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FIBROLIPOMATOUS HAMARTOMA OF ULNAR NERVE: A RARE CASE REPORT.

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Abstract: *Nervous fibrolipomatous hamartoma is said to be a rare tumor-like condition involving the peripheral nerves,in which the epineurium and perineurium are enlarged and distorted by excess of fatty and fibrous tissue s that infiltrate between and around nerve boundaries. The median nerve is more likely to develop a hamartoma than other nerves with a predilection for the carpal tunnel.*
A fibrolipomatous hamartoma – is a rare, benign, congenital lesion most commonly found in the median nerve, usually at the level of the wrist or hand.
We report a case of this rare condition in ulnar nerve.

I. Introduction:

Neural fibrolipoma, also known as fibrolipomatous hamartoma (FLH), is a rare, slow growing and benign tumor of a peripheral nerve, most often affecting the median nerve of younger patients.
The lesion is more common below age of 40years as revealed in previous reports, some cases in older patients h ave also been noted.
In lipofibromatous hamartoma (LH), the epineurium and perineurium are enlarged and distorted by excess matur e fat along with fibrous tissue which infiltrate between and around nerve boundaries. LH of a nerve has also bee n reported as lipomatosis, fibrolipomatous nerve enlargement, lipofibroma, fibrofatty overgrowth, fatty infiltrati on of the nerve and neurolipoma. According to WHO classification of tumors, it is called as nervous lipomatosis .[1]
Although FLH was first described in 1953, less than 100 cases have been documented so far in the available ind exed English literature. Although it is considered of congenital origin, the exact etiology of fibrolipomatous ham artoma of nerve still remains unknown. Most cases of FLH occur in first three decades of life. [2]
Intramural lipoma and fibrolipomatous hamartoma of the nerve are rare soft tissue tumors that most commonly occur in the fore-arm. [3]

II. Case Report:

A 50 year old male patient came with complaints of swelling near elbow joint since 3 months. It was not associa ted with pain and was gradually increasing in size. There was no history of any other swelling in the body. The p ast and family history was insignificant. On examination, the swelling was 2x2cms, mobile, soft, nontender with normal skin surface. The clinical diagnosis of lipoma was made and excision of the swelling was done. Per-ope ratively, the swelling was attached to ulnar nerve.
Grossly, we received a soft fibrofatty well circumscribed, unencapsulated mass, 2x2cm. Cut section showed fatt y areas.
Microscopically, the unencapsulated tumor was composed of intimate admixture of fibrous tissue and mature ad ipocytes in a haphazard manner. There was no evidence of necrosis/ atypia/ malignancy.
Nerve tissue was not seen. Based on these features, the diagnosis of fibrolipomatous hamartoma of ulnar nerve was made.

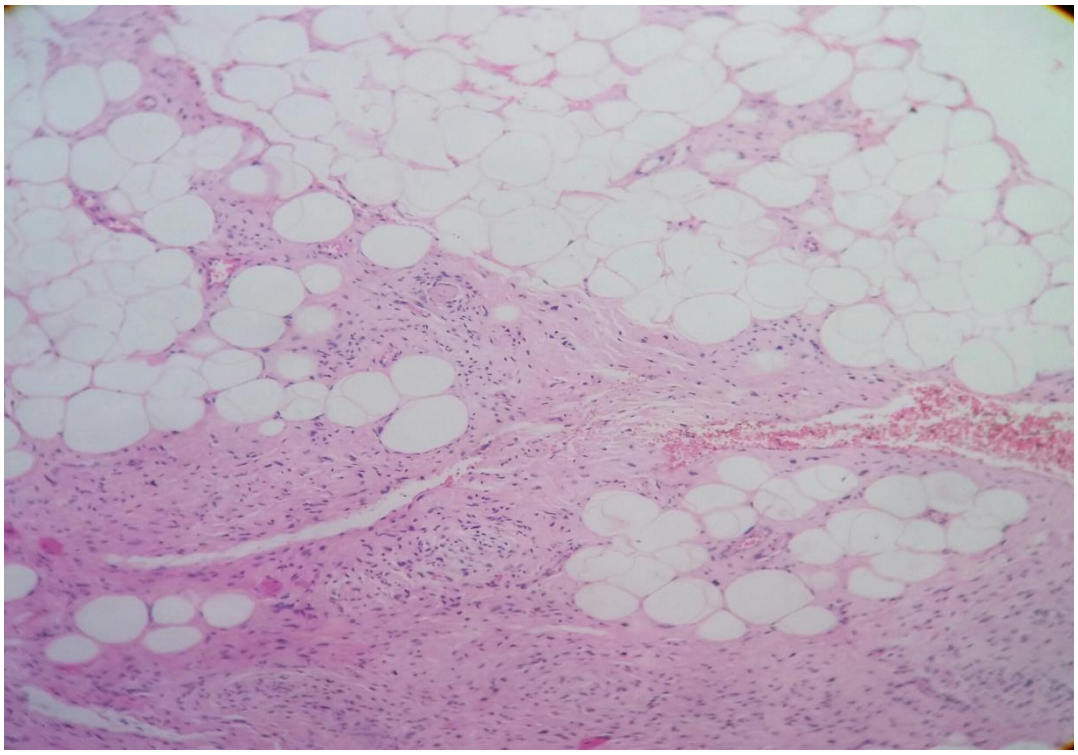

Figure:
10x, H & E: showing intimate admixture of fibrous and adipose tissue.

III. Discussion:

Intraneural lipoma, also known as neural fibrolipoma, lipofibromatous hamartoma, or perineural lipoma, is a be nign mass composed of hypertrophied fibrofatty tissue intermixed with nerve tissue. The condition is described by the term intraneural lipoma, used by Morley in 1964.

Histo-pathologically, the tumor is characterised by fibrofatty enlargement of the ulnar nerve, usually confined b y the nerve sheeth. There is massive epineural and perineural fibrosis is seen surrounding and compressing indiv idual nerve bundles. Individual nerve fibres are usually normal. The nerves and surrounding fibrosis are interspe rsed with hamartomatous fatty tissue, usually confined to the nerve sheeth.[1]

The majority of the cases present with a long standing painless mass. Neurological symptoms may occur after se veral years, mainly due to nerve compression. Compression neuropathy leads to sensory and motor symptoms in cluding pain, tenderness, diminished sensation, paresthesia and weakness. These neurological symptoms are ofte n slowly progressive.

The characteristic histological features may sometimes show compression of nerve by extensive fibrosis.

The tumor can be misdiagnosed as intraneural lipoma, ganglionar cyst, traumatic neuroma and vascular malform ations along with other intraneural tumors like intraneural lipoma, neurilemmomas and neurofibromas. Intraneur al fibromas are encapsulated and easily detached from the nerve fascicles and show proliferation of mature adip ocytes without intermingled nerve fibres. Neurilemmomas and neurofibromas are tumors of Schwann cells. Neu rilemmomas are encapsulated tumors, extrinsic to the nerve fibres and show the characteristic patterns of Antoni A and B areas. Neurofibromas are not encapsulated and small neuritis are present within the tumor. FLH, in con trast, consists of fibrofatty tissue surrounding and infiltrating the epi- and perineurium of nerve trunk.

It is said that complete excision of the tumor involving the major nerve trunk should not be done because it may lead to severe sensory or motor loss. Surgical intervention is done symptomatically. The main surgical options a re said to be decompression of the nerve or microsurgically dissecting the neural elements or the excision of the nerve with or without grafting. [2]

However in our case, ulnar nerve was not excised and patient had no sensory or motor disturbances.

The maximum size of the intraneural lipoma was reported to be 200mm x 100mm and the minimum to be 25 x 4 0mm. a history of trauma is usually absent. The pathology of fibrolipomatous hamartoma of the nerve usually sh ows a mixture of collagenous fibres, fatty tissue and nerve fibres. The etiology of FHL of the nerve is reportedly unknown, and several other etiologic factors have been postulated like abnormal development of flexor retinacu lum in children, history of trauma and chronic nerve inflammation. It has been reported that FHL tend to arise in younger age groups before the third decade and have no sexual pre dominance. [3]

Our case was unusual in that patient was in 5^{th} decade and had no clinical features related to ulnar nerve.

IV. Conclusion:

Eventhough rare, fibrolipomatous hamartoma should be considered in the differential diagnosis of peripheral ner ve tumors.

References:

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Legends:

10x, H & E: showing intimate admixture of fibrous and adipose tissue.