Canadian Association of Neuropathologists L'Association Canadienne des Neuropathologistes

CASE No 1

F. Al Sufiani, H. Desai and L.C. Ang

London Health Sciences Centre, University of Western Ontario, London, Ontario, Canada; Windsor Health Center, Windsor, Ontario, Canada.

A 67-year-old woman presented with a two year history of droopy eyelids, which was not fluctuating in severity with fatigue. There were no complaints of diplopia, dysphagia, dysarthria, shortness of breath, headache, nausea and vomiting, focal sensory disturbance in the limbs or bladder/bowel incontinence. She was diagnosed previously as a case of polymyalgia rheumatica. She also complained of symptoms of bradycardia and Raynaud's phenomena. In the past, she had right sided weakness and was diagnosed as having a stroke. She was on Plavix since and her other medications are Ogen, Accupril, Novohydrazide, Restoril, Imovane, Valium, Fentanyl patches, Prednisone and Aspirin. There was also previous history of closed head injury, fracture of T12 vertebra and collapsed clavicle. No known family history of similar symptoms was reported.

On examination, she was awake and alert. She had bilateral ptosis but the extraocular movements were full. Her pupils were symmetrical. There was marked temporal wasting but mild wasting in the limbs. She was able to offer resistance to testing of all muscle groups. The reflexes were hypoactive. Plantar responses were down going.

Serum CK level was mildly elevated. The EMG studies did not show any evidence of myotonia. Myopathic motor units were seen in a few muscles in the lower extremities.

Tensilon test and muscle biopsy were ordered.

Material Submitted: Plastic section stained with toluidine blue.

Question: What is the diagnosis?