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Case Report Open Access

Chronic Demyelinating Inflammatory Polyradiculoneuropathy Associated With Sarcoidosis

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Abstract

Sarcoidosis is an inflammatory multisystem disorder, usually involving the lung, the skin, the lymph nodes, and the eyes. The prevalence of clinical involvement of the nervous system is estimated to be about 5% to 15% [1]. Both central and peripheral nervous system can be affected. Sarcoid polyneuropathy is a rare and clinically heterogeneous disorder that may be the initial presentation of sarcoidosis [2]. Only a few cases have been reported of chronic demyelinating inflammatory polyradiculoneuropathy (CIDP) associated with sarcoidosis. We report a clinical course of a patient presented with a sensorimotor demyelinating polyneuropathy with secondary axonal loss few years after being diagnosed with lung sarcoidosis.

Keywords: Sarcoidosis; Polyneuropathy; CIDP; Nerve biopsy

Case Report

A 60-year-old woman, with medical history of lung sarcoidosis diagnosed in 2003 treated by 40 mg prednisolone daily, was admitted in our neurological department in March 2011 for paresthesia and burning sensation in her two hands evolving for five months.

Neurological examination revealed mild distal limb weakness, generalized hyporeflexia, impairment of touch, pain, and temperature sensations in both hands. Vibration sense was severely affected in the higher limbs, especially in the left side. Patient was ataxic with a positive Romberg's sign.

Electrophysiological study demonstrated bilateral elongated distal latencies, reduced nerve conduction velocities, and conduction blocks respectively in the median, the ulnar and the peroneal nerves (Table 1). It also objectified delayed F response in the median, the ulnar and the tibial nerves (Table 2) fulfilling the EFNS/PNS diagnostic criteria for CIDP.

Cerebrospinal fluid (CSF) analysis revealed an elevated protein level (0.54 g/L; normal range 0.16 – 0.4 g/L) without pleocytosis. Magnetic resonance imaging (MRI) of the brain and the spinal cord was normal. Nerve biopsy was not performed because of patient's refusal. Serologies of Lyme, hepatitis B and C and human immunodeficiency virus were negatives. Levels of thyroid hormones, glycaemia and Vitamin B12 were within normal limits. Screening tests for autoantibodies, including antinuclear antibody (Ab), anti-SS A/SS B Ab, anti-cardiolipin Ab, antineutrophil cytoplasmic Abs (P-ANCA and C-ANCA) and tumor markers were also unrevealing.

The patient received initially IV methylprednisolone for 3 days (1gr/day), then oral prednisolone of 60 mg/day for 2 months followed by a very gradual reduction. Thereafter, the patient was maintained on oral prednisolone (40 mg daily) in association with azathioprine (100 mg daily). Slight improvement in the muscle strength was observed.

Nerve/Sites	Lat. (ms)	Amplitude (mV)	Amp.1-2 (%)	Intensity Stim.	Dist. (cm)	Vit. (m/s)
R Median						
Wrist	6.00	1.4	100	60 mA	-	-
Elbow	18.60	0.6	42.85	100 mA	27	21.4
Erb	-	-	-	-	-	-
L Median						
Wrist	8.85	0.9	100	50 mA	-	-
Elbow	43.30	0.1	11	50 mA	-	-
3. Erb		-	-	-	-	-
R Ulnar						
Wrist	4.45	2.4	100	59 mA	-	-

Elbow	14.9	0.7	26.16	59 mA	4	-
Axilla	10.05	0.6	85.71	100 mA	29	27,8
L Ulnar						
Wrist	3.30	1.2	100	84 mA	-	-
Elbow	14.35	0.7	58.33	84 mA	24	21.7
Axilla	19.60	0.4	57.14	100 mA	12	22.9
L Tibial						
Internal malleole	9.90	0.4	100	51 mA	-	-
R Tibial						
Internal malleole	8.25	0.9	100	61 mA	-	-
L Peroneal						
Ankle	5.35	2.2	100	51 mA	-	-
Collar	13.15	1.8	81.81	73 mA	33	42.3
R Peroneal						
Ankle	5.50	0.5	100	55 mA	-	-
Collar	12.05	0.5	100	55 mA	30	45.8

Table 1: Motor conduction velocities.

Nerf	Lat F (Ms)	F-M (ms)
L Tibial	BLOC	-
R Tibial	50.90	52.43
L Median	23.65	24.64
R Median	BLOC	-
L Ulnar	27.30	27.80
R Ulnar	BLOC	-

Table 2: F wave.

Discussion

Peripheral nervous system involvement in sarcoidosis is rare, affecting 2% of patients [3]. Its patterns are varied and include multiple mononeuropathies, focal or multifocal neuropathy, and CIDP as in our case [1].

Typically, systemic symptoms of sarcoidosis precede the neuropathy. In some cases, nervous system involvement can be the presenting form of sarcoidosis. Ducray et al. [4] reported a case of a 36-year-old-man who first presented two relapses of chronic inflammatory demyelinating polyneuropathy (CIDP) before the diagnosis of sarcoidosis was made. In 2015, Singhal et al. [5], reported also the clinical, electrophysiological, and pathological findings of a patient who carried a diagnosis of sensory-predominant CIDP for over a decade but was ultimately found to have sarcoidosis. In cases of suspected peripheral nerve sarcoidosis, biopsy of both nerve and muscle tissue may increase the diagnostic yield and help rule out other rare inflammatory, infectious, or neoplastic causes of neuropathies.

Several explanations for the pathogenesis of neuropathy associated with sarcoidois have been suggested: such as endoneural granuloma, perivascular granuloma and necrotizing epineural vasculitis [5].

Evidence-based guidance for treatment of neurosarcoidosis is limited by its rarity. Glucocorticoids are considered the first-line treatment. In refractory cases a second immune modulator is often added. Our patient did not well respond to steroid but showed considerable improvement with azathiopirine. Fewer than 10% of patients do not respond to any immune modulatory therapy.

Conclusion

Sarcoid polyneuropathy is not only rare, but it also presents with a variety of clinical syndromes which often delay the diagnosis of this challenging disorder. This diagnosis should be considered in every patient with CIDP even in the absence of systemic signs. In cases of suspected peripheral nerve sarcoidosis, nerve biopsy may help diagnose this treatable disorder.

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