Hip dysplasia associated with a hereditary sensorimotor polyneuropathy mimics a myopathic process

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Abstract

Some orthopedic complications have been reported in the hereditary neuropathies. However, the association of the hip dysplasia with this category of neuropathy is rarely recognized. We present a 13-year-old boy with the progressive weakness of the lower extremities, difficulty in walking, climbing stairs, and rising from floor; a wide-based, hyper-extended and waddling gait similar to a myopathic process. Hip radiography showed dysplastic acetabulae with hip subluxation, broken Shenton's lines, and valgus femoral necks. In electrodiagnosis, there was a significant neuropathic process (absent all evoked sensory potentials, abnormal evoked motor responses, and neurogenic electeromyography) which eventually was found to be a hereditary mixed axonal and demyelinating sensorimotor polyneuropathy with concomitant hip dysplasia confirmed with thorough physical examination and the electrodiagnostic study. In patients with gait difficulties such as waddling gait mimicking a myopathic process, hereditary polyneuropathy complicated with hip dysplasia should be considered as well.

Key Words

Charcot-Marie-Tooth disease, hereditary motor and sensory neuropathy, hip dysplasia

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Introduction

Hereditary neuropathies account for approximately half of the undiagnosed peripheral neuropathies referred to major medical centers. The most common hereditary neuropathy falls into the category of Charcot-Marie-Tooth disease. Some orthopedic complications have been reported in hereditary neuropathies such as scoliosis, pes cavus, and recurrent dislocation of the patella. However, the association of the hip dysplasia with hereditary neuropathies has been rarely recognized.[1-5]

Here, we present a boy with the waddling gait, difficulty in climbing stairs, and rising from floor. He was initially referred with the suspicion of a myopathic process but was eventually diagnosed as a hereditary mixed axonal and demyelinating sensorimotor polyneuropathy complicated with hip dysplasia.

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Case Report

A 13-year-old boy was referred to an electrodiagnostic clinic for the evaluation of a probable myopathic process. He presented with the progressive weakness of the lower extremities which was initiated 8 years prior to our examination. He had difficulty in walking, climbing stairs, and rising from floor. His family history had no significant finding.

With a physical examination, he exhibited a wide based, hyper-extended and waddling gait. He was also unable to jump or perform heel walking, hopping, and toe walking. The Trendelenburg and Gower signs were positive. Additionally, atrophy of the pelvic girdle muscles, limitation of motion of hip joints (especially in extension and abduction), achilles tendon shortening, and slight flexion contracture of hip joints were present. Manual muscle testing was normal except for the bilateral pelvic girdle muscles, bilateral ankle dorsiflexors, and big toe extensors that were 2/5 (the patient has the partial range of motion in the gravity eliminated position). Impaired vibration and light touch were found in the sensory exam.

Routine laboratory tests such as complete blood count, liver function test, fasting blood sugar, urinalysis, and creatine phosphokinase were within normal limits. Only, lactate dehydrogenase was slightly elevated. The result of biopsy

Table 1: The summary of electrodiagnosis

Test	Result
Sensory examination	Absent sensory nerve action potential with the stimulation of the sural, superficial peroneal, median, ulnar, and superficial radial nerves
Motor examination	Absent compound muscle action potential with the stimulation of the Tibial and common peroneal nerves.
	Left median nerve (recorded from Abductor pollicis brevis muscle): Wrist latency: 10.1 ms, elbow latency: 15.3 ms, amplitude (base to peak): 0.4 mv, conduction velocity: 29 m/s
	Right ulnar nerve (recorded from Adductor digiti minimi muscle): Wrist latency: 7.3 ms, below elbow latency: 12.1 ms, amplitude (base to peak): 0.3 mv, conduction velocity: 30.4 m/s
F-wave	Absent with the stimulation of the Median, Ulnar, Tibial, and common peroneal nerves.
H- reflex	Absent with both tibial nerves stimulation
Needle electromyography	Right and left peroneous longus, left gastrocnemious, right tibialis anterior, right gluteous medius, right gluteous maximus, left first dorsal interosseous, and right abductor pollicis brevis muscles:
	No spontaneous potentials, highly polyphasic motor unit action potentials with high amplitude and long duration and discrete recruitment pattern.
	Left quadriceps, right deltoid and left biceps muscles:
	no spontaneous potentials, slight increased polyphasic motor unit action potentials with high amplitude and long duration and decreased recruitment pattern.

ms = Millisecond; mv = Millivolt; m/s: Meter per second

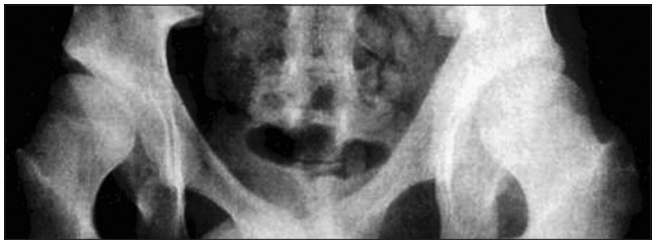

Figure 1: The pelvic radiograph showed dysplastic acetabulae with hip subluxation, broken Shenton's lines and valgus femoral necks

from quadriceps muscle was not significant and showed mild random atrophy of fibers. An ATPase reaction revealed affection of both type I and type II fibers. The periodic acid-Schiff stain did not show any significant change in stainable glycogen. Brain CT-Scan was also normal.

In our electrodiagnostic study, all evoked sensory potentials were absent. A motor nerves evaluation showed absent-evoked potentials of all routine distal lower limb muscles. Other evoked motor responses had significant increased distal latencies, decreased amplitudes, and decreased nerve conduction velocities with no sign of temporal dispersion or conduction block. All F-waves (F from foot) and H-reflexes (Hoffmann-reflexes) were absent. In the needle electromyography, there were polyphasic motor unit action potentials with high amplitude and long duration especially in the distal muscles [Table 1].

The above findings suggested the impression of a hereditary mixed axonal and demyelinating sensorimotor polyneuropathy (such as X-linked Charcot-Marie-Tooth disease). Additionally, a requested plain pelvic radiography showed bilateral hip dysplasia [Figure 1].

Discussion

The association of hip dysplasia with the hereditary sensorimotor polyneuropathy was initially described by Kumar *et al*.[2] Later, investigators have found that hip dysplasia has a prevalence of about 10% in this category of neuropathy.[3,4,6]

The reason of this association is unclear. However, muscle weakness around the hip joint has been suggested as a cause. It usually presents in the second or third decade of life and is initially asymptomatic but may later present with pain and gait abnormalities.[7,8]

Interestingly, our patient had the signs and symptoms resembling a myopathic process and was referred for its confirmation with an electrodiagnostic study. However, these problems were eventually found to be related to a hereditary mixed axonal and demyelinating sensorimotor polyneuropathy with complicated hip dysplasia.

Therefore, this presentation reveals that the neuropathy may not always be an exclusively distal process as is often supposed.[4]

Therefore, in patients with gait difficulties such as wadding gait and Trendelenburg lurch, besides a myopathic process hereditary polyneuropathy complicated with hip dysplasia should be considered as well. Early diagnosis is essential to avoid serious morbidity associated with the condition.

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