Differential Diagnosis of Post-Polio Syndrome and the Role of Muscle Imaging

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Characteristic clinical picture

- Polio at age 7 years.
- Sequelae R lower leg.
- Over the past few years increasing weakness R leg.

Thigh muscles also affected
Post polio syndrome criteria

- Prior episode of paralytic polio
- Period of neurologic and functional stability
- Onset of new muscle weakness
- Exclusion of other conditions
Diagnostic dilemma’s

1. Patients with neurological condition which is found to be related to previous polio during the diagnostic process
e.g., PPS

2. Mimic of PPS
e.g., SMAIII, CMT

3. PPS-patients who develop a neurological condition indirectly related to previous polio
e.g., radiculopathy, entrapment neuropathy

4. PPS-patients who develop a neurological condition which is unrelated to polio
e.g., syringomyelia
Mr vD, 58 years old
seen in 2009

Referral History

- Swallowing difficulty
- Increasingly sensation of choking which is experienced as frightening. Initially during the night but now also during daytime.
- In addition difficulty eating.
- Bulbar polio in 1956 (following tonsillectomy).
- Iron lung, tracheostomy, tube feeding, myotomy upper sphincter oesophagus

Previous hx
Mr vD cont’d

• The history was suggestive of laryngeal stridor, a diagnosis of PPS was made and the patient underwent tracheostomy and also was given a PEG.

• This patient developed his signs and symptoms in the musculature that had previously been affected, had regained strength and subsequently deteriorated.
Diagnostic dilemma’s

1. Patients with neurological condition which is found to be related to previous polio during the diagnostic process, e.g., PPS

2. Mimic of PPS
   e.g., SMAIII, CMT, IBM, …..

3. PPS-patients who develop a neurological condition indirectly related to previous polio
   e.g., radiculopathy, entrapment neuropathy

4. PPS-patients who develop a neurological condition which is unrelated to polio
   e.g., syringomyelia
Mimic: Mr S, 73 years old (d.o.b. 1938) at time of examination

Referral

History

- Spinal muscular atrophy type 3?
- 1956 decrease in strength L arm and both legs + fever. Diagnosis?
- 1958 diagnosed as SMA3.
- Gradual improvement of strength.
- Sequelae: proximal leg muscle weakness (difficulty getting out of a chair).
- 1999: R patella #. Since then diminishing strength leg muscles
Atrophy of the thigh muscles, R>>L
Positive Gowers’ sign.
**Mimic:** Male, 70 years at examination
Progressive walking difficulty.
Guillain-Barré syndrome at age 4 years (?)
Ex/ Generalized mild muscle weakness both legs.
EMG: neurogenic
Weakness and wasting of distal legs can be seen in PPS and in hereditary neuropathies.
Diagnostic dilemma’s

1. Patients with neurological condition which is found to be related to previous polio during the diagnostic process,
   e.g., PPS

2. Mimic of PPS
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   e.g., syringomyelia
Ms R, 56 years old at time of examination

History
- In 1955 polio (at age 1 yrs).
- Sequelae: paralysis of both legs.
- She now walks with crutches.
- CTS surgery in 1994, diabetes mellitus since about 4 years.
- Since about a year complaints about sensory disturbances in both arms irradiating to ring and little finger.

Diagnosis
- Entrapment of the ulnar nerve
MRI revealed a ganglion at the site of EMG entrapment. The ganglion was surgically removed and the sensory disturbances have resolved.
Diagnostic dilemma’s

1. Patients with neurological condition which is found to be related to previous polio during the diagnostic process, e.g., PPS

2. Mimic of PPS e.g., SMAIII, CMT

3. PPS-patients who develop a neurological condition indirectly related to previous polio e.g., radiculopathy, entrapment neuropathy

4. PPS-patients who develop a neurological condition which is unrelated to polio e.g., syringomyelia, muscle amyloidosis
Mrs de B, 64 years old
seen by us in 2009

History
• Difficulty walking since about 10 years, uses crutches for outdoor activities.

Previous hx
• Polio at age 1.5 yrs. Sequelae L leg. Surgery left ankle and knee.
• In 2007 decrease in strength in R thigh muscles and since 2008 of the arms.
• She is now wheelchair dependent, needs home care.
• Atrophy and shorter left leg, no fasciculations
• Generalised weakness arms (MRC 4),
• R leg dist MRC 4, prox 2, L leg MRC 1-2.
• Areflexia.
EMG: spontaneous muscle activity in various muscles of R leg and paraspinal muscle and signs of reinnervation in those muscles.

DD?
- Progressive spinal muscular atrophy
- Post polio syndrome
Ancillary investigations

• M-protein, IgG κ (serum, urine)
• Bone marrow puncture showed monoclonal plasma cells IgG kappa.
• Lip biopsy: amyloidosis, monoclonal kappa +-ve
• Echo heart: consistent with amyloidosis
• Muscle biopsy …..
Mr N, 40 years old at time of examination

History

• Since 1 year slowly progressive weakness R thigh, and sensory changes in the sole of his R foot and palm of his R hand.

Previous hx

• 1978: polio. Both legs, abdominal muscles and L triceps brachii muscle paralysed.
  • Sequelae: scoliosis and weakness and wasting of L leg
  • 1983: cervical neurofibroma, partial resection.
• Horner syndrome
• Weakness L upper arm and shoulder girdle muscles
• Diffuse weakness legs, L>R, proximal > distal
• Sensory changes distal legs
• Bilateral extensor plantar responses
SYRINGOMYELIA
In November/December 2011 the patient noticed pain in his right shoulder irradiating to the elbow. The pain disappeared in a few weeks and then he noticed decreased dexterity of his right hand. NO/ sequelae of polio in the left leg. Atrophy of mm. interossei, m. abductor pollicis brevis and m. opponens pollicis. 
Dx: Plexus brachialis neuropathy (lower plexus)
Plexus brachialis neuropathy

- Excruciating pain lasting a few days-weeks
- Followed by mostly unilateral muscle weakness
- Usually a good prognosis (up to 2-3 years)
CT/MRI is useful in PPS

- Can visualize sequelae polio
- Can detect subclinical involvement in asymptomatic muscle
- Can be reassuring, showing no replacement of muscle by fat in weak leg
- Can be helpful in differential diagnosis
Female, 53 yrs at examination. Polio at age 3 years with sequelae in right leg. Increasing weakness R thigh muscles
Male, age at examination 60 years - polio at age 7 years. Both legs were involved, sequelae left leg. Over the years increasing weakness L leg. No complaints about R leg.
Female, 51 years at examination – polio at age 2 - Sequelae R leg. Increasing weakness R leg, but also weakness L leg.
Differential diagnosis of PPS

- ALS/PMA
- Hereditary SMA
- Entrapment neuropathies
- Multifocal motor neuropathy
- Hereditary neuropathy (CMT)
- Plexus brachialis neuropathy
- Radiculopathy (cervical/lumbar)
- Inclusion body myositis
- Muscle amyloidosis
Differential diagnosis PPS

Key questions

• Is there a credible history of polio?
• Are there sequelae?
• If pain is a prominent complaint, consider radiculopathies or entrapment neuropathies and perform EMG/imaging.
• A patient with PPS may also acquire disorders not related to PPS, e.g., neuralgic amyotrophy, syringomyelia, etc. If there is a rapid disease course, consider other diagnoses, e.g. amyloidosis, motor neuron disease.