Erythromelalgia due to Autoimmune Small Fiber Neuropathy: A Case Report

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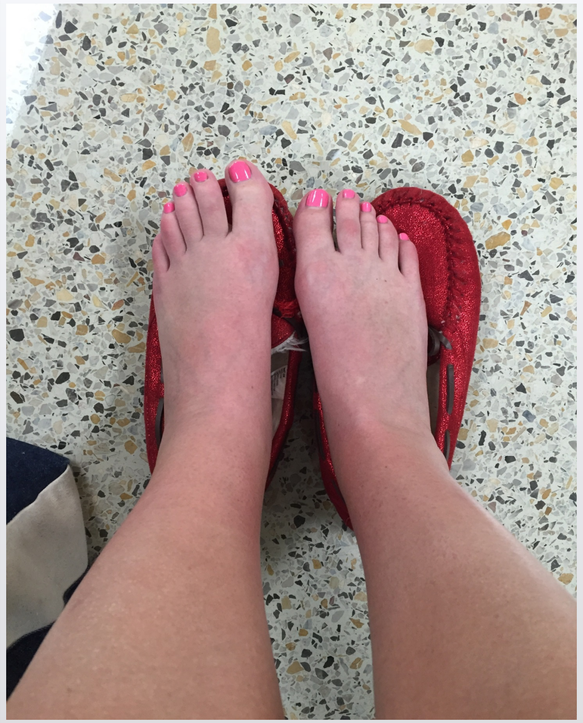

Figure 1: Erythema of lower extemities

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INTRODUCTION

The classic presentation of GBS is that of AIDP, presents with progressive back pain, paresthesia, symmetric ascending weakness and hyporeflexia preceded days to weeks by an illness is easily made and treated. Nerve conductions studies (NCS) can help confirm the diagnosis, but can also detect variants from classical AIDP that may be useful in prognosis and need for further work. Most notably the difference between axonal and demyelination variants.

Sometimes diagnosis is not always forthcoming when it comes to GBS, especially if it does not present as the classic AIDP. In the case below we describe an uncommon presentation of an emerging variant of GBS and the successful treatment of a 31-year-old with acute erythromelalgia thought to be caused by a small fiber neuropathy variant of GBS. The presentation, clinical findings and work up of this case is similar to a few cases found in recent literature review, some calling this a variant of GBS [9] and others labeling this a autoimmune small fiber axonopathy.

METHODS

Case report with supporting laboratory and electrophysiological data.

CASE DESCRIPTION

A 31 year old woman presented with eight weeks of progressive, symmetric painful burning, swelling, and erythema of the bilateral distal lower extremities, exacerbated by hot water, sock or shoes, and relieved by submerging feet in a cold bath. Gabapentin was ineffective. Physical exam disclosed erythema and edema of the feet in areas covered by footwear, with severe allodynia to mid-calf without hypesthesia and with normal strength and reflexes. Nerve conduction studies demonstrated patchy mild sensory axon loss. Extensive evaluation including serum and CSF laboratory studies and body imaging revealed no evidence of secondary causes of erythromelalgia. The patient was diagnosed with SFN. Given the time-course and the absence of treatable secondary causes of her disorder, she was offered treatment with IVIg for presumed autoimmune neuropathy. Marked improvement in pain, allodynia, and erythema was noted during treatment, which has persisted for 2 months of ongoing treatment with IVIg, gabapentin, and duloxetine.

DISCUSSION

Erythromelalgia is an uncommon presentation of acute SFN, particularly in the absence of underlying secondary cause. Prior reports have suggested an autoimmune etiology for this presentation; robust improvement after initiation of immunomodulatory therapy is supportive of an autoimmune etiology and raises the possibility of acute SFN as another primarily axonal variant of Guillain Barre Syndrome.

TABLE 1: Results of significant laboratory studies

Test	Results
-***Initial Work Labs:*** *, ESR, C-Reactive Protein, Rheumatoid Factor, Iron, Ferritin, Urine Protein Electrophoresis W/Reflex IFE, Methylmalonate, Hemoglobin A1c, Thyroid Stimulating Hormone, Thyrotropin, Vitamin B12, Folate, HIV-1/O/2 Ab Porphobilinogen, Porphobilinogen/Creatinine, Tryptase,*	(-)
-***Autoimmune***: Neuronal nuclear Type 1 Ab, Purkinje Cell Type 1 Ab, Neuronal Nuclear Type 2 Ab, Myeloperoxidase Proteinase 3 Ab, Neutrophil Cytoplasmic Ab C-ANCA, Neutrophil Cytoplasmic Ab Perinuclear, Neutrophil Cytoplasmic Ab Atypical, Extractable Nuclear Ab Panel, Ribonucleoprotein Extractable Nuclear Ab,Smith Extractable Nuclear A, SS-A Ab, SS-B Ab, SCL-70 Extractable Nuclear Ab,	(-)
-***Infectious:*** Cytomegalovirus Ab IgM, Hepatitis Virus Panel Chronic and Acute), HIV	(-)
-CSF WBC -CSF RBC -CSF Glucose -CSF Protein	0 0 62 mg/dl (40-70 mg/dl) 22 mg/dl (15-45 mg/dl)
-***Antiganglioside Antibodies***: IgG Monos. GM1, IgM Monos. GM1, IgG Asialo. GM1 , IgM Asialo. GM1, IgG Disialo. GD1b, IgM Disialo. GD1b , Anti-GQ1b (IgG)	(-)

TABLE 2: Results of significant radiology studies

Imaging & EDX	Results
Nerve conduction study	evidence suggestive of possible sensory axonal loss, mostly normal
Electromyography	unremarkable
MRI Brain with and without contrast	Unremarkable
MRI C/T/L-spine without contrast	Unremarkable
CT Chest/Abdomen/Pelvis with/without contrast	No evidence of malignancy

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