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A RARE CASE OF LONGITUDINALLY EXTENSIVE TRANSVERSE MYELITIS ASSOCIATED WITH STRESS CARDIOMYOPATHY IN YOUNG.

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final Manuscript

Abstract

A typical sign of neuromyelitis optica is longitudinally extensive transverse myelitis (LETM) (NMO). Not all LETM patients, nevertheless, have antibodies against aquaporin-4 (AQP4). Idiopathic isolated N-LETM is not that uncommon among first-ever LETM and differs greatly from P-LETM in which astrocytic damage is clearly present in many ways. Although neuromyelitis optica is the most common cause of longitudinally widespread transverse myelitis, there are several alternative possibilities. Infection, cancer, malignancy, and metabolic disruption are a few of these. Some of these can be treated easily. To distinguish between these reasons, laboratory and radiological examinations can be useful. Differentiating between inflammatory and non-inflammatory aetiologies and identifying individuals who are at high risk of a recurring course are key components of treating longitudinally widespread transverse myelitis. We present you a case idiopathic longitudianally extensive transverse myelitis in a young male the treatment of which caused steroid induced cardiomyopathy.

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Case Report

The patient is a 34-year-old male with no comorbidities who presented to the emergency department on day 15 of his complains of inability to walk, diminision of vision and headache. He developed fever during the first 48 h of admission, which subsequently resolved. From day five onwards, he complained of myalgia of his lower limbs, especially upon walking up stairs. There was a further onset of fever with progressive bilateral lower limb weakness on day 10. By day 15, he was unable to stand and developed urinary retention. There were no respiratory or gastrointestinal symptoms. On examination, he was afebrile with normal vital signs and had numbness and allodynia below the T10 spinal level. In the lower limbs, there was bilateral hypertonia with reduced power (0/5 proximally and distally) along with exaggerated deep tendon reflexes at the knees and ankles with upgoing plantar. Upper limb and cranial nerve examinations were normal, although there was evidence papilledema on fundoscopic examination and sinus bradycardia on ECG. No history of recent or remote COVID vaccination or infection. Blood investigations revealed haemoglobin of 13.0 g/dL with total white cells count 15000 which came to normal range within 3 days. Renal and liver functions test were normal. Cultures of blood and urine showed no growth. Suspecting meningitis, a lumbar puncture was done which showed very high protein levels 167mg/l, glucose 42mg/l, total leucocyte counts 460 predominantly lymphocytes 90%. CSF ADA was 8.23 and culture showed no growth, no acid-fast bacilli seen. Cardiac studies by 2D echo were normal with ejection fraction of 60% and normal chambers. An MRI whole spine screening was done which was evident of longitudinally extensive transverse myelitis.

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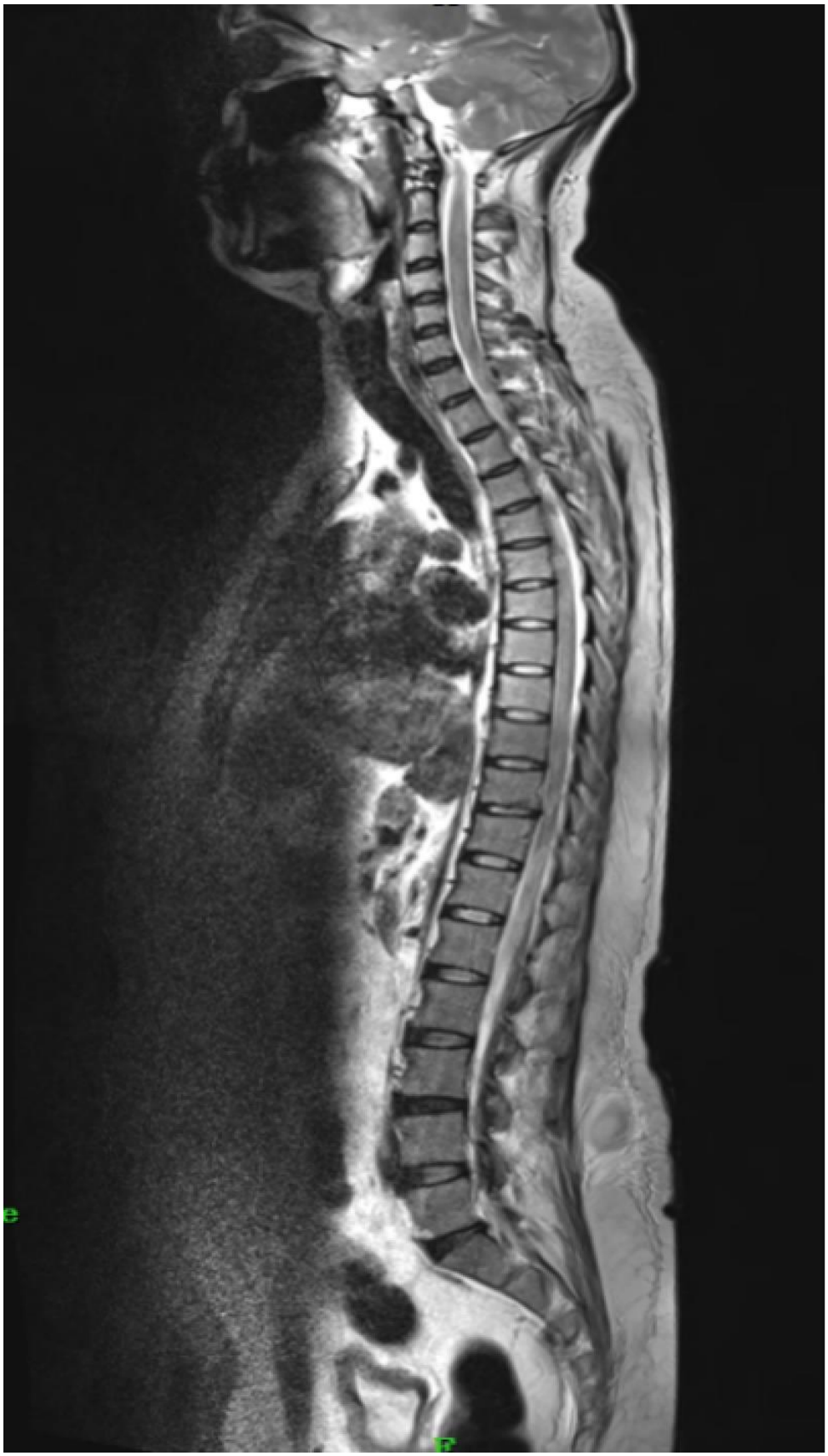

FIGURE 1: showing T2 hyperintensities across the spine

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Due to financial issues further, extensive investigation could not be done in this patient. He was diagnosed with LETM and started with plasmapheresis with antibiotics. After the second cycle, patient was started on IV methylprednisolone 1gm per day for 3 days followed by oral shift to steroids on day 4. After the 5th cycle of plasmapheresis and day 3 of iv methylprednisolone, patient developed sudden onset hypotension and breathlessness. Chest Xray suggestive of flash pulmonary edema and patient was immediately started on diuretics and ionotropic support along with oxygen requirement of 10 litres O2 to maintain saturation of 96%. His 2D echo revealed an ejection fraction of 15% and global LV hypokinesia. Heart failure was attributed to the high dose steroid given to the patient. However, within a week The patient responded to the treatment and eventually ejection fraction improved to 55%. Upon discharge, the patient was able to walk with support but was still limited given ongoing pain. Four weeks later, the patient was able to walk without assistance. He will continue to be on a regular follow-up to observe the recurrence of symptoms and monitor recovery.

Discussion

With an estimated prevalence of 1.34 to 4.6 occurrences per million yearly, TM is a relatively rare illness.[1] When three or more vertebral segments of the spine MRI show contiguous central cord lesions with varying contrast enhancement, LETM is diagnosed [9]. Depending on where the lesion is on the spinal cord, patients generally arrive with a dramatic presentation of acute or subacute paraparesis or tetraparesis, sensory problems, altered gait, bladder, bowel, and sexual dysfunction.[2] Our case showed no relationship between sudden onset of LETM and a stress or cause or family history. However, differentials like multiple sclerosis, neuromyelitis optic spectrum disorder and infectious diseases especially viral were kept in mind. Additionally, the lack of CSF pleocytosis, a classical MS lesion in the spine (usually patchy involving one or two vertebrae with peripheral enhancement) did not relate to MS. Viral infections supported by the CSF study lymphocyte predominant would be the first differential for our patient. However, an autoimmune transverse myelitis was also considered. It is frequently associated with infectious or systemic autoimmune diseases, but its etiology remains unknown in a substantial portion of cases, which are classified as idiopathic.[1]

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Non-compressive myelopathies have a variety of aetiologies, including ischemic disorders, delayed radiation effects, paraneoplastic diseases, infectious or parainfectious conditions, and systemic autoimmune diseases. Among the latter, SLE, Sjögren's syndrome, sarcoidosis, Behçet's disease, various connective tissue disorders, and the antiphospholipid syndrome (APS), either primary or subsequent to SS, can all be linked to ATM. Additionally, neuromyelitis optica (NMO), also known as Devic's syndrome and defined as the triad of neuromyelitis optica (ATM) and optic neuritis, can present as the first symptom of multiple sclerosis (MS) and ATM, respectively (ON).[1] In 2002, the Transverse Myelitis Consortium Working Group (TMCWG) proposed diagnostic criteria and nosology of ATM.[3] The finding that sera from more than 70% of NMO patients carried an antibody that was only seen in a small percentage of MS patients (10%) was a turning point in establishing the differences between MS and NMO.[4] Longitudinally extensive TM lesions, which are defined as lesions on the spinal MRI that span three or more contiguous segments, are a hallmark of NMO (LETM). The seropositivity rate was 50% (70/139) in the pooled data from 7 US and European studies[5] and 67% in Hong Kong Chinese[6]. NMO-IgG has also been discovered in individuals with LETM who do not have clinical or preclinical ON. Be aware that patients with recurrent LETM exhibit higher rates of NMO-IgG detection than patients with isolated episodes.[7] We demonstrate that the severity of myelitis in AQP4-Ab illness correlates with the size of the lesion and remaining impairment. First myelitis attacks have spinal cord lesions that resemble those seen later in the disease's progression, with a tendency to affect the central grey matter and a high frequency of cord oedema and T1 hypointensity.[8] The corpus callosum is typically involved in the cerebellar, periventricular, and cortical/juxtacortical lesions of MS, whereas the dorsal brainstem (area postrema), the periependymal region of the diencephalon, the hemispheric white matter, and regions close to the lateral ventricles are typically involved in the lesions of NMO. Because of the differences in their respective prognoses and treatments, NMO must be distinguished from MS and other demyelinating diseases.[9]

Approximately 250,000 people globally and 4,000 persons in the US are thought to have NMO. 5 When compared to SLE myelitis, the prognosis of NMO, which manifests as a monophasic or relapsing type, is dismal.[10] To ensure accurate identification of this small subset of patients with NMO and SLE and to improve early recognition of NMO for patients presenting with a

first central nervous system (CNS) event, especially LETM, multidisciplinary, longitudinal care coordination that includes neurology, rheumatology, and physical and rehabilitation medicine is necessary. 22 recorded cases of SLE with LETM were found in a recent comprehensive analysis of the literature between 1966 and 2008, despite the fact that the prevalence of LETM in SLE patients remains unknown.[11]

Immunosuppressive treatments, most frequently azathioprine, rituximab, or mycophenolate, that suppress B cells or antibody production are usually used to treat patients with established NMO or relapsing NMO spectrum disorder.[12] Intensive immunosuppressive therapy combined with concurrent high-dose corticosteroids is the cornerstone of treatment for autoimmune transverse myelitis (eg, cyclophosphamide). Early, aggressive therapy is essential for a positive outcome.[13] LETM is the main contributor to disability, followed by ON. NMO alone has very poor clinical outcomes, with the majority of patients experiencing severe, irreversible neurologic impairment. NMO-IgG antibodies may be linked to a higher risk (> 60%) of LETM recurrence and the emergence of ON, according to research.[14] In a prospective research, patients with an isolated LETM who were NMO-IgG seropositive had a risk of more than 50% for having recurrent myelitis or new-onset ON, compared to 0% for patients who were NMO-IgG seronegative.[15] To identify patients with NMO who are at high risk for relapse and a worse prognosis, an early search for AQP4-Ab should always be conducted in patients with SLE who exhibit symptoms of demyelination.[16]

Controlled investigations and the creation of a standardised registry for SLE patients that includes LETM and NMO could aid in the creation of an efficient treatment plan and focused management approach for this crippling condition.

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