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Pediatric Chronic Inflammatory Demyelinating Polyradiculoneuropathy: A Case Report of a Rare Entity

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CASE DESCRIPTION

Our patient is a 12-year-old male with no past medical history, normal birth history, and no significant family history who presented to our outpatient clinic with generalized weakness of four months duration. He developed sudden onset numbness and weakness of both hands to where he could not open soda bottles or hold a cup. The weakness progressed proximally in the upper extremities and also in the distal lower extremities. He complained of associated dysesthesias, double vision, and gait ataxia. He denied pain of the extremities or difficulty breathing, swallowing or speaking. He never experienced bowel or bladder incontinence.

He was hospitalized after initial onset of symptoms. Nerve conduction study/electromyography (NCS/EMG) performed in the hospital showed largely normal amplitudes, but some mildly slowed conduction velocities and delayed distal latencies, with increased fibrillations on EMG. He was diagnosed with Guillain-Barre Syndrome (GBS) and received 2g/kg of intravenous immunoglobulin (IVIG) therapy. He noticed improvement after treatment and discharged home at a functionally independent level. His hand grip strength improved, however his gait imbalance persisted.

Three months later, he noticed worsening double vision and progressive upper worse than lower extremity weakness. He was started on pulse steroids and monthly IVIG therapy outpatient. He experienced minimal improvement despite treatment and was referred for a follow-up EMG.

ASSESSMENT/RESULTS

He presented for follow-up EMG study four months after initial onset of symptoms. Physical exam revealed cranial nerve sixth palsy on the right. Muscle strength testing revealed 5/5 bilateral shoulder shrug, 3+/5 in the bilateral upper extremities, and 4/5 in the bilateral lower extremities. Deep tendon reflexes were trace in the knees and ankles, 1/4 at the brachioradialis and triceps, and absent in the biceps bilaterally. Sensation to light touch was intact throughout. Romberg test was positive.

Repeat NCS/EMG showed the following: There was slowing noted of the sural sensory responses with poor persistence. The right radial, median, and ulnar sensory responses were delayed with slowed velocities. The median and ulnar motor responses revealed slowed velocities with significant axonal block noted in both nerves. The right peroneal and tibial nerves also revealed slowed velocities. F-waves were absent for the tibial and peroneal nerves. Ulnar nerve F-wave showed slowed latency with very poor persistence. EMG revealed dense fibrillations and decreased recruitment in the right upper extremity muscles. The proximal right lower extremity had normal insertional activity and recruitment. However, the distal muscles revealed some fibrillations with decreased recruitment. Evidence of demyelination and conduction blocks seen in both upper and lower extremities, along with fibrillations and decreased recruitment, was most consistent with the diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP).

DISCUSSION

CIDP classically presents with neurologic symptoms, including progressive weakness, pain, paresthesias, absent tendon reflexes, cranial nerve involvement, and ataxia. This autoimmune disease has a predilection for older males, with onset gradually developing over an eight-week course. Pediatric CIDP is rare, however it is the most common acquired treatable polyneuropathy in children. Uniquely, children have variable presentations, which can cause delay in diagnosis and treatment. Limited case reports document presentations of acute, relapsing-remitting courses, with symptoms being more severe than compared to adults. Treatment options include immunoglobulin therapy, corticosteroids, plasma exchange, immunosuppressives, and immunomodulatory agents.

Our patient exhibited an atypical course of CIPD. This was complicated by CIDP refractory to IVIG therapy. Literature review shows patients with IVIG refractory CIDP will show some response to alternative treatment, such as steroids. However, our patient also received pulse steroids without improvement. There are case reports documenting presence of anti-neurofascin antibodies in patients with IVIG refractory CIDP. Interestingly, some of these patients presented with acute onset of symptoms resembling GBS, much like our patient. This unique patient population has been shown to be responsive to rituximab. Our patient is currently being tested for anti-neurofascin antibodies and was empirically started on rituximab with close clinical monitoring, along with continuation of steroid and IVIG therapy.

CONCLUSION

This patient presented with rapid onset of symptoms, significant weakness of the upper greater than lower extremities, and minimal improvement over time. Diagnosis was challenging and ultimately confirmed four months later on EMG/NCS. Early recognition of CIDP is key as rapid initiation of treatment can prevent irreversible nerve damage and functional disability. This case serves to highlight a rare case of pediatric CIDP and aid clinicians in early diagnosis of this disease.

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