

Canadian Association of Neuropathologists
L'Association Canadienne des Neuropathologistes

CASE No 8

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This 22-year-old woman of Jamaican descent first came to medical attention when she was 12 years-old. She had skin lesions that were treated in 2003 with no recurrence. She was found to be positive for sickle cell trait. In November 2004 she developed congestive heart failure and a cardiomyopathy. She then developed progressive shortness of breath and was found to have hypoxic respiratory disease. A CT lung scan showed diffuse ground glass opacities bilaterally with interlobular septal thickening. This was initially thought to be due to heart failure but a diagnosis of sarcoidosis was also entertained. An open lung biopsy was considered to be too risky, given her low pulmonary reserve and pulmonary hypertension. Since November 2004, she required nasal prong oxygen 24 hrs/day at 3-4 litres/min. Over the last 6-9 months, she noted progressive foot drops bilaterally which made it difficult to ambulate. On examination there was obvious diffuse proximal and distal muscle wasting as well as skin changes in both forearms, wrists and ankles. Loss of power was documented in various muscle groups of both extremities. Vasculitis was considered as a cause of the myopathy. Nerve conduction studies showed multifocal motor and sensory involvement of an axonal nature. Muscle and nerve biopsies were done.

Specimen: Sural Nerve Biopsy

Stain: Hematoxylin-Eosin

Questions:

1. What is the differential diagnosis ?
2. What laboratory aids would you use to reach a diagnosis ?