

### CASE 1997-4

Submitted by: Dr Jeanne E Bell, Neuropathology Laboratory, Department of Pathology, Western General Hospital, Crewe Road, Edinburgh EH4 2XU.

Clinical History: 29 year old woman with a 17 month history of a progressive dementing illness associated with ataxia and choreiform movements. She deteriorated despite supportive therapy and died. Permission for autopsy was restricted to the brain and internal organs.

Necropsy findings: Emaciated young woman with muscle wasting and fixed flexion deformities of the limbs. The brain (weight 1122 grammes) showed global atrophy. No focal abnormality was noted on section of the fixed brain.

Material submitted: H&E section of cerebrum.

Points for discussion:

1. Diagnosis.
2. Pathogenesis