Case study

Neurofibroma of the median nerve managed with ultrasound guided nerve block: a rare case report.

Abstract

Background: Median nerve neurofibroma can mimic carpal tunnel syndrome. In unilateral carpal tunnel syndrome, wrist imaging is suggested to exclude tumours. Diagnosis and management of peripheral nerve sheath tumours can be challenging.

Case: This is a case report of 42year old female with median nerve neurofibroma who presented with excruciating pain as the main symptom. Though surgical excision is the preferred treatment, it is associated with the risk of functional deficit. In this case, patient was unwilling for surgery. This report highlights the evaluation of neurofibroma and the role of median nerve block in the symptomatic as well as functional improvement.

Conclusion: Nerve block can be effective if neurofibroma presents with pain as the main symptom without any impairment of motor function or daily activities.

Keywords

Benign tumours; carpal tunnel syndrome; median nerve; median nerve tumour; nerve blocks; neurofibroma; peripheral nerve tumours; ultrasound guided.

1. **Introduction:** Peripheral nerve sheath tumors [PNSTs] are rare primary neurogenic tumors that arise from nerve sheath outside central nervous system, neurilemomas/schwannomas and neurofibromas being the commonest [1]. Neurofibromas, rarely encapsulated, originate from the endoneurium. They involve nerve fibrils, mast cells, perineural cells, Schwann cells and fibroblasts [2]. Carpal tunnel syndrome [CTS] is the commonest entrapment neuropathy with incidence of 2.7% (5.8% in women). Median nerve involvement is common in CTS. PNSTs can have similar presentation to CTS.
   Neurofibromas occur in 20-40year age group with male: female incidence 1:1 and has predilection for trunk, head and neck and extremities. It can be solitary, sporadic (90%) with low malignancy risk or inherited (10%). The plexiform type with incidence rate of 1 in 2600 to 3000 individuals is pathognomonic of neurofibromatosis - 1 (NF-1 gene mutation) and has implicit possibility of malignancy [2,3].
   Surgical removal of the tumor is difficult because of the unclear margins of the tumor and risk of functional deficit. The diagnosis and management of PNSTs can be challenging. This case report of 42year old female with median nerve neurofibroma, an uncommon presentation mimicking CTS, highlights the

evaluation of neurofibroma and the role of median nerve block in the symptomatic as well as functional improvement.

2. **Case Report:** A 42year old female noticed a small sized swelling of insidious onset over the palmar radial aspect of right hand 6 months ago, not associated with any pain or discharge initially. It was of progressive in nature and enlarged up to the present size of 5 x 4 cm in 6months duration. She developed pain and burning sensation along the right hand, radial aspect involving the index, middle and ring finger for about 4 weeks before she presented to us. The pain was of continuous excruciating nature. Hand and finger movements were restricted due to pain affecting her daily activities and was not relieved by analgesics and co-analgesics. No history of fever, trauma or previous surgeries. No known co-morbidities.

   On examination, the swelling was of approximately 5 x 4 cm size with well-defined borders, tenderness, firm consistency, restricted mobility and no discharge. Phalen sign, reverse Phalen sign were negative and Tinel sign positive. There was hyperalgesic response to pin-prick and cold. Motor examination was normal. Systemic examination was insignificant.

   Ultrasonogram local site showed large multi-lobulated lesion in the subcutaneous plane of palmar aspect of right hand with high vascularity on color doppler study. MRI of the right wrist revealed large well defined multiseptated lesion with few cystic areas most likely to be peripheral nerve sheath tumor of benign etiology.

   Decision was made to treat the tumor conservatively as the main symptom of the benign tumor was excruciating pain hampering the daily activities with no motor involvement and also, as the patient was unwilling for surgery. After obtaining informed written consent and proper patient positioning, a successful median nerve block was performed under all aseptic precautions using 22 G needle by depositing 2 ml of 1% lignocaine and 10 mg injection methylprednisolone acetate to the tumor by ultrasound visualization.

3. **Discussion:** Most median nerve tumours are benign arising in the peripheral neural sheath (schwannoma and neurofibroma) or intraneural (lipoma, haemangioma, or hamartomas) or causing extrinsic compression (lipomas or cysts).[4] Median nerve neurofibromas are often asymptomatic but rarely cause symptoms like CTS (pain, numbness, mass effects, pruritus or paresthesia) [5].

   Literature shows that, surgical excision was the preferred treatment with extremely rare local recurrence, though it was associated with the risk of functional deficit and only partial resection was possible in most cases [6]. This case report reveals that nerve block can be effective if neurofibroma presents with pain as the main symptom without any impairment of motor function or daily activities.

   CTS occurs in association with hypothyroidism, rheumatoid arthritis, synovitis, pregnancy, diabetes, wrist trauma, tumors and prolonged working with vibrating tools [7]. Zoe H. Dailiana et al described tumors mimicking CTS [8]. K. Nakamichi and S. Tachibana reported 7 case series of tumors causing CTS [9]. In unilateral CTS, wrist imaging is suggested to exclude tumors [9]. The ‘target sign’ on the T2

sequence in MRI being 100% specific and 59% sensitive of PNSTs is due to peripheral myxomatous tissue(hyperintense) and central fibro-collagenous tissue(hypointense) [10].

Nerve blocks can be associated with complications like infection, bleeding, nerve injury and local anesthetic toxicity. Median nerve can be confused with tendons [11]. The ultra sound guided nerve block is a potentially feasible technique to administer block at the most convenient and safest position for significant pain reduction within a short block performance time (time required to place the ultrasound probe, acquire the desired images and perform the perineural injection of nerve) with good patient satisfaction and outcome [12, 13].

Strict aseptic precautions should be followed to avoid any possible risk of infection. The use of ultrasound decreases the risk of vascular and nerve injuries as compared to the anatomic landmark technique. Considering the lesser volumes of local anesthetics used, the risk of toxicity is less. No complications occurred in our case.

The procedure was followed by immediate pain relief within 4 to 5 minutes of drug infiltration. Patient was followed up after 1 month and thereafter 3 months. Patient had good pain relief and great improvement in daily activities.

4. **Conclusion:** This case report describes the use of ultra sound guided median nerve block effectively and safely in managing the uncommon presentation of neurofibroma of median nerve.

   Clinical symptoms, growth and nature of tumor are the most important factors to be considered during the treatment. The interventional pain medicine using nerve block, by reducing the pain and preserving the nerve function, helped in improving the quality of life providing patient satisfaction.

5. References

1. Barfred and Lis Zachariae. Neurofibroma in the median nerve treated with resection and free nerve transplantation-Case Reports. Scandinavian Journal of Plastic and Reconstructive Surgery 9: 245-248, 1975.
2. Engy Abdellatid, Dia Kamel. Soft tissue peripheral nerve neurofibroma – general. Pathology outlines.com, Inc. 2002-2021.
3. Lynn Messersmith, Kevin Krauland Neurofibroma. [Updated 2020 August 10]. Stat Pearls[internet]. Treasure Island [FL]: Stat Pearls Publishing;2021 Jan.
4. Kang HJ, Shin SJ, Kang ES. Schwannomas of the upper extremity. Journal of Hand Surgery British. 2000;25(6):604–7.
5. Staser K, Yang FC et al. pathogenesis of plexiform neurofibroma: tumor-stromal/hematopoietic interactions in tumor progression. Annuals Rev Pathol.2012; 7:469-95.
6. Allaway RJ, Gosline SJC et al. Cutaneous neurofibromas in the genomics era: current understanding and open questions. British Journal of Cancer. 2018 Jun;118(12): 1539-1548.

7. Gbriel Costa, Katia Torres Batista, Ulises Prieto y Schwartzman et al. Benign tumors affecting the median nerve case series reports of diagnostic and surgical strategies. Published on Revista Brasileira Ortopedia.2018;53(2):192-199.
8. Dailiana Z.H., Bougioukli S, Vritimidis S et al. Tumors and tumor like lesions mimicking carpal tunnel syndrome. Archives of Orthopedic and Trauma Surgery 2014;134(1):139-144. [pub med] [Google scholar].
9. Nakamichi K, Tachibana S. Unilateral carpal tunnel syndrome and space occupying lesions. Journal of Hand Surgery British.1993:18(6):748-749.
10. Ajaya Kumar Ayyappan Unnithan, Anna Joseph. Schwannoma of median nerve: Case report and review of literature. *Interdisciplinary Neurosurgery 17 (2019) 75–78*. Published by Elsevier B.V.
11. McCartney CJ, Xu D, Constantinescu C, Abbas S, Chan VW. Ultra-sound examination of peripheral nerves in the forearm. Regional Anesthesia and Pain Medicine. 2007 Sep-Oct;32(5):434–439. Erratum in: Regional Anesthesia and Pain Med. 2008 Mar-Apr;33(2):188.
12. Liebmann O, Price D, Mills C, Gardner R, Wang R, Wilson S, et al. Feasibility of forearm ultrasonography-guided nerve blocks of the radial, ulnar, and median nerves for hand procedures in the emergency department. Annals of Emergency Medicine. 2006;48(5):558–62. E-pub 2006 Jun 14.
13. Jose R Soberon, Neil R Bhatt, Bobby D. Nossaman et al. Distal peripheral nerve blockade for patients undergoing hand surgery: a pilot study. American Association for Hand Surgery2014. https://doi.org/10.1007/s11552-014-9680-4. Published on September 30 2014.