis (2). However, our limitation on thoroughly investigate infection panel with CSF polymerase chain reaction or serology given limited availability for these investigations and cannot rule out the possibility of post-infectious LETM being the aetiology.

To date, there was no case report associating LETM to injuries inflicted by sea urchins and their venoms. Toxic substances identified in sea urchin venoms are steroids, glycosides, histamine, serotonin, cholinergic substances, and bradykinin-mimic substances, which may induce immunological responses (1). Puncture injuries are known to cause local and systemic inflammatory reactions. Local reactions may range from pain and swelling around the puncture wound, while systemic inflammatory reactions include paresthesia, fever, malaise, muscular weakness, systemic arthritis, and dyspnea (1). Even the tattooing of the skins by its pigments, as in our case, can simulate fragment retentions, which lead to delayed complications in the form of granuloma formation, chronic arthropathy, neuropathy, osteomyelitis and vesicular reaction (1).

Our patient recovered on high dose steroid therapy without the institution of antibiotics may suggest underlying immunological pathophysiology, e.g., delayed hypersensitivity reaction, which also common in the envenomation of other sea creatures, including jellyfish (4). Delayed immunological manifestation presenting as transverse myelitis were also being reported post-vaccination, albeit rare (5). Pathophysiology begins from the deposition of venom, which acts as molecular mimicry, which may set off a complicated system of cellular and cytokines interaction term epitope spreading that initiates immune response via dendritic and T-cells activation evoking delayed hypersensitivity reaction (4,5). This induced autoimmunity may set off an inflammatory reaction at the spinal cord, which leads to LETM. Our index case developed LETM after ten days of sea-urchin stings which fit the postulated pathophysiology of delayed immunological reaction. However, there is no report linking sea urchin venom to neuromuscular conduction disturbances leading to muscle weakness that has seen among jellyfish's sting. The pragmatic approach in treating immune-mediated cause of LETM is with high dose steroids (2,3). Other therapeutic options include immunosuppressive therapy with azathioprine, mycophenolate mofetil, cyclophosphamide, and even plasma exchange in unresponsive cases (2,3). If infectious causes are suspected, empirical treatment with a third-generation cephalosporin, ampicillin, and acyclovir should be commenced (2,3).

This case emphasised on the importance of early recognition of transverse myelitis by clinical suspicion coupled with urgent MRI and prompt institution of therapy in aiding fast recovery. In our case, methylprednisolone was administered early after diagnosis and the patient showed dramatic improvement even after two days of treatment where the patient eventually had a full recovery.

CONCLUSION

LETM is an immune-mediated inflammatory spinal cord disease, which may be associated with delayed immunological hypersensitivity from sea-urchin venom. This case demonstrates the potential serious neurological sequelae post-sea-urchin sting and the importance of prompt recognition and management in aiding faster recovery.

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