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 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 40 years as revealed in previous reports, some cases in older patients have also been noted.

In lipofibromatous hamartoma (LH), the epineurium and perineurium are enlarged and distorted by excess mature fat along with fibrous tissue which infiltrate between and around nerve boundaries. LH of a nerve has also been reported as lipomatosis, fibrolipomatous nerve enlargement, lipofibroma, fibrofatty overgrowth, fatty infiltration of the nerve and neurolipoma. According to WHO classification of tumors, it is called as nervous lipomatosis.

Although FLH was first described in 1953, less than 100 cases have been documented so far in the available indexed English literature. Although it is considered of congenital origin, the exact etiology of fibrolipomatous hamartoma of nerve still remains unknown. Most cases of FLH occur in first three decades of life.

Intramural lipoma and fibrolipomatous hamartoma of the nerve are rare soft tissue tumors that most commonly occur in the forearm.

II. Case Report:

A 50 year old male patient came with complaints of swelling near elbow joint since 3 months. It was not associated with pain and was gradually increasing in size. There was no history of any other swelling in the body. The past and family history was insignificant. On examination, the swelling was 2x2cm, mobile, soft, non-tender with normal skin surface. The clinical diagnosis of lipoma was made and excision of the swelling was done. Per-operatively, the swelling was attached to ulnar nerve.

Grossly, we received a soft fibrofatty well circumscribed, unencapsulated mass, 2x2cm. Cut section showed fatty areas.

Microscopically, the unencapsulated tumor was composed of intimate admixture of fibrous tissue and mature adipocytes 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.
III. Discussion:

Intraneural lipoma, also known as neural fibrolipoma, lipofibromatous hamartoma, or perineural lipoma, is a benign mass composed of hypertrophied fibrofatty tissue intermixed with nerve tissue. The condition is described by the term intraneural lipoma, used by Morley in 1964. Histopathologically, the tumor is characterised by fibrofatty enlargement of the ulnar nerve, usually confined by the nerve sheath. There is massive epineural and perineural fibrosis seen surrounding and compressing individual nerve bundles. Individual nerve fibres are usually normal. The nerves and surrounding fibrosis are interspersed with hamartomatous fatty tissue, usually confined to the nerve sheath. The majority of the cases present with a long standing painless mass. Neurological symptoms may occur after several years, mainly due to nerve compression. Compression neuropathy leads to sensory and motor symptoms including pain, tenderness, diminished sensation, paresthesia and weakness. These neurological symptoms are often slowly progressive.

The characteristic histological features may sometimes show compression of nerve by extensive fibrosis. The tumor can be misdiagnosed as intraneural lipoma, ganglionic cyst, traumatic neuroma and vascular malformations along with other intraneural tumors like intraneural lipoma, neurilemmomas and neurofibromas. Intraneurial fibromas are encapsulated and easily detached from the nerve fascicles and show proliferation of mature adipocytes without intermingled nerve fibres. Neurilemmomas and neurofibromas are tumors of Schwann cells. Neurilemmomas 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 contrast, consists of fibrofatty tissue surrounding and infiltrating the epineural 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 are said to be decompression of the nerve or microsurgically dissecting the neural elements or the excision of the nerve with or without grafting. 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 40mm. A history of trauma is usually absent. The pathology of fibrolipomatous hamartoma of the nerve usually shows 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 retinaculum 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 predominance.

Our case was unusual in that patient was in 5th 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 nerve tumors.

References:

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