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FIBRO-LIPOMATOUS HAMARTOMA.

Orthopaedics

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KEYWORDS

Fibro lipomatous hamartoma, Neural fibro lipoma, LFH, Peri neural lipoma

INTRODUCTION :

Fibro-lipomatous hamartomas are non-malignant tumors that are commonly seen in infants and rarely affect young adults and children.[1] Neural fibro-lipoma, lipo-fibromatous hamartoma, peri-neural lipoma and intra-neural lipoma are the other terms used for fibrolipomatous hamartoma. Among all, the median nerve is by far the most commonly affected(80% of cases) and less commonly affected are the and radial nerves and the brachial plexus. [2]

Although median nerve is the commonly affected nerve, lesions are also found in the radial, sciatic, plantar and ulnar nerves. Further, the median nerve is most commonly involved at the level of hand and wrist.[3]

Clinical findings:

This 16 year old male, presented with an asymptomatic mass, on forearm / volar aspect of wrist, which was soft and slow growing. Pain, paraesthesia and other features suggesting carpel tunnel syndrome were seen due to compression of the median nerve. Fibro-lipomatous hamartomas affects both the sexes equally.[4]Although the exact pathogenesis of this tumour is not known, it is assumed that, it could be due to a congenital abnormality, in the fibro fatty tissue growth that results in fusiform enlargement of the nerve due to infiltration of the endoneurium, perineurium and epineurium.[5]

CT / MRI FINDINGS:

Due to the thickening of the nerve bundles and the co-existing fibrous proliferation, the hamartoma showed a fusiform enlargement of the nerve that resembled a tubular or a serpentine structure. The MRI features of fibromatous hamartomas are pathognomonic, presenting with high signal intensity, from the fat on T1 and T2-weighted sequences which will get dropped out on a fat suppressed image, surrounding the bands of enlarged nerve fascicles.[6]

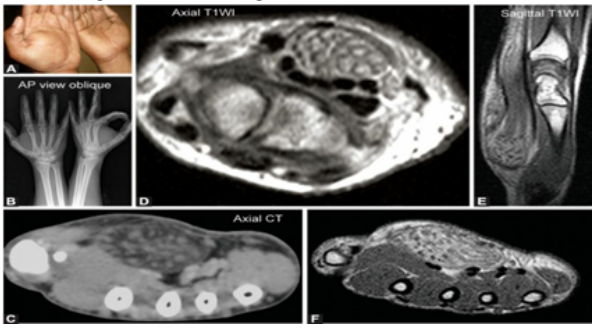

Figure 1(A to F) :

Fig 1 (A) : Hands show swelling of the left thenar eminence.
Fig 1(B) : X-ray AP and oblique views of left hand shows a swelling of the soft tissue in the region of thenar eminence.

Fig 1(C) : Axial CT section shows a well-defined heterogeneous lesion seen in ventral aspect distal to wrist, anterior to the flexor tendons having fatty density within the lesion.

Fig 1 (D to F) : Axial T1WI and sagittal T1WI (D and E) shows well-defined oval lesion on ventral aspect just distal to wrist appearing hyper intense with flow voids within it. In the fatty lesion, flow voids are seen abutting the flexor tendons with polka dot appearance (D to F) because of enlarged median nerve with fat infiltration in between fascicles of median nerve.

TREATMENT :

Tumor excision is not necessary in all cases. Neurogenic pain is by far the most important drawback in case of a resection. The other drawback is that, it is difficult to identify the margins for resection as LFH involves the entire nerve till the plexus.[7] Decompressing the peripheral nerves in case of LFH without macrodactyly, can relieve pain and paraesthesia, instead of motor and sensory sequelae permanently.

Debulking or amputation of the finger is the choice of treatment in case of LFH with macrodactyly. However amputation of finger is not frequently done in case of children. Joint debridement, tendon transfer and osseous overgrowth excision are the other reconstructive procedures , done in selected patients to improve hand function.

CONCLUSION :

LFH is asymptomatic, unless it is present for many years. The symptoms are mainly due to nerve compression, rather than intra-neural tumour involvement. Asymptomatic patients do not need any intervention. In case of high morbidity or impossible decompression, excision surgery is contraindicated. Good outcome can be achieved by careful micro-surgical dissection.

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