Idiopathic Median Mononeuropathy in a Previously Healthy Child: The Usefulness of Magnetic Resonance Imaging in Making the Diagnosis

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Mononeuropathy is rare among children. Deymeer and Jones [1] reported that 6% of children (78 out of 1,319) referred to their pediatric electromyography (EMG) laboratory within an 11-year-period (1979 to 1990) were diagnosed with mononeuropathy, and 17 patients were diagnosed with pediatric median mononeuropathy over 14 years (1979 to 1994). A more recent paper from Davis and Vedanarayanan [2] reported their experience diagnosing 13 pediatric patients with carpal tunnel syndrome, or median neuropathy at the wrist, within an 11-year-period, from 1,800 children who underwent EMG-nerve conduction study (NCS). Carpal tunnel syndrome is a clinical syndrome where the patient experiences a multitude of symptoms including tingling or numbness of the hand, pain in hand, wrist, or forearm, and reduced grip strength, believed to be caused by compression of the median nerve in the carpal tunnel [3]. Causes of carpal tunnel syndrome in pediatric patients include lysosomal storage diseases like: mucopolysaccharidosis and mucopolipidosis, congenital bone anomalies, hypothyroidism, or myopathic contractures [4].

A previously healthy 33-month-old girl came to the hospital complaining of weakness in her right hand. Her mother noticed the weakness a

week prior when she held the patient's hand while walking. In some photos taken of her at daycare, it was identified that the patient had transitioned to primary usage of her left hand, despite her right-handedness; suggesting that symptoms, like weakness, may have been prevalent before the prior week. When asked by clinicians, the patient denied symptoms of weakness in other parts of her body. She was unable to use her left hand properly, suggesting her having pathological left-handedness was less likely. No history of trauma or recent infections was found, rather, the patient had been in good condition, sleeping and eating well. Her immunization records were up-to-date and development normal. She didn't receive vaccination recently.

A physical examination at the time of admission revealed weakness of the right hand. She was unable to grasp and perform thumb opposition motions. The resulting motor grades for her thumb, second finger, and third finger were rated as grade I, and her fourth and fifth finger were rated as grade V, whereas the motor strengths of all other extremities were reported normal. Sensitivity to pain was not impaired in either hand, although other sensory testing was limited due to her age. Reflex testing revealed normal for bilateral biceps, brachioradialis and triceps deep

tendon reflexes, and Babinski reflex was negative. Other physical examinations were unremarkable.

Simple radiograph imaging was performed on her bilateral forearm and revealed no significant bony abnormalities. NCS of the right arm generated a normal response for both the sensory and motor components in the right median and ulnar nerves, although F-waves were not observed when stimulating the right median nerve. Magnetic resonance imaging (MRI) of the right wrist with contrast enhancement showed diffuse T2 signal alterations, with swelling of the distal median nerve from distal radius level, and no evidence of extrinsic nerve compression in the covered range. This suggests a median nerve neuropathy of inflammatory origin. Furthermore, increased T2 signal intensity in the pronator quadratus, abductor pollicis brevis, flexor pollicis brevis, opponens pollicis, and 1st, 2nd, and 3rd lumbricals muscles was observed without any definite atrophic changes, suggesting acute denervation changes of the median nerve.

Diagnosis of idiopathic median nerve mononeuropathy was established given the absence of both trauma history and evidence of

extrinsic nerve compression. She received steroid pulse therapy with intravenous methylprednisolone (30 mg/kg/day) for 3 days. There were no reported side effects and weakness improved on the 4th day of treatment. She was discharged the following day and followed up on at an outpatient clinic a week later. During the outpatient visit, motor strengths of all digits of her right hand were rated grade V, and she showed no other symptoms. (Fig. 1).

Even though the terms carpal tunnel syndrome and median mononeuropathy in children are used interchangeably in much of the literature, carpal tunnel syndrome is a specific form of median nerve mononeuropathy, which affects the distal portion of median nerve and is caused by increased pressure on the carpal tunnel. Causes of carpal tunnel syndrome are in most cases unknown, and suspected risk factors include diabetes mellitus, hypothyroidism, obesity, and arthritis [3].

Anatomically, the distal median nerve was involved in this case; however, MRI indicated no evidence of extrinsic nerve compression, making a carpal tunnel syndrome diagnosis unlikely. Since there was no history of infection or any preceding trauma, the pa-

Fig. 1. Magnetic resonance imaging T2 fat saturated sequence with contrast enhancement of left wrist showing (A) swelling of the distal median nerve (arrow) and (B) increased T2 signal intensity in the pronator quadratus (arrowheads), and (C, D) abnormal T2 hyperintensity without definite atrophic changes on thenar muscles (arrow and arrowheads).

tient was diagnosed with idiopathic pediatric median mononeuropathy.

In contrast to carpal tunnel syndrome, where sensory symptoms such as numbness, tingling, burning, or pain are considered to be characteristic symptoms, the patient's chief complaint was weakness of her right hand. Since the patient appeared to be irritated until she was with methylprednisone pulse therapy, this makes it hard to rule out the possibility of her not being able to express her sensory symptoms. A pediatric patient's inability to effectively communicate their symptoms decreases pediatricians' ability to detect rare conditions that benefit from early detection and appropriate management.

Traditionally, the gold standard for diagnosing median mononeuropathy is NCS. The sensitivity and specificity of NCSs of carpal tunnel syndrome patients vary from 75% to 96.7% depending on the study [5]. Since these studies are based on patient data from an adult population, there are clear limitations to extrapolating the results found in these studies to pediatric cases. With technological improvements to MRI technology, MRIs are used increasingly for diagnosing carpal tunnel syndrome, especially to elucidate any anatomical abnormalities. In this case, NCS failed to reveal any abnormalities in the patient's median nerve. This may have been a false negative due to the acute nature of the illness. However, a MRI of the wrist revealed evidence of median nerve neuropathy. Currently, there is no research that directly compares the sensitivity and specificity of MRI and NCS examinations as a means of diagnosing neuropathies, but as shown by this case, an MRI might be a useful tool for diagnosing neuropathies more than has been previously reported.

In summary, this case represents a pediatric patient with idiopathic mononeuropathy of the median nerve and who was diag-

nosed with clinical features and imaging studies (standard X-ray and MRI), and responded well to methylprednisone pulse therapy.

This study was conducted with verbal informed consent from her parents.

Conflicts of interest

No potential conflicts of interest relevant to this article was reported.

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