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Case 3

Submitted by Antje Bornemann, MD, Hans H Goebel, MD, Jürgen Bohl, MD, Romain Gheradi MD

Departments of Neuropathology, Universities of Tübingen, Mainz, and Creteil/Paris

Case Reference Number: 14320-N1018/01

Clinical history

A 63-year-old lady physician in private practice had experienced backache for the last 15 years owing to a pseudo-spondylolisthesis at L4 to S1. For the past 10 years, she developed muscular atrophy in her arms and, 3 ½ years ago, weakness in her back muscles ensued. At the time of examination her walking distance had decreased to 500 metres requiring a cane, and she had difficulties in climbing stairs and walking downhill. Over the last year she developed paraesthesia of the right thigh. Furthermore, she complained of arthralgia in large joints. Neurological examination revealed bilateral weakness of her face, her shoulder girdle, and her right arm, as well as scapula alata. She showed muscle atrophy in both thighs. Walking on heels was impaired, but not on toes. She showed Gowers sign and had hyperlordosis. Her deep tendon reflexes were weak. Electromyography revealed myopathic patterns but no response in the paraspinal muscles. Electroneurography revealed nerve conduction velocities of 47.8 m/sec in the right tibial nerve with an amplitude of 3.9 m/V and of 50.9 m/sec in the right sural nerve. Her creatine kinase was at upper borderline level of 100 U/L. She had an erythrocyte sedimentation rate of 2/23 mm for the first two hours.

No other family members were known to be affected by any neuromuscular disease.

The patient had undergone repetitive vaccination against hepatitis B, in 1970, 1980, and 1990. In 2000 she decided on having a combined hepatitis A and B vaccination. Injection site was the left deltoid muscle without any immediate localized and generalized discomfort or abnormal findings.

Material submitted:

1. One semithin section – methylene blue-stained (Richardson). – from the biopsied left deltoid muscle
2. One electron micrograph from an interstitial cell

Points for Discussion:

1. Diagnosis
2. Significance of findings