ASYMMETRICAL MULTIFOCAL GIANT CELL TUMOR OF THE TENDON SHEATH - AN UNCOMMON ENTITY

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INTRODUCTION

Giant cell tumor of the tendon sheath is an uncommon tumor which can be clinically confused with more common conditions. Histopathological findings are characteristic. The tumor is usually solitary. Multiple lesions are extremely rare. We report a case of multi focal bilaterally asymmetrical giant cell tumor of the tendon sheath occurring on both hands of a 60 years old lady which was clinically misdiagnosed as implantation epidermoid cyst.

CASE REPORT

A fifty five years old lady presented with a slowly growing firm, asymptomatic lobulated nodular swelling on the lateral side of the distal inter phalangeal joint of left little finger for the last three years. For the last one year she noticed another swelling with similar characters on the lateral side of right index finger just proximal to the meta-carpo phalangeal joint (fig. 1). There was no other complaint. Clinical diagnosis at that time included implantation epidermoid cysts. Both of the lesions were excised under local anesthesia and submitted for histopathology. The histopathology from both specimens revealed plump histiocytes laden with hemosiderin and fat. Multiple giant cells were seen with haphazardly arranged nuclei (fig. 2). The tumor was surrounded by a capsule. The findings were consistent with a diagnosis of giant cell tumor of the tendon sheath. The patient was lost to follow up.

DISCUSSION

Giant cell tumor of the tendon sheath also called pigmented villo-nodular synovitis, is a rare benign synovial mesenchymal tumor of unknown etiology. It is generally related to tendon sheaths, capsular ligaments and joints or its origin may be unknown.

It occurs most commonly in the flexor tendons of the hands, followed by ankles, toes, and knees. In a study of 207 cases, the tumor was situated on the digits in 182 cases where as in 25 cases the location was in large joints [1]. The tumor occurs more commonly in adult women. The age ranged from 6 years to 77 years (mean age 37.5 years) [2]. It is quiet uncommon in children.

On the hands volar aspect of the proximal phalanx of second and third fingers are mostly involved. Dorsal involvement is less common. Wrist may be involved in about 17% of cases [2]. Rarely palms may be involved. Feet may also be affected uncommonly with involvement of the toes and ankle. The tumor rarely present as an intra articular nodule (localized nodular synovitis). Unusual rare locations include spine, tendon of the superior oblique muscle, periungual and subungual areas.

The most common presentation is a solitary palpable, painless mass present for several weeks to years. About one fourth of the cases may develop symptoms due to pressure effect on the adjacent bone in form of cortical sclerosis, cortical erosion or medullary extension by the tumor of the bone [2]. Other symptoms include mild pain (21%), loss of sensation (8%) and joint restriction (6%) [2].

The tumor is most of the time solitary. Multiple lesions are extremely rare. Large studies show that less than 1% of cases have a multicentric origin. In a study of 118 cases only one patient had two distinct lesions [3]. In another series of 141 patients, 3.5% had multicentric lesions [2]. Bilaterally symmetrical involvement has been described

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by Khanduja in one patient [4]. Our case also had two tumors but situated at different locations on different hands. Such asymmetrical distribution has not been described before.

Incidence of malignant lesions is extremely rare and only eight cases have been reported [5].

Microscopically the tumor consists of a mixture of round to polygonal histiocytes, foam cells, hemosiderin laden macrophages and multinucleate giant cells. There may be large clefts or pseudo glandular spaces lined by synovial cells. The lesion may be capsulated or uncapsulated.

Differential diagnosis on the digit includes ganglion, Herbeden nodule, implantation epidermoid cyst, bursitis, gout, hematoma osteoma, exostosis, xanthoma and haematoma.

The treatment of choice is surgical excision which has 45% local recurrence [6]. The risk factors associated with recurrence include presence of adjacent degenerative joint disease, location at the distal interphalangeal joint of the finger or interphalangeal joint of the thumb, presence of an osseous erosion and histologically absence of a capsule. Excision followed by radiotherapy will reduce the incidence of recurrence in these patients.

CONCLUSION

Giant cell tumor of the hand is a rare entity and mostly presents as a solitary lesion. However it should be kept in differential diagnosis of asymmetrical multiple nodular lesions presenting on the hands.

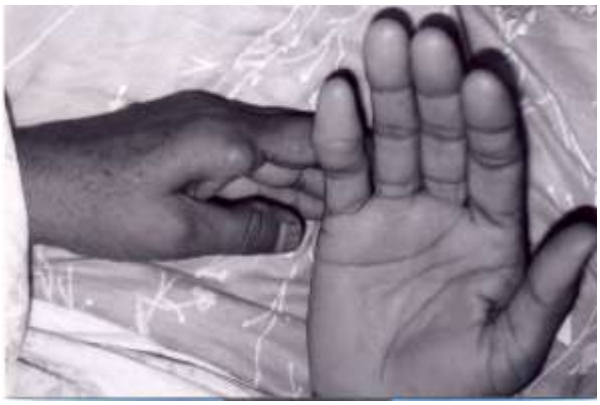

Fig. 1: Two lesions distributed asymmetrically on the base of index finger of right hand and of little finger of left hand.

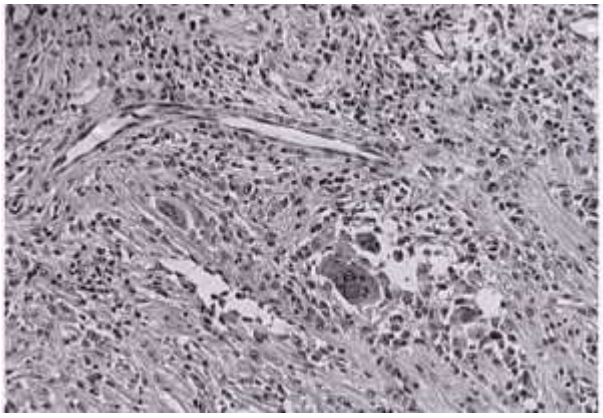

Fig. 1: Multinucleate giant cells along with plump histiocytes (H&E X 40).

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