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CASE REPORT

Sciatic nerve enlargement in the Klippel-Trenaunay–Weber syndrome

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Summary The case of a 35-year-old woman with Klippel–Trenaunay–Weber syndrome (KTWS) showing clinical symptoms of a peroneal nerve lesion is presented. An immense nerve enlargement along most of the sciatic, peroneal and tibial nerve was found to be due to a lipoma arising from the epi- and perineurium. Treatment consisted of extensive microsurgical neurolysis and excision of the tumor resulting in decompression of the affected nerves. Although rare, a perineural lipoma should be kept in mind in patients with KTWS showing neurological abnormalities.

The Klippel–Trenaunay–Weber Syndrome (KTWS) is a symptom complex consisting of cutaneous vascular nevi (port-wine stains, hemangiomas, or lymphangiomas) of one extremity, venous varices which appear early on the affected limb and local hypertrophy of the bone and soft tissues of the affected part.[15] To a much lesser degree visceral hemangiomas involving organs such as the gastro intestinal tract, liver, spleen, bladder, kidney, lung, and heart also occur. The syndrome is thought to result from in utero insult early in gestation,[16] although the exact cause of KTWS still remains unknown. A genetic basis has been confirmed and many authors have grouped KTWS with neurocutaneous inherited disorders of phakomatosis, including von Hippel-Lindau disease, neurofibromatosis, Sturge-Weber syndrome, and tuberous sclerosis. Nerve involvement is rarely seen in this disorder, although it has been described in association with the KTWS.[2,3,7] The current report should be of interest, since it documents for the first time an extensive enlargement of several nerves namely, the sciatic, peroneal and tibial on the affected limb in a patient with KTW syndrome.

Case report

A 35-year-old woman was referred to our clinic because of progressive weakness of her right lower leg, which began in 1995 and slowly progressed until

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she had severe troubles in walking. Past history revealed the stigmata of KTWS. In 1987 lipectomy had been performed on her right lower leg because of progressive enlargement, and a reduction of the fourth toe, which had shown signs of bony and soft tissue hypertrophy. In addition, in 1995 she was operated on, because of a melanoma on the right lower leg, which was excised in toto, postoperative had been uneventful. Clinical examination revealed the signs of a complete peroneal lesion. The patient was unable to lift the toes of her right leg and showed reduced sensibility in the first interdigital space.

A MRI revealed a significant swelling of the right sciatic, peroneal and tibial nerve throughout the course in the upper and lower leg (Fig. 1). The diameter of the right sciatic nerve and its branches was found to be three times that of the contralateral sciatic nerve. An additional nerve sonography confirmed these findings (Fig. 2).

Motor nerve conduction studies were performed on the right peroneal and tibial nerve using conventional procedures. For the tibial nerve motor nerve conduction velocity was 34 m/s and the amplitude of compound muscle action potentials was 2.3 mV. No conduction parameters could be detected from the right peroneal nerve; likewise, the antidromic sensory nerve conduction studies performed on the right sural nerve showed no response.

Surgery consisted of extensive microsurgical neurolysis of the sciatic nerve and its branches starting in the proximal upper leg proceeding to the proximal third of the lower leg. The immense nerve enlargement (Fig. 3) was found to be due to a tumor-like fatty growth arising from the epi- and perineurium. Histological examination of the biopsies taken was consistent with the findings at surgery and revealed a lipoma.

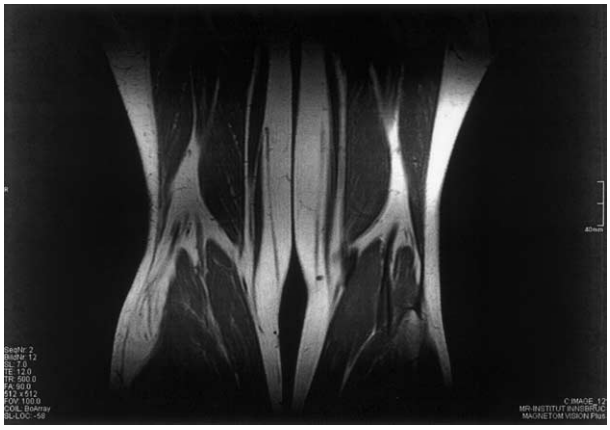

Figure 1 MRI revealed a significant swelling of the right sciatic, peroneal and tibial nerve.

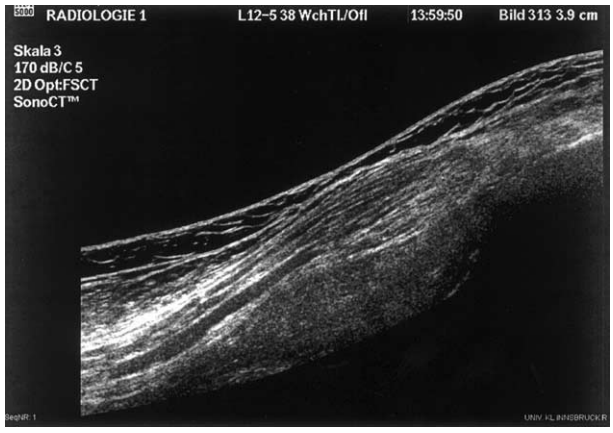

Figure 2 Nerve sonography confirmed a swelling of the sciatic nerve.

Postoperatively the patient recovered well and showed improved function just days after surgery with less difficulty in walking. However, the follow-up period is yet too short for a final nerve conduction study and a conclusive clinical evaluation.

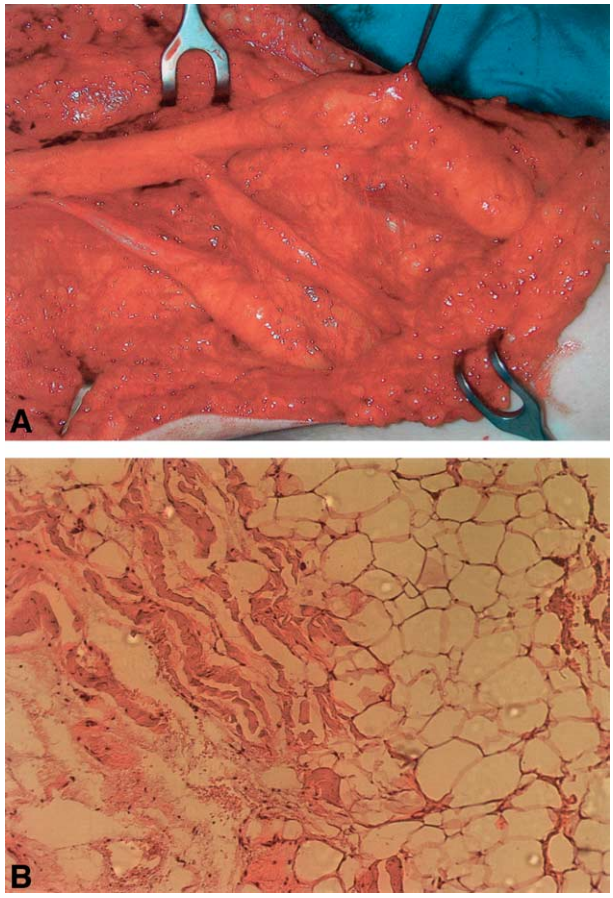

Figure 3 Massive enlargement of the sciatic nerve and its branches in the popliteal fossa.