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Case Study

Brachial Plexus Palsy Induced by Charcot Marie Tooth Disease

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Abstract

Charcot-marie-tooth disease (CMT) is an inherited neurological disorder, also known as hereditary motor and sensory neuropathy (HMSN) or peroneal muscular atrophy, comprises a group of disorders that affect peripheral nerves. There is a mutation in the gene that produce a protein which involved in the axon structure or myelin sheath. This results in abnormality in signal transfer through the nerve which causes impairment in the sensomotor function of the peripheral nerves. There are different subtypes of the disease according to the type of mutation. Some types like cmt3 causes severe neuropathy which begins in infancy and has poor prognosis. other types start in adolescence and has variable prognosis. We would like to report a case of brachial plexus palsy induced by Charcot marie tooth disease type CMT1A which is known as hereditary neuropathy with predisposition to pressure palsy (HNPP).

Introduction

28 years old male (soldier) presented with sudden loss of sensation of his left arm, forearm and hand in addition to loss of shoulder abduction, elbow flexion and severe weakness of his hand, wrist and finger flexion/extension.

The symptoms started immediately while he was in training for shooting targets at military using gun which should be stabilized on his left shoulder while shooting. The symptoms got severe few hours after the training finished. The patient doesn't have any similar history before and none of his family members had any similar disease or problems as per his knowledge.

Clinical Examination

He presented to emergency department with the following examination

- C5 root palsy (absent shoulder abduction)
- C6 root palsy (absent elbow flexion)
- C7 root partial palsy (grade 3 elbow extension/weak finger extension grade 3)
- C8/T1 roots partial palsy (weak wrist /fingers flexion grade 3)

He had as well numbness of the whole upper limb with loss of pain sensation mainly in the arm and forearm. No abnormality detected in right upper limb or lower limbs neurological exam.

Work Up

The patient had shoulder x-ray to rule out any shoulder abnormality which came normal a CT scan of the shoulder was requested as well and showed: There is a small soft tissue linear calcific density at the anterior inferior aspect of the humeral head measures 5 mm. Differential include a small loose body or related to the anterior inferior glen-humeral ligament humeral avulsion. No depressed or displaced fracture noted. Glen humeral and AC joint articulations are congruent

We requested also for EMG which showed: Bilateral median, ulnar, peroneal and radial nerves showed prolonged distal latencies with normal CMAP amplitudes WITH SLOW CVs along with prolonged minimal F latencies and waveforms.

And since the palsy was induced by pressure we requested microarray analysis for the Charcot Marie Tooth disease which came as following: Microarray analysis revealed a gain of the short arm of chromosome 17. The deletion in this individual is 1.3 mega bases (Mb) in size. The duplication and deletion of PMP22 causes charcot marie tooth disease type 1A (CMT1A) and hereditary liability to pressure palsies (HNPP), respectively.

Comment

Hereditary Neuropathy with liability to Pressure Palsies or HNPP is a slowly progressive, hereditary, neuromuscular disorder which makes an individual very susceptible to nerve injury from pressure.

Recommendation

Clinical correlation between this result and the patient's phenotype is recommended. Genetic counseling and genetic study on family members recommended to discuss the implications of this result.

Management

The patient was referred to the physical therapy unit for rehabilitation and he gained full function and sensation in two months' time. after that he had similar episode caused by military training as well but it lasted only for less than 24 hours. The patient was given recommendation for mechanical pressure free work and regular follow up every 6 months.