Perineuriomas

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Perineurioma is a rare tumor entity. Children and young adults are particularly affected [1]. There is no gender difference in the frequency of occurrence [2]. The tumor mainly affects the large nerves of both the upper and lower extremities with equal distribution [2]. Due to the gradual course of the disease, the diagnosis is usually made late, if at all [3].

17.1 Symptoms

Typically, a motor mononeuropathy exists. The patients show a slowly progressive paralysis with muscular atrophy (see Fig. 17.1), but only in rare cases is there a sensory deficit.

Pain is often absent. The symptoms are said to be caused by compression of the axons due to the increase in neoplastic tissue [4].

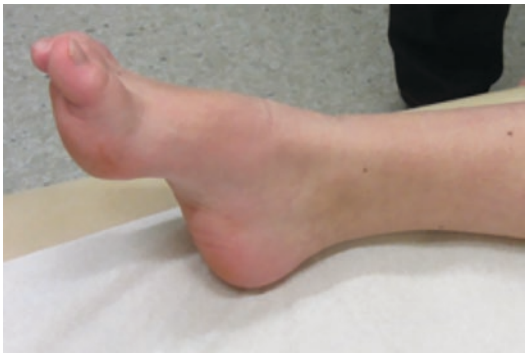

Fig. 17.1 Neurogenic clubfoot as a result of a perineurioma of the right sciatic nerve

17.2 Pathology

Perineuriomas are considered benign nerve tumors. They can occur within a nerve (intraneural) or manifest extraneurally as a soft tissue tumor. However, these rarely have a direct relationship to the nerve. Subtypes are the sclerosing and the reticular perineurioma. The malignant perineurioma is very rare and originates exclusively from extraneural perineuriomas [1]. Intraneural perineuriomas, on the other hand, do not undergo malignant transformation and are very slow growing. Perineuriomas are associated with anomalies of chromosome 22, in particular monosomy or deletion of the 22q11–q13.1 bands [5].

Macroscopically, perineurioma appears as a distinct hardening and spindle-like thickening

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of the affected nerve section. Histological findings are long, thin tumor cells with bipolar cytoplasmic processes with wavy or tapering nuclei [1]. The perineurioma cells infiltrate the endoneurium of the affected fascicle and form concentric whorls of perineural cells (pseudo-onion bulbs) around the nerve fiber. Immunohistochemically, a positive reaction to the epithelial membrane antigen (EMA) is, by definition, observed [6]. The perineurioma cells show no expression of the Schwann cell marker S-100. Claudin-1 and GLUT, however, are often expressed. Mitosis can occur, but necrosis is absent, as a rule [1].

17.3 Diagnostic

The gold standard in perineurioma diagnostics is MRI (ideally 3 Tesla). MRI shows long-distance enlargement of fascicles within the affected nerves and an intense, homogeneous gadolinium enhancement [3, 7, 8] (see Figs. 17.2 and 17.3). Affected fascicles appear isointense on T1- and hyperintense on T2-weighted images [3, 7, 8]. Due to its characteristic appearance, perineurioma can be distinguished from the most common differential diagnosis (chronic inflammatory demyelinating polyneuropathy/mononeuropathy) on MRI [3]. The high-resolution ultrasound can describe the extent of the tumor relatively precisely just like the MRI [9]. Both methods are also used in follow-up care [10]. In electrophysiological studies, amplitude reductions, conduction blocks, as well as denervation signs may occur in EMG.

17.4 Therapy Options

The therapy of perineuriomas is not undisputed. Due to the small number of cases, general recommendations are not available. As there is usually no complete loss of function of the affected nerve, there is a risk of functional impairment through surgery.

However, the risk of malignancy does not seem to exist [4].

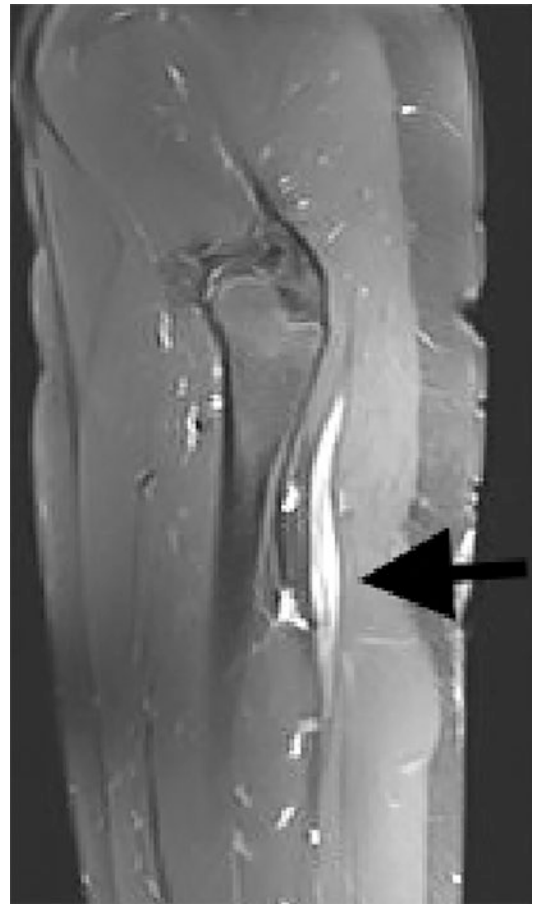

Fig. 17.2 Example of an intraneural perineurioma of the right sciatic nerve (arrow) in MRI (sagittal) after application of gadolinium

In addition to the possibility of merely observing the tumor by performing regular imaging and electrophysiological examinations, there is the alternative of surgical therapy. Here, too, various surgical strategies are available.

In order to confirm the diagnosis, at least one biopsy of an affected and non-functional fascicle should be taken [11] (see Figs. 17.4 and 17.5). Based on the theory that the axons are compressed by the tumor cells, a decompression of the fascicle by an epineurotomy makes sense, even though there are not yet studies to date able to demonstrate its effectiveness.

Another possibility is the complete resection of the tumor with subsequent reconstruction by nerve grafting [3, 4, 10, 12]. The resection

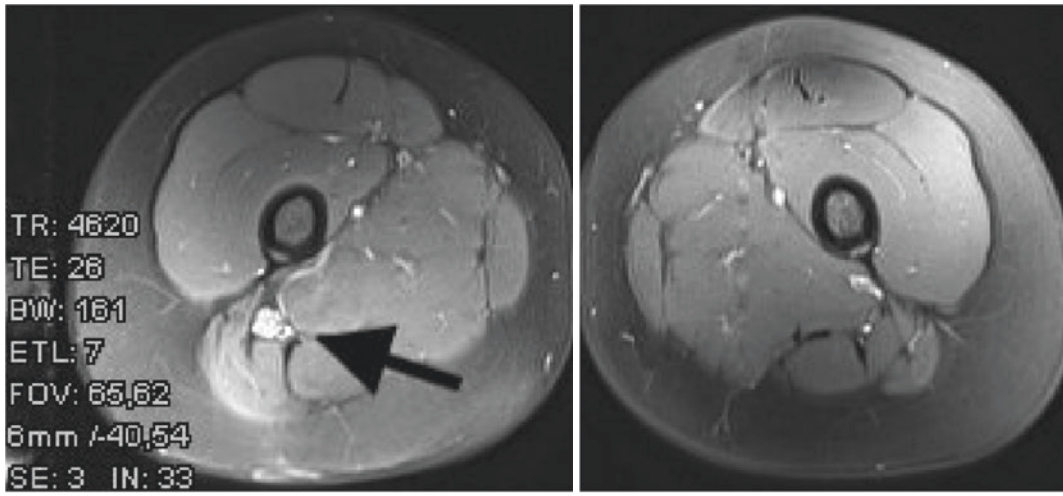

Fig. 17.3 Intraneural perineurioma of the right sciatic nerve (arrow) in MRI (axial) after application of gadolinium

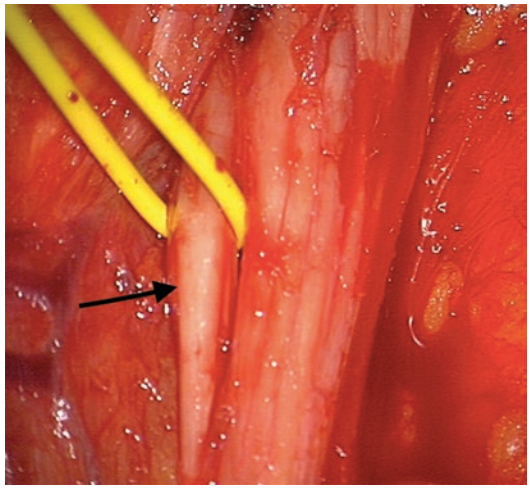

Fig. 17.4 Thickened, non-functional fascicle of the sciatic nerve (arrow) before biopsy

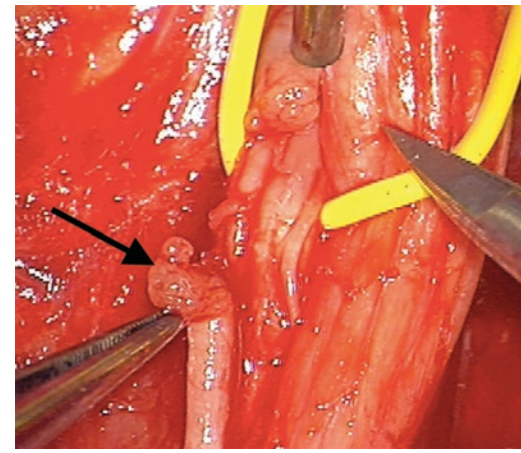

Fig. 17.5 Thickened, non-functional fascicle of the sciatic nerve (arrow) after biopsy

margins must be tumor-free to prevent respreading. For this, the possibility of a frozen section analysis must be available [10]. However, the presence of further tumor cells at other sites within the nerve cannot be ruled out either intraoperatively or by imaging. A complete resection of the tumor can therefore not be guaranteed. In addition, it should be noted that a complete loss of function after resection of the tumor and subsequent transplantation is initially very likely.

A third option is a tendon transfer to improve the reduced motor function. The tendon transfer can be performed in combination with or without tumor resection. Also, in certain cases, a distal nerve transfer might be possible to restore function, for example, distal anterior interosseous nerve to ulnar motor transfer if the tumor involves the proximal ulnar nerve.

Regardless of the surgical method, the surgical microscope and intraoperative nerve stimulation as well as micro-instruments should be used intraoperatively. The high-resolution ultrasound for planning the skin incision and showing the tumor extent can be helpful. The determination of the nerve conduction velocity can also be useful.

As a rule, it is not necessary to install a drainage system. We recommend elastic wrapping of the corresponding extremity. The bandage should be changed after 2 h. If no transplantation has been performed, the extremity can be mobilized immediately [10].

17.5 Results

Long-term data are not available due to the small number of studies with low case numbers. Mauermann et al. describe in a study of 23 patients with confirmed perineurioma that no significant progress was observed within 45 months. Based on these data, they favor a conservative approach [11]. In a retrospective study with 20 patients, Wilson et al. were able to show that intraneural perineuriomas rarely gain length on MRI. In addition, they do not spread to other nerves [8]. Restrepo et al. do not consider a biopsy to be necessary in the case of characteristic MRI findings with a suitable clinical course and symptoms and recommend regular follow-up examinations [3]. Gruen et al., however, performed a complete resection of the tumor with subsequent transplantation in 15 patients [13]. Ultimately, a therapy concept must be worked out individually after weighing all risks with the patient concerned.

17.6 Follow-Up Treatment

After diagnosis, we recommend the next checkup 3 months later. In addition to a current 3 Tesla MRI with gadolinium, an electrophysiological examination should be performed. If worsening of symptoms should occur, the therapy regime must be re-evaluated.

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