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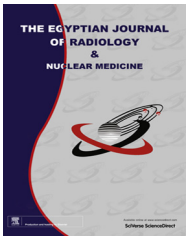

Hirayama disease in unilateral and bilateral forms-3 case reports

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KEYWORDS

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Abstract Hirayama disease is a benign, nonprogressive motor neuron disease affecting the upper limbs. It is secondary to an abnormal anterior displacement of the posterior dura with secondary compression of the lower cervical spinal cord. It should be suspected in young male patients with a chronic history of weakness and atrophy involving the upper extremities followed by clinical stability in few years. The involvement is usually unilateral but may be bilateral. MR imaging is the best way to make the diagnosis but it necessitates the use of both extension/flexion and post contrast studies.

1. Introduction

Hirayama disease, also known as monomelic amyotrophy or benign focal amyotrophy, is a rare disease affecting primarily males of Southeast Asian decent in their second to third decades (1). It differs from the other motor neuron diseases because of its nonprogressive nature and pathological findings of focal ischemic changes in the anterior horn cells of the localized lower cervical cord. It is characterized by an insidious onset of distal muscular atrophy of the upper limb involving the hand and the forearm. The disease process is unilateral in most patients, asymmetrically bilateral in some and rarely symmetric. This is a lower motor neuron disease causing both acute and chronic denervation identified on EMG in the C7-T2 myotomes. There are generally no sensory deficits although paresthesias may be seen. The gradual onset of purely motor disturbances may mimic early amyotrophic lateral sclerosis. This latter diagnosis may be excluded as it involves a pyramidal tract and is progressive without clinical stabilization. The

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clinician should be aware of this disease and treat it promptly in order to prevent progression of the cord atrophy. We present three cases of Hirayama disease with unilateral and bilateral involvement.

2. Case report 1

An 18-year-old man presented with severe disability of the left hand. He had a four year history of weakness of the left hand which was gradually progressive. He had muscular atrophy in the intrinsic muscles of the left hand and in the distal muscles of the left forearm. A clinical diagnosis of amyotrophic lateral sclerosis was made earlier keeping in view the progressive nature of the disease. However, the patient had spontaneous stabilization of the symptoms and the characteristic MR findings changed the diagnosis to Hirayama disease.

3. Case report 2

A 20-year-old man presented with weakness and a gradual loss of muscle bulk of the right distal upper limb for the past 5 years. It was earlier progressive but now is stable for the past one year. He denied weakness of any other limb. On clinical examination, atrophy of small muscles of the hand and distal forearm was appreciable. No conduction block was seen on EMG. MR confirmed the clinical suspicion of Hirayama disease.

4. Case report 3

A 28-year-old man complained of progressive weakness and atrophy of both distal upper limbs for the last 8 years. The involvement was bilateral, however, more on the right side. There was muscular atrophy. Clawing of the right hand was also noticed. On EMG, there was a low amplitude of bilateral median and ulnar nerves.

In all these three cases, there was no neck pain or radicular symptoms. No sensory signs were seen. Results of routine blood analysis were normal. Family history was negative for neuromuscular disorder.

MR imaging was done in the neutral position and neck flexion in all these three cases. Post contrast study was also done. Straightening of the cervical cord, localized cord atrophy at C5–C7 level, T2-weighted hyperintensity due to myelomalacia, widened epidural space on neck flexion with T2-weighted flow voids and post contrast epidural venous congestion involving the lower cervical cord were seen in all the three patients (Figs. 1–3). Case 1 had involvement of the left upper limb and the imaging showed the left sided pear shaped cervical cord on axial images (Fig. 1B). Case 2 showed bilateral asymmetric cord flattening (right > left), although clinically the weakness was limited to the right side only (Fig. 2B). High signal intensity on T2-weighted imaging was best appreciated in this case (Fig. 2A). Case 3 showed cord flattening which was symmetrical (Fig. 3C). Here the patient had bilateral asymmetric involvement of distal upperlimbs.

All the three patients were managed conservatively with cervical collar, muscle strengthening exercises and training in hand-coordination. The two patients having unilateral involvement benefited the most. However, the third patient having bilateral asymmetric involvement has to undergo tendon reconstruction for the clawing of hand. This emphasizes the importance of early recognition of the disease so that clinical progression can be halted early.

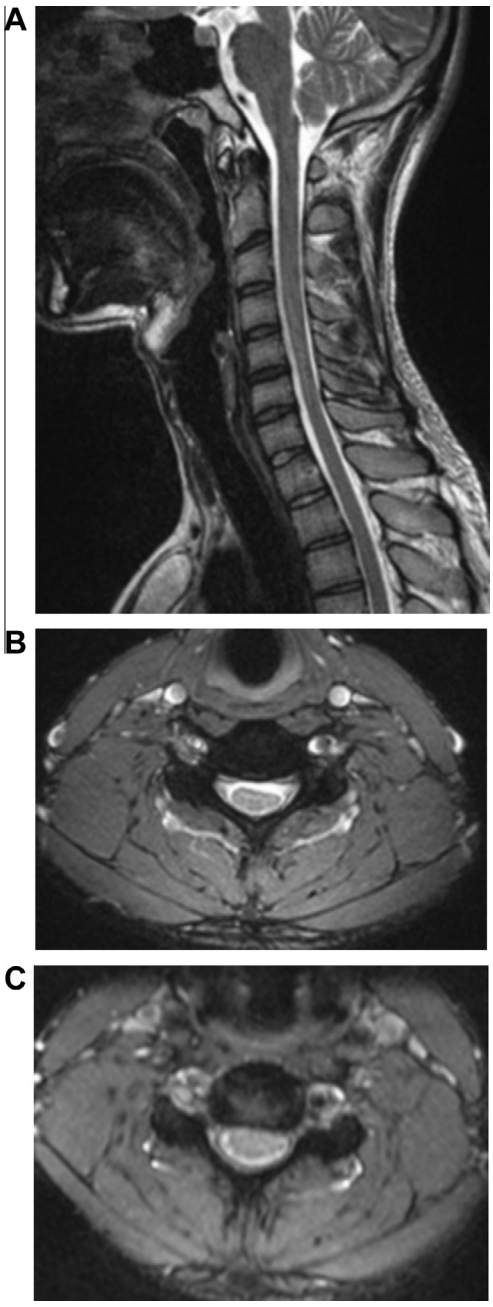

Fig. 1 An 18-year-old male with Hirayama disease having weakness and muscle atrophy of the distal forearm and hand on the left side. (A). Sagittal T2-weighted image in neutral position shows straightening of cervical curvature with focal cord atrophy at C5 and C6 levels. (B). Axial T2-weighted image in neutral position at C6 level shows asymmetric anterior cord flattening on the left side. High T2 signal intensity of the cord is also seen secondary to cord compression. (C). Axial T2-weighted image in neck flexion shows forward displacement of the posterior dural wall, more prominent at C5 and C6, with widening of the posterior epidural space.