SOLITARY INTEROSSEOUS NEUROFIBROMA OF DISTAL FOREARM: AN UNUSUAL PRESENTATION
P. Ashok Kumar1, Ch. V. Murali Krishna2, K. Srinivas Rao3 P. Rambabu4, V. Krishnaswamy5

ABSTRACT: Solitary Neurofibromas are uncommon nerve tumors and are still rarer in interosseous locations. They are not detected until they cause a significant damage to the neighbouring tissues. We present a case of a solitary interosseous neurofibroma of the right distal forearm, without neurological deficit. It was diagnosed by Radiology and histopathology and was excised.

KEYWORDS: Solitary, Neurofibroma, Interosseous.

INTRODUCTION: Solitary Neurofibromas are benign tumors of nerve sheath origin and may arise from the Schwann cells, perineural cells, and fibroblasts. By definition they are confined and are seen in patients who do not suffer from Von Recklinghausen’s Disease. According to Geschickter, 90% are true solitary tumors, only 10 % are associated with Von Recklinghausen’s Disease.

We present a case of a solitary interosseous neurofibroma of the right lower forearm which caused erosion of the distal radius and ulna and lytic lesion of distal radius on plain roentgenogram.

CASE PRESENTATION: A 58-year-old woman presented with a painless swelling of 1 year duration on the right lower forearm and wrist predominantly on the volar surface, slowly progressive and attained a size of 8.9×4.9×4.0cm without neurological symptoms. There is no family history of neurofibromatosis or similar swellings elsewhere in the body. On clinical examination, the tumor was deep seated arising from interosseous space of distal forearm encircling the entire distal radius and encroaching on to the wrist. A clinical diagnosis of benign soft tissue tumor was made. X-ray showed erosions and scalloping of the radius and ulna. MRI showed well encapsulated mass in distal forearm with pressure effects and scalloping over distal radius and ulna.

Detailed examination of the patient for the evidence of Neurofibromatosis revealed no neurofibromas in other locations, no “café au lait” spots, Lisch nodules, axillary or inguinal freckling, sphenoid wing dysplasia or thinning of the cortex of long bone, and optic glioma. At least two or more lesions are required for the diagnosis of neurofibromatosis.

The tumor was excised and measured 8.9×4.9×4.0 cm located between the forearm muscles and arising from the median nerve. Cut section was ivory white and glistening was relatively circumscribed but unencapsulated and showed no evidence of hemorrhage or necrosis.

HISTOPATHOLOGY: Histopathological examination showed a tumor with the cells arranged in lobules of anastomosing cords, strands and nests against a background of myxoid material and intervening fibrous septae. Section also revealed soft tissue tumor mass with spindle cells arranged in irregular fascicles having elongated wavy nuclei, minimal nuclear atypia. No giant cells, mitotic figures or necrosis. Tumor cells are separated by hyalinated collagenous tissue.
DISCUSSION: True nerve tumors are of neuroectodermal origin. They are completely encapsulated by epineurium and thus have a true capsule. The 2 types of benign nerve tumors representing the vast majority of all benign nerve tumors are schwannoma and neurofibroma. Benign tumors of peripheral nerves have been documented to be relatively rare in the hand and forearm, representing less than 5% of tumors in this area.\(^4\)

Neurofibromas occur in 2 basic disease patterns: sporadic and in association with neurofibromatosis. Localized neurofibromas are thought to be solitary lesions not associated with systemic disease. Approximately 90% of neurofibromas are localized lesions;\(^2,4\) the rest are associated with neurofibromatosis. This distinction becomes important because of the higher likelihood of malignancy of neurofibromas in neurofibromatosis.

Neurofibromas (NF) are rare tumors\(^1,5\) and are further rare in intermuscular locations. All the cases are not associated with neurofibromatosis (NF1). The diffuse and the plexiform patterns have a close relation with neurofibromatosis. The solitary (sporadic) form occurs in those who do not have neurofibromatosis.\(^5\) Neurofibromas frequently affect patients aged 20–30 years. MPNSTs are significantly more likely to be located on the limbs compared with benign nerve sheath.\(^6\) Whereas malignant transformation is rare in isolated neurofibromas, it has been reported to occur in 2% to 13% cases of von Recklinghausen disease.\(^1,4\) When symptoms warrant, treatment for isolated neurofibromas is excision. However, when the diagnosis is clear and excision of nerve fibers is necessary, a small amount of tumor may be left behind, given the low potential for malignant transformation and recurrence, as suggested by Healey and McCormack.\(^4\)

CONCLUSION: Benign nerve tumors, such as schwannomas and neurofibromas are relatively uncommon entities, representing approximately 11.5% of benign soft-tissue tumors in one surgeon’s 16-year experience. Schwannomas can be distinguished from neurofibromas by the former’s lack of potential for malignant degeneration, plexiform growth or multiplicity of growth sites (Characteristic of neurofibromatosis).\(^4\) Tumours. Benign nerve tumors most commonly present with pain or paresthesia.\(^1,4\)

Although the most common location of benign nerve tumors is the volar surface of the hand, 25% of the lesions were on the dorsal surface of the fingers. MRI or fine-needle aspiration should be used in evaluation. In the majority of cases, once a diagnosis is clear, the entity can be treated with simple excision.

REFERENCES:
1. Solitary Giant Intramuscular Myxoid Neurofibroma Resulting in an above Elbow Amputation- Hindawi Publishing Corporation Case Reports in Pathology Volume 2012, Article ID 353215, 3 pages.
2. Solitary neurofibroma of Kidney; clinical histological and chromosomal appearance, Athanasios Kostakopoulos, Maria Chorti, Vassilios Protogerou and Stavroula Kokkinou. International Urology and Nephrology 35; 11-13, 2003.
3. Solitary Benign Periphearl-Nerve Tumors : Review of 32years experience JBJS(Br),1995-77B‘497-500.Nicholas Js KEHOE, Robin P Reid, J Campbell sempere From western infirmary, Glasgow, Scotland.
4. Benign Nerve Tumors of the Hand and the Forearm - Chris J. Lincoski, MD, G. Dean Harter, MD, and David C. Bush, MD; Am J Orthop. 2007; 36 (3): E32-E36. Copyright 2007, Quadrant HealthCom Inc.

5. Solitary neurofibroma of the gingiva with prominent differentiation of Meissner bodies: A case report Jun Ohno*, Teruaki Iwahashi, Ryuki Ozasa, Kazuhiko Okamura, Kunihisa aniguchi

6. Early-stage malignant peripheral nerve sheath tumour arising from a solitary eurorfibroma- The British Journal of Radiology, February 2012 DOI: 10.1259/bjr/55588872 T Hyodo, MD, Y Sugawara, MD, K Sakayama, MD and K Kito, MD.
CASE REPORT

AUTHORS:
1. P. Ashok Kumar
2. Ch. V. Murali Krishna
3. K. Srinivas Rao
4. P. Rambabu
5. V. Krishnaswamy

PARTICULARS OF CONTRIBUTORS:
1. Incharge Professor, Department of Orthopaedics, King George Hospital, Andhra Medical College, Visakhapatnam, Andhra Pradesh.
2. Assistant Professor, Department of Orthopaedics, King George Hospital, Andhra Medical College, Visakhapatnam, Andhra Pradesh.
3. Assistant Professor, Department of Orthopaedics, King George Hospital, Andhra Medical College, Visakhapatnam, Andhra Pradesh.
4. Assistant Professor, Department of Orthopaedics, King George Hospital, Andhra Medical College, Visakhapatnam, Andhra Pradesh.
5. Junior Resident, Department of Orthopaedics, King George Hospital, Andhra Medical College, Visakhapatnam, Andhra Pradesh.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Dr. P. Ashok Kumar,
# B-98, Dayal Nagar,
Visakhapatnam-530043,
Andhra Pradesh.
E-mail: ashok_ortho59@rediffmail.com

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