

# Post-polio syndrome and amyotrophic lateral sclerosis - similarities, differences and diagnostic dilemmas

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Post-polio syndrome/post-polio muscular atrophy (PPS/PPMA) and amyotrophic lateral sclerosis (ALS) are progressive neurodegenerative disorders characterized by motor neurons (MNs) loss in the spinal cord, brain stem and motor cortex. PPS/PPMA is regarded as ALS-mimic syndrome. About 1% of patients with history of paralytic polio have been reported to develop ALS as coincidental findings.

## PPS/PPMA

## ALS

### Etiopathogenesis

- ◆ **Unclear**, not fully understood
- ◆ **Multifactorial**: various pathological factors connected with progressive MNs stress, accompanied by age-dependent risk factors.

- ◆ Largely **unclear**
- ◆ **Multifactorial**: various endogenous and/or environmental factors implicated in progressive MNs stress

### Clinical symptoms

- ◆ **Progressive weakness and muscular atrophy** (mainly due to lower MNs involvement)
- ◆ Occasionally **bulbar symptoms** (dysarthria, dysphagia respiratory problems)
- ◆ **Muscle pain** and joint pain
- ◆ Often **fear, anxiety**, rarely depression

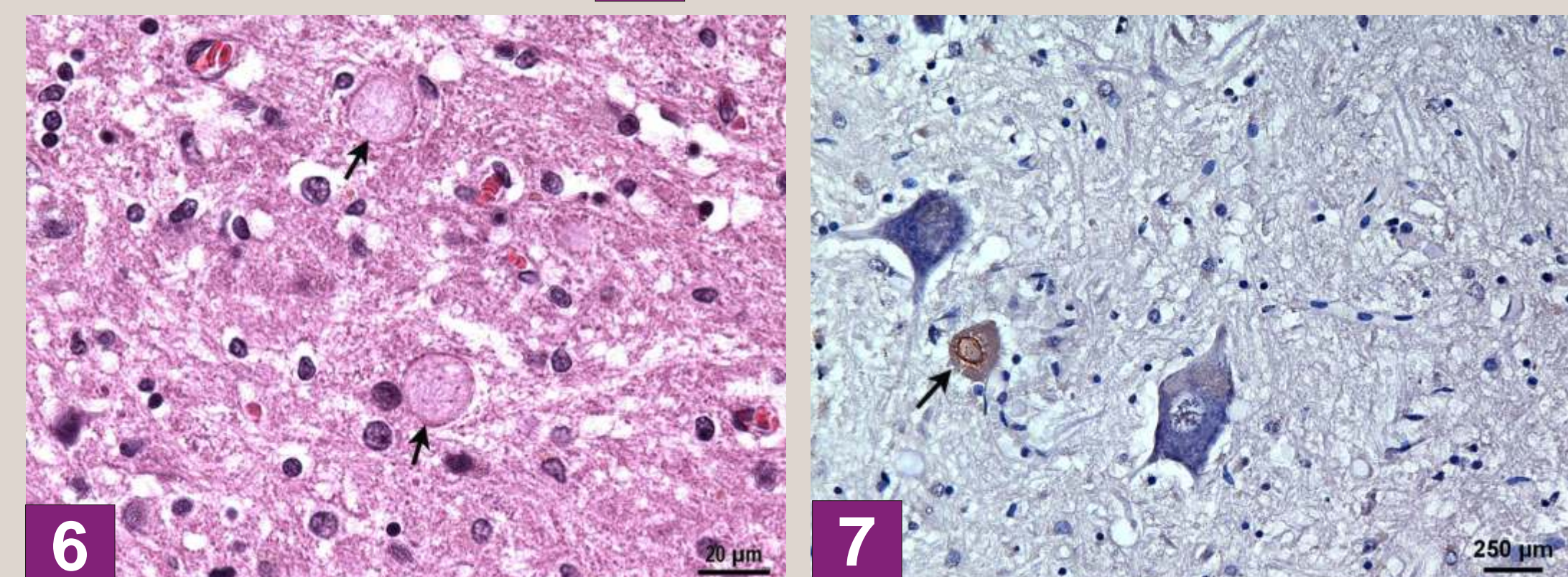
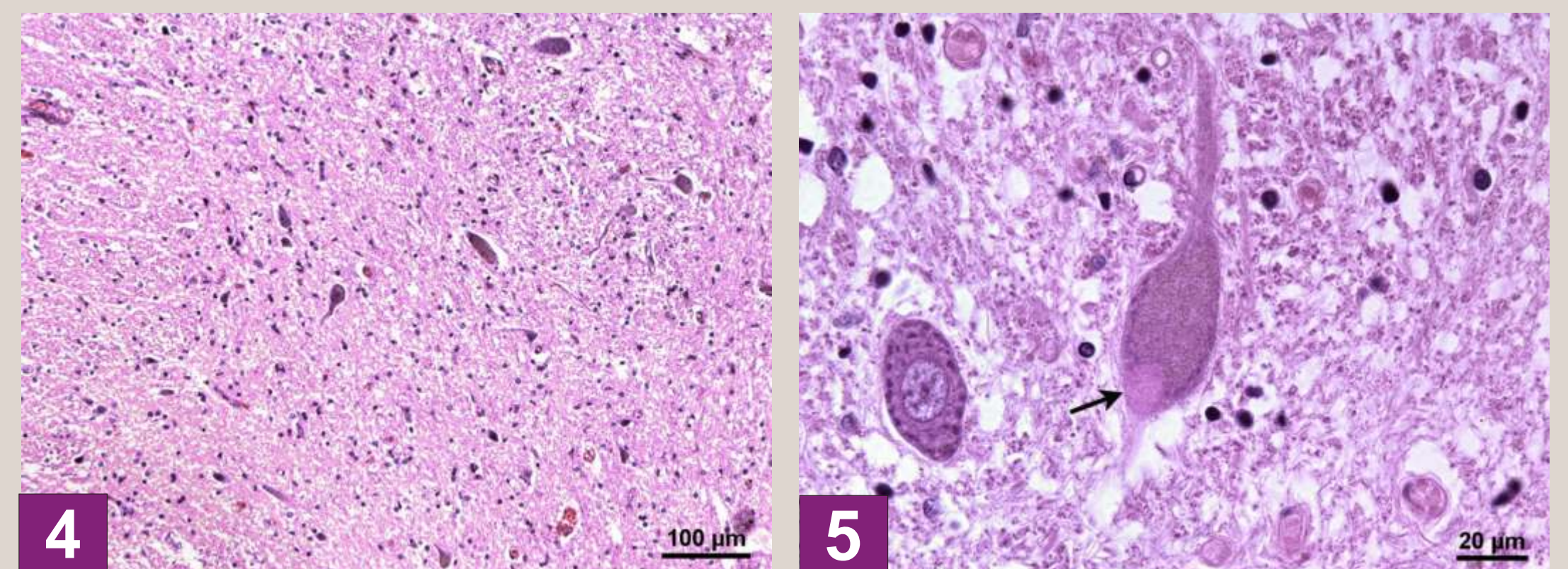
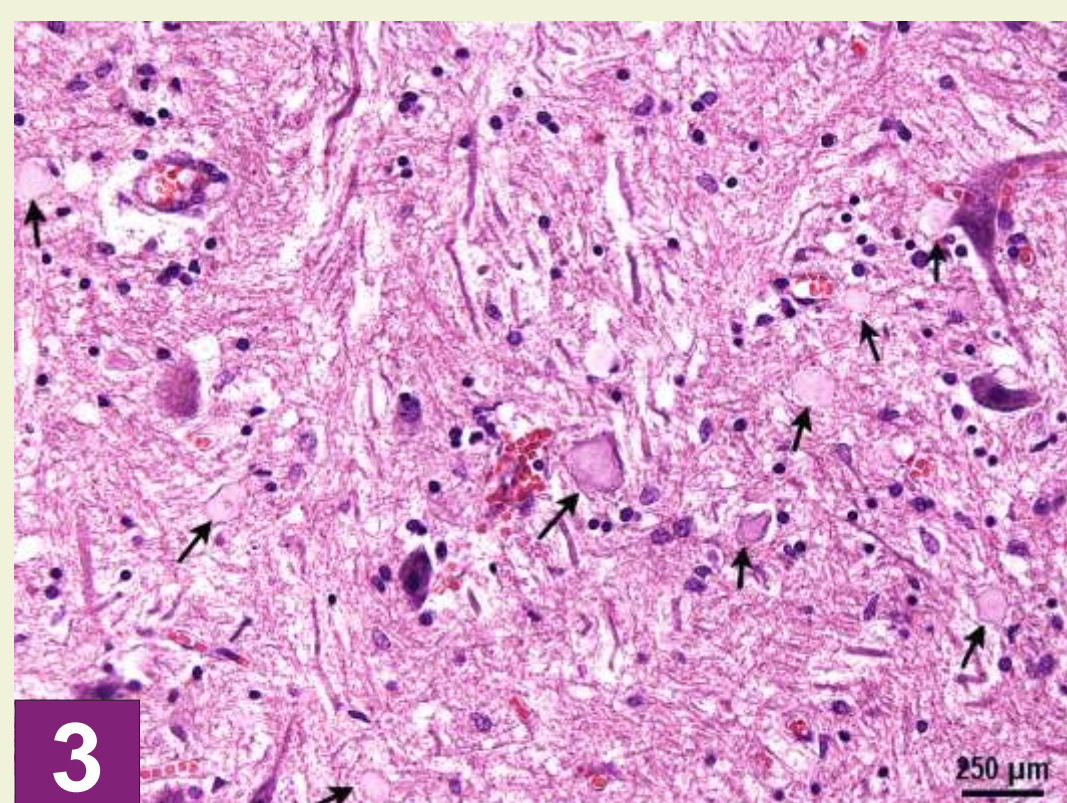
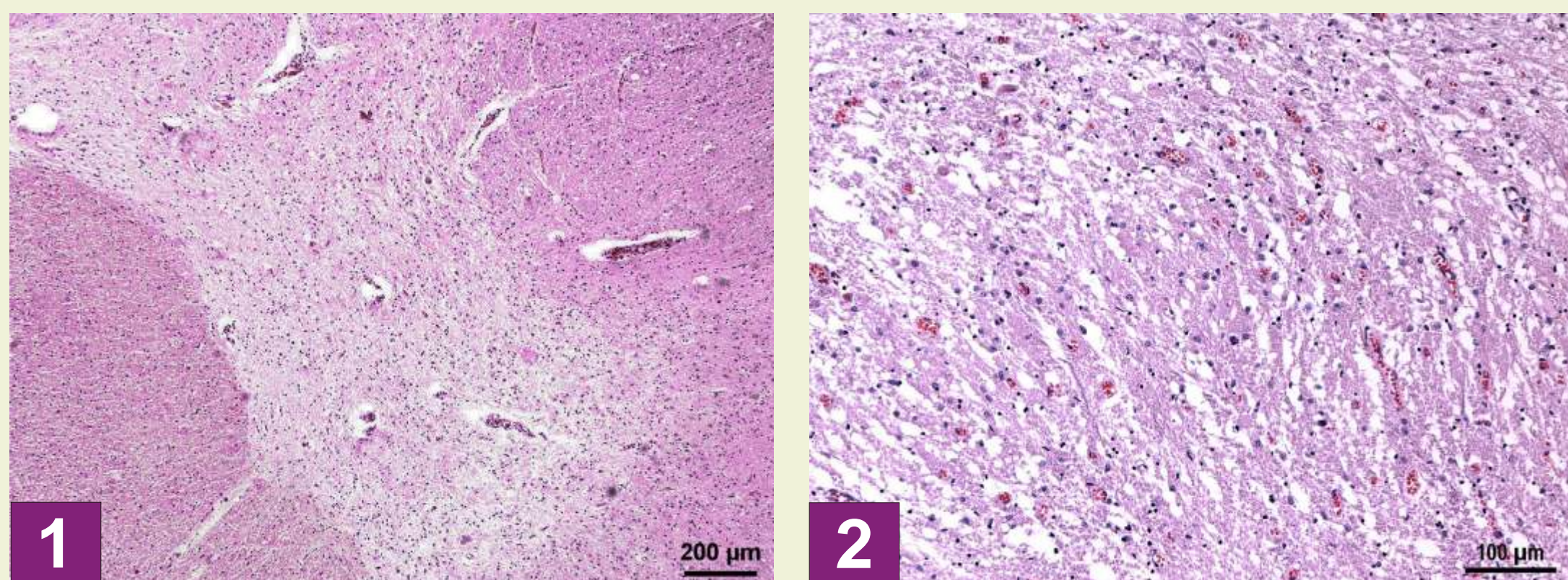
- ◆ **Progressive weakness and muscular atrophy** (due to upper and lower MNs involvement)
- ◆ **Bulbar symptoms** (dysarthria, dysphagia, respiratory failure)
- ◆ **Muscle pain** and muscle cramps
- ◆ Often **fear, anxiety** and depression

### Neuropathology

- (Only a few autopsy findings so far reported)
- ◆ **Loss of MNs in the anterior horn** mainly of lower spinal cord (1), rarely upper spinal cord or/and brain stem
  - ◆ Lack of degeneration of corticospinal tract
  - ◆ **Gliosis** (2)
  - ◆ Inflammatory B cells infiltrates, often perivascular
  - ◆ Neuronal inclusions are not typical features, spheroids were seen occasionally (3)

- (Autopsy findings represent the terminal stage of disease)
- ◆ **Loss of MNs in the anterior horn** of lower and upper spinal cord (4), brain stem, motor cortex
  - ◆ Degeneration of corticospinal tract
  - ◆ Widespread **gliosis**
  - ◆ Lack of inflammatory changes
  - ◆ A variety of neuronal inclusions i.e. spheroids, Lewy-body-like inclusions (5), Bunina bodies, hyaline inclusions (6) and skein-like, ubiquitin or TDP43-immunoreactive inclusions (7)

The main pathology is similar to late period of poliomyelitis



### Diagnosis

- ◆ **Exclusion of other neurological conditions with similar symptoms.**
- ◆ There are **not specific tests** and biomarkers to confirm the diagnosis

- ◆ **Exclusion of other neurological conditions with similar symptoms.**
- ◆ There are not **diagnostic tests** or procedures useful to confirm or exclude the diagnosis

### Prognosis

- ◆ Usually **slowly** progressive course

- ◆ **Rapidly progressive, devastating, fatal disease**
- ◆ Life expectancy is usually 3-5 years after diagnosis

### Treatment

- **Lack of effective drug treatment** (only IVIG)
- **Supportive care - multidisciplinary teams of healthcare**
- **Individually tailored training programs**
- Occasionally ventilatory assistance

- **Lack of effective drug treatment** (only riluzole)
- **Supportive care - multidisciplinary teams of healthcare**
- **Individually tailored training programs**
- Respiratory assistance and supplemental nutrition at early stages of the disease.

The distinction between PPS and ALS bears important prognostic implications. Both these motor neuron diseases still remain a challenge for clinical and scientific community.