

## 64<sup>TH</sup> ANNUAL DIAGNOSTIC SLIDE SESSION 2023

### CASE 2023-3

Submitted by:

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Clinical History: 18-year-old male who achieved developmental milestones during early infancy but then failed to develop normal walking skills by the end of the first year. This was followed by cognitive developmental delay, plateau in motor skills and subsequent progressive loss of cognitive and motor skills.

Brain MRI showed distinctive increased iron in the globus pallidus and substantia nigra and a distinctive hyperintense halo in the cerebral peduncles.

Autopsy is limited to the brain:

The brain showed mild cortical atrophy involving the frontal, parietal, and temporal lobes in addition to dark-brown discoloration of globus pallidus and substantia nigra, more prominent in the latter (fixed brain weight, 887 g).

Material submitted:

1. H&E stained slide of a representative midbrain with substantia nigra discoloration
2. Gross images of brain autopsy findings

Key diagnostic points:

1. What is the differential diagnosis for this type of lesion that involves the globus pallidus and substantia nigra.
2. What other stains would be helpful to establish a specific diagnosis of this disease.