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Median Nerve Malformation at the Carpal Tunnel, a Case Report

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1. Abstract

Several anatomical variations of the median nerve within the carpal tunnel have been described. In this review, we report on a three-year-old girl who underwent surgery for a left palm mass and was diagnosed with a median nerve malformation. The median nerve branches lengthened, tangled, wrapped around each other, and were enclosed in a sac before continuing distally. Proximal neurolysis in the distal forearm was required to relocate the median nerve and close the wound, and branches were pulled proximally and located in the carpal tunnel.

2. Introduction

Peripheral nerves can exhibit multiple anatomical variations and be affected by many external anomalies along the nerve course. Several anatomical variations of the median nerve within the carpal tunnel have been described. Although many nerve pathologies can be diagnosed based on clinical and electrophysiological studies, imaging studies such as ultrasound and MRI are essential to confirm many peripheral nerve pathologies.

3. Case Presentation

A three-year-old girl presented with a left palm mass which a mother first noticed at the age of five months. The mass was painless, with no fluctuation in size. The child used her hand normally, but the mother occasionally noticed that her daughter could not tolerate holding cold objects. However, the family sought medical advice at the age of first notice of the lump, and they were reassured then they desired advice again at the age of three.

The child's history revealed that she was full-term and developed well according to age. There were no other lumps or other medical conditions. The parents denied a history of trauma. The child's examination revealed an isolated soft left palm oval mass, measuring 2*1.5 cm, with a smooth surface, non-tender, non-pulsatile, and no fluid thrill. No skin rash or other pathology on the exam.

Ultrasonography of the lump demonstrated a well-defined superficial oval shape hyperechoic mass lesion measuring 3.2*1 cm with a negative color flow, mostly representing lipoma. Accordingly, the decision was to do an excisional biopsy without performing a hand MRI, as the clinical exam and ultrasound revealed a superficial mass. The surgery was performed under general anesthesia, and a tourniquet was applied. A curved incision was performed directly over the lump. Dissection revealed a well-defined superficial sac with a nerve crossing its volar surface; circumferential dissection was performed, (Figure 1). However, the sac's proximal part was continuously proximally toward the carpal tunnel. Therefore, the decision was to extend the wound proximally and open the carpal tunnel. The recurrent branch of the median nerve is aroused from the radial side of the sac and crosses the superficial to the transverse carpal ligament. The sac was engulfing the median nerve at the carpal tunnel. Therefore, the sac was opened and revealed tortuous, elongated, tangled median nerve branches. Internal neurolysis was performed, and no definitive mass was found. However, multiple specimens were taken from the sac, surrounding soft tissue, and epineurium.

The median nerve at the carpal tunnel was thickened, and the branches were elongated 3-4 times and tangled and rolled around each other before continuing distally, (Figure 2). Therefore, the mass represents a malformation of the median nerve branches encircled by a sack. After the neurolysis of each branch, the nerve became larger as the enclosing sac was reducing its size. Therefore, we could not reposition the nerves to their position because of the increased size after the neurolysis, and the branches were bulging from the wound. So we extended the wound proximally and did median nerve neurolysis to pull the nerve to the forearm to reduce the amount of nerve in the palm. However, the median nerve

branches were positioned at the carpal tunnel, and the wound was closed over the nerve. Histopathology revealed normal epineural tissues, and no pathology was found. Postoperative neurological examination at two weeks revealed normal motor and sensory function. The wound healed at two weeks, the child was initiated into a rehabilitation program, and the family was instructed about protective measures.

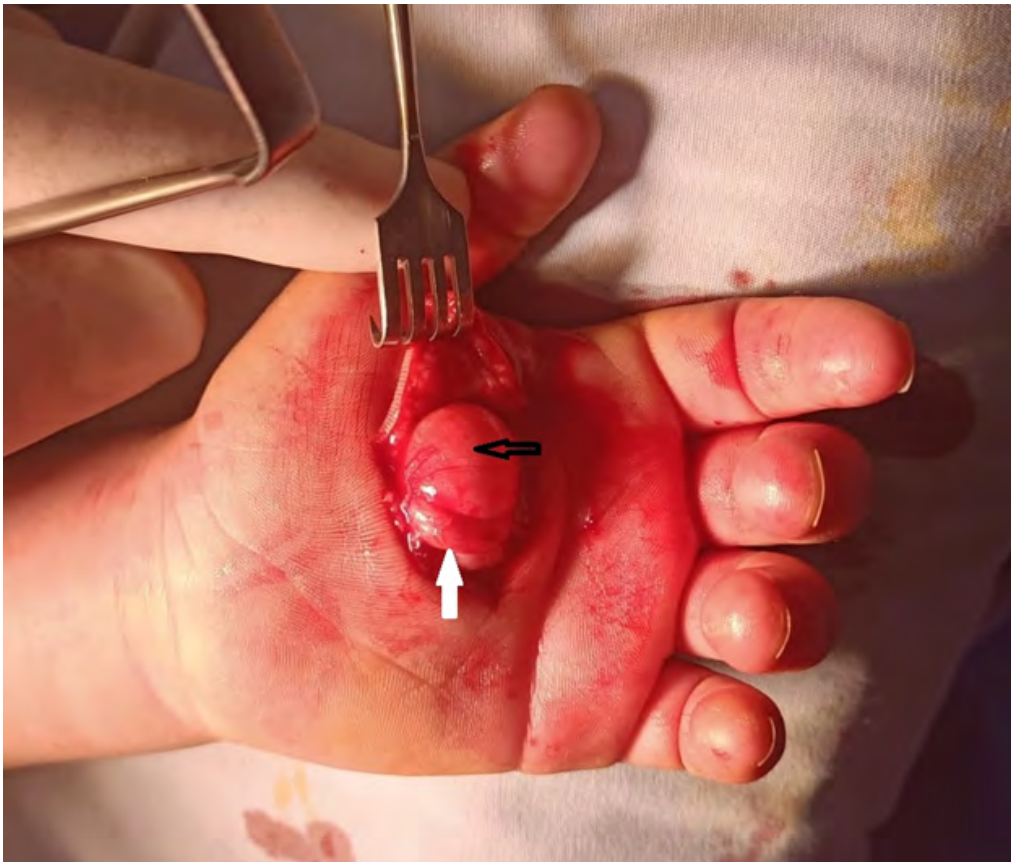

Figure 1: Intraoperative finding showed a sac (black arrow) with a nerve overlying it (white arrow).

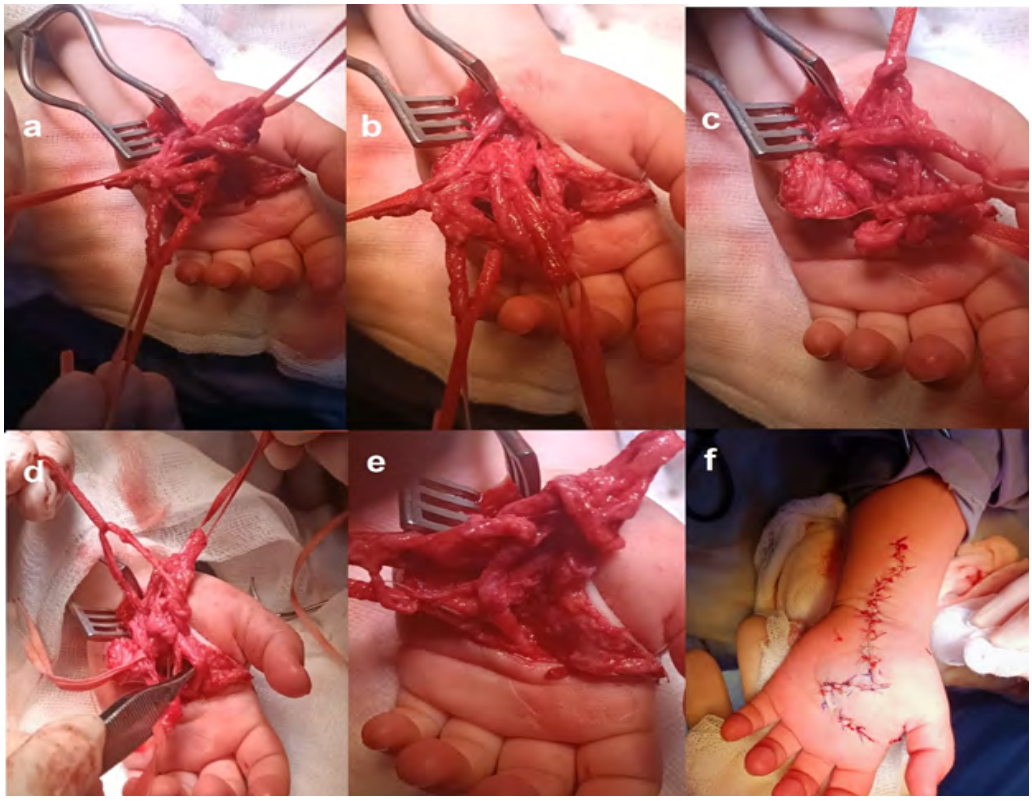

Figure 2: a-e): Intraoperative findings showed dilated tortuous, tangled branches of the median nerve within the carpal tunnel that are continuous distally. f) proximal extension of the wound to do neurolysis and pull the nerve proximally to allow wound closure.

4. Discussion

Peripheral nerves may have several anatomic variations and be affected by many extrinsic abnormalities along the course of the nerve, which may expose the nerve to compression [1]. Several anatomic variations for the median nerve within the carpal tunnel were described. However, Lanz [2] categorized them into four groups: Group I: Variations of the course of the thenar branch (recurrent motor branch) of the median nerve, which distributed in relation to transverse carpal ligaments in the following incidence: extraligamentous (46%), subligamentous (31%), and transligamentous (23%) [3]. Group II: Accessory median nerve branches at the distal carpal tunnel. Group III: High division of the median nerve may be associated with the median artery, where the size of both nerves is approximately the same diameter while the artery is highly variable [4-7]. An accessory lumbrical muscle between the two branches is sometimes found [8]. A persistent median artery has a superficial course close to the transverse carpal ligament; therefore, preoperative diagnosis of this anatomic variation may be of clinical importance [9]. Group IV: Accessory branches proximal to the carpal tunnel commonly pierce the transverse carpal ligament to join the nerve distally. Lanz confirms the necessity for approaching the median nerve from the ulnar side when opening the carpal tunnel to avoid lacerations of variant branches.

Some inherited and developmental anomalies of the peripheral nervous system, such as fibrolipomatous hamartoma, Charcot-Marie-Tooth disease, and hereditary neuropathy can present with neuroimaging abnormalities [10]. In addition to clinical and electrophysiological studies, imaging is essential in many peripheral nerve pathologies. Imaging can detect changes in nerve diameter and continuity and identify peripheral nerve tumors, traumatic neuromas, entrapment, demyelination, and infection. The most commonly used modalities are ultrasound and Magnetic Resonance Imaging. However, MRI visualizes the nerves and surrounding soft tissue features and can describe nerve lesions in an area difficult to visualize with ultrasound. [11-12].

Ultrasound can identify many nerves and nerve-related pathologies and provides inexpensive and accurate morphological information on various nerve pathologies, including hereditary and developmental disorders, nerve involvement in common diseases, entrapment syndromes, traumatic injuries, and neurogenic masses. Ultrasound is an excellent adjunct to clinical and electrophysiological tests to diagnose peripheral neuropathies [13]. Zaidman et al. reviewed Ultrasound and MRI results for suspected brachial plexopathy or mononeuropathies, excluding carpal and cubital tunnel syndromes, and compared the results of imaging studies to the intraoperative findings, clinical and electrophysiological evaluation; Zaidman et al. found that ultrasound is more accurate than conventional MRI for evaluating suspected peripheral nervous system lesions and identifying multifocal pathology. However, ultrasound is less expensive and can be done at the bedside. Therefore, they recommended it as an initial imaging study adjunctive to clinical and neurophysiological studies in a scenario that requires the availability of clinical differential diagnosis and experience in performing ultrasound [14].

MRI was not done in our case because such a study on a child requires anesthesia. Therefore, we depend on clinical and ultrasound which indicate a presence of a well-defined superficial mass. So we decided to do an excisional biopsy instead of exposing the child to anesthesia twice. However, MRI study in this child may add more anatomical details considering that ultrasound study is operator-dependent. The mass represents median nerve malformation where the branches are elongated 3-4 times, tangled and rolled around each other, and contained within an epineural sac before continuing distally. Internal neurolysis increased the size of the median nerve branches, which could not be repositioned to the same cavity containing the mass. Therefore, proximal neurolysis of the median nerve at the distal forearm was performed to pull the tortuous branches proximally. Although allocating median nerve branches within the carpal tunnel might expose the nerve to compression, this was necessary to reduce the nerve and close the wound. However, the superficial position of the nerve in the palm may put the nerve at higher risk for injury. Therefore, the family was instructed about complementing protective measures.

5. Conclusion

Several anatomical variations of the median nerve within the carpal tunnel have been described. In this review, we report on a three-year-old girl who underwent surgery for a palm mass. An intraoperative finding determined that the mass was a median nerve malformation due to the long, tortuous, and knotted branches of the median nerve. We recommend the use of imaging techniques such as ultrasound or MRI as an adjunct to the clinical assessment of masses in pediatric age groups.

6. Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient father has given consent for his daughter's images and other clinical information to be reported in the journal. The patient father understands that his daughter's name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

7. Conflict of Interest

None.

8. Source of Support

None.

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