Histopathological Study of neurofibrolipoma in a tertiary hospital

Dr. Vaibhav Mane *, Dr. Vishrabdha Pawar **
(Associate Prof. of Pathology*, Professor. of Pathology**)
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/neurilemmoma was done. The patient underwent surgery and the mass was removed. SCO fibrofatty tissue bit m.11.5 x 3 x 1 cms. Postoperatively no complaints 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. Microscopically 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 2nd 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 [1,2,4,5,10]

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 lipomatous hamartoma of the nerve is a rare benign tumour of unknown origin frequently seen in first 3 decades of life [1,2,3,4,5,11,12]

Neurofibrolipoma is also called as Fibrolipomatosis of nerve , Fibrolipomatous hamartoma of nerve , Lipofibroma of nerve , Lipomatosis of nerve , Neural lipomatous 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. [1,2,3,4,5,6] Macroactyly 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 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 pseudo ‘onion-bulb’ like change (can be confused with intraneural perineurioma) [1,2,3,4]. May have associated bone overgrowth causing macroactyly [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 [1,2,4,5,10,11,12].

The histogenesis of fibrofatty overgrowth of nerve is controversial. 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 macroactyly 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 [1,2,3,4,5]. 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 . [1,2,3,4,5,10,11,12] Frozen section examination of the mass should be performed if there is any doubt about the
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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.

Bibliography


Tables:

Table 1. Distribution as per Gender

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<th>Diagnosis</th>
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<tr>
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<td>4</td>
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<td>Total</td>
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<td>2</td>
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Table 2: Age and sex wise distribution of the lesions

<table>
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</tr>
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Table No.3 : Clinical symptoms of patients

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<td>Painful swelling</td>
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Table no.4 Location of lesion

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<td>Right ring Finger</td>
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<td>Left Little Finger</td>
<td>Ulnar Nerve</td>
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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)