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Review article

Motor neurone disease-associated neck pain misdiagnosed as cervical spondylosis: A case report and literature review

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ABSTRACT

Background: Motor neurone disease (MND) is a chronic, progressive and currently incurable neurodegenerative disorder. Although pain as a symptom appears in many patients with MND, it is often misdiagnosed as other diseases when occurs before the onset of weakness. Patients are often assigned to non-neurological departments due to the atypical symptoms, which can lead to diagnostic delay and inappropriate treatment.
Objective: To analyze the causes of misdiagnosis and improve the clinician's understanding of neck pain in patients with MND.
Methods: We reviewed relevant literature and retrospectively reported a misdiagnosis case of MND-associated neck pain.
Results: A case of MND presenting prominently as neck pain was suspected of suffering from cervical spondylosis and wrongly assigned to orthopedic clinic. When eventually being diagnosed as MND, his neck pain was found to be caused by intracranial hypertension (ICH) resulting from hypoxia via insidious respiratory failure through ventilator insufficiency.
Conclusion: Careful evaluation of the clinical progression of the symptoms, extensive EMG and nerve conduction study, as well as the establishment of better clinical approach to the diagnosis and higher public awareness allow a reduction of misdiagnosis.

1. Introduction

Motor neurone disease (MND), as a rare disease of nervous system, was often misdiagnosed because it may mimic several disorders at onset, such as cervical myelopathy. As MND shows almost invariably a subtle onset, early diagnosis can be difficult. The diagnostic delay in MND have been reported to range between eight and more than fifteen months, and the median time from onset to diagnosis was 12 months in spinal-onset patients [1]. Therefore, more comprehensive understanding the symptoms of MND can help early diagnosis and bear significant implications in order to access to an appropriate management and care.

Neck ache, especially chronic – pain is the most commonly complaint in clinical practice. It is the common clinical picture of cervical spondylosis or cervical muscle injury [2,3]. Yet, pain has been a widely neglected symptom in patients with MND. Although recent studies indicate that pain is a prevalent symptom in MND, affecting 46% of patients [4], pain that occurs before the onset of weakness is often misdiagnosed as other diseases and referred to non-neurological departments, resulting in serious diagnostic delay. Therefore, improving the recognition of pain in patients with MND has important clinical implications for reducing diagnostic delay and providing appropriate treatment. We herein report a misdiagnosis case of MND-associated neck pain. His initial symptom was muscle soreness in the neck, which gradually worsened and was eventually found to be due to intracranial hypertension (ICH) resulting from hypoxia via insidious respiratory failure. The delay time from onset to correct diagnosis of this patient is two years. In this report, we reviewed previous literatures to analyze the causes of misdiagnosis and further enhance the understanding of the pain in patients with MND.

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2. Results

A 73-year-old man with serious neck pain and developed progressive weakness was referred to our hospital. His medical history included milde mphysema, well-controlled hypertension and carotid artery plaque, for which he took nifedipine sustained release tablets, valsartan, and atorvastatin calcium. He was a nonsmoker, and had no exposure to any toxic and no significant family history of note.

About three and a half years ago, he suffered from soreness in the neck and felt weak when raised arms without any clear causes. Symptoms got progressively worse and extended to the waist in the following three months. As the obvious neck and waist soreness, he was admitted to the department of orthopedics. Magnetic resonance imaging (MRI) of cervical and lumbar spine showed degenerate, and he was treated with analgesics to relieve the ache. However, the soreness continued.

Three years ago, his hands became thinner, accompanied with muscular tremors. Electromyogram (EMG) revealed neurogenic injury: long-duration and high-amplitude of motor unit action potentials (MUAPs) and simple-mixed phase in bilateral C5, C6 innervated muscles. He was given symptomatic treatment and advised to follow up EMG.

Two years ago, he felt shortness of breath and was admitted to the department of respiration for chronic lung disease. At that time, his SaO_2 fluctuated between 83% and 88% and he was advised to take long-term domiciliary oxygen therapy (LTDOT).

Dysarthrosis and dysphagia occurred approximately one and a half years ago, with difficulty in writing and walking. Meanwhile, the neck ache aggravated, and he felt like a rock on the neck to be crushed to death. He began to feel amazingly sleepy and drowsy during the day. Once again, he visited his doctor in department of orthopedics and wanted to have cervical operation to solve his pain. But, his dysphagia and dysarthria could not be explained by cervical spondylosis, and he was referred to neurology. Physical examination revealed dysarthrosis and a little shortness of breath. Atrophy was seen in both hands, shoulders, hips, and lower extremities. Muscular tension was normal in the four limbs. Reflexes were not elicited in bilateral upper limbs, but were present and symmetric in both lower limbs. He had muscle weakness affecting all limbs. Proximal and distal muscle strengths were graded 3 and 4 respectively in the upper limbs, and were graded 4 and 5-respectively in the lower limbs. Bilateral pathological signs were negative and coordination intact was elicited. He was suspected of suffering from MND and admitted to our Neurology Department. The repeated EMG examination revealed abnormal spontaneous activities (the fibrillation and positive sharp wave), long-duration and high-amplitude of MUAPs and simple phase in finger extensors, wrist flexors, sternocleidomastoid, anterior, trapezius, tibialis, gastrocnemius, cervical and thoracic paraspinal muscles. Cranial (MRI) showed lacunar cerebral infarction in the basal ganglia. Cervical MRI at this time disclosed degenerate and bulging of disk between C3 and C4, C4 and C5, as well as C5 and C6 (Fig. 1). Chest computed tomography (CT) showed mild emphysema, small amount of inflammation in the bilateral lower lung and pleural effusion. Laboratory findings showed that complete blood count, creatine kinase, liver functions, renal function, tumour markers and the immune system were normal. Electrolyte investigations showed low serum sodium and chloride levels (sodium:122 mmol/L; chloride:80.7 mmol/L) on admission (day 1). Three days after admission, he was given a lumbar puncture in order to identify chronic inflammatory demyelinating polyradiculoneuropathy (CIDP). The routine examination of cerebrospinal fluid (CSF) was normal, but, the intracranial pressure (ICP) was surprisingly elevated (300cmH_2O). We use mannitol, glycerol and fructose injection to reduce intracranial pressure and supplement saline to improve hyponatremia. On day 4, when we were looking for the cause of intracranial hypertension, he became unresponsive, and was in a confused state of mind. Just at that moment, the arterial blood gas analysis revealed hypoxemia (PaO_2:34.20 mmHg, SaO2:53.40%), hypercapnia ($PaCO_2$:122.20 mmHg) and respiratory acidosis (PH:7.19). After the initiation of noninvasive positive pressure ventilation (NPPV), his hypoxemia improved (PaO2: 87.10 mmHg; PaCO2:50.00 mmHg; SaO2:90%). He became conscious. Yet, PaO2 dropped again to 44 mmHg 8 h later and PaCO2 was 120.09 mmHg. He was again confusion. The endotracheal intubation was taken after obtaining written consent from his families (day 6). Subsequently, his hypoxemia and hypercapnia improved (PaO2:92 mmHg, PaCO2:44 mmHg; PH:7.42), he became alert, the neck pain disappeared and ICP returned tonormal (120 cm H_2O). Therefore, we confirmed that the ICP was caused by hypercapnia, and eventually resulted in neck pain. Unfortunately, pulmonary function tests cannot be performed due to the severe condition of the patient. At day 8, he has a fever and his blood laboratory data indicated the increase of white blood cell (WBC). CT of chest showed inflammation in the bilateral lower lung and pleural effusion were aggravated (Fig. 2). We initiated antibiotic to treat the patient's pneumonia. His son recalled that about one

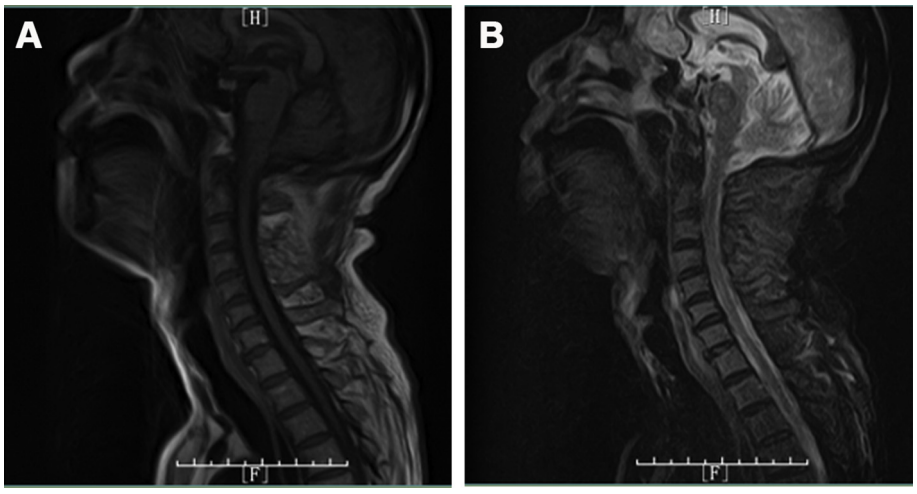

Fig. 1. Cervical MRI: no obvious cervical spinal cord lesions. T1-weighted (A) and T2-weighted (B) MRI images of cervical showed degenerate and bulging of disk between C3 and C4, C4 and C5, as well as C5 and C6, but no obvious cervical spinal cord lesions.

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