ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 05 | May - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A RARE CASE OF MYXOID NEUROFIBROMA OF THE FINGER

Plastic Surgery

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ABSTRACT

Myxoid neurofibromas are uncommon benign tumours of perineural origin which are rare in the acral regions of upper extremities. They are generally asymptomatic unless they cause a significant damage to the neighbouring tissues. We present a case of a myxoid neurofibroma of the right index finger tip which was excised and diagnosed by histopathology and confirmed by immunohistochemistry.

KEYWORDS

Benign, Digital, Myxoid, Perineural, Rare, Surgery

INTRODUCTION

Neurofibromas are tumours of the nerve sheath origin and may arise from the Schwann cells, perineural cells, and fibroblasts.[1] Myxoid neurofibroma is a tumor that also originates from Schwann cells. The tumor has a uniform myxoid stroma with moderate cellularity and sinusoidal spindled nuclei.[2] The presence of intratumor axons is an important distinguishing feature from nerve sheath myxomas and can be demonstrated with silver stains or with immunohistochemical staining for neurofilament protein.

CASE REPORT

A 32 year old housewife presented to us with a painful swelling in the distal of the right index finger since 6 months. It started spontaneously and grew gradually to the present size. She complains of sharp pricking type of pain for the last 2 months which has increased since past 2 weeks. There was no history of trauma or any infection of the right index finger. There was no ulceration or bleeding from the swelling. She had no other swellings elsewhere in the body. On examination, a 1 x 1 cm swelling was present on the radio-dorsal side of the distal phalanx at the base of the nail plate complex of the right index finger. **(Fig. 1)**

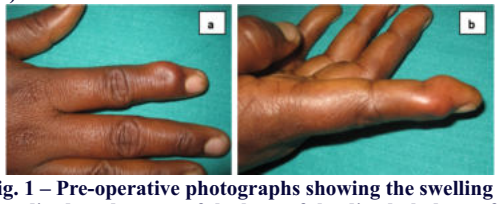

Fig. 1 – Pre-operative photographs showing the swelling in the radio-dorsal aspect of the base of the distal phalanx of the right index finger, dorsal (a) and lateral (b) views

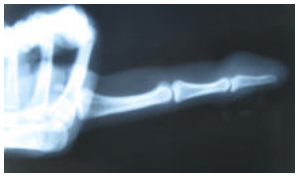

Fig. 2 – X ray showing a soft tissue shadow in the base of the distal phalanx of the index finger

The swelling was not warm or tender, firm in consistency with free from underlying distal phalanx and the overlying skin was free. The swelling was not compressible and non-pulsatile. A provisional diagnosis of an implantation dermoid was made. X-ray revealed a soft tissue swelling along the base of the distal base not involving the bone or the distal interphalangeal joint. **(Fig. 2)** MRI showed the lesion to be well-circumscribed, hypo-intense on T1- and hyper-intense on T2-weighted images suggestive of a neurofibroma. **(Fig. 3)**

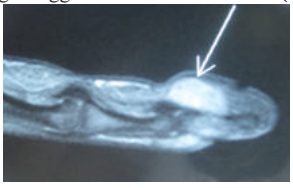

Fig. 3 – MRI showing a well-circumscribed, hypo-intense on T1- and hyper-intense on T2-weighted image

We proceeded for excision biopsy of the lesion. Under digital block with plain 2% lignocaine and glove tourniquet, a mid lateral incision was made over the swelling and deepened in layers. With sharp dissection the lesion was excised in toto and sent for histopathological examination. Tourniquet was released and haemostasis was secured. The incision was closed with 4-0 nylon sutures and a finger bandage was applied. **(Fig. 4a & b)**

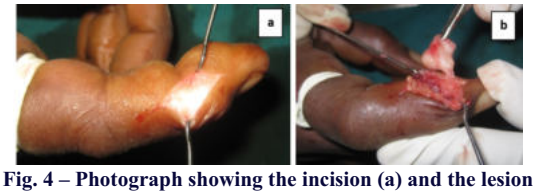

Fig. 4 – Photograph showing the incision (a) and the lesion being dissected out (b)

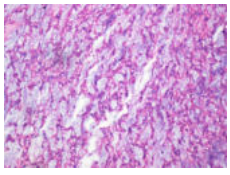

Fig. 5 – Microscopy showing spindle shaped cells with uniform wavy elongated nuclei pointing in various directions separated by abundant myxoid stroma with no mitoses.

Post-operative period was uneventful. Microscopic examination

revealed a benign neoplasm composed of spindle shaped cells with uniform wavy elongated nuclei pointing in various directions separated by abundant myxoid stroma and no mitoses were seen. The features were suggestive of myxoid neurofibroma confirmed by immunohistochemistry showed positivity for S100 protein by the tumour cells. **(Fig. 5)**

DISCUSSION

Neurofibromas are benign, heterogeneous tumors arising from the connective tissue of peripheral nerve sheath[1]. Neurofibromas are rare tumours and all the cases are not associated with neurofibromatosis (NF1)[2]. The diffuse and the plexiform patterns have a close relation with neurofibromatosis. The solitary (sporadic) form occurs in those who do not have neurofibromatosis.[3] Myxoid neurofibroma (MN) is a benign tumour of perineural origin, which is demonstrated by a positive immunohistochemical staining for S100 protein. MN typically occurs in younger adults with a predilection to the face, shoulders, arms and periungual regions.[2,4]Myxoid Neurofibroma is a relatively uncommon variant of solitary neurofibroma characterized histologically by abundant myxoid matrix and small spindle cells with variably tapering nuclei, condensed chromatin, inconspicuous nucleoli and scant cytoplasm[3]. These tumours are confused often clinically and histopathologically with myxomas. Myxomas have no nerve involvement and are S100 negative.[4]Myxoid neurofibromas can cause problems resulting from involving adjacent structures. The presence of mitotic activity in neurofibroma is indicative of malignancy and is seen even in schwannoma.[5] The differential diagnosis of myxoid neurofibroma includes intramuscular myxoma, nerve sheath myxoma (NSM) and myxoid dermatofibrosarcoma protuberans (DFSP).[6,7] The distinction is made largely from histopathological and immunohistochemical analysis. Myxoid neurofibroma stains positively for S-100, though intramuscular myxoma does not. Myxoid neurofibroma typically has collagen bundles within the myxoid stroma, is not mitotic and lacks a lobular structure.[7] In addition, there are often nerve fibers or axons present within the tumor. NSM on the other hand, commonly demonstrates a multi-lobulated architecture and no collagen bundles within the stroma and may show mitoses.[7] Finally, myxoid DFSP is often poorly-circumscribed with infiltrative margins and a storiform architecture and will stain negative for S-100 and positive for CD-34. The mainstay of surgical treatment is standard excision.

CONCLUSION

Digital myxoid neurofibromas are unusual tumours of the perineurium which are generally asymptomatic. Hence a thorough physical and radiological examination is necessary to diagnose these lesions. Surgical excision is the treatment of choice and these lesions are characterized by positive immunohistochemical staining for S100 protein.

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