www.jcimcr.org

JCIMCR OPEN ACCESS Journal of Clinical Images and Medical Case Reports

ISSN 2766-7820

Case Report

Open Access, Volume 3

Rare recurrent solitary plexiform neurofibroma of the dorsum of the finger: A case report

Xiao-Lan Ou; Rui Li; Wen-Rui Qu; Heng Tian*

Department of Hand Surgery, The Second Hospital of Jilin University, No. 218 Lane Ziqiang, Nanguan District, Changchun 130041, Jinlin Province, China.

***Corresponding Author: Heng Tian**
Department of Hand Surgery, The Second Hospital of Jilin University, Jinlin Province, China.
Email: rzlmengtong@163.com

Received: May 05, 2022
Accepted: Jun 03, 2022
Published: Jun 10, 2022
Archived: www.jcimcr.org

DOI: www.doi.org/10.52768/2766-7820/1882

Abstract

Background: Plexiform neurofibroma (PN) is a subtype of neurofibroma that is rather uncommon in the clinic. Plexiform neurofibroma is usually associated with neurofibromatosis type I (NF1). Only very few patients have isolated PN, which means they have neither NF1 signs nor relevant family history. Solitary PN is extremely rare. There have been no instances of finger back PN or solitary PN all around the world, which could lead to misdiagnosis, missed diagnosis, and mistreatment.

Case presentation: We present a rare case of a 50-year-old middle-aged woman who experienced numerous postoperative recurrences of painful tumors on the dorsal side of her proximal middle finger on her right hand. Following surgery, the pathology diagnosis was PN. The pathology diagnosis after surgery was PN. After the operation, the wound healed in stage I, and the outcomes were satisfactory.

Conclusion: In the dorsal finger nerve, solitary PN can occur. As a result, the differential diagnosis of NF1, PN, and isolated PN as hand masses is crucial for early diagnosis and rational treatment, especially for masses with pain associated with neuropathy should be considered as a possibility of this disease. The lesion may be invasive or multiple in character, Keep a close eye on cautious probing throughout the surgery. To reduce recurrence, to avoid further deterioration, and to alleviate patients' agony and psychological strain, the lesion tissue must be entirely removed.

Keywords: Plexiform neurofibroma; Solitary plexiform neurofibroma; Neurofibromas; Recurrence; Case report.

Introduction

NF1 is an autosomal dominant genetic disease. PN is considered to be a characteristic manifestation of NF1 and even thought almost exclusively seen in patients with NF1 [1,2]. However, the study discovered that PN can rarely appear in patients without a family history or clinical signs associated with NF1, which is called solitary PN [3]. So far, solitary PN of the dorsal finger nerve has not been reported in the literature. We report a case of isolated PN of the dorsal finger nerve. The patient and his family members have no history of NF1 and no features of NF1.

Case presentation

The patient, a 50 years old female, was admitted to the hospital with a progressive mass of the right middle finger for 30 years, after multiple operations and recurrence with enlargement for 1 year. The patient developed a tumor on the dorsal side of the proximal segment of the right middle finger 30 years ago. The tumor gradually increased and was painful and uncomfortable. He had undergone multiple tumor resections at the local hospital, and each one recurred following resection (no pathological examination). One year ago, the tumor recurred with pain after resection, so he came to our department. Physi-

cal examination: the dorsal side of the proximal segment of the middle finger of the right hand is locally bump, about 2 cm × 1 cm in size, the healed surgical scar can be seen on the surface (Figure 1a,1b), which is tough, the boundary with the skin is unclear, the mobility is poor, there is no pulsation, tenderness (+), the skin on the dorsal side of the proximal segment feels numb, and there is no obvious abnormality in the activities of interphalangeal joint and metacarpophalangeal joint. Local ultrasound examination of the tumor revealed that the dorsal skin of the right middle finger was subdermal and 1.1 cm × 0.4 cm × 0.7 cm, 0.4 cm × 0.2 cm × 0.5 cm, 0.26 cm × 0.19 cm slightly hyperechoic light mass, arranged from far to near, with a clear boundary and regular shape, and no blood flow signal (Figure 2). Under local anesthesia, a shuttle incision was designed with the dorsal finger tumor as the center, and the resection scope was prolonged to 2 mm at the edge of the tumor; the tumor and the scarred skin on it were removed. The mass was seen to be white and tough, located on the superficial side of the extensor tendon, with mild adhesion to the extensor tendon membrane and tight adhesion to the excised skin, the extensor tendon membrane was excised, and the incision was extended to the distal and proximal and explored: the mass was bead-shaped and grew to proximally along the dorsal finger nerve in a bead-like pattern, and the nerve was thicker than normal; the proximal end of the mass reached the metacarpophalangeal joint. the mass and the dorsal finger nerve were excised, and the distal and proximal nerves were expanded removed to the normal nerve (Figure 1 c). Postoperative pathological diagnosis: plexiform neurofibroma (Figure 3), immunohistochemical staining: S-100 (+), SMA (-), Desmin (-), CD34 (-), Ki67 (1%), NF (-), MUC4 (-), CD10 (foci +), ERG (-), (Figure 4). Postoperative anti-inflammatory dressing change was given and the wound healed well. Regular and long-term follow-up is recommended after discharge. There is no recurrence after 18 months of follow-up.

Discussion

Plexiform neurofibroma (PN), also known as "von Recklinghausen disease", [4,5] is a rare autosomal dominant disease, which is usually related to neurofibromatosis (NF). Up to 30% - 50% of patients with the neurofibromatosis type I (NF1) also have PN.1,6 There are scattered reports of the absence of a family history or clinical characteristics associated with NF1, called isolated PN, which may be caused by a sporadic mutation of the NF1 tumor suppressor gene [4,7]. The disease mostly occurs in children aged 2 ~ 12, often in the trunk, head and neck, and limbsm [4,8]. A few occur in the liver, stomach, and other internal organs [5,9]. It is characterized by diffuse hypertrophy of the involved nerves with preservation of the nerve bundle structure, as well as hyperplasia and hypertrophy of the associated skin and subcutaneous tissue. The severity of symptoms depends on the location of the lesion growth and the degree of damage to the peripheral nerve. Tenderness is often not accompanied in the early stage. With the growth of the tumor, pain and nerve compression can occur, causing corresponding symptoms and signs [10,11]. PN of brachial plexus and radial nerve has been reported in the relevant literature [12]. There are no reports of dorsal finger nerve PN. It is rare to have PN of the fingers, and isolated PN of the fingers is extremely rare.

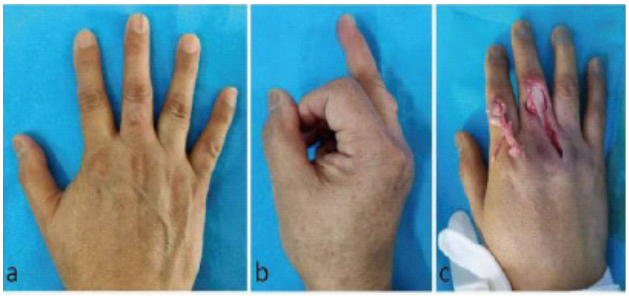

Figure 1: Right-hand middle finger proximal finger dorsal mass. (a) Frontal. (b) Lateral. (c) Resected beaded mass. and diseased nerve.

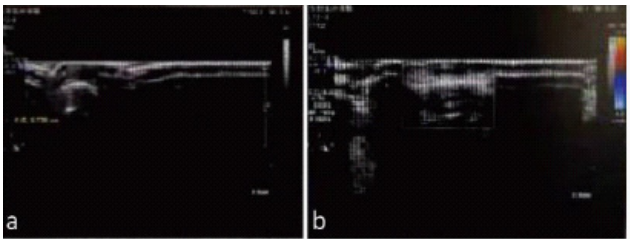

Figure 2: Ultrasound showed that slightly hyperechoic subcutaneously mass under the dorsal of the right middle finger, unclear boundary, regular shape.

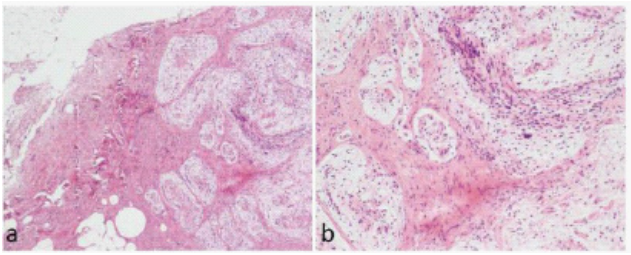

Figure 3: Histological features of a plexiform neurofibroma. (Hematoxylin-Eosin staining (HE), magnification × 40/100).

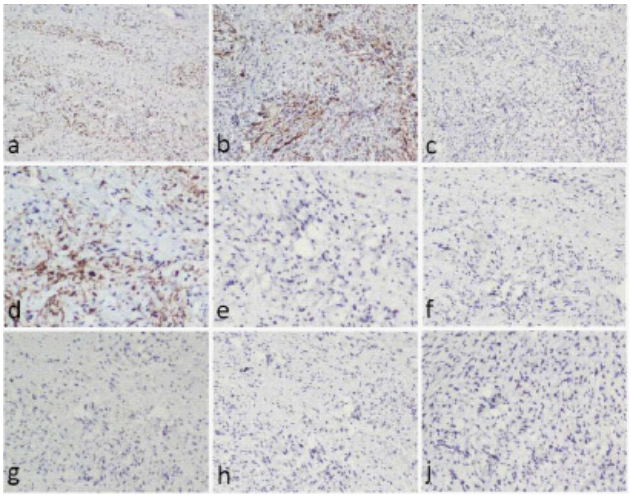

Figure 4: Immunohistochemistry: (a) s-100(+), (b) CD10 (Foci +), (c) Ki67 (1%), (d) CD34 (-), (e) SMA (-), (f) Desmin (-), (g) NF (-), (h) MUC4 (-), (j) ERG(-).

Imaging findings of PN: Under ultrasound, it shows single or multiple hypoechoic solid masses with a clear boundary and regular shape, with uniform internal echo and no blood flow signal [13,14]. Plain magnetic resonance imaging (MRI) revealed a low signal on T1WI and high signal on T2WI, On T2-weighted imaging was significantly enhanced in varying degrees, the signal of

small tumor is uniform, and the signal of large tumor is uneven [15,16]. Ultrasound and MRI can help clarify the shape, size, and location of the tumor and its connection to surrounding tissues, and provide guiding significance for the formulation of a surgical strategy. The ultrasound findings of the patient were consistent with the above characteristics. Pathological diagnosis is the gold standard. The general manifestations of pathological examination: the boundary of tumor tissue is slightly clear, in strip or bead shape, grayish white, tough, along the nerve direction of the tumor or connected to the nerve bundle at the proximal and distal. Microscopically: it is mainly composed of Schwann cells, fibroblasts, perineurial cells, and mast cells, which together form a typical plexus structure [3,14]. Immunohistochemical analysis showed that the tumor cells were positive for S-100 protein, and the Ki-67 proliferation index (PI) was 0% - 5% [17,18]. PN is usually diagnosed in children and rarely after puberty [4,8]. It is mainly distinguished from plexiform schwannoma [19]. Plexiform schwannoma often exists alone, has less recurrence, generally does not involve nerve fibers, the tumor body envelope is complete, the nerve sheath forms part of the envelope, and there is no nerve axon in the tumor body, and bleeding and necrosis are common [12,20]. In addition, PN needs to be differentiated from liposarcoma, hemangioma, and myxoma. The most common clinical manifestation is painless mass, which is rarely related to neurological symptoms. This case of PN has typical features: the tumor is largely beaded, white in color, and the tumor grows along the dorsal nerve of the finger with pain, while immunohistochemistry: S-100 (+), Ki67 (positive rate 1%), CD10 (focal +), which is consistent with the above presentation.

Early diagnosis and surgical resection are the mainstays of treatment of PN. PN is non encapsulated and diffuse. At the same time, the low radiosensitivity of PN makes it difficult to remove and easier to relapse [21]. It is necessary to expand the scope of resection. Recurrence and leaving corresponding nerve injury symptoms are the two major difficulties in PN treatment. Complete excision without compromising the parent nerve function is the best choice for treatment, but it is difficult to achieve [22,23]. However, some studies have pointed out that the recurrence of isolated PN after surgical resection is rare.4 PN is a benign lesion with invasive growth, after 10-20 years of incubation, it can develop into malignant peripheral nerve sheath tumors (MPNST), with a risk of malignant transformation rate of approximately 2% ~ 5% [18,21]. It has even been reported that the risk of malignant transformation of PN is increased to 10% [10,24]. When the patient increases rapidly in a short time, the pain intensifies, and there is cystic degeneration and necrosis in the tumor, which indicates malignant transformation, poor prognosis, and high mortality [20]. Therefore, regular follow-up of PN patients is particularly important. The dorsal finger nerve was involved in this case. The nerve involved in PN was small and could not be partially removed. Moreover, the sensory damage range after the resection of the dorsal finger nerve was small. The tumor body and damaged nerve were completely removed to avoid postoperative recurrence. The patient had a history of 30 years and recurred many times after the operation, but there was no tendency of malignant transformation. Characteristics of the patient: ① The patient has no family history of NF1, no so-called skin “café-au-lait” spots, axillary or groin freckling, and other typical clinical manifestations. PN in fingers is extremely rare, which increases the difficulty of preoperative diagnosis. ② Preoperative physical examination and ultrasound showed that the tumor was beaded and painful along the peripheral nerve, suggesting that the tumor was related to the peripheral nerve. ③ Multiple operations and postoperative recurrence indicate that multiple tumors are multiple or invasive, and conventional resection cannot be completely removed. Therefore, we used extended resection, carefully found all the tumors along the diseased nerve, and resected the diseased nerve together to avoid local recurrence.

Conclusion

This is the first reported case of isolated PN on the dorsum of the finger. This disease should be considered in the differential diagnosis of all hand masses. Hand tissues are diverse, and a variety of bone and soft tissue tumors can occur in the hand, resulting in a variety of hand tumors and difficult preoperative diagnosis. Clinicians need to master the characteristics of various tumors. Careful physical examination before the operation, combined with a medical history and auxiliary examination, preliminarily judges the nature and scope of the tumor, searches for the source of the tumor, and complete resection during operation, to reduce the recurrence rate of the tumor as much as possible.

Declarations

Ethics approval and consent to participate: This study was approved by the Research Ethics Committee of the Second Hospital of Jilin University.

Consent for publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests: The authors declare that they have no competing interests.

Funding: There is no funding for disclosure.

Authors’ contributions: XLO wrote the manuscript and reviewed the clinical notes, RL and WRQ revised the manuscript and confirmed the histopathological examination results, HT analyzed and interpreted the patient data an. all authors read and approved the final manuscript.

Acknowledgments: We would like to thank the patient for agreeing to the publication of this case report.

References

1. Selumetinib in Children with Inoperable Plexiform Neurofibromas. New England Journal of Medicine. 2020; 382.
2. Ros PR, Eshaghi N. Plexiform neurofibroma of the pelvis: CT and MRI findings. Magnetic Resonance Imaging. 1991; 9: 463-465.
3. Beert E. et al. Biallelic inactivation of NF1 in a sporadic plexiform neurofibroma. Genes, Chromosomes and Cancer. 2012; 51.
4. Zwane, N. P., Noffke, C. & Raubenheimer, E. J. Solitary oral plexiform neurofibroma: Review of literature and report of a case. Oral Oncology. 2011; 47: 449-451.
5. Shi, L. et al. Solitary plexiform neurofibroma of the stomach: A case report. World Journal of Gastroenterology. 2014.
6. Tchernev G, Chokoeva AA, Patterson JW, Bakardzhiev I, Tana C. Plexiform Neurofibroma: A Case Report. Medicine. 2016; 95: e2663.
7. Ji G, Wang K, Jiao C, Lu Z, Li, X. Solitary Plexiform Neurofibroma of the Hepatic Artery. Journal of Gastrointestinal Surgery Official Journal of the Society for Surgery of the Alimentary Tract. 2017.

8. Mauro et al. Plexiform neurofibroma in the ear canal of a patient with Type I Neurofibromatosis. Brazilian Journal of Otorhinolaryngology. 2009.

9. Nebiki H, Hiramatsu S, Sakata Y, Suekane T, Inoue T. A rare case of plexiform neurofibroma of the liver in a patient without neurofibromatosis type 1. Clinical Journal of Gastroenterology. 2020.

10. Khajavi et al. A brief report of plexiform neurofibroma. CURRENT PROBLEMS IN CANCER. 2018; 42: 256-260.

11. Murphey, M., Smith, W. S., Smith, S. E., Kransdorf, M. J. & Temple, H. T. Imaging of Synovial Sarcoma with Radiologic-Pathologic Correlation1. Radiographics. 1999; 26: 1253-1280.

12. Spinner RJ. et al. Colocalized cellular schwannoma and plexiform neurofibroma in the absence of neurofibromatosis. Case report. Journal of Neurosurgery. 2007; 107: 435-439.

13. Wortsman X, Lobos N, Parra R, Carreño L. Multidimensional Ultrasound and Computed Tomography Imaging Support in Bleeding Plexiform Neurofibromatosis of the Scalp: A Case Report and Literature Review. Indian Journal of Dermatology. 2015; 60: 421.

14. Atkins NK, Stensby JD, Gaballah AH. Lumbosacral plexiform neurofibroma: a rare case in an adult without neurofibromatosis type I. Skeletal Radiology. 2020; 49: 321-330.

15. Eoghan MB, Ngan BY, Navarro OM. Pediatric Soft-Tissue Tumors and Pseudotumors: MR Imaging Features with Pathologic Correlation. Radiographics. 2009; 29: 1-35.

16. Lin J, Martel W. Cross-sectional imaging of peripheral nerve sheath tumors: characteristic signs on CT, MR imaging, and sonography. AJR. 2001; 176: 75-82.

17. Cholakkal SVCS, Nachiketh G. Large Solitary Plexiform Neurofibroma of the Penis Without Erectile Dysfunction: a Case Report from Kerala. Indian Journal of Surgery. 2018.

18. Fujiwara M, Isaka F, Honjo G, Ozawa T, Tachibana T. Solitary plexiform neurofibroma: recurrence 32 years after excision. J Eur Acad Dermatol. 2006; 20.

19. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: diagnostic overview and update on selected diagnostic problems. Acta Neuropathologica. 2012; 123: 295-319.

20. Murphey M, Arcara LK, Fanburg-Smith J. Imaging of Musculoskeletal Liposarcoma with Radiologic-Pathologic Correlation1. Radiographics. 2005; 25: 1371-1395.

21. Ferner RE, O Doherty MJ. Neurofibroma and schwannoma. Current Opinion in Neurology. 2002; 15: 679-684.

22. Needle MN, Cnaan A, Dattilo J, Chatten J, Molloy PT. Prognostic signs in the surgical management of plexiform neurofibroma: the Children's Hospital of Philadelphia experience, 1974-1994. Journal of Pediatrics. 1997; 131: 678-682.

23. Hadi FU. Solitary plexiform neurofibroma of the nasal tip: Case report and review of the literature. International Journal of Pediatric Otorhinolaryngology Extra. 2007.

24. Listed, N. A. National Institutes of Health Consensus Development Conference Statement: neurofibromatosis. Bethesda, Md., USA, July 13-15, 1987. Neurofibromatosis 1988; 1: 172-178.