Spider Bite Monoparesis

Carrell-Krusen Neuromuscular Symposium
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and Ian Butler, MBBS
HPI

- 12 yr female with right arm cellulitis 1 month prior to presentation with acute onset of left arm monoparesis, swelling, and decreased sensation.
- Awoke with circumscribed erythematous patches on left arm and left cheek that were draining purulent material
Patient noted multiple small, black fuzzy spiders in her room the night prior to presentation.

Review of systems significant for 3 episodes of non-bloody vomiting on the morning of admission and **left arm pain of 2 days duration.**
Other Relevant History

PSH: None

Medications: None  Allergies: NKDA  Immunizations: Up-to-date

Birth History: Good prenatal care with no pregnancy complications. Born at term by SVD.

Social History: Lives with mother, step-father, brother and sister. No recent travel, no sick contacts. Has straight A grades in 7th grade.

Family History: Mother had prior ectopic pregnancy. No neurologic disorders.

Developmental History: No developmental delays
Neurological Exam on Admission

**General:** Active, alert, well-developed, well-nourished female

**Cranial Nerves:** CN II-XII intact. PERRLA, EOMI.

**Motor:** RUE 5/5 strength, **LUE 0/5 strength**

**Sensory:** hypoesthesia of left arm compared to right arm
- Right arm: light-touch/pain/temperature modalities intact.
- **Left arm:** pinprick and temperature absent over arm, light-touch intact on left shoulder.

**DTRs:** 2/4 except areflexia of left arm

**Coordination and Gait:** No ataxia, normal gait.
MRI (STIR Protocol) - Increased T2 signal in left lower neck and left superior thoracic cavity
Enhancing soft tissue mass within left inferior neck between scalene muscles, extending to lateral chest wall.
Brachial Plexus MRI
Humerus MRI w/wo contrast showing edema and inflammation of the perimysium
Preliminary DDX

- Thoracic outlet syndrome
- Inflammatory disorder
- Infection i.e Lyme disease
- Malignancy
- Brachial neuritis
- Spider bite
- Epidural hematoma
Treated with clindamycin, vancomycin, ceftriaxone, lovenox, triamcinolone cream, and amitriptyline.

Consulted infectious disease, cardiology, rheumatology, dermatology, interventional radiology, and surgery.
EMG

Muscles showing increased insertional activity, fibrillations, and positive waves:
- Flexor carpi ulnaris, abductor pollicis brevis, pronator teres, brachioradialis, extensor indicis, biceps, deltoids.

- Median, radial, ulnar, and axillary nerves + lateral/medial/posterior cords.
  (No C5 nerve root involvement as rhomboid muscle was normal)

- EMG suggestive of brachial plexus injury
Histopathology
Left deltoid muscle

H&E x 100

Gomori trichrome x 100
Muscle and fascia biopsy Gomori trichrome x 100

Edematous fascia with histiocytic infiltration
Edematous fascia with histiocytic infiltration
Acid phosphatase x 100
Skin and subcutaneous biopsy with histiocytic infiltration, H&E x 100
Proposed Mechanism

- Immune mediated allergic response
- Spider venom contains mucopolysaccharides, hyaluronidase, phospholipase, serotonin, histamine, and neurotoxins. Neurotoxins are the most important components of the venom. These low-molecular weight polypeptides cause severe adrenergic and cholinergic activities and affect sodium, potassium, and chloride channels of various cells.
- 1\textsuperscript{st} - IL-1 and TNF-\(\alpha\) mediated inflammation
- 2\textsuperscript{nd} - TH1 cells secrete mainly INF, IL-2, and TNF-\(\alpha\). These cells are responsible for macrophage activation, TH2 induce and activate mast cells and eosinophils in the tissue

Discussion and Conclusion

- Spider venom → systemic inflammatory response → immune mediated reaction leading to brachial plexitis and focal myositis
- Follow up at 7 months shows 75% clinical improvement
MRI on admission

1 week later
Acknowledgments

● Dr. Ian Butler
● Dr. Shade Moody (electromyography)
● Dr. Meenakshi Bhattacharjee (neuropathology)
● Dr. Rajan Patel (neuroradiology)
The End

Any Questions?
References


Extra Slides
## Motor Nerve Conduction:

<table>
<thead>
<tr>
<th>Nerve and Site</th>
<th>Latency</th>
<th>Amplitude</th>
<th>Segment</th>
<th>Latency Difference</th>
<th>Distance</th>
<th>Conduction Velocity</th>
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</thead>
<tbody>
<tr>
<td><strong>Median.L</strong></td>
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<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Wrist</td>
<td>3.2 ms</td>
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<td>APB-Wrist</td>
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<td>Elbow</td>
<td>6.8 ms</td>
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<td>Wrist-Elbow</td>
<td>3.6 ms</td>
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<tr>
<td>Wrist</td>
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<td>ADM-Wrist</td>
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<tr>
<td>Below elbow</td>
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<td>1.9 mV</td>
<td>Wrist-Below elbow</td>
<td>3.3 ms</td>
<td>190 mm</td>
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<tr>
<td>Above elbow</td>
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<td>Below elbow-Above elbow</td>
<td>2.6 ms</td>
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<tr>
<td>Forearm</td>
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<td>Extensor indicis proprius-Forearm</td>
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<td>mm</td>
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<tr>
<td>Lateral brachium</td>
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<td>NR mV</td>
<td>Forearm-Lateral brachium</td>
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<td>m/s</td>
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### Sensory Nerve Conduction:

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<tr>
<th>Nerve and Site</th>
<th>Onset Latency</th>
<th>Peak Latency</th>
<th>Amplitude</th>
<th>Segment</th>
<th>Latency Difference</th>
<th>Distance</th>
<th>Conduction Velocity</th>
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<tbody>
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<td><strong>Median.L</strong></td>
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<tr>
<td>Wrist</td>
<td>2.2 ms</td>
<td>2.7 ms</td>
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<td>µV</td>
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<tr>
<td>Forearm</td>
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<td>15 µV</td>
<td>Snuff box-Forearm</td>
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<td>15 µV</td>
<td>Forearm-A2</td>
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<td>m/s</td>
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<tr>
<td>Elbow</td>
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<td>2.2 ms</td>
<td>6 µV</td>
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<td>2.4 ms</td>
<td>6 µV</td>
<td>Elbow-A2</td>
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<td>mm</td>
<td>m/s</td>
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<td><strong>Lateral antebrachial cutaneous.L</strong></td>
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<tr>
<td></td>
<td>1.9 ms</td>
<td>2.6 ms</td>
<td>10 µV</td>
<td>Elbow-A2</td>
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# Needle EMG Examination:

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<thead>
<tr>
<th>Muscle</th>
<th>Insertional</th>
<th>Spontaneous Activity</th>
<th>Volitional MUAPs</th>
<th>Max Volitional Activity</th>
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<tr>
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<tr>
<td>First dorsal interosseous L</td>
<td>Normal</td>
<td>None</td>
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<td>Reduced</td>
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<tr>
<td>Flexor carpi ulnaris L</td>
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<td>Normal</td>
<td>Reduced</td>
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<td>Abductor pollicis brevis L</td>
<td>Increased</td>
<td>1+</td>
<td>Normal</td>
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<tr>
<td>Pronator teres L</td>
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<td>Brachioradialis L</td>
<td>Increased</td>
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<td>Reduced</td>
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<tr>
<td>Triceps brachii L</td>
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<td>None</td>
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<td>Pronator quadratus L</td>
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<td>Biceps brachii L</td>
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<td>Deltoid L</td>
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<td>Normal</td>
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<td>Reduced</td>
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<tr>
<td>C7 paraspinal L</td>
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# F-Wave Studies

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<thead>
<tr>
<th>Nerve</th>
<th>M-Latency</th>
<th>F-Latency</th>
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<tbody>
<tr>
<td>Median L</td>
<td>6.8</td>
<td>23.1</td>
</tr>
<tr>
<td>Ulnar L</td>
<td>2.3</td>
<td>28.1</td>
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Brachial Neuritis
Brachial Plexus Anatomy
Brachial neuritis: Background

- Inflammatory change involving the brachial plexus
- Affects mainly the lower motor neurons of the brachial plexus, individual nerves, or nerve branches
- Characterized by acute onset of excruciating unilateral shoulder pain, followed by flaccid paralysis and weakness of the shoulder and arm muscles
Brachial neuritis: Etiology

- Traumatic
- Neoplastic and radiation induced plexopathy
- Viral
- Vaccinations
- Systemic
- Immunologic
- Hereditary
- Idiopathic
Brachial neuritis: Pathophysiology

- Brachial neuritis exists in an inherited and an idiopathic form.
- In the idiopathic form, the pathophysiology is unknown, but is thought to be an immune system mediated inflammatory reaction against nerve fibers of the brachial plexus.
  - Herpes simplex, varicella zoster virus\(^9\)
  - Altered lymphocyte subsets\(^10\)
- The inherited form is autosomal dominant and has been linked to mutations in the SEPT9 gene on chromosome 17q25\(^11\)
  - Septins involved in the formation of the cytoskeleton and cell division.
Brachial neuritis: Epidemiology

- Brachial plexitis has been reported in individuals from age 3 months to 74 years, most commonly presenting in the 3rd-7th decades of life
- Prevalence is highest in men (M:F 2:1 to 11.5:1)
- Incidence of brachial plexitis 1.6 cases per 100,000 person-years in the United States\textsuperscript{12}
Brachial neuritis: Presentation

- Abrupt onset of pain at shoulder and upper arm
- Pain may be bilateral in 10-30% of patients
- A minority of patients do not experience the initial painful stage
- Pain intensity is usually very high, lasting from a few hours to several weeks
- As the pain subsides, weakness becomes apparent.
- Affects a wide variety of muscles, usually those innervated by upper trunk (supraspinatus, infraspinatus, deltoid, biceps, serratus anterior)
- Atrophy and reduced muscle strength of affected muscles become prominent after approximately 2 weeks
- Can also present with numbness, reduced reflexes, and sensory loss
- Course of the neuritis is usually gradual improvement and recovery of muscle strength in 3-4 months, although some patients may have several years of muscle weakness or slight permanent weakness
DDx of brachial neuritis

- Cervical spondylosis
- Cervical radiculopathy
- Suprascapular notch masses
- Rotator cuff deficiency
- Shoulder impingement
- Calcific tendonitis
- Adhesive capsulitis
- Disuse atrophy of shoulder girdle muscles
- Quadrilateral space syndrome
- Trauma

- Acute poliomyelitis
- Cervical disc disease
- Mononeuritis multiplex
- Neoplastic brachial plexopathy
- Pediatric HIV infection
- Polymyalgia rheumatica
- Thoracic outlet syndrome
Brachial neuritis: Workup

- Laboratory studies usually within the reference range
- Electrodiagnostic testing (EMG, NCS)
  - Show abnormalities c/w brachial plexus lesions (axon loss)
  - Fibrillation potentials and positive waves suggestive of muscle denervation
- MRI
  - Denervation changes in muscles
  - Muscle initially appears normal
  - Over the next few weeks, high T2 signal develops
  - Gradually, atrophy and fatty infiltration will develop with the increase in T1 signal and decreased muscle bulk
- Ultrasound
BN: Treatment and Management

- No specific treatment
- Typically **self-limiting**, requiring only supportive therapy and conservative management
- Majority of patients have a slow but steady recovery of motor function over 6-18 months
- 90% of patients have excellent recovery by 3 years
- **Physical therapy** to maintain strength and mobility
- **Occupational therapy** for functional conditioning of the upper extremity
- Medical pain management: corticosteroids?\(^{13}\)
- Surgical intervention: nerve grafting or tendon transfers

Focal Myositis

- Rare and benign dysimmune disease that presents as isolated intramuscular mass, or pseudotumor.

- First described in 1977 by Heffner et al.\textsuperscript{7}

“... myopathy affecting a single skeletal muscle without systemic manifestation with a histologically proven inflammatory myositis process” - Flaisler
Etiology Unknown
Proposed mechanisms:

1. Neurologic: denervation in setting of genetic susceptibility, radiculopathy, or nerve lesion.

2. Immune: triggered by dysimmune/autoimmune diseases i.e. Behcet.$^1$

Other causes:

1. Trauma/mechanical lesions

2. Infection: associated with Lyme, coxsackie, TB, influenza, etc. Suspect if fever, diarrhea, or skin lesion present.

3. Iatrogenic: associated with statin use.$^2$
FM: Epidemiology

- Affects any age, preferably middle-aged adults.
- Affects males and females equally.
- Only 200 cases reported as of 11/2016.\textsuperscript{3}
FM: Clinical Presentation

- **Intramuscular mass or swelling** within one muscle. Grows slowly. Can be **painful**.

- Usually in lower extremities

- **No systemic symptoms** usually.

- No weakness or sensory impairments if no nerve association.

- No recurrence in 2-6 years.
FM: Differential Diagnoses

- Benign tumors: similar MRI as FM
- Malignant tumors: invade adjacent structures.
- Myositis ossificans: calcification of a muscle.
- Proliferative myositis: presence of basophilic fibroblasts.
- Polymyositis: symmetric involvement of proximal muscles.
- Muscular dystrophy: assess clinical history.
FM: Workup

- CK and ESR (usually moderately elevated), CRP normal
- Order MRI
- Order EMG
- Confirm with muscle biopsy
MRI Triceps

Triceps muscle appears hyperintense on T2 weighted images (A) and T2 STIR images (C). A patchy hyperintense signal is observed after gadolinium injection on T1 weighted images (B, D).
FM: EMG

Exclusively in affected muscle

- Spontaneous activity associated with positive sharp wave or fibrillation
- Small amplitude polyphasic motor unit potentials

http://www.iomonitoring.org/imgemg/Fig-5.jpg
FM: Muscle Biopsy

- Macrophage (arrow) and inflammatory cell infiltration (arrow head).³
- Hypertrophic muscle fiber regeneration.
- Interstitial fibrosis (star).
FM: Treatment

Self-limited. Usually regresses within one-two weeks.

Relapse occurs in 1-18% of cases.\textsuperscript{4}

Steroids recommended IF associated with elevated CK/ESR, nerve lesion, or autoimmune process.

- 0.75 mg/kg/day for 4–12 weeks