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Case Report

Intraneural Ulnar Nerve Ganglion: A Surgical Case Report of a 10-cm-Long Recurring Ganglion Cyst in the Forearm

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Keywords
Ganglion · Ulnar nerve · Surgery

Abstract
Introduction: Intraneural ganglions are benign and rare mucinous cysts that originate within peripheral nerves and typically can lead to symptoms and signs of peripheral neuropathy. The most common location is the peroneal nerve, and the second most common location is the ulnar nerve. ***Case Presentation:*** We present a case of a 53-year-old man who presented with increasing numbness in the ulnar aspect of the left hand and decreasing hand strength. MRI showed an intraneural ganglion, and as the patient had clinically progressive symptoms, a decision was made for surgical excision. The patient was symptom-free after the procedure and had no neurological deficits. Eighteen months later, the patient contacted us again as his symptoms had returned. A new MRI showed ganglion recurrence. Due to progressive clinical symptoms, another attempt was made to remove the ganglion surgically. Paraffin immunostains excluded other diagnoses like synovial cyst, posttraumatic neuronal cyst, Tarlov cyst, mesothelial cyst, and cystic lymphangioma. At follow-up 3 months postoperatively, the patient was symptom-free and had normal neurological findings. ***Conclusion:*** Intraneural ganglion should be considered as a differential diagnosis of a cystic mass close to a nerve. For surgery, we favor less radical methods, such as simple decompression.

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Introduction

Intraneural ganglions are benign mucinous cysts that originate within peripheral nerve epineurium and typically can lead to symptoms and signs of peripheral neuropathy. They are uncommon and occur within the epineurium of nerves. The most common location is the peroneal nerve, and the second most common location is the ulnar nerve. The pathogenesis is controversial and poorly understood. There are three major theories to explain formation of intraneural ganglion cysts: the degenerative theory, the tumoral theory, and the synovial (articular) theory [1, 2]. The de novo model of cyst formation attributes the formation of intraneural ganglions to degenerative changes within the epineurium or perineurium, leading to cyst formation within the nerve sheath. Spinner and colleagues introduced the unifying articular/synovial theory of formation of intraneural ganglion [1]. They showed that peroneal intraneural ganglia originate from the superior tibiofibular joint and that the articular branch of the peroneal nerve can serve as a conduit for the cyst fluid to pass from a capsular defect in the joint. Desy et al. [3] reviewed 417 articles describing 645 cases of intraneural ganglia. Approximately half of the cases had an articular branch to the nearby joint. Six of the ulnar nerve ganglion in the hand or wrist had a branch (38%). There was a statistically significant association between cyst recurrence and percutaneous aspiration as well as failure to disconnect the articular branch or address the joint. Spinner found that only 23% cases had joint connections identified for intraneural ganglion cysts at the wrist [1].

Different surgical approaches have been recommended to remove the intraneural ganglion cyst. Outcomes have been disappointing, and the recurrence rate is high and probably underreported. Historically, some surgeons have suggested resection of the nerve and nerve grafting as an option for treating these lesions. These days, most authors recommend a simple excision of the articular branch and decompression of the cyst [1, 4, 5], arguably an easier and equally effective procedure. Considering the fact that on average only one-third of the ulnar intraneural ganglions have a branch, this poses difficulties for the surgeon [3]. The fact that a high rate of recurrence has also been described complicates matters and should be included in the decision-making process when choosing the intervention [3]. A ganglion cyst is really a pseudocyst because it has no true lining. It should be distinguished from cystic lesions with lining, such as posttraumatic neuroma, cystic lymphangioma, perineurial/Tarlov cyst, and cystic metastasis of neuroendocrine tumor [6].

Case Presentation

We present a case of a 53-year-old man who was referred to our unit with increasing numbness on the ulnar aspect of the left hand and decreasing hand strength. He had initially been treated at another hospital for suspected compression of the ulnar nerve in Guyon's canal with open release of the nerve. During this surgery, cystic changes to the ulnar nerve were noted, and the wound was closed. The patient thereafter underwent an MRI before being referred to our unit for further treatment. There was a clear difference to the unaffected right hand in terms of 2PD and strength, with the muscles of the left hand innervated by the ulnar nerve being particularly affected.

MRI showed an intraneural ganglion (Fig. 1a). The radiology report was somewhat ambiguous regarding a possible branch to the distal radioulnar joint but could not rule out that there could be an articular branch at the level of the branching superficial sensory branch of the ulnar nerve – main ulnar nerve. As the patient had clinically progressive symptoms, the decision was made for surgical excision.

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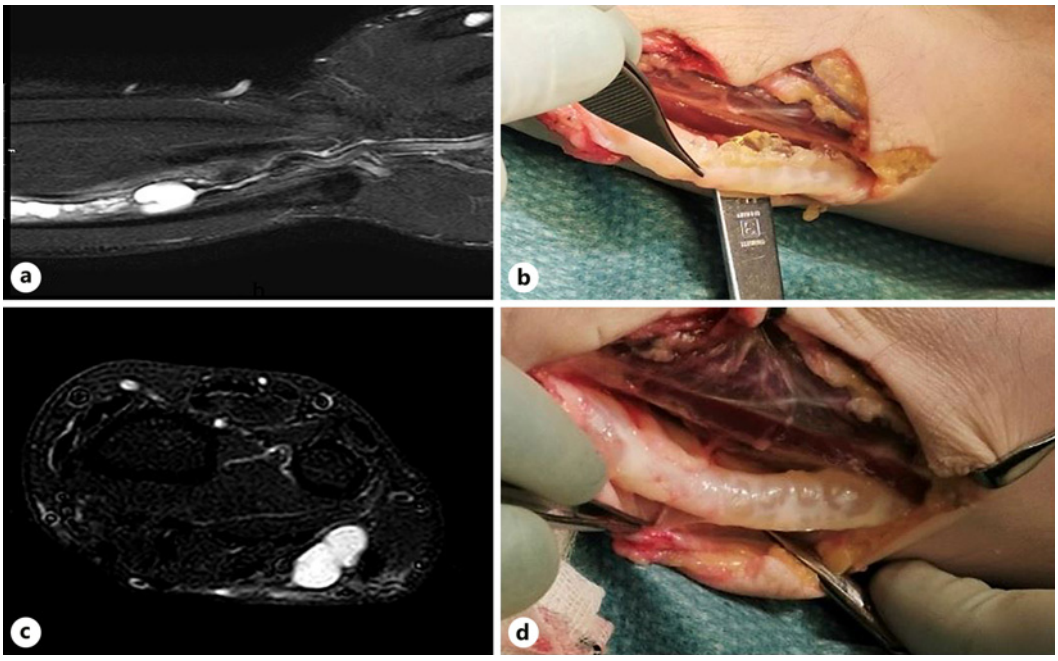

Fig. 1. a, **c** Preoperative MRI. **b**, **d** Findings during primary operation at our unit.

The intraoperative finding showed an intraneural ganglion more than 10 cm long, which ran in the ulnar nerve in the forearm (Fig. 1b, c). The distal end was located 3.5 cm proximal from the wrist. The ganglion was excised surgically under loupe magnification after decompression of the ganglion under the premise of not damaging nerve tissue and therefore not necessarily radical. Intraoperatively, we could not identify an articular branch, despite a careful exploration of the area around the branching of the superficial sensory branch – main ulnar nerve. The patient was symptom-free after the procedure and had no neurological deficits. Microscopic analysis showed dense collagenous wall surrounding foci of myxoid degeneration admixed with stellate myofibroblasts. The cysts had no true epithelial lining. No cytologic atypia and/or mitotic activity were noted (Fig. 1d).

Eighteen months later, the patient contacted us again as his symptoms had returned. A new MRI showed ganglion recurrence. No articular branch could be identified on the MRI.

Due to progressive clinical symptoms, another attempt was made to remove the ganglion surgically. The incision was extended distally and proximally beyond the area of the actual intraneural pathology. As was the case during the primary operation, no articular branch of the nerve into the joint capsule could be identified. We removed the cyst as thoroughly as possible without damaging nerve tissue after decompression (Fig. 2a, b). Tissue from the ganglion was sent to pathology and showed a nerve with residual myxoid degeneration and cyst formation (Fig. 2c, d). Paraffin immunostains were performed. The synovial marker CD68 was negative (Fig. 3a), ruling out a synovial cyst. The S100 stain (neuronal marker) was normal (Fig. 3b), arguing against posttraumatic neuronal cyst. The mesothelial and perineural marker EMA was positive in the cyst wall but not in the cyst surface (Fig. 3c), ruling out Tarlov cyst and mesothelial cyst. Lastly, the lymphendothelial marker D2–40 was also negative in the cyst surface (Fig. 3d), excluding cystic lymphangioma. The patient was followed clinically for 3 months postoperatively. He had normal neurological findings, including equal 2-point discrimination in all digits and improved strength with clinically no signs for a recurrence.

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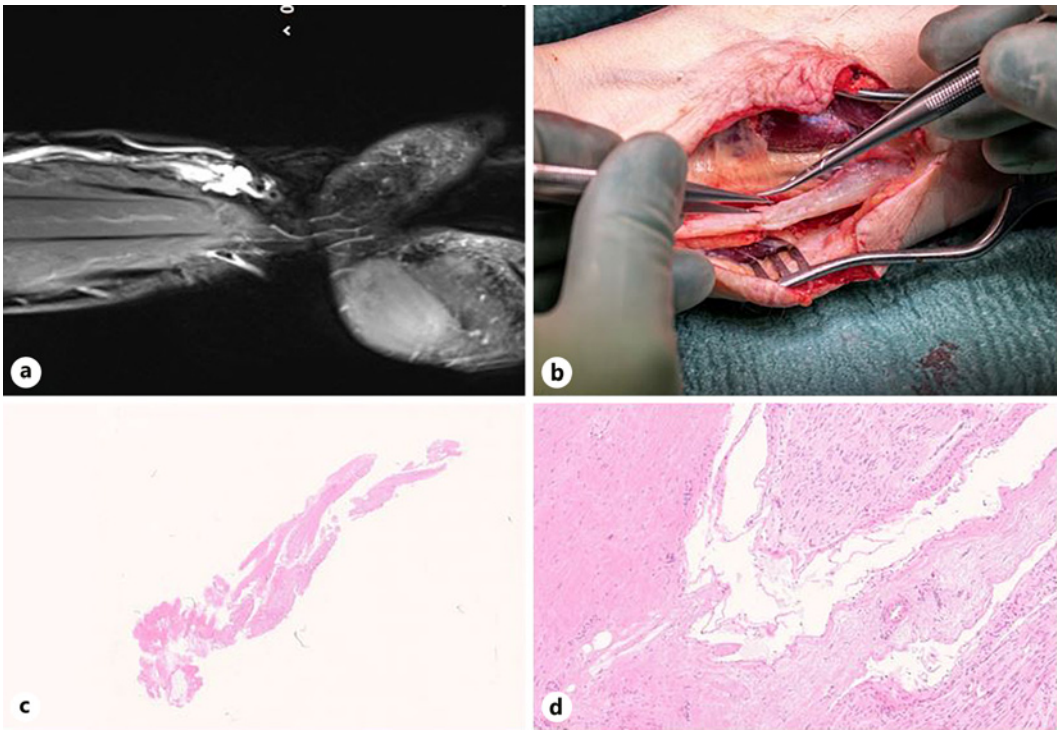

Fig. 2. a MRI after recurrence. **b** Findings during reoperation. **c** Low power view of the whole specimen. **d** Myxoid degeneration (ganglion cyst).

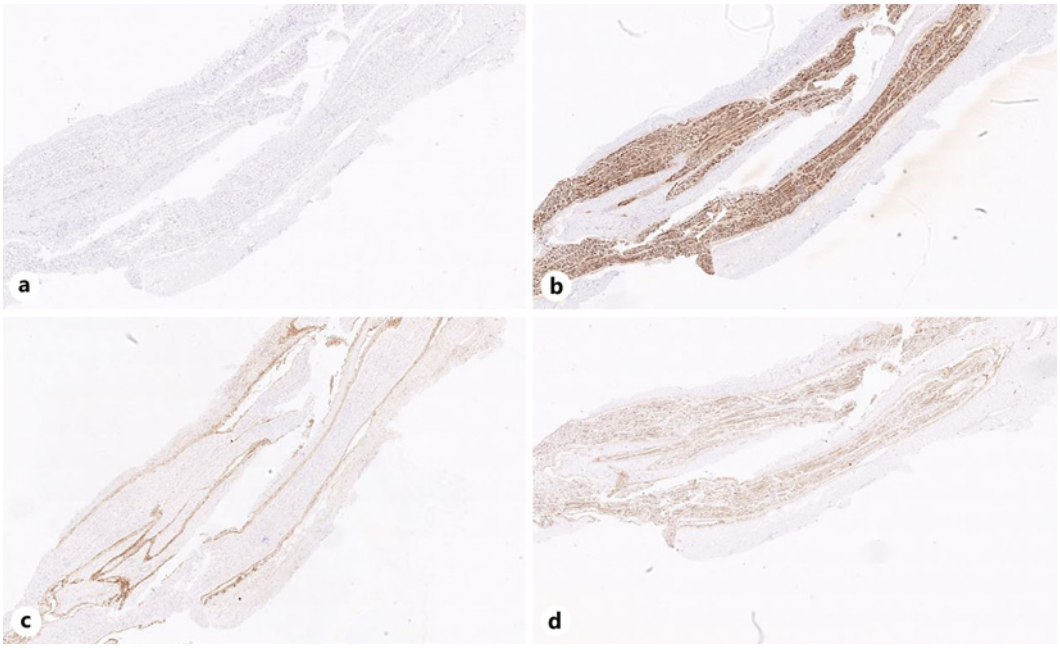

Fig. 3. a CD68. **b** S100. **c** EMA. **d** D2–40.

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Discussion

According to the synovial (articular) theory, many authors recommend dissection of the articular branch of the nerve and decompression to treat the cyst, assuming that the branch is the origin of the cyst [4, 5]. Others have found no difference in outcome between patients who underwent cyst excision and those who underwent simple articular branch excision and cyst decompression [1, 3]. Other options include surgical treatment as simple excision or ligation of the articular branch leaving the cyst in place to resorb on its own. It has been described that the risk of a recurrence increases if the articular branch is not addressed [4]. Nonoperative treatment has been mentioned as an option in asymptomatic/mildly symptomatic cases [3].

In our patient, the intraneural ganglion was approximately 10 cm in size with the distal end located 3.5 cm proximal from the wrist. The preoperative MRI findings indicated that a distal intra-articular branch could connect to the distal radioulnar joint. There may be a possibility that a distal intraneural branch was connected to the superficial branch of the ulnar nerve according to the MRI. During the first operation, a branch to the joint was looked for in the region described in the MRI, but none was found. At the reoperation, the incision was widened to ensure that no articular branch was missed. Again, no branch could be found. A small articular branch may have been unknowingly resected, but it may be that the branch was absent in this case. The reason for the recurrence is somewhat unclear. There may have been an articular connection after all, as there was scarring in the area from the initial surgery at the referring hospital. However, the surgery at our unit was undertaken by an experienced senior consultant. When cyst decompression is part of surgery plan, iatrogenic nerve damage can be avoided by simultaneous simple cyst incision and evacuation of the mucosal fluid to decompress the cyst instead of more radical interventions, such as complete cyst removal. We used this technique for both the first and second intervention and were able to avoid neurological deficits. No electromyography was undertaken, but it could be an option should the patient's symptoms return.

The case numbers of the studies in the literature are too small to statistically ensure that surgical targeting of the articular branch is a sufficient measure to treat the intraneural ganglion in the ulnar nerve. However, it can be concluded from the literature that only a minority of cases of intraneural ganglions in the ulnar nerve have an articular branch, despite the fact that most surgeons nowadays probably look closely for it. This may indicate that the majority of cases must be treated differently. Ganglion cyst is a diagnosis of exclusion; it is a pseudocyst with no lining. When no lining can be recognized in the H&E-stained sections, a presumptive diagnosis of ganglion cyst is made [7]. Paraffin immunostains are not routinely performed in these cases. Cystic lesions within the nerve include posttraumatic neural cyst, Tarlov cyst, mesothelial cysts, and cystic lymphangioma. For example, Gonzalez reported a case of intraneural lymphatic cyst of the ulnar nerve in the canal of Guyon [8]. These different forms of cysts not only have different pathogenesis, but they can also show different clinical courses and risks of recurrence. Not all intraneural cysts have articular branches since not all intraneural cysts are intraneural ganglion. Immunostains can be useful to verify the diagnosis of intraneural ganglion cyst.

Conclusion

Intraneural ganglion should be considered as a differential diagnosis of a cystic mass close to a nerve. For surgery, we favor less radical methods, such as simple decompression. Even if the risk of recurrence may be increased, the advantages with a probably lower risk of neurological deficits in our experience outweigh the possible increased risk of recurrence. Minimal surgical interventions also simplify any reoperations, as pronounced scar tissue

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increases the risk of nerve damage during reoperations. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000535613).

Statement of Ethics

This retrospective review of patient data did not require ethical approval in accordance with local guidelines. The patient has given his oral and written informed consent for publishing data and images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

D.R. treated the patient, conducted the investigation, and wrote the original draft. A.S. performed the immunostainings and microscopical analysis and reviewed the manuscript. M.S. contributed to project administration and review and editing.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author.

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