



# **DSS 2015**

## **Case 7**

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# Financial Disclosures

- None

# Clinical

- **55-year-old man**

- 3 months of progressive gait unsteadiness; numbness and tingling in feet and fingertips

- **Neuro Exam:**

- Ataxic gait, positive Romberg, loss of vibration, proprioception, pinprick in feet, reflexes brisk in UEs absent in LEs

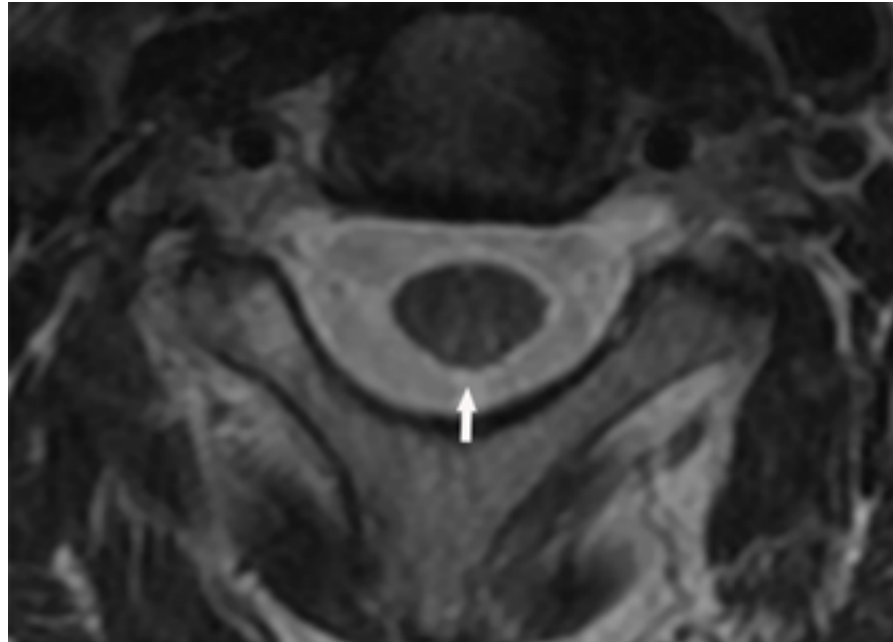
- **Microcytic anemia**

- **Biopsy confirmation of celiac disease**

- **Negative laboratory tests:**

- vitamin B12, thiamin, vitamin E, SPEP, glucose, Hb A1C, LFTs, TSH, ANA, paraneoplastic, PET-CT, serologies (Anaplasma, Erlichia, Babesiosis, B. burgdorferi)

# MR Image

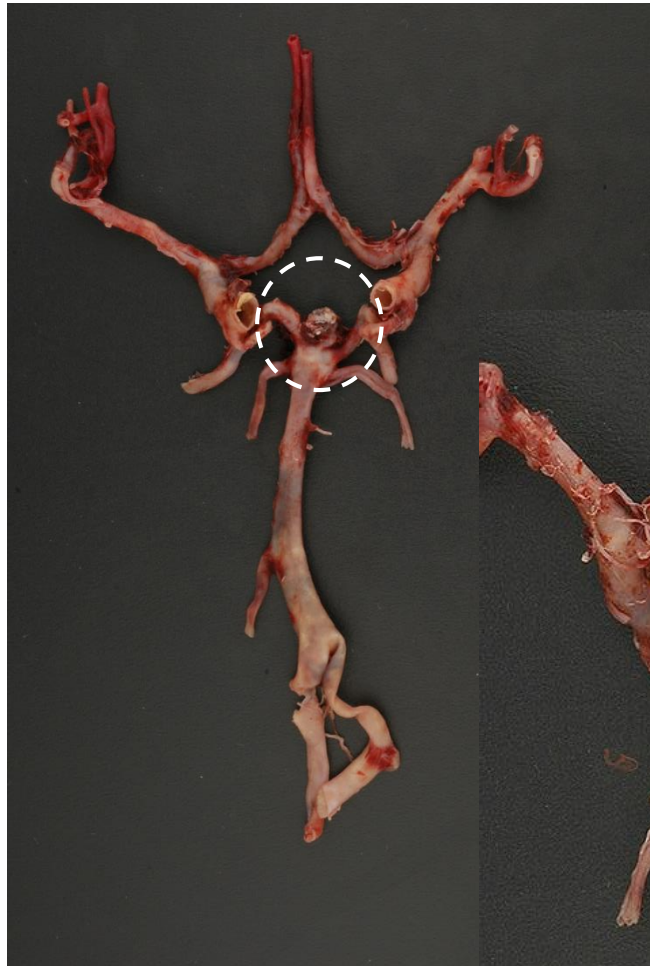


Very subtle T2 signal hyperintensity in the dorsal columns

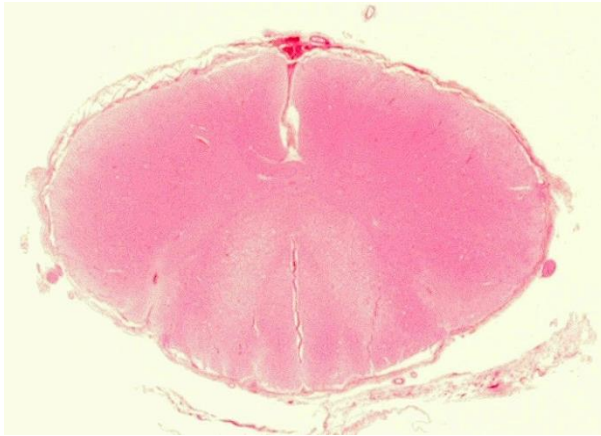
# Clinical History

- Additional laboratory testing established a diagnosis
- Five months later, he collapsed suddenly and died
- Autopsy disclosed a ruptured basilar tip aneurysm with marked subarachnoid hemorrhage

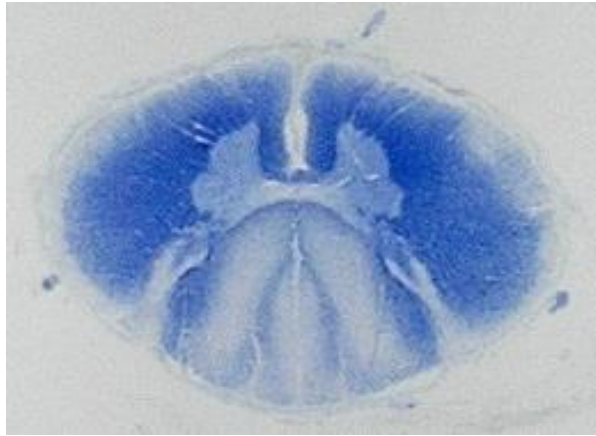
# Autopsy Findings



# Spinal Cord Histology



**H&E**

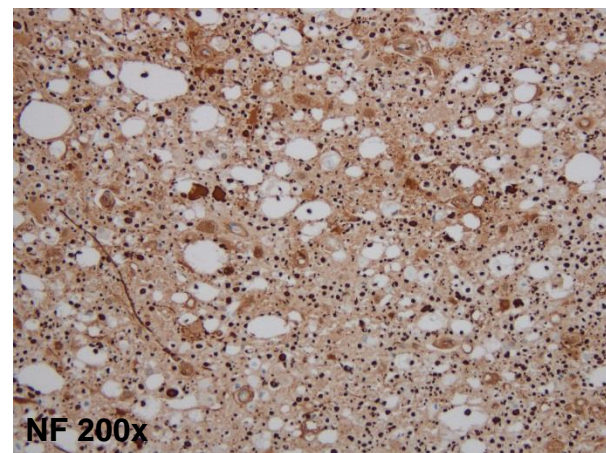
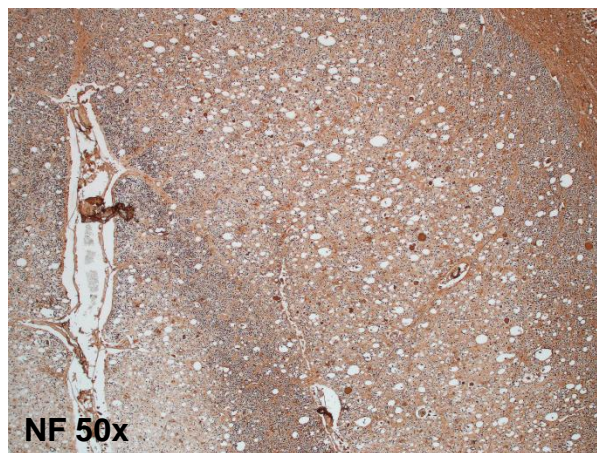
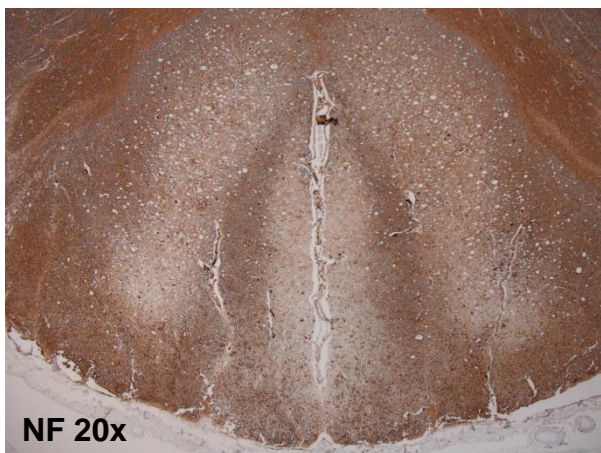
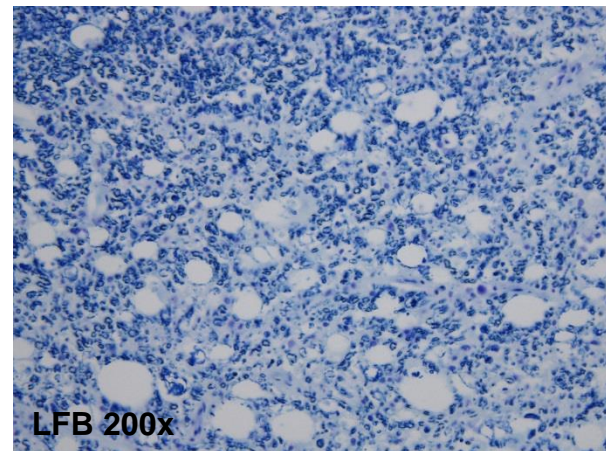
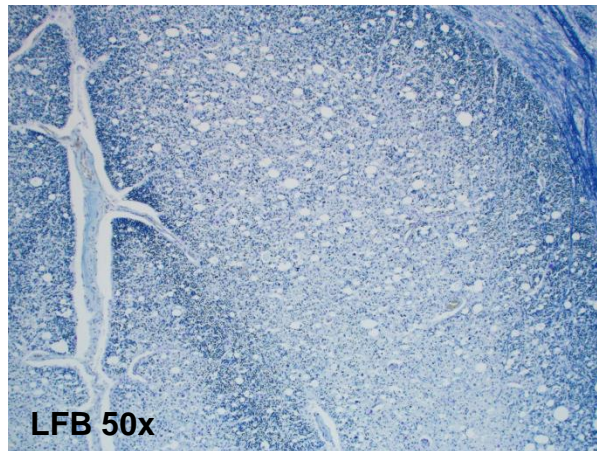
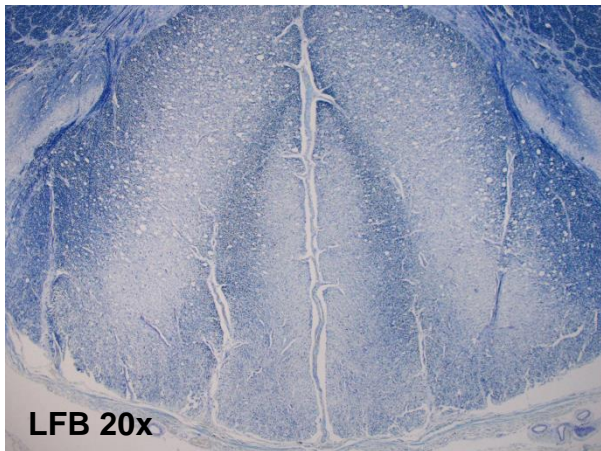
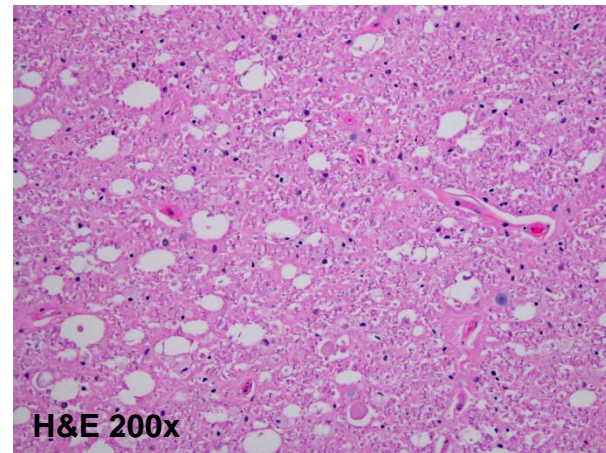
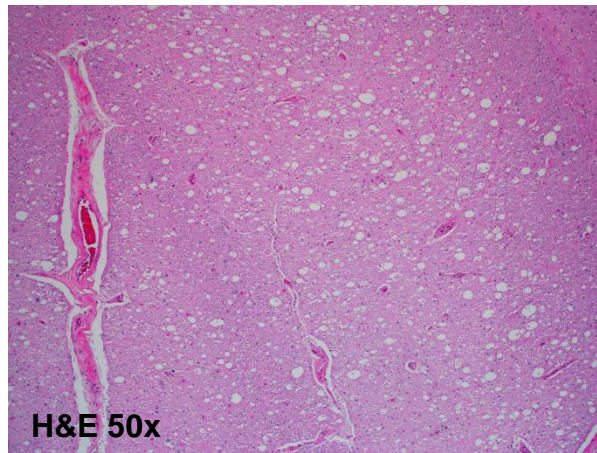
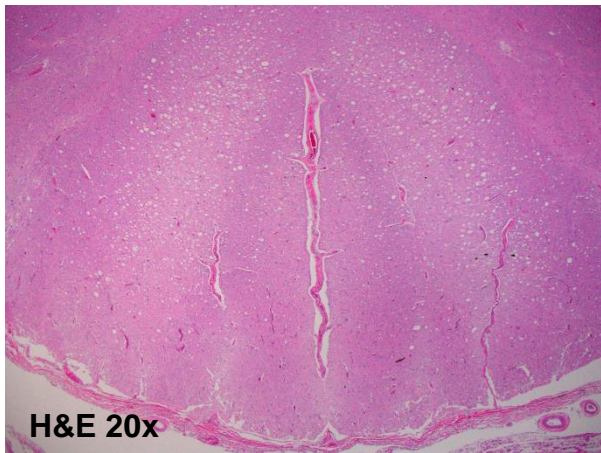


**Luxol Fast Blue**



**Neurofilament**







# Discussion

## Diagnosis?

# Diagnosis

## Copper Deficiency Myelopathy

- Serum copper markedly decreased at 0.27  $\mu\text{g/mL}$  (normal range 0.75-1.45)
- Ceruloplasmin decreased at 9.8 mg/dL (normal range 15-30)
- Serum zinc normal

# Copper Deficiency Myelopathy

- Long recognized to occur in ruminant animals, termed “swayback” or “enzootic ataxia”
- Identical presentation and pathology to subacute combined degeneration due to B12 deficiency
  - Gait disturbances and paresthesias
  - Vacuolating myelin loss of the dorsal columns
  - Cervical spinal cord typically most severely affected
- Hematologic manifestations often also present (anemia and neutropenia)



# Most Common Etiologies

- Upper GI surgery (including bariatric surgery)
- Celiac Disease
- Zinc overload, often from denture cream

## Treatment and Patient Outcome

- Copper supplementation
  - Resolution of copper deficiency and hematologic abnormalities
  - Stabilization or minimal improvement of neurologic symptoms





# Questions & Discussion