Images in neuroscience

Weakness and sensory disturbance of the left extremities

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1. Clinical background

A 71-year-old woman presented with progressive weakness and paresthesia of the left extremities that had started 2 months earlier. Physical examination showed left palm rash. Fig.1 is the X-ray of the chest on admission. Neurological examination revealed weakness and sensory disturbance of the left extremities, hyporeflexia of the lower extremities, neurogenic bladder, and constipation without headache or nuchal stiffness. Nerve conduction studies of her left extremities were nearly normal with slightly low amplitude and those of her right extremities were normal. Laboratory tests revealed pleocytosis and elevated protein levels in cerebrospinal fluid (CSF), and elevated serum angiotensin converting enzyme (ACE) levels. Cervical MRI revealed intradural-extramedullary nerve root swelling and enhancement predominantly in the left region (Fig. 2).

2. What is the most likely diagnosis?

1. Chronic inflammatory demyelinating polyradiculoneuropathy
2. Sarcoidosis
3. Malignant lymphoma
4. Leukemia
5. Tuberculosis
1. Answer

2. Sarcoidosis

2. Discussion

Chest X-ray of Fig. 1 shows bilateral hilar lymphadenopathy and the patient was diagnosed with sarcoidosis based on biopsy of lymph node in the lung. After the diagnosis was made, high-dose oral steroid therapy was administered. Neurological symptoms ameliorated gradually with diminishing of nerve root swelling and disappearance of gadolinium-enhancement (Fig. 3). Steroids improved both neurological symptoms and MRI findings. These consequences suggests that nerve root swelling was not a tumor but rather an inflammatory changes come from sarcoidosis.

Neurological involvements occur 5 to 10% of systemic sarcoidosis. Facial nerve paralysis, visual loss, and meningitis are commonly evolved as first symptoms. Intradural involvements in spinal lesions are infrequent in neurosarcoidosis. In particular, polyradiculopathy is considered a rare symptom in neurosarcoidosis and commonly affects the thoracolumbar or lumbosacral roots. MRI findings in some reports have revealed intramedullary involvements or mass-like extramedullary lesions. Typically, spinal cord lesions in sarcoidosis reveal patchy intramedullary enhancements along with leptomeningeal enhancements in MRI. This finding is considered to be the reason why direct invasion of the leptomeninges is a major cause of spinal lesions in sarcoidosis. Our case
demonstrated an intradural-extramedullary, non-mass-like nerve root involvement that was detected with MRI. In addition, the nerve conduction study excluded peripheral neuropathy. It suggested that inflammation of the nerve root was caused by limited hematogenous spread without direct invasion or axonal damage of peripheral nerves.

References


Figure legend

Fig. 1.
Chest X-ray on admission.

Fig. 2.
MRI of the intrathecal nerve roots before therapy show intrathecal nerve root swelling and enhancement predominantly in the left region at C6 level mainly. 
(A)-(C) Axial MR images at C6 level. (D)-(F) Sagittal MR images. (A) (600/15[TR/TE]) and (D) (630/14[TR/TE]) are T1-weighted image; (B) and (E) are contrast-enhanced T1-weighted images; (C) (4700/108[TR/TE]) and (F) (4000/115[TR/TE]) are T2-weighted images.

Fig. 3.
MRI of the intrathecal nerve roots after therapy. Nerve root swelling and enhancement improved with oral steroid therapy. T1-weighted MR image (700/13[TR/TE]) (A). Gd-enhanced T1-weighted image (700/13[TR/TE]) (B).