Parsonage-Turner Syndrome: Update on Presentations/Pathology/Treatment

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Objectives:
1. Describe the common and uncommon presentations of Parsonage-Turner Syndrome
2. Describe treatment to markedly improve the long-term outcome of Parsonage-Turner Syndrome

Patient #1, 37 yo NP
9/11/17 MyChart msg to PCP

I am writing because I injured my left shoulder on 8/30/17 in the morning (7am) playing with my kids. I heard a popping sound and had mild tenderness. I went to work and around noon, began to have significant pain and could not raise my left arm and had radiating pain down to my fingers. On 8/31/17, it felt a little better and pain was off and on so I decided to give it time and stretch, ice etc. After approx. two weeks of conservative tx, I am still experiencing come and go pain with a number of range of motion positions (more with extension when reaching, lifting shoulder and internal rotation). What is weird is I do have full ROM. Pain has been daily since onset and also wakes me at night. It is not as bad as that first day, yet constant enough to be bothersome and impact activities. I have been taking ibuprofen, helps a little. Would you recommend an app with you, sports therapy, or should I just go to PT? Thank you!

What did they find?
• 66 of the 136 pts were in the hospital with other conditions when the syndrome started, others had recently recovered from illness
• 98 of the cases had a precipitating factor
• Other-rapid development of weakness after a period of pain, 56/136 sensory changes on exam, ~1/3 were bilateral, all but the most severe cases improved over months

1/22/2019
AKA
Neuralgic Amyotrophy
Brachial Neuritis
Brachial Plexitis
Autoimmune Plexopathy

Most prefer the eponym, Parsonage-Turner Syndrome

Where?
Brachial Plexus

Wikipedia BP Diagram Nerve Root Colors

Why?
Each motor neuron is headed to a SPECIFIC GROUP OF MUSCLE FIBERS (motor unit) in a SPECIFIC muscle.
Each sensory neuron is destined for a SPECIFIC AREA OF TISSUE (skin, joint/tendon) to provide sensory supply.
Myelin is tightly wrapped lipid bilayers with specialized protein constituent pattern, very susceptible to stress-induced autoimmune attack.

“So you mean, it’s like Bell’s palsy of the brachial plexus?”

Bell’s Palsy-autoimmune attack on facial nerve

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Pain</th>
<th>Paresis</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Steroids</td>
<td>0-7/10</td>
<td>0-5/5</td>
<td>0-10/10</td>
</tr>
<tr>
<td>Pain meds</td>
<td>0-5/10</td>
<td>0-5/5</td>
<td>0-5/10</td>
</tr>
</tbody>
</table>

Largest study of Parsonage-Turner (PTS): 246 Cases

The clinical spectrum of neuralgic amyotrophy in 246 cases

- National referral center, all PTS pts in the Netherlands, 85% seen at center, 80% prospective, 20% retrospective
- All pts with an attack of acute, painful, patchy brachial and/or LS plexopathy that followed a monophasic course, without other specific etiology
- All included pts with paresis before pain, painless but w/t typical attacks, and attacks with nerves outside the brachial plexus
- Pain assessed with 10 pt VAS
- Severity of paresis MRC grade 4-5 mild; 3-4 mod; <3 severe
- Distribution of paresis
- Pain relief meds
- Subjective overall recovery
- Function outcome
- Ability to work
- Rankin scale
Pt Characteristics

68% male - unusual for autoimmune
87% right handed
20% family history of PTS*
Mean age onset 41 yo

Mean number of attacks 1.5
Mean time to diagnosis 44 weeks* 75% within 28 weeks
62% initially received another diagnosis, usually glenohumeral pathology or cervical radiculopathy

Characteristics of PTS Attacks

• 90% started with typical pain, onset usually over a few hours (contrast with radic)
• When absent initially, 2/3 of those developed pain later
• 61% started at night
• 72% one arm, 28% both arms

Pain Characteristics

• 65% reported subsequent MSK-type pain, esp periscapular region
• 17% developed glenohumeral adhesive capsulitis
• Glenohumeral subluxation/luxation developed in 9%
• 10% developed contractures of the wrist or fingers

• Location of Pain
  • 40% radiated from shoulder to arm
  • 35% radiated from cervical spine
  • 20% radiated from scapular or chest into arm***
  • 5% radiated in lower plexus distribution - median arm or hand, axilla

• Most effective pain relief from NSAID/opiate combination, diclofenac 100 bid/MSO4 10 bid

Weakness

• First sign of Weakness
  • 34% within 24 hrs of pain onset
  • After 1-7 days in 40%
  • After 1-2 weeks in 15%
  • Mean time to onset ~ 12-days

• Distribution of Weakness
  • Middle/Lower plexus 23% F, 10% M*
  • Upper part of plexus most common, in 71%
  • With long thoracic nerve/err ant in 71%
  • Without long thoracic nerve in 21%

* Nerves outside of brachial plexus in 17%
  • 8% LS plexus
  • 7% phrenic nerve*
  • 2% recurrent laryngeal nerve
  • 3% other

• Attacks reoccurred in 26% of pts
• Median time to reoccurrence ~ 2 yrs

Weakness Distribution

• Middle/Lower plexus 23% F, 10% M*
• Upper part of plexus most common, in 71%
• With long thoracic nerve/err ant in 71%
• Without long thoracic nerve in 21%
Weakness-take home exam

- Check Infraspinatus weakness

- Take shirt off and look for inferior angle scapular winging secondary to long thoracic nerve/serratus anterior... esp good to differentiate from rotator cuff/supraspin tendon

Infraspinatus force testing

Unilateral vs bilateral

Sensory Symptoms

- Reported by 69% of pts
- On clinical exam 78% had a sensory disturbance
  - Hypaesthesia most common complaint in 46%
  - Parasthesia and hypaesthesia 39%
  - Parasthesia only 14%
  - Hypaesthesia and/or paresthesia over deltoid/lateral arm most common (c/w rotator cuff)

Predisposing Factors/Antecedent Events

- 53% reported antecedent event
- In 52%, event occurred the week before the attack
- 17% it occurred hours before

### Table 5 Distribution of sensory symptoms regrouped into anatomical regions

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Percentage</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lower back/upper arm</td>
<td>13%</td>
<td>37</td>
</tr>
<tr>
<td>Upper arm/hand only</td>
<td>28%</td>
<td>77</td>
</tr>
<tr>
<td>Head/forehead</td>
<td>7%</td>
<td>20</td>
</tr>
<tr>
<td>Neck, back and scalp</td>
<td>5%</td>
<td>15</td>
</tr>
<tr>
<td>Other</td>
<td>6.7%</td>
<td>18</td>
</tr>
</tbody>
</table>

### Table 6 Antecedent events

<table>
<thead>
<tr>
<th>Antecedent event</th>
<th>Percentage</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Illness</td>
<td>4.8%</td>
<td>10</td>
</tr>
<tr>
<td>Spring</td>
<td>17.4%</td>
<td>36</td>
</tr>
<tr>
<td>Surgery</td>
<td>15.2%</td>
<td>31</td>
</tr>
<tr>
<td>Injury</td>
<td>12.4%</td>
<td>25</td>
</tr>
<tr>
<td>Vaccination</td>
<td>4.3%</td>
<td>9</td>
</tr>
<tr>
<td>Headache/myalgia</td>
<td>6.2%</td>
<td>12</td>
</tr>
<tr>
<td>Trauma</td>
<td>4.2%</td>
<td>9</td>
</tr>
<tr>
<td>Other</td>
<td>7.0%</td>
<td>14</td>
</tr>
</tbody>
</table>
Lab Tests

**EMG**
- Best at ≥3 wks after symptom onset
- Performed in 87% of patients
- Abnormal in 96%
- Important to carefully check uncommon muscles - infraspinatus/supraspinatus/serratus anterior

**Imaging Studies**
- 54% of patients had a CXR
  - About 4% had elevated of one/both diaphragms due to phrenic nerve involvement
- 44% had MRI of cervical spine
  - 51% of scans showed degen changes
- 20% (50 pts) had brachial plexus MRI
  - Only 3 were abnormal

Functional Recovery

**Persistent Pain**

**Residual Weakness**

![Graph showing percentage of patients with persistent pain for each of the five follow-up periods.](image1)

![Graph showing percentage of patients with residual weakness for each of the five follow-up periods.](image2)
Subjective Overall Recovery...not great?

Netherlands Disability Benefits ≇ Outcome?

Government spending on social security decreased after the system was reformed, but the number of benefit claimants continued to rise. By the end of the 1980s, the proportion of non-native members of the labour force had increased substantially, as more and more workers claimed incapacity benefits. Now, along with income protection, prevention and reintegration were being incorporated into the system. Another important new goal was activation (i.e. encouraging participation in the workforce), which reduced social exclusion and strengthened the income distribution. The new welfare state was characterized by a more active and participatory approach, which involved the active participation of the people in the design and implementation of the system. The outcome is not different than the reform of 1987. The key issue now is to reverse each party's current interest in reducing social security benefits claims.

There is no legal maximum for levels of benefit. Some schemes result in a maximum of 70–80% of former salary but these are exceptional. The disadvantage of these schemes which result in maximum of 100% of former salary is that they do not provide for continuation of rights to old age benefits during periods of disability.

Shanghai 1988

Hedgehogs, revolutions, and wandering Dutch who don't work...

Bell’s Palsy-autoimmune attack on facial nerve

<table>
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<tr>
<th>Effect of early treatment of Bell's palsy with corticosteroids</th>
<th></th>
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<tbody>
<tr>
<td>Early treatment</td>
<td></td>
<td></td>
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<tr>
<td>Improvement</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Usually a 2-3 day course of steroids</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Myelin is tightly wrapped lipid bilayers with specialized protein constituent pattern, very susceptible to stress-induced autoimmune attack.

2009 paper J Neuro Neurosurg Psychiatry

**Study of steroid treatment for PTS**

- Retrospective study
- 50 patients
- Treated within one month of symptom onset
- Simple 13 day course prednisolone
  - 60mg/day x 7 days
  - 50mg x 1 day
  - 40mg x 1 day
  - 30mg x 1 day
  - 20mg x 1 day
  - 10mg x 1 day
  - 5mg x 1 day

**Is There A Difference Between Prednisone And Prednisolone?**

Both prednisone and prednisolone are in the same chemical family, they are used to treat similar conditions and are generally considered equally effective. However, in people with rheumatoid arthritis, prednisone is usually preferred. This is because prednisone is converted to prednisolone in the body, and it is this prednisolone that is the active molecule.

Prednisone is usually much cheaper than prednisolone.

**Median time until pain relief, 20.5 vs 12.5 days**

- Treated
- Untreated

Days to Pain Relief
Recovery of strength within one month, 18.0% vs 6.3%

Full functional recovery within 1st year, 12% vs 1%

Anterior interosseous nerve
Where is the AIN compression?
Will you see Parsonage-Turner? Not if you don’t look...

Incidence of Neuralgic Amyotrophy (Parsonage-Turner Syndrome) in a Primary Care Setting - A Prospective Cohort Study.

Abstract

Objective: This is a prospective, observational study of patients aged 18-70 years who consulted a family physician about possible neuralgic amyotrophy (NA) from January 2014 to December 2018. The primary outcome was the evaluation of the annual incidence rate of NA in the primary care setting.

Methods: We conducted a prospective cohort study of patients aged 18-70 years who consulted a family physician about possible NA from January 2014 to December 2018. NA was diagnosed by a clinical examination and confirmed by electromyography. The annual incidence rate of NA was calculated using the formula: Number of newly diagnosed cases / Total population at risk x 1000

Results: During the study period, 50 patients were identified as having NA. The annual incidence rate of NA was 1/1000. Compared with MI, the annual incidence rate of NA was 1/410. Much more common than we realize. Medical and/or surgical treatment possible.

Why care?

In a population of nearly 15,000

Annual incidence rate 1/1000

Compared with MI: Annual incidence rate 1/410

Much more common than we realize

Medical and/or surgical treatment possible

Interfascicular Microneurolysis
Summary:

Think of Parsonage-Turner Syndrome early and often

When in doubt, short trigger for 13 day steroid course-very effective, try to start in 30 days from onset of symptoms (is this window necessary?)

Future for interfascicular microneurolysis likely very bright