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Case Report

Neurofibroma Involving a Bifid Median Nerve in a Pediatric Patient: A Case Report

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Neurofibromas are benign peripheral nerve sheath tumors that typically develop within cutaneous nerve branches but can involve major nerves as well. They can be sporadic or associated with neurofibromatosis type 1. In this report, we describe the surgical treatment of a pediatric patient with neurofibromatosis type 1 presenting with a neurofibroma of a bifid median nerve. Involvement of the median nerve was not evident on preoperative examination or imaging, therefore altering the risk-benefit ratio of the procedure. After bifid nerve involvement was identified intraoperatively, the patient's parents were counseled on the risks and benefits of surgical excision before resuming the case. Ultimately, the neurofibroma was resected, and the patient experienced no neurological deficits after surgery.

Neurofibromas are one of the most common peripheral nerve sheath tumors (PNSTs).[1] They can occur sporadically but are often associated with neurofibromatosis type 1(NF-1), an autosomal dominant neurocutaneous disorder.[2] When presenting in the hand, they most often involve cutaneous nerve branches but can on occasion involve major nerves. These lesions are usually asymptomatic but can lead to localized pain or neurologic deficits. In this report, we describe a pediatric patient with NF-1 who presented with a neurofibroma of a bifid median nerve. Initially, involvement of the median nerve was not evident due to the atypical location of the lesion, absence of localizing neurological symptoms, and a nondiagnostic ultrasound.

Case Report

A 6-year-old girl with a medical history of NF-1 was referred to our clinic with three soft tissue masses as follows: a tender 6 mm mass on the dorsal/ulnar aspect of her right ring finger at the level of the proximal interphalangeal joint, a 13 mm mass on her right dorsal/ulnar wrist, and a tender, well-defined 15 mm mass on the volar/ulnar aspect of her left wrist, located midway between palmaris longus and flexor carpi ulnaris. Ultrasound demonstrated findings consistent with neurofibromas with suspected subcutaneous nerve branch involvement. No left median or ulnar nerve involvement was identified, and no anatomic variation was noted. During her preoperative visit, the patient experienced no peripheral sensory changes or motor weakness. In the setting of these ultrasound findings, her physical examination, and the absence of localizing neurological symptoms, it was believed that the masses were unlikely to involve any major nerve. The decision was made to surgically remove the masses from the right ring finger and the left volar wrist because these were the most symptomatic and were believed to carry a low risk of neurological sequelae.

Operative procedure

The procedure was performed with the patient in a supine position, with two hand tables and bilateral upper arm tourniquets. The right dorsal ring finger mass was addressed first. The neurofibroma was encountered in the subcutaneous plane and was noted to completely engulf the dorsal branch of the ulnar digital nerve. The nerve branch could not be dissected out from the lesion and was therefore sacrificed during excision.

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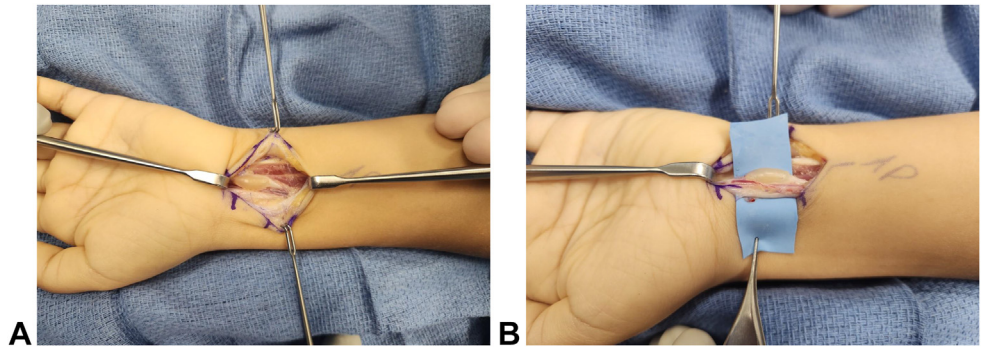

Figure 1. A, B Intraoperative visualization of neurofibroma of a bifid median nerve, with associated persistent median artery.

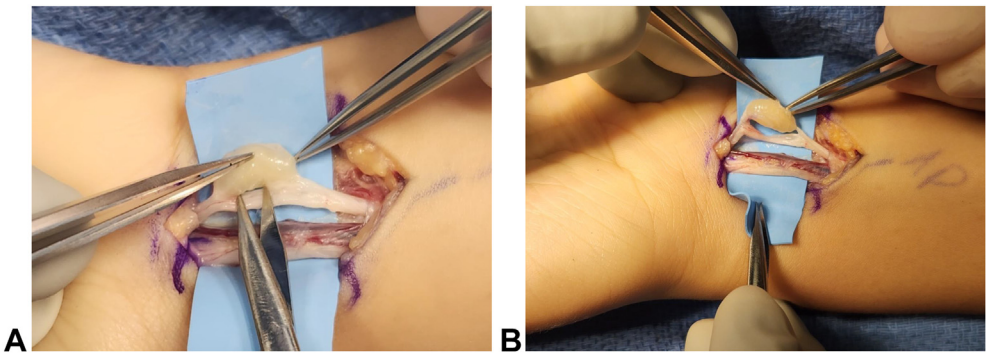

Figure 2. A Intraneural dissection of the neurofibroma. **B** Dissected neurofibroma enveloping ulnar-most fascicle of bifid median nerve.

Attention was then turned to the left volar wrist mass. A longitudinal incision was made over the volar distal forearm. A white, shiny, and firm nodule was visualized deep into the subcutaneous layer (Fig. 1A). The median nerve proper was identified radially, but the presence of a persistent median artery and bifid median nerve were subsequently appreciated. The mass involved the ulnar branch of the bifid median nerve, which accounted for approximately 40% of the entire width of the nerve (Fig. 1B).

After encountering this, the attending surgeon temporarily paused further dissection and held an extensive discussion with the patient's mother (in person) and father (over the phone) to review this unanticipated intraoperative finding. They were informed that the mass involved a portion of the median nerve itself and that neurofibromas often infiltrate nerves in a fashion that could make full excision difficult or impossible. The parents were presented with the option of continued observation, with the understanding that the mass could grow and become more symptomatic. Alternatively, the surgery could proceed with resection of the mass, with a not-insignificant risk of nerve injury and resulting residual deficit. The parents ultimately decided to proceed with resection due to concerns about the progression of the tumor. They deferred to the surgeon's judgment to abort the attempted excision if the probability of significant nerve deficit was deemed to be too high.

Intraneural dissection under 4× loupe magnification was initiated from the proximal to the distal aspect of the mass. The epineurium was incised, and the lesion was carefully dissected from the healthy nerve fascicles (Fig. 2A). At its distal aspect, the lesion was inseparable from the median nerve's most ulnar fascicle, which was sacrificed (Figs. 2B, 3A).

Both lesions were sent for evaluation by pathology, and histological findings were noted to be consistent with intraneural neurofibroma (Fig. 3B).

At the 2-week postoperative visit, the patient demonstrated minimal pain at the incision sites, and normal sensibility throughout the median nerve distribution was maintained. At her 2-month follow-up, the patient had no subjective complaints, and sensation in the left median nerve distribution remained preserved. Sensation in the fourth webspace was specifically confirmed to be intact. The remaining lesions will be monitored, with the patient reevaluated annually.

Discussion

Neurofibromas are common PNSTs that are often associated with NF-1. Neurofibromas can be histologically identified by the fusiform shape of the tumor cells and can be classified into three subtypes associated with peripheral nerve involvement. Localized cutaneous neurofibromas are the most common, arising from dermal Schwann cells and leading to cutaneous plaques that are present in nearly 100% of the patients with NF-1.[3] Localized intraneural neurofibromas, as diagnosed in this case, lead to fusiform, segmental lesions that develop within nerve fascicles and envelop axons. Finally, plexiform neurofibromas often involve multiple fascicles, have a distinct gross pattern of longitudinal, multinodular fusiform deformation of nerves, and can be highly infiltrative.[4]

Schwannoma (aka neurilemmoma) is a common PNST that must be differentiated from neurofibroma. It may occur spontaneously or be associated with neurofibromatosis type 2. Although the cell of origin for these tumors is also the Schwann cell, schwannomas initiate from a single fascicle within the main nerve, often have a well-defined capsule, and displace the rest of the nerve.[4] This is an important distinction when considering resection, as neurofibromas often engulf involved nerves and can blend into surrounding tissues, whereas schwannomas are generally well-

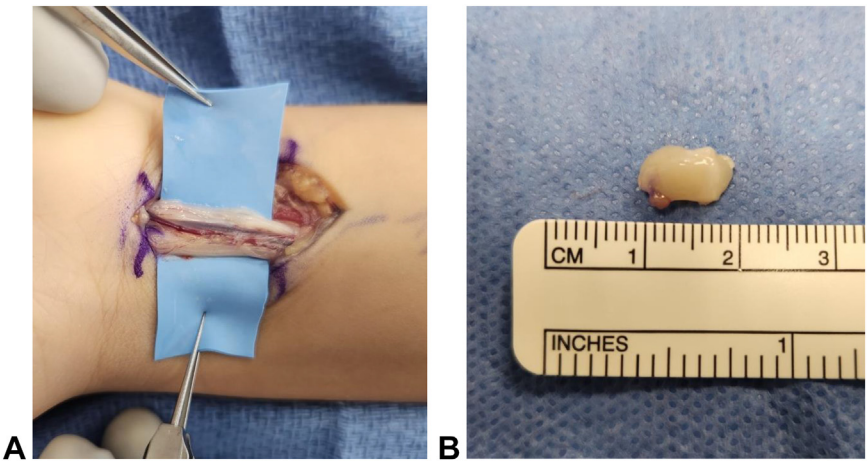

Figure 3. **A** Postresection visualization of bifid median nerve and persistent median artery. **B** Neurofibroma *ex vivo*.

encapsulated, located eccentrically to nerves, and therefore easily resected.[5]

This case presents a rare confluence of intraneural neurofibroma involving a bifid median nerve which, per the authors' knowledge, has not been previously described in the literature. Kougioumtzis et al[1] reported resection of a 6.5 cm neurofibroma involving a (non-bifid) median nerve where marginal dissection was possible due to the eccentric location of the tumor. Batista et al reported a 4 cm plexiform neurofibroma in the median nerve of a 3-year-old girl that was causing pain and motor and sensory deficits. After resection, the patient's sensation remained unchanged, but motor function was restored.[6] Gandhi and Bozentka describe a patient with bilateral plexiform neurofibromas of the PIN at the level of the wrist that were both resected with no resulting sensory deficit. The masses were minimally symptomatic and approximately 1.5 cm at initial presentation but enlarged to 3.5 cm and 5.5 cm before resection.[7] Successful resection in these cases, as well as our patient, demonstrate that surgical management of neurofibromas in major nerves can relieve pain and limit symptoms of tumor progression. The lack of new postoperative neurological deficits in these patients suggests that enveloped nerve fascicles can often be safely sacrificed during resection because they are clinically nonfunctional. This also implies that functional loss already sustained may not improve unless it is caused by the compression of adjacent fibers.

Regardless, surgeons and patients must weigh the risk of iatrogenic nerve injury against the advantage of preventing progression due to tumor size or transformation. It should be noted that NF-1 patients have an 8% to 13% chance of developing a malignant PNST in their lifetime.[8] Multiple factors including lesion location, size, symptomatology, and malignant potential should be considered when evaluating a patient for surgical intervention. In addition, preparedness for anatomical variation when resecting hand lesions can inform preoperative planning. When a major nerve or branch is involved, conservative management should be considered in the case of an asymptomatic lesion.

As evidenced by this case, ultrasound is a nonspecific modality for identifying neurofibromas and may be unreliable in identifying a bifid median nerve.[9] Despite this, the authors believe that it is a useful diagnostic tool that can be used to better characterize individual patient anatomy and pathology and usually differentiate nerve sheath tumors from other hand masses. MRI can also be considered, although it also cannot definitively differentiate neurofibromas from other nerve sheath tumors such as schwannoma.[10]

Standard surgical consent at our institution specifically includes an allowance for the surgeon to make decisions and perform additional procedures as needed based on intraoperative findings per their best judgment. In this case, however, counseling the parents during the surgery was believed to be prudent, given the significant shift in the risk-benefit ratio presented by the unanticipated intraoperative finding of median nerve involvement. This provided an opportunity for shared decision-making and increased transparency regarding the likelihood of neurological sequelae after surgery.

In conclusion, this case presents the resection of a neurofibroma of a bifid median nerve, the involvement of which was not identified preoperatively despite ultrasound evaluation. A high index of suspicion for neurofibroma should be held in patients with NF-1 who present with soft tissue masses. Surgeons should be aware of the possibility of neurofibroma involvement of major nerves and consider possible anatomic variations when planning the resection of nerve tumors of the hand. A candid discussion should be held with patients and their families regarding the risks and benefits of surgery in such circumstances.

Informed consent was obtained from the patient's parents for the preparation and submission of this case report.

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