Migratory neuropathy: recognizing the snowflake in a sea of anchors

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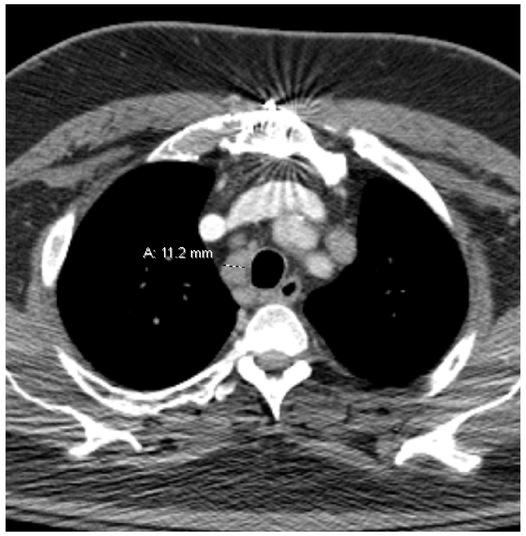
CASE HISTORY

A 55-year-old man with a history of congestive heart failure (EF 35%) and insulin dependent diabetes mellitus presented with 12 weeks of progressive sensory and motor polyneuropathy involving lower extremities, with concomitant Bells palsy. His condition had worsened at home to the point that he was unable to ambulate or care for himself. He was admitted and given a presumptive diagnosis of AIDP. He was treated with 7 days of plasmapheresis without improvement in symptoms, but improved after a short course of steroids. He was discharged to rehab where his bilateral distal lower extremity weakness improved, but his hamstring weakness worsened. He was readmitted after he had sudden onset left upper extremity weakness, at which time the diagnosis of AIDP was reconsidered.

WORKUP

- CSF cell count: 38 WBC, 93% lymphocytes.
- CSF glucose 100, protein 289
- CSF cultures and pathogen panel negative
- Treponema pallidum Ab negative
- No evidence of vitamin B12, B1, folate deficiency
- Autoimmune encephalitis panel negative
- Borrelia burgdorferi Ab negative
- ACE level negative
- MRI brain negative for acute pathology
- MRI C/T/L spine showed disc protrusion with central canal stenosis at C6-7 without cord compression or enhancement
- EMG/NCS: severe peripheral asymmetric neuropathy with axon loss greater than demyelination.
- Hilar lymph node biopsy: granulomatous lymphadenitis consistent with sarcoidosis
- L gastrocnemius biopsy: active neurogenic atrophy
- L sural nerve biopsy: loss of myelinated axons w/axonal degeneration

IMAGING



CT chest abdomen pelvis with contrast: nonspecific mediastinal and bilateral hilar lymphadenopathy

CASE CONCLUSION

Diagnosis: neurosarcoidosis

- Bilateral hilar/mediastinal lymphadenopathy found on chest CT
- Lymph node biopsy consistent with sarcoidosis
- Muscle/nerve biopsy was noncontributory
- Patient was started on steroids with improvement in symptoms
- Rheumatology was consulted, started patient on steroid-sparing therapy
- With treatment, the patient regained his ability to walk

REFERENCES

1. Stern, BJ. Neurologic Sarcoidosis. In: UpToDate, Post, TW (ed), UpToDate, Waltham, MA, 2020.

DISCUSSION

The diagnosis of neurosarcoidosis is supported by:

- Improvement with steroids but not with plasmapheresis
- Patchy, asymmetric peripheral nerve involvement
- Pathology evidence of sarcoidosis elsewhere in body
- Elevated protein on CSF studies
- Improvement with steroids

The diagnosis of neurosarcoidosis is not refuted by negative peripheral muscle/nerve biopsy.

It is important to identify cognitive biases at work in this case:

- Anchoring bias
- Premature closure bias

Sarcoidosis is phenotypically heterogenous, each presentation as unique as a snowflake.

- 5-10% of sarcoidosis presents with neurologic symptoms.
- Most common neurologic presentation is Bells palsy.
- Can affect any part of neurologic axis, causing peripheral neuropathy, myopathy, radiculopathy, seizures, meningitis, hydrocephalus, vasculitis, and hypothalamic dysfunction
- Patchy involvement of disease makes diagnosis by biopsy difficult. Usually a clinical diagnosis.

TAKE HOME POINTS

- Consider sarcoidosis in your differential for a wide variety of neurologic symptoms, especially if more common etiologies have been excluded.
- Constantly re-evaluate your original diagnosis, especially if it no longer fits the evidence available.
- Beware anchoring and premature closure biases.
- Neurologic sarcoidosis is difficult to diagnose on nerve/muscle biopsy and is typically a clinical diagnosis.

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