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Histopathological Study of neurofibrolipoma in a tertiary hospital

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Abstract:
Introduction: *Four cases of neurofibrolipoma (lipofibromatous hamartoma of the nerve) were presented. Neurological examination was normal except to minimal pain on palpation. Surgical exploration revealed that nerve and its digital branches were infiltrated by fibrofatty tissues. Fibrofatty tissues were dissected and removed from the nerve by microsurgical technique. Histological examination confirmed the diagnosis as a Neural neurofibrolipoma or lipofibromatous hamartoma of the nerve. The result of surgical debulking was satisfactory.*
Objectives*: To study the histopathological spectrum of neurofibroilipoma in a tertiary hospital.*
Material and methods*: This is a three years retrospective analysis of patients in a tertiary hospital from Aug.2012 to Aug. 2015*
Results *: Total 4 neurofibrolipoma cases are reported in our institute Most of the patients were in 1 st to 2 nd decade and with equal sex distribution Surgical excision is the treatment .*
Key words*: fingers; hamartoma; nerve tissue, lipofibromatous hamartoma, neural fibrolipoma*

I. Introduction

Most soft-tissue lesions of the hand are benign, and about 5% originate from peripheral nerves. [1,2,3,4,5,7,8,]

Neurofibrolipoma or lipofibromatous hamartoma is a rare, benign, nerve-related tumour that most commonly originates from the median nerve. [1,2,3,4]

Fibrolipomatous hamartomas of nerve are rare, benign, fibrofatty malformations of peripheral nerves. [2,4,7,8,10]Other terms applied to this condition are neural fibrolipoma, lipofibromatous hamartoma of nerves and neurolipomatosis. [1,2,3,]

Most cases occur in the first 3 decades of life (often at birth or early childhood) [1,2,3,5,6] and a third are associated with macrodactyly which is referred to as macrodystrophia lipomatosa. [1,2,3,4,5,]The upper extremity is commonly involved with a marked predilection for the median nerve. [1,2,3,4,5,7,8,9,10]Other sites include the lower extremity, ulnar nerve, radial nerve and brachial plexus. [1,2,3,4,]

It usually presents as a soft,slowly growing fusiform swelling consisting of fibrofatty tissue surrounding and infiltrating major nerve and its branches (most often median and rarely ulnar nerve).[1,2,3,7,8,9,10] Swelling may be accompanied by increasing pain, tenderness and diminished sensation.[2,5,6,7,] There may be symptoms of compression neuropathy similar to carpal tunnel syndrome in some of the lesions.[1,2,]

Grossly the affected nerve is diffusely enlarged.[1,2,3,] Soft ,tan-yellow ,fusiform mass diffusely infiltrate and replace portions of the nerve.[1,2,3,5,6,].Microscopically nerve trunk is surrounded and infiltrated by fibrofatty tissue. The epineurium of the affected nerve is expanded by fibrofatty tissue. The lesion has a diffuse infiltrative character. Nerve fascicles are well preserved. [1,2,3,4,5,6,7,]Perineural fibrosis may be present. Prolonged compression of nerves by fibrofatty tissue may result in neural degeneration and atrophy. [6,9,11,12]

II. Aim and Objectives :

1)To study the histopathological features of neurofibrolipoma .
2)To correlate them with demographic and clinical features.

III. Material And Methods :

A retrospective study carried out in the Department of Pathology in a tertiary hospital. 4 cases received in the period of Aug.2012 to Aug.2015 were studied. The histopathological reports of these cases were collected from the records of histopathology section. The clinical details, radiological findings and provisional diagnosis procured from the medical record department. Corresponding slides were collected and reevaluated for confirmation of the diagnosis.

IV. Observations :

Case 1. 35 /M presented with painless swelling over ring finger of left hand .Clinically complains of mobile ,mass over left ring finger for 1 .6 yrs .Radiological and haematological investigations were normal. Clinical diagnosis of neurofibroma /neurilemomma was done. The patient underwent surgery and the mass was removed.SCO fibrofatty tissue bit m.11.5 x 3 x 1cms. Postoperatively no complains as well as recurrence.

Case 2.16/M presented with painless swelling right little finger for 9 months.SCO elongated skin covered tissue bit m.12 x 2.5 x 1 cm Histopathology shows a lesion showing fibrofatty and fibromuscular tissue growing along epineurium, perineurium and surrounding and infiltrating nerve trunk.Focal thickening of perineurium is also noted.

Case 3. 4 /F painful swelling over right ring finger since 6 months SCO fibrofatty tissue bits m 7 x 1.2 x 0.5 cm .Microscopically diagnosis of neurofibrolipoma is given.

Case 4. 13 /F painless swelling over left ring finger since 10 months .SCO fibrofatty tissue bits m 6.5 x 1.5 x 0.6 cm .Microsopically the diagnosis of neurofibrolipoma is given.

V. Results

4 cases of the Neurofibrolipoma specimens studied were confirmed on histopathological diagnosis. There was a equal distribution in both sexes (Table 1).

Further the age and sex wise distribution of the lesions was done and overall the affected the age group is 1st and 2 nd decade (Table 2).

The clinical presentation is painless swelling in 3 cases while one is with painful swelling (Table 3). Ring finger is commonly affected in our study followed by little finger and median nerve is commonly affected . (Table 4)

VI. Discussion :

Ganglion cysts, giant cell tumours of the tendon sheath, and hemangiomas are the common soft tissue lesions of the hand .[1,2,4,6,7,10]

Schwannomas and neurofibromas are the commonest peripheral nerve tumours seen in hand. [1,2,3,4,]The peripheral nerve tumours are very less and comprise <5% of all tumours of the hand. [1,2,6,7,8,]

Neural fibrolipoma or lipofibromatous hamartoma of the nerve is a rare benign tumour of unknown origin frequently seen in first 3 decades of life.[1,2,3,8,9,11,12]

Neurofibrolipoma is also called as Fibrolipomatosis of nerve ,Fibrolipomatous hamartoma of nerve ,Lipofibroma of nerve,, Lipomatosis of nerve,Neural lipofibromatous hamartoma [1,2,3,4,5,6,7,8,]

The median nerve is commonly affected in hand , but involvement of various nerves has been reported. [1,2,3,4,5,7,9,11,12].Macrodactyly has been associated in one third of patients with median nerve involvement.[1,2,4,5,6,]

On histological examination, neural fibrolipoma is characterised by fibrofatty tissue proliferation with infiltration of the epineurium and perineurium.

This tumour may cause thickening of the perineurium and perivascular fibrous tissues. [1,2,3,4,]

Following gross and histopathological criteria should be seen to confirm the diagnosis of neural fibrolipoma lesion which surrounds and infiltrates a segment of a major nerve and causes and sausage shaped expansion.The epineurium is expanded by mature adipose tissue with a fibrous component with concentric perineurial thickening which are EMA and CD34 positive with separates nerve bundles. [1,2,3,4,5,]Sometimes metaplastic bone has been reported Affected nerve may show pseduo-'onion-bulb' like change (can be confused with intraneural perineurioma).[1,2,3,4]May have associated bone overgrowth causing macrodactyly.[1,2,3,5,] No association with neurofibromatosis

Complete excision of the fibrofatty growth is contraindicated because it may cause severe sensory or motor disturbances.[1,2,9,10,11,12]

The histogenesis of fibrofatty overgrowth of nerve is contraversial. Mature fat cells have been described within the normal nerve sheath, and it is thought that proliferation of these cells leads to the fatty enlargement of the nerve and its coverings. [1,2,3,4]The relationship of these neural changes to the development of macrodactyly is not exactly known.[1,2,3,4]

MR imaging demonstrated fusiform nerve enlargement that was caused by fatty proliferation and thickening of nerve bundles. Nerve bundles appeared as serpentine tubular structures, hypointense on both T1- and T2-weighted images.[1,2,7,8,9,] The degree of fatty proliferation varied among patients.

. The variation of fatty proliferation among patients and involved nerves as well as the tendency of the abnormalities to follow the branching pattern of the nerves is well demonstrated with MR imaging.[1,2,3,4,]

While doing microsurgical dissection on soft-tissue masses of fingers and hands utmost care should be taken.[1,2,8,9,10,11,12]Frozen section examination of the mass should be performed if there is any doubt about the

diagnosis before carrying out any reconstructive surgery[1,2,4,5,]. Based on our experience, microsurgical dissection is associated with good recovery, low recurrence, and preservation of neural function..

VII. Conclusion

A neural fibrolipoma of the nerve (also known as a fibrolipomatous hamartoma) is a benign neoplasm of nerves, resulting from anomalous growth of fibroadipose tissue of the nerve sheath. In conclusion, the recommended treatment for this lesion is limited excision.

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Tables:

Table 1. Distribution as per Gender

Diagnosis	Male	Female	Total
Neurofibrolipoma	**2**	**2**	**4**
Total	**2**	**2**	**4**

Table 2 : Age and sex wise distribution of the lesions

Age (years)	M	F	Total
1 -10		**1**	**1**
11- 20	1	**1**	**2**
21 -30	-	-	-
31- 40	**1**	-	**1**
Total	**2**	**2**	**4**

Table No.3 : Clinical symptoms of patients

Clinical symptom	Number of cases
Painless Swelling	**3**
Painlful swelling	**1**

Table no.4 Location of lesion

Location	Nerve Involved	Number of cases
Left Ring Finger	**Median Nerve**	**2**
Right ring finger	**Median Nerve**	**1**
Left Little Finger	**Ulnar Nerve**	**1**

Legends:

Figure 1: Neurofibrolipoma Gross : Diffuse enlargement of nerve

Figure 2:– Neurofibrolipoma Gross : Diffuse enlargement of nerve

Figure 3: Thickened nerve bundle with intervening adipose tissue (H and E, 10 X)

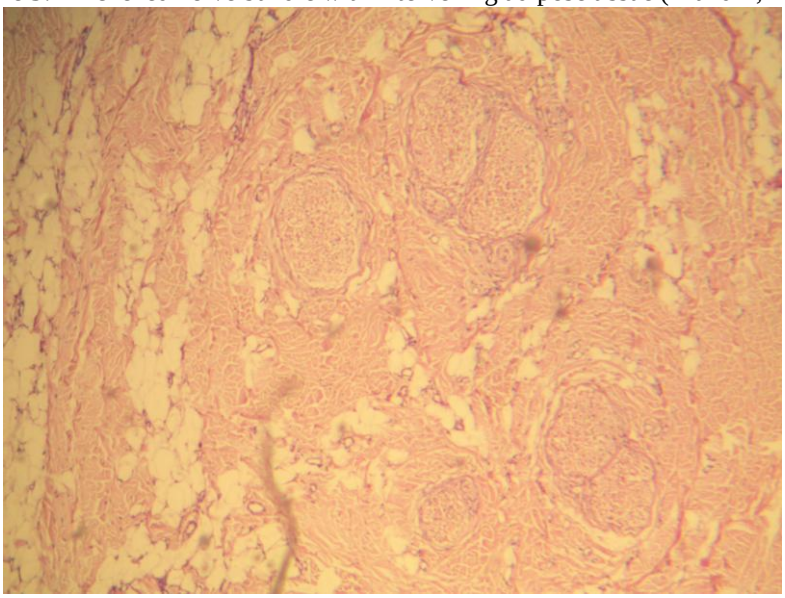

Figure 4: Nerve bundles with perineural thickening (H and E, 40 X)