Schwannomatosis

Figure T2-weighted fat-saturated MR images showing multiple bilateral masses along the cervical, brachial (A), and sciatic nerves (B)

A 41-year-old man with a long history of von Recklinghausen disease presented with progressive bilateral leg pain. Clinical examination showed a severe bilateral paresis of ankle dorsiflexion. Achilles tendon reflexes were absent, while the patellar tendon reflex responses were weak. Furthermore, atrophy of the small muscles of the left hand and forearm was identified, accompanied by a mild to moderate paresis of the finger movements and hand flexion. The biceps tendon and radial periosteal reflexes were obtained weakly while the triceps tendon reflexes and Tromner were absent. Finally, multiple typical cutaneous neurofibromas and café-au-lait spots were seen.

MRI scans revealed ubiquitous solid masses, especially along the cervical, brachial, and sciatic nerves (figure).

The classic form of von Recklinghausen disease includes the development of neurofibromas.¹ In our case these were innumerable, so that the term schwannomatosis seems to be justified.

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