Giant cell tumor of the tendon sheath mimicking a plexiform neurofibroma

Swagata Arvind Tambe, Mayur Bhobe, Hemangi R. Jerajani

Department of Dermatology, MGM Medical College and Hospital, Kamothe, Navi Mumbai, Maharashtra, India

ABSTRACT

Giant-cell tumor of the tendon sheath (GCTTS) is a benign soft tissue tumor of the limbs arising from the complex of the tendon sheath and periarticular soft tissues of small joints. It is the second most common benign space occupying lesion in the hand and usually presents as a painless soft tissue mass, which grows slowly in size for many years. We present an interesting case of an enormous GCTTS presenting as a slowly growing mass over left sole of a 52-year-old woman. The duration of GCTTS may range from a few weeks to 30 years but in our case the duration of tumor was almost 48 years, which could be the longest reported duration of GCTTS.

Key words: Giant cell tumor of tendon sheath, sole, unusual presentation

INTRODUCTION

Giant-cell tumor of the tendon sheath (GCTTS) is the second most common benign space-occupying lesion of the hand (2–5%), after ganglion.[1,2] It usually occurs in the third to fifth decades of life. It occurs most commonly in the flexor tendons of the hands, followed by the ankles, toes, and knees.[2] This tumor usually presents as a painless soft tissue mass. The duration of the tumor can be as long as 30 years.

CASE REPORT

A 52-year-old female, farmer by occupation presented with an asymptomatic swelling over the instep of her left sole since three years of age. The swelling gradually increased in size over a period of many years. Since the last few years, patient started developing discomfort on weight bearing or standing for prolonged duration. There was difficulty in walking with footwear. There was no history of preceding trauma to the foot or history of discharging sinuses from the lesion or other symptoms suggestive of genodermatoses like Proteus syndrome or neurofibromatosis. The patient also denied personal or family history suggestive of tuberculosis.

General examination of the patient did not reveal any abnormality. Cutaneous examination revealed a single ovoid swelling of 6 cm × 7 cm with well-defined edges over the instep of left sole with overlying normal skin [Figure 1a and b]. On palpation, the swelling was minimally tender, and the temperature over the swelling was normal. Swelling was soft, lobular with a bag of worms feel. It was noncompressible, nonreducible, nonpulsatile and was not fixed to the skin. It was less mobile, slightly more prominent on contracting underlying muscle. There was no evidence of regional lymphadenopathy. The differential diagnoses of plexiform neurofibroma, angioleiomyoma, Proteus syndrome, striated muscle hamartoma and lipoma were considered.

Radiographs of left foot showed a soft tissue swelling on the medial aspect of the left sole without evidence of any cortical erosion. Ultrasonography of swelling showed a hypoechoic nonvascular subcutaneous mass probably of neural origin.

On performing a punch biopsy, frank white discharge was seen. Gram stain of the discharge showed numerous neutrophils, but pus culture examination was sterile. Histopathological examination revealed a partially circumcised tumor consisting of diffuse infiltration of mononuclear cells, epithelioid cells and osteoclast-like giant cells in a dense collagenous stroma with few mitotic figures [Figure 2a and b]. A final diagnosis of GCTTS was

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Address for correspondence:
Dr. Swagata Arvind Tambe,
19/558, Udyan Society, Nehru Nagar, Kurla East, Mumbai - 400 024, Maharashtra, India.
E-mail: swagatatambe@gmail.com

made. Excision of the swelling was performed. The swelling resolved completely without any recurrence after two years of follow-up [Figure 3].

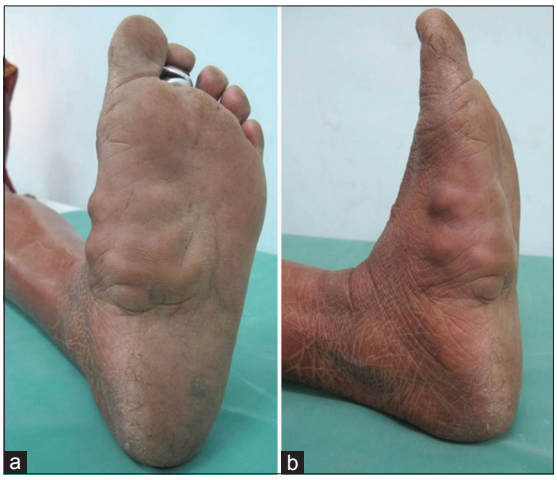

Figure 1: (a) Cutaneous examination of the patient revealed a single ovoid swelling of 6 cm × 7 cm with a lobulated surface over the instep of left sole with overlying normal skin (b) extension of the swelling on the medial aspect of the left foot

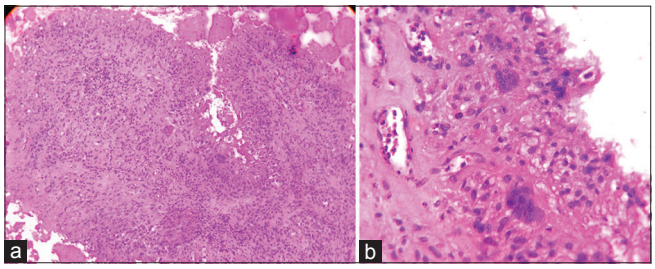

Figure 2: (a) Histopathological examination from the lobulated mass (H and E, x 40) revealed a partially circumscribed tumor consisting of diffuse infiltration of mononuclear cells, epithelioid cells in a dense collagenous stroma. (b) Higher magnification (H and E, x 100) revealed diffuse infiltration of stroma with mononuclear cells, epithelioid cells and osteoclast like giant cells in a dense collagenous stroma with few mitotic figures

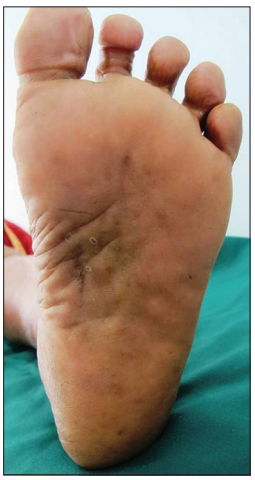

Figure 3: Complete resolution of the swelling after surgical excision

DISCUSSION

Giant-cell tumors of the tendon sheath are benign soft tissue tumors of the limbs that arise from the complex of the tendon sheath and periarticular soft tissues of the small joints. It was first described by Chassaignac in 1852.[3] Numerous terms have been used to designate this pathological condition, including benign synovioma, localized nodular tenosynovitis, tenosynovial giant-cell tumor, fibrous histocytoma of synovium, histiocytic giant-cell, xanthomatous giant-cell tumor, xanthoma of the synovium, xanthogranuloma, xanthosarcoma, fibrous xanthoma, fibroma of tendon, myeloid endothelioma, endothelioma, villous arthritis, sclerosing hemangioma, fibrohemosideric sarcoma, giant-cell fibrohemangioma, and localized nodular synovitis.[2-6]

It affects individuals between 30 to 50 years of age with a female preponderance.[2] The proposed pathogenic theories include role of trauma, disturbed lipid metabolism, osteoclastic proliferation, infection, vascular disturbances, immune mechanisms, inflammation, neoplasia, and metabolic disturbances.[2,3,7]

It is the second most common tumor of the hand, with simple ganglion cysts being the most common.[2] The hand is the most common site (77%) with predominant involvement of the volar aspect and fingers adjacent to the distal interphalangeal joint. They are very rarely seen on the foot (3%).[6] Though the involvement of feet is rare, it is the second most common site. In the feet, toe tips, ankle and large joints of the foot are commonly affected.[8] Rarely other sites including the vertebral column may be affected.

Usually, the lesions are asymptomatic and present with a palpable, often painless, soft tissue mass that gradually increases in size over a long period.[2-4] Duration ranges from weeks to as long as 30 years. In our case, the swelling was noticed at the age of 3 years; thus the total duration of the tumor at presentation was 48 years, which could be the longest duration reported in the literature. The tumor mass may vary from solitary to multiple discrete soft tissue nodules.[1] Usually, GCTTS are tendon-based, well circumscribed and localized. However, some of them may grow diffusely or expand, as in our case. Al-Qattan presented the most popular classification of the GCTTS into two types, determined by the presence of a pseudo capsule surrounding it. Each type was also sub-classified depending on the thickness of the capsule, lobulation of the tumor, the presence of satellite lesions, and the diffuse or multicentric growth of the tumor.[4]

The newer classification for GCTTS, is also proposed by Al-Qattan where he classifies Type I-as single tumor which is round or multilobulated, and Type II, where there are two

or more distinct tumors which are not joined together. This classification is useful in predicting the recurrence as satellite lesions are often missed.

Conventional radiographs commonly present a completely normal radiological view or may show a soft tissue mass.[5,9] Rarely, radiographs may present cystic changes without cortical expansion, bony pressure erosion or indentation, periosteal reaction, calcification, and degenerative changes of the adjacent joints.[3,5,9] Magnetic resonance imaging, computed tomography, ultrasonography and color Doppler sonography images may reveal the precise topography of the tumor and are very useful in preoperative planning.[2,3,9] Sonographic appearance reveals hypoechoic mass and may show vascularity on color Doppler sonography.[10]

Histopathology is usually diagnostic, but cytodiagnosis has been found useful.[11] Histopathology reveals multinucleated giant cells, polyhedral histiocytes, fibrosis and hemosiderin deposits.[12] Histological features of cellularity and mitoses, though previously found significant, were not found to influence recurrence.

Complete surgical excision remains the mainstay of treatment, assisted either with an operating microscope or a magnifying loupe. Meticulous dissection and exploration are essential because satellite lesions are common. Possible complications of GCTTS include local recurrence, numbness, joint stiffness, painful scar, and skin necrosis.[2] Local recurrence has been reported in up to 45% of cases.[13] The tumor has extensions that go around and under critical structures, including the neurovascular bundle. Difficulties excising the lesion and using sparing surgery to maintain function have been cited by many authors as factors contributing to recurrence. Postoperative radiotherapy has been advocated by some authors to reduce the recurrence rate.[13]

This case has been reported because of the unusual site of presentation on the sole, long duration of the tumor and the large size of the lesion that initially suggested a diagnosis of plexiform neurofibroma, but on investigation the diagnosis of GCTTS was confirmed.

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