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CASE REPORT

Neurological impairment in a patient with concurrent cervical disc herniation and POEMS syndrome

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Abstract

Purpose POEMS syndrome is a rare clonal plasma cell disease characterized by polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes. We report a rare case of neurological impairment in patients with concurrent cervical disc herniation and POEMS syndrome.

Methods A patient presented to a local hospital with C3/4 and C4/5 disc herniation, apparent spinal cord compression concomitant with neurological signs, and concurrent POEMS syndrome. Anterior cervical discectomy and fusion was performed.

Results The limb numbness was only slightly alleviated, and 10 days postoperatively the patient complained of muscle weakness of the extremities and was referred to our hospital. The patients exhibited non-typical neurological signs and an enlarged liver and spleen that could not be explained. Electroneuromyography and immunofixation electrophoresis produced abnormal results. We diagnosed concurrent POEMS syndrome, for which drug therapy was prescribed. The patient's symptoms receded.

Conclusion Patients presenting with cervical spondylopathy and non-typical neurological signs and symptoms or other systemic problems should be evaluated for the presence of concurrent disease and ruled out differential diagnoses.

Keywords Neurological impairment · POMES syndrome · Cervical disc herniation · Cervical spondylosis

Introduction

Cervical disc herniation is a common spinal disorder that can lead to radiculopathy and myelopathy. If conservative treatments for cervical disc herniation fail, anterior cervical discectomy and fusion (ACDF) is standard treatment. The clinical presentation of cervical spondylotic myelopathy includes numbness, paresthesias, sensory deficits, muscle weakness of extremities, gait disturbances, and/or bladder or bowel dysfunction [1]. The Hoffmann sign is usually positive. The symptoms are likely to be relieved by decompression surgery.

Some patients with cervical spondylotic myelopathy, however, may have a concurrent disease that affects the central nervous system. Multiple sclerosis (MS), for example, is a chronic demyelinating autoimmune disease that has symptoms similar to those associated with myelopathy: spasticity, sensory disturbances, gait ataxia, weakness. It is thus difficult to differentiate neurological signs due to cervical spondylosis from those of demyelinating autoimmune disease. Their treatments are also different and thus complicated for patients with multiple myelopathic pathologies. Lubelski et al. [1] reported the surgical results of patients who had both MS and cervical stenosis. They found that the modified Japanese Orthopaedic Association scores that had improved at the short-term postoperative follow-up worsened at the long-term follow-up.

POEMS syndrome is a rare clonal plasma cell disease characterized by polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes [2]. Because of demyelinating polyneuropathy with multiorgan involvement, POEMS syndrome often presents as peripheral neuropathy [3]. We found that neurological impairment in patients with concurrent cervical disc herniation and POEMS syndrome is rarely reported. Therefore, we report a case of C3/4 and C4/5 disc herniation with neurological signs that was diagnosed as POEMS syndrome after decompression surgery.

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Case report

A 39-year-old man presented at a local hospital with a 3 month history of leg numbness and gait disturbances and 1 month of upper limb numbness. On physical examination, the limb muscle strength and muscular tension were normal. He had upper limb tendon reflex weakness, no lower extremity tendon reflex, and hypesthesia of both hands and feet. Hoffmann sign and Babinski reflex were negative. Deep sensation was normal. Magnetic resonance imaging revealed C3/C4 and C4/C5 dural sac and spinal cord compression. The cervical spinal canal behind C3/4 and C4/5 disc showed a herniated disc at the centre and left side on the T2-weighted axial image (Fig. 1a). However, the spinal cord signal was normal. Three-dimensional computed tomography (CT) of the cervical spine showed no calcification of the posterior longitudinal ligament or ligamentum flavum (Fig. 1b).

Routine blood examination (RBE) showed white blood cells at 13.44×109/L (neutrophils 86.2%). Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were elevated. Nevertheless, C3–C5 ACDF was performed at local hospital (Fig. 2). The limb numbness was slightly diminished and the other symptoms unchanged. Ten days postoperatively, his muscle weakness increased accompanied by hyperpyrexia. He was referred to our hospital.

A comprehensive examination was performed. Urine tests demonstrated proteinuria. Abdominal ultrasonography showed enlarged liver and spleen. The neurologist suggested electroneuromyography, immunofixation electrophoresis, and a urine kappa (κ) light chain test. Electroneuromyography showed upper and lower limb peripheral neuropathy. Immunofixation electrophoresis showed a low blood κ light chain level (5.31 g/L, reference value 6.89–13.00), elevated blood lambda (λ) light chain level (11.7 g/L, reference value 3.80–6.50), and low κ/λ light chain ratio. The urine κ light chain concentration had apparently elevated (0.157 g/L, reference value < 0.02). In addition, the patient appeared emaciated with swarthy skin.

The 2003 diagnostic criteria for POEMS included two major criteria (polyneuropathy and monoclonal plasma proliferative disorder) and seven minor criteria (bone lesion, Castleman disease, organomegaly, oedema, endocrinopathy, skin changes, papilledema) [4]. Two major criteria and at least one minor criterion were necessary for diagnosis. According to our comprehensive analysis of symptoms, physical signs, and anomaly indexes, the diagnosis was POEMS syndrome. The rising blood λ light chain and abnormal blood κ/λ light chain ratio indicated the presence of M protein. Therefore, with the neuropathy and the presence of some minor criteria, the diagnosis of POEMS syndrome was confirmed. The patient underwent comprehensive medical therapy including rehabilitation exercise and drug therapy (melphalan combined with dexamethasone). The symptoms of limb numbness and muscle weakness were improved after the treatment. Then, this patient was followed up by neurologist.

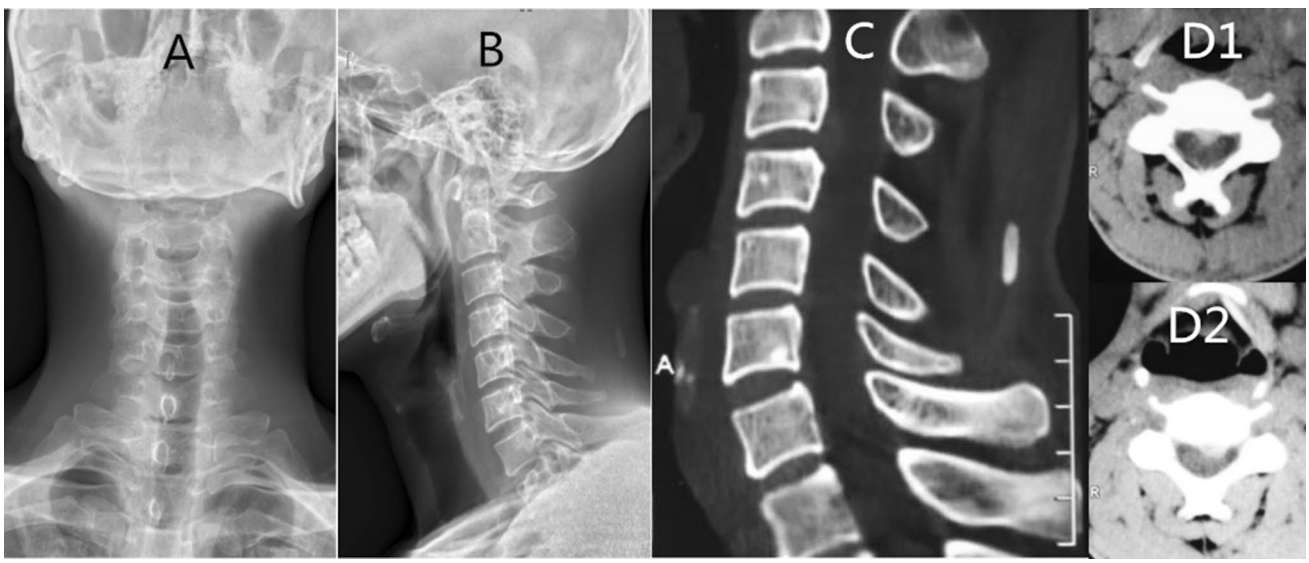

Fig. 1 **a**, **b** Cervical radiography shows normal alignment. **c** Sagittal three-dimensional computed tomography (CT) shows no calcification of the posterior longitudinal ligament or the ligamentum flavum. **d** Transverse sections of cervical CT show an intraspinal soft tissue shadow at the centre at the C3/4 level (**d1**) and on the left side of the C4/5 level (**d2**)

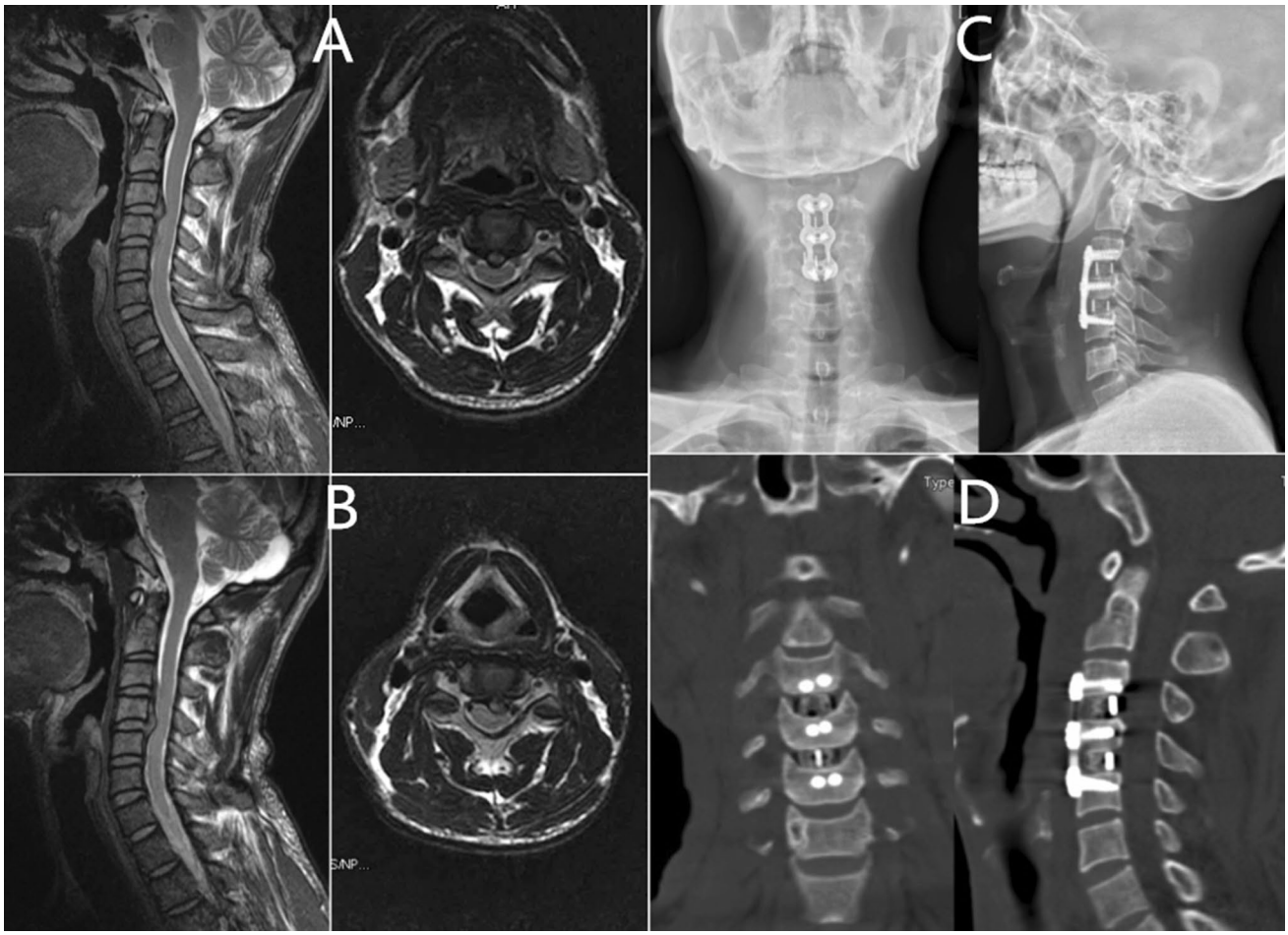

Fig. 2 **a**, **b** Magnetic resonance imaging reveals C3/C4 and C4/C5 dural sacs and spinal cord with apparent compression. The cervical spinal canal behind C3/4 (**a**) and C4/5 (**b**) discs has a herniated disc at the centre and left side on this T2-weighted axial image. **c**, **d** Anterior cervical discectomy and fusion was performed at C3–C5. Postoperative imaging shows good internal fixation. Herniated discs were completely resected

Discussion

Because of the rarity and complicated clinical manifestations of POEMS syndrome, its diagnosis may be missed. POEMS syndrome involves many organs and bodily systems [2]. Misdiagnoses may include other peripheral neuropathies, tuberculosis, diabetes, chronic nephritis, various skin diseases, and multiple myeloma [5]. Dispenzieri et al. [4] reported that polyneuropathy was an essential element for diagnosing POEMS syndrome. Therefore, neurological symptoms are among the most important clinical manifestations. Li et al. [2] found 100% of the patients with POEMS syndrome had peripheral neuropathy. Other specific features of POEMS syndrome include the high level of serum vascular endothelial growth factor, extravascular volume overload, organomegaly, endocrinopathy, monoclonal plasma cell dyscrasia, skin changes, papilledema, bone lesions, and hemangioma [6, 7].

After reviewing the preoperative clinical manifestations and examination results of this patient, we identified several abnormalities different from those of cervical spondylotic myelopathy due to intervertebral disc herniation. First, the neurological signs were different. Polyneuropathy, one of the essential manifestations of POEMS syndrome, may include polyneuropathy, peripheral neuropathy, segmental demyelination alone, axonal loss alone, or a mixture of axonal loss and segmental demyelination [2]. Adams et al. [3] speculated that the neural pathological change is due to immune-mediated nerve injury. The neuropathy of POEMS syndrome is symmetrical and ascending, with either insidious or rapidly progressing onset. Patients often describe numbness and dysesthesias followed by a progressively ascending weakness that overshadows the sensory impairment [4]. In contrast, cervical spondylotic myelopathy is due to spinal cord compression. Reflexes at the biceps, triceps, patellar, and Achilles tendons are usually hyperactive. Pathological

reflexes, including the Hoffman sign and Babinski sign, are usually positive. The patient often presents with asymmetrical extremity weakness and numbness and feels that his/her legs are stepping in cotton (as did our patient). The neurological signs of our patient—symmetrical distal extremity numbness, normal muscle strength, muscular tension—and negative pathological signs were different from those of cervical spondylotic myelopathy alone. Second, our patient showed an unexplained enlarged liver and spleen as well as abnormal electroneuromyography and immunofixation electrophoresis results, which could not be explained by simple cervical spondylotic myelopathy.

Cervical spondylopathy may be accompanied by a disease that affects the nervous system (e.g. MS, Castleman disease, POEMS syndrome) [1, 8, 9]. The overlapping symptoms make identification difficult. When these pathologies converge, the diagnosis and treatment are complicated as the natural histories and therapies are vastly different. Therefore, when the differential diagnosis is cervical spondylopathy but the patient presents with non-typical neurological signs and symptoms, the diagnosing physician should be vigilant for other possibilities.

Lubelski et al. [1] found that preoperative MRI findings were associated with postoperative outcomes in cohorts of either MS or cervical stenosis patients but not with those having *concurrent* MS and cervical stenosis. (The postoperative outcomes in concurrent MS and cervical stenosis patients worsened at the long-term follow-up, possibly related to MS progression.) MS is a chronic demyelinating autoimmune disease with neurological symptoms similar to those of myelopathy, thereby leading to an incorrect diagnosis and treatment.

Castleman disease is a rare lymphoproliferative disorder that may be associated with peripheral neuropathy [10]. Naddaf et al. reported 105 patients with Castleman disease, 27 (27.5%) of whom had peripheral neuropathy [8]. They found that Castleman disease was an additional cause of a demyelinating neuropathy and characteristically presented with mild, predominantly sensory deficits in a duration-dependent pattern, involving mainly the distal lower limbs, but rarely with motor deficits. Therefore, the neurological signs of cervical spondylopathy must be distinguished with those of Castleman disease. If a patient with cervical disc herniation or spinal stenosis has neurological signs and presents with unexplained lymphadenectasis, Castleman disease should be considered. In addition, some haematological disorders (e.g. monoclonal gammopathy of undetermined significance, Waldenström macroglobulinemia) may also be associated with peripheral neuropathy with demyelinating features [11, 12]. The related similarity should be noted.

Reports of neurological impairment concurrent with cervical disc herniation and POEMS syndrome are rare. Because of the aggravated neurological signs postoperatively in our patient, he was referred to our hospital, where POEMS syndrome was finally diagnosed. Because limb numbness only slightly diminished postoperatively, we thought that the neurological symptoms in this case were mainly derived from POEMS syndrome. Thus, spinal cord compression might have explained some of his symptoms but was not the main cause. Although severe disc herniation caused spinal cord compression, the spinal cord signal of this patient shown in MRI was normal (Fig. 1a). In this situation, the diagnosis of cervical spondylotic myelopathy is mainly based on symptoms, neurological examination, and laboratory. However, neurological symptoms and pathological signs of this patient were non-typical for the diagnosis of cervical spondylotic myelopathy. In addition, RBE, ESR, and CRP showed obvious signs of inflammation. Therefore, it was mandatory to perform extensive examination in order to rule out frequent differential diagnoses. The surgery should not be performed without extensive tests for standard differential diagnoses. The preoperative management in this patient was not proper. Medical treatment for POEMS syndrome should be prioritized. If the neurological symptoms are relieved after medical treatment, surgery probably can be avoided. In conclusion, when a cervical spondylopathy patient presents with non-typical neurological signs and symptoms or other systemic problems, concurrent disease may be present and differential diagnoses must be ruled out.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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