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A Girl with Atypical Chronic Inflammatory Demyelinating Polyneuropathy

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Chronic inflammatory demyelinating polyneuropathy (CIDP) is a chronic, acquired immune and inflammatory disorder that targets the peripheral nerves. The cardinal features include a progressive or a relapsing-remitting course, predominant motor symptoms and signs, symmetrical involvement of arms and legs, proximal muscles involvement along with distal muscles, and decrease or absence of deep tendon reflexes. The diagnosis is confirmed by cerebrospinal fluid (CSF) protein elevation without pleocytosis, and nerve conduction evidence of a primary demyelinating polyneuropathy.

A 17-year-old girl was admitted with increase in falling and progressive difficulty in raising arms for 6 months. The weakness had a waxing and waning course for the past few months. One month before admission, she noticed diurnal variation of weakness with most severe weakness in the morning that usually got better in the afternoon. She complained of frequent shoulder pain, fluctuating limb weakness and chronic fatigability. Examination showed multiple sites of tenderness, neck and shoulder stiffness and fatigability. Rapid fluctuation of muscle weakness within the same day or within 1 to 2 days were observed. She was initially suspected to have fibromyalgia and she had slightly elevated erythrocyte sedimentation rate and positive for anti-dsDNA. Subsequent nerve conduction study confirmed demyelinating sensorimotor polyneuropathy with sparing of sural nerves. Lumbar puncture showed raised protein level and protein-cytological dissociation. MRI spine demonstrated gadolinium contrast enhanced nerve roots at the cauda equina. Chronic inflammatory demyelinating polyneuropathy was diagnosed. She was started on intravenous immunoglobulin with rapid clinical improvement.

Our patient demonstrated a close relationship between fibromyalgia and CIDP. The predominant presentation of fibromyalgia highlighting that neuropathic nature of pain and morning stiffness can be the atypical presentation at some stage of CIDP.