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Selective Surgical Treatment of the Intraneural Perineurioma of the Femoral Nerve by Tumor Resection and Graft Repair

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Abstract

Background: The perineurioma is a rare nerve tumor. The etiology is unknown and only a small number of case reports can be found in the literature, which includes proposals of conservative as well as surgical means of treatment. The current objective is to present an effective therapeutic strategy in treating this disease, which led to complete recovery.

Case: A 90% lesion of the femoral nerve of an 18-year-old female was diagnosed as perineurioma, because after immunohistochemical staining, EMA-positive and S100 protein-negative cells were detected in the pseudo onion. Then, under intraoperative neurography, the pathological fascicles were selectively resected, while the healthy fascicles were left in situ. The nerve defect was reconstructed by sural nerve grafts.

Three years after treatment, neither clinical nor electroneurographical evidence of recurrence or progression of the perineurioma was observed. The patient was pain-free and the nerve worked well.

Conclusion: This case indicates that selective surgical resection of the perineurioma guided by intraoperative electroneurography with simultaneous reconstruction of the nerve enables reinnervation of the femoral muscles.

Introduction

The intraneural perineurioma, known as hypertrophic neuropathy [1], is a rare [2] benign nerve tumor of unknown pathogenesis [3], which might be caused by toxic or traumatic factors [4]. Its progression favors [5] more and more real neoplastic lesions [6,7], but reactive processes such as infections have never been observed [3]. The tumor shows a characteristic cellular morphology. Proliferating cells are Epithelial Membrane Antigen (EMA)-positive perineurial cells and are not derived from Schwann cells.

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Until now, reliable guidelines have not been established for treating this nerve pathology [1,8], which affects the peripheral as well as, though more rarely, the cranial nerves in a localized area destroying them within a few years. More than 120 perineuriomas have been reported in the literature [9,10]. Typically, they show onion bulb formations in areas of main nerves such as in brachial plexus [11,12], peroneal nerve [1,13], ulnar nerve[1,14], median nerve [15,16], radial nerve [1,17], sciatic nerve [18,19], tibial nerve [20,21] and trigeminal nerve [2,15,22-24]. Only four publications have reported a perineurioma in the femoral nerve [1,25-27].

Thus far, most of the authors described individual cases. Gruen et al. [18] published a follow-up report examining 15 patients. Nine of these patients were treated by resecting the tumor of the nerve and reconstructing the nerve using sural nerve grafts. One year after treatment, seven of these nine patients showed either the same or better nerve function than before. Also, Emory et al. [1] published the treatment of eight patients. Six of these patients were managed by a conservative strategy. The remaining two patients were treated by resection of the nerve and reconstruction using nerve grafts; in both cases, there was no recovery of nerve function. Therefore, in the present case it was decided to resect the pathological fascicles selectively and to reconstruct the nerve defect (N. femoralis) with sural nerve grafts. The long-term follow-up showed complete recovery of the femoral nerve and no evidence of recurrence of the perineurioma.

Case Report

History and physical examination

According to the 18-year-old female patient's statement, the first signs of the disease were indicated by a sudden but transient pain-free weakness of the left leg. The knee joint gave way without disturbance of sensibility, but the weakness dissipated within two days. Following this episode, the patient occasionally experienced a mild pain in the left thigh, radiating to the knee after prolonged sitting or while rising. During this period, the knee gave way intermittently. Especially after aerobic exercises or when climbing stairs, weakness of longer duration was felt in the left leg. In the later course of the disease, paraesthesia and pain in the groin radiating to the calf developed as well. Two years after the first feeling of leg weakness, the patient had to walk on crutches-her quadriceps muscle had become too weak and the pain too intensive. The clinical examination showed obvious atrophy of the left quadriceps muscle; the circumference of the left thigh was 5.2 cm smaller than that of the right one. The patellar tendon reflex was absent, and the left knee joint could not be extended. Strength of the quadriceps femoris muscle was M1. Electromyographically positive waves, fibrillation potentials, and a poor number of motor unit potentials were recorded. Herniation of an intervertebral disc and other spinal lesions were excluded. MRI showed the femoral nerve in the T2 weighted sections to be hyperintensive over a distance of about 3 cm above the inguinal ligament and slightly thickened as compared to the other side (Figure 1). The quadriceps muscle was atrophied and showed fatty degeneration.

Surgery

The femoral nerve was exposed retroperitoneally in its whole length from the muscular lacuna to the iliopectineal arcus and further to the division between the iliac muscle and the psoas muscle. Macroscopically, a small thickening with hypervascularity (Figure 2) was visible 6 cm above the inguinal ligament. Three small vessels crossing the nerve at a 90° angle appeared on the ventrolateral side, and a 1 cm long lesion could be observed 6 cm proximal to the inguinal ligament. During surgery, neurography revealed temporal dispersion of the excitation potential of the nerve proximal to the visible changes. The dispersion increased considerably in the area of thickening. After longitudinal epineuriotomy and microscopic assessment, 2 thickened fascicles were identified beside others of normal caliber. The selective neurography of these fascicles disclosed a complete loss of fast conducting nerve fibers and a dispersed nerve action potential of slowly conducting fibers, indicating a demyelinating process (Figure 3). Another two fascicles, however, appeared macroscopically normal and showed normal nerve potentials. 4 cm of the pathological fascicles were resected in order to ensure resection of as much of the tumor as necessary to avoid recurrence. The resection was controlled macroscopically and neurographically. The two healthy fascicles were left in place. After resection, the defect was bridged by two sural nerve grafts of 4.2 cm; each of the grafted nerves were coapted tension free and secured with a 11/0 nylon suture (Figure 4).

Tumor characterization

Microscopically, the whole length of the excised nerve tumor showed endo and epineural areas of hypervascularization and edema. Myelinated nerve fibers were considerably reduced in number, and many onion bulb like formations were seen in the thickened area of the nerve. Rare Schwann cells were found in the center of the onion bulb like whorls. Immunohistochemical examination showed the pseudo onion bulb consisting of EMA-positive and S-100 protein-negative cells (Figure 5) [5]. Electron microscopy supported the diagnosis of a perineurioma of the femoral nerve, as it revealed a high content of pinocytic vesicles [5,7] and the common occurrence of tight junctions. Despite normal gross appearance under the operating microscope and normal findings on palpation, the ends of the resected nerve were not healthy, as predicted by the intraoperative neurography of the proximal nerve edge.

Postoperative course for three years

Postoperatively, the femoral nerve exhibited complete paresis as compared to a 90% preoperative function. Pain was increased initially, depending on the position of the leg. This pain, however, was completely resolved after four months. Two months after surgery, the Hoffmann Tinel sign was found just below the inguinal ligament. Five months after the surgery, the patient was able to walk without crutches. After 18 months, she was able to extend the leg against gravity and resistance. The gait was normal and she resumed aerobic activities. Three years after the operation, there are no clinical and neurographical signs of recurrence of the perineurioma. MRI examination of the left femoral nerve showed no abnormalities, neither two nor three years after surgery.

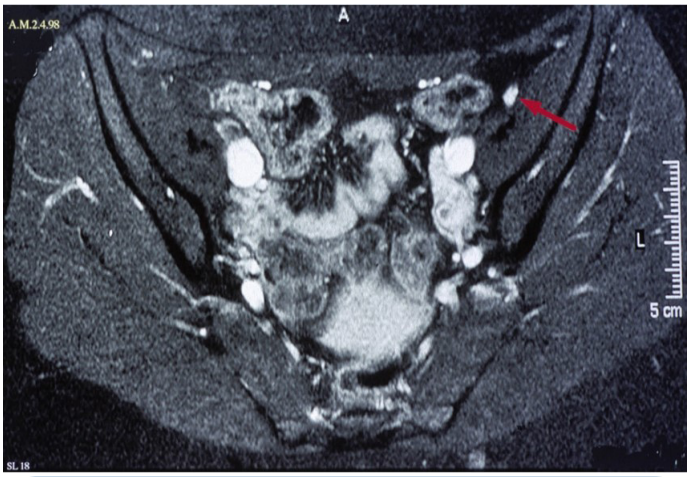

Figure 1: MRI shows the femoral nerve in the T2 weighted sections to be hyperintensive for roughly 3 cm above the inguinal ligament and slightly thickened (red arrow).

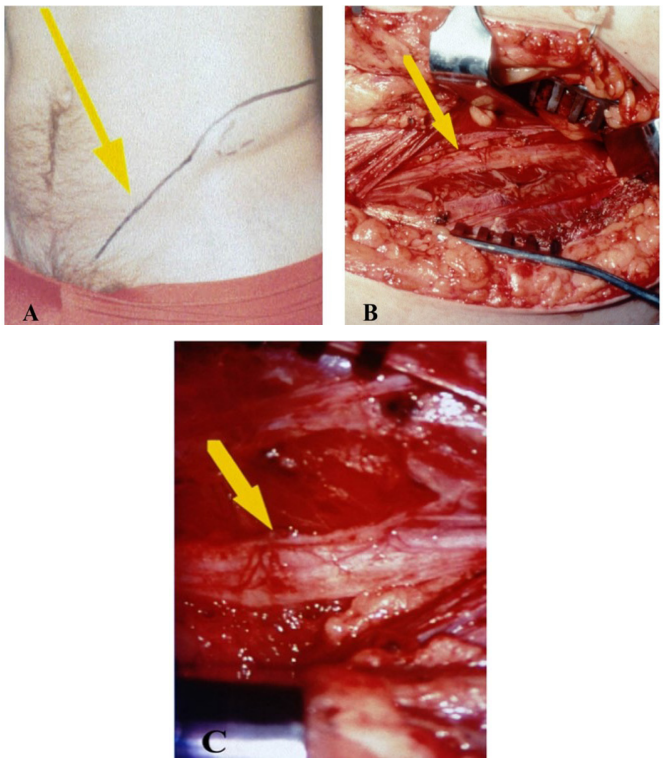

Figure 2: Surgical incision for N.femoralis (yellow arrow) perineurioma (A), epineural areas of hypervascularization and edema of perineurioma (B, C higher magnification).

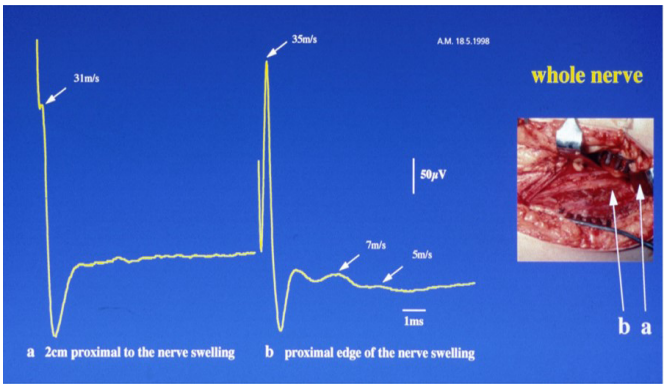

Figure 3: Intraoperative neurography, a helpful method, which gives the surgeon valuable and guiding hints.

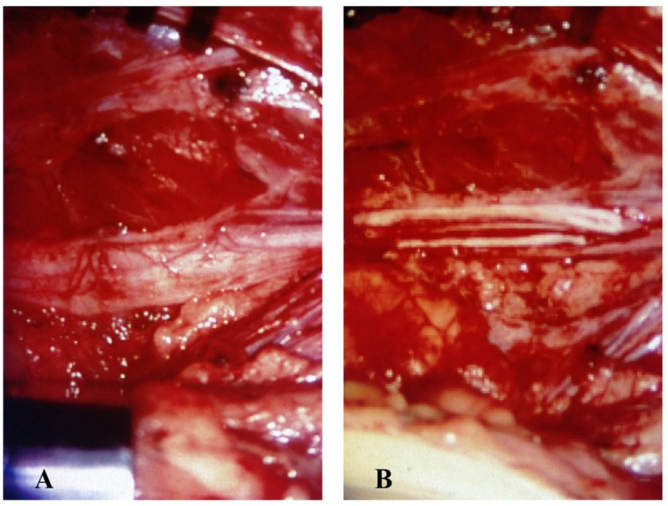

Figure 4: The pathological fascicles **(A)** were selectively resected, while the healthy fascicles were left in situ. The nerve was reconstructed by transplanting the nervus suralis **(B)**.

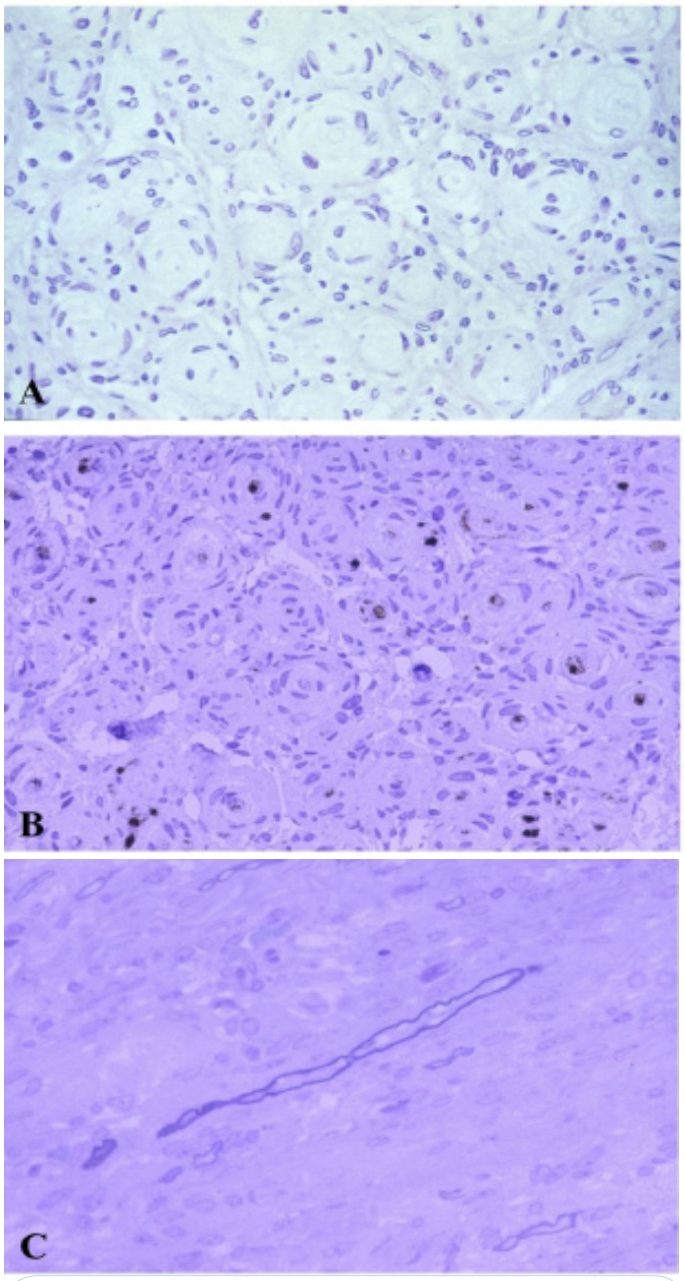

Figure 5: EMA-positive **(A)** and S100 protein-negative cells **(B)** were detected in the pseudoonion (A, B cross section, C longitudinal section).

Discussion

Until now, there has been no recorded evidence for spontaneous regression or standstill of a perineurioma. Perineuriomas do not metastasize. Their recurrence has been noted up to six years [5] after surgical treatment.

Various possibilities of treatment of perineuriomas are currently being discussed. Some authors [3,8] estrict their treatment to neurolysis and diagnostic nerve biopsy as long as nerve function is observed. Emory et al. [1] as well as Gruen et al. [18] have treated this tumor using nerve transplantation. The first group did not obtain functional improvement of the nerve in the two patients treated using nerve resection. The second group reported partial success, as three of seven patients' conditions improved and four remained unchanged. The authors concluded that resecting the nerve and substituting it by a transplant is superior to biopsy or neurolysis, which usually induces a complete loss of the nerve function. It is recommended to identify the involved fascicles and determine the necessary extent of their resection by intraoperative Electro Nuro Graphy (ENG).

This report confirms that the tumor slowly progresses over years if it is not, or incorrectly, treated [2,18]. As observed during surgery, the tumor had grown up to macroscopic visibility. However, only two of four fascicles were involved [18], this was demonstrated by intraoperative neurography, a very helpful method which gives the surgeon valuable and guiding hints. Defining the tumor's border was difficult because of a considerable subclinical spread of the tumor. After 4 cm of the nerve bearing the 1 cm long tumor had been resected, a subclinical spread had no longer exhibited influence on the postoperative result. Whether or not a too narrow resection of the nerve bearing the tumor is the reason for the poor results reported in some cases of nerve resection and transplantation remains open [1,3,6,8,24].

A significant and useful reinnervation of the femoral nerve after selective fascicle resection and transplantation had also been achieved. The patient became pain-free and, following nerve regeneration, recovered from the paresis of the quadriceps muscle within 3 years. Thus, the present case confirms the view of Gruen et al. [18] that tumor resection and grafting is the best treatment of perineurioma. If the nerve is only partially affected, selective resection of the affected fascicles may be performed.

Perineuriomas must be precisely distinguished from other neuropathies. The differential diagnosis must be established with great care, taking into account the patient's history, clinical findings, concurrent or generalized diseases, as well as MRI investigations. Intraoperative neurographic evaluation is recommended.

Conclusions

An untreated perineurioma may lead to a complete functional loss of the involved nerve. In the present case, this could be prevented by selective resection of the tumorous nerve fascicles and restoration of the nerve function by nerve transplantation. Unsuccessful cases after surgical treatment as reported in literature may be due to insufficient resection of the involved nerve segments.

For the future treatment of this disease, intraoperative neurography to identify affected nerve tissue and extension of the changes of the nerve is suggested.

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