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Case Report / *Olgu Sunumu*

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Multifocal giant cell tumor of the tendon sheath occuring at different localizations of the same tendon of a finger: a case report and review of the literature

Aynı parmak tendonunun farklı tendon kılıfı ve odaklarında oluşan çoklu dev hücreli tümör: Olgu sunumu ve literatür incelemesi

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The giant cell tumor of the tendon sheath is regarded as one of the most common neoplasms of the hand. This tumor usually manifests itself as a localized, solitary, painless and palpable subcutaneous nodule on the palmar aspect of a digit. A multifocal origin of the tumor has rarely been reported in the literature before. In this article we present a case of a giant cell tumor of the tendon sheath, in which two separate lesions developed simultaneously on the same tendon (flexor digitorum superficialis) of the little finger of the right hand together with a literature review about multifocal cases.

Key words: Multifocality; giant cell tumor of tendon sheath; recurrence.

Tendon kılıfının dev hücreli tümörü elin en sık görülen tümörlerinden biri olarak kabul edilir. Bu tümör çoğunlukla parmağın palmar yüzünde izole, soliter, ağrısız ve ele gelen derialtı yerleşimli bir nodül şeklinde görülür. Bu tümörün çok odaktan köken aldığı literatürde nadiren bildirilmiştir. Bu makalede literatür incelemesi eşliğinde, sağ elin küçük parmağının aynı tendonu üzerinde (fleksör dijitorum superfisiyalis) eş zamanlı iki ayrı lezyon gelişen bir dev hücreli tendon kılıfı olgusu sunuldu.

Anahtar sözcükler: Multifokalite; dev hücreli tendon kılıfı tümörü; rekürens.

The giant cell tumor of the tendon sheath (GCTTS) is the second most frequent soft tissue tumor of the hand encountered after ganglion cysts.[1] The giant cell tumor of the tendon sheath typically affects adults in the 3rd and 4th decades of life and is more often seen in women than men.[1,2] The tumor usually manifests itself as a slow-growing, localized, usually painless and palpable subcutaneous mass on the palmar aspect of a digit.[2,3] Involvement of the foot, wrists, ankles and elbows are rare.[4] They are usually relatively small ranging in size from 0.5 to 4 cm in diameter.[1]

The etiology of GCTTS remains unknown. It has been considered to be an inflammatory process arising as a result of chronic antigenic stimulation[5] or a history of trauma,[1] Hirohata[6] proposed a localized disturbance of lipid metabolism with lipid-laden foamy cells as the likely cause. Despite the frequency of large lipid deposits, elevated serum lipid levels were not described.[2,3] On the other hand, GCTTS was considered to be a neoplasm arising from the lining and sublining cells of the tendon sheath.[3,7] Recent studies demonstrating cytogenetic abnormalities;[8] the natural behaviour of local recurrences and multifocality[3] have raised the idea that GCTTS is a neoplasm. Clonal chromosomal aberrations were observed suggesting a neoplastic etiology in one study.[9] Overall, most

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investigators now believe GCTTS to be a neoplastic process.[9] Despite a well-recognized propensity for local recurrence, few reports of malignant GCTTS have also been documented.[10]

To the best of our knowledge, multifocal lesions of GCTTS have rarely been described in the literature.[11-13] Therefore, we would like to report a case of GCTTS in which two separated lesions developed on the tendon of the flexor digitorum superficialis of the right hand.

CASE REPORT

A 26-year-old man operated three years ago on a mass at the 5th metacarpo-phalangeal level of the palmar aspect of the right hand at another center was admitted to our out-patient clinic for a recurrent tumor at the distal interphalangeal level of the palmar aspect of the little finger. On the physical examination, the mass was well-defined, slightly tender, stiff and located at the subcutaneous level. The plain radiographs showed no bony abnormality. The patient had no history of antecedent trauma or systemic illness.

The tumoral lesion at the distal interphalangeal level of the palmar aspect of the little finger, originating from the flexor digitorum superficialis (FDS) tendon and invading the A4 pulley, was excised as well as the pulley (Figure 1a). An A4 pulley reconstruction was planned to be performed on the transverse carpal ligament (TCL) at the wrist crease. During the TCL graft harvesting, a second mass on the tendon of the FDS at the level of the ¼ distal forearm was incidently detected (Figure 1b). Intraoperatively, both masses were found to originate from the FDS tendon. No continuity between the masses was observed (Figure 2). Grossly, the mass at the distal interphalangeal joint was measuring approximately 1 cm in diameter, yellowish tan to orange colored; while the other one at the level of the ¼ distal forearm was approximately 2 cm in diameter; lobulated and also yellowish tan to orange colored (Figure 3). The mass on the right distal forearm has also been excised with invaded particular muscle-tendon part portion.

Histopathological examination of both masses revealed mononuclear cells in a hyalinized collagenous matrix with scattered multinucleated giant cells. Hemosiderin granules were observed in the cytoplasm of the mononuclear cells (Figure 4). These findings were consistent with GCTTS.

Six months after surgery, hand functions were preserved without any recurrences (Figure 5a, b).

DISCUSSION

Although local recurrences after operative excision have been reported in up to 44% of cases,[14] multifocal lesions of GCTTS are extremely unusual. In a series of 207 cases with GCTTS, Ushijima et al.[1] recorded only one patient with two tumors at different sides, one on the right ring finger, the other on the left great toe. Phalen et al.[3] also described three patients with two tumors: First patient had bilateral tumors on each hand while the other two patients had two tumors on the same finger but not on the same tendon. In another study[2] with 117 cases, one patient had two distinct lesions. Furthermore, Reilly et al.[14] also reported three patients with multifocal involvement, with one

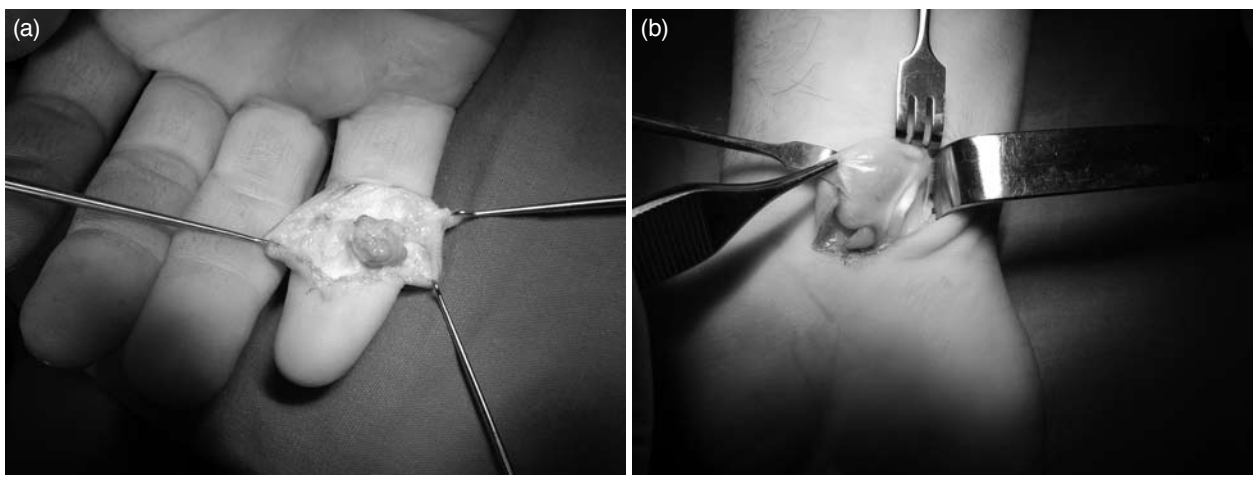

Figure 1. (a) The mass at distal interphalangeal level of the palmar aspect of the little finger. **(b)** The mass at the level of ¼ distal forearm.

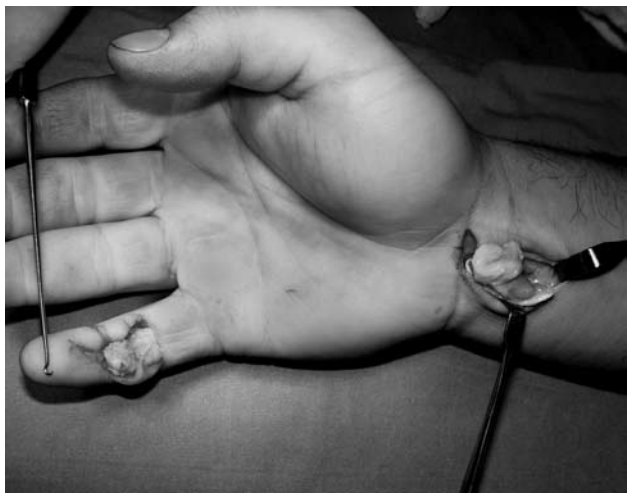

Figure 2. Operative view showing two nodular lesion on the same tendon.

having bilateral tumors. Moreover, a case with bilateral symmetrical involvement of the thumb[11] and five multifocal cases on the hand[15] were reported.

Despite these rare multifocal cases, we could only find two reports on patients who had multifocal lesions on the same tendon of the hand like the case presented here. Park[13] described a patient with GCTTS who had two separated masses on the flexor pollicis tendon sheath of the thumb. Hitora et al.[12] also presented a case with two tumors on the little finger: one at the metacarpophalangeal level, the other at the interphalangeal level; both on the palmar aspect. Table I summarizes the multifocal cases in the literature.

As the etiology and pathogenesis of GCTTS remain controversial, the cause of multifocality is also unidentified. Insufficient excision of the mass was related with policentricity in one study.[15] Similarly, in the case presented here, the lesion at the level of the ¼ distal right forearm could have appeared in collaboration with the recurrent lesion at the distal interphalangeal level due to insufficient excision. On the other hand, it is possible that these recurrent lesions could not have been identified during the first surgery session performed on the mass at metacarpophalangeal joint level.

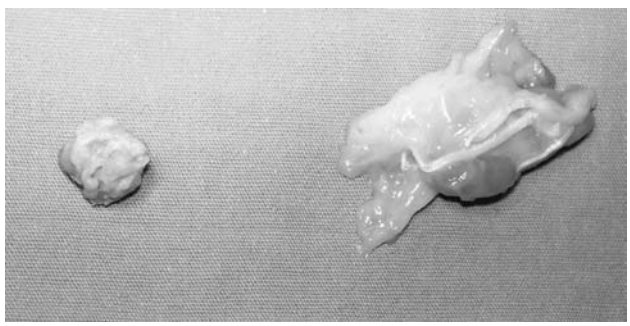

Figure 3. Gross photographs of excised tumors, intraoperatively.

The differential diagnosis of a subcutaneous tumor on the hand is an extensive task. The diffuse form of GCTTS has to be differentiated from the localized form of GCTTS. This diffuse type of GCTTS can be differentiated by a lesser frequency, through its localization such as on large joints, the invasion to the surrounding tissue, its aggressiveness and malign transformation.[1] Myxoid cysts, dermatofibroma, glomus tumors, neurofibroma, sweat gland tumors, giant cell tumors of the soft tissue, giant cell tumors of the bone, fibromas and rheumatiod nodules and primitive neuroectodermal tumors should also be considered in the clinical differential diagnosis of the GCTTS. These tumors are usually differentiated by the help of histopathological examinations.[1,3,16,17]

Our case is especially interesting because the recurrent tumor at the distal interphalangeal level of the right little finger was detected preoperatively, while the mass at the level of the ¼ distal right forearm was detected peroperatively and presented no symptoms or signs before the surgery.

Since the GCTTS is a non-malignant, encapsulated, slow-growing neoplasm; the treatment of choice is a complete local excision. In our case, after

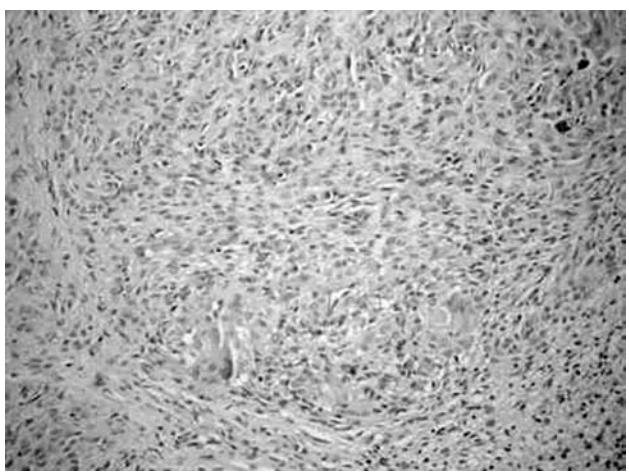

Figure 4. Histopathological examination of the excised nodule on the distal interphalangeal level revealing hemosiderin granules in cytoplasm of mononuclear cells, scattered multinucleated giant cells and mononuclear cells in a hyalinized collagenous matrix (H-E x 200).

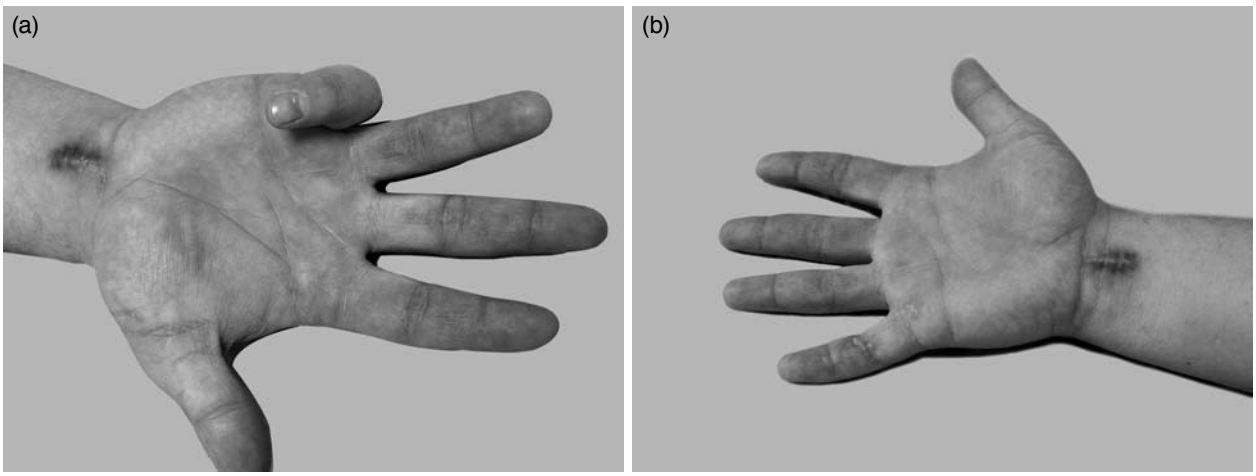

Figure 5. (a, b) Clinical photographs of the hand showing preservation of the hand functions six months after surgery.

the excision of the tumors, a pulley system reconstruction was performed additionally. The pulley system reconstruction, although not reaching to preinjury levels, yields better results than leaving the member unrepaired. The A2 and A4 pulleys are accepted to be the most important pulleys of the hand and it has been reported that the hand has a near-normal function if the A2 and A4 pulleys are intact.[18] To maintain normal hand functions, it is recommended to repair at least these two pulley systems.[19] There have been various techniques developed for the reconstruction, however, the ideal method is yet unknown. For pulley reconstructions, synovial lined grafts are recommended due to their lower degree of resistance development during the tendon/pulley system work and the lesser scar formation with faster healing. As autologous grafts, the flexor tendon tail, tendon graft or extensor retinaculum can be used.[19] We used the technique that had been described by

TABLE I

Literature review of multifocal cases

References	Number of the patient	Gender/age (year-old)	Localization
Ushijima et al.[1]	1	Not reported	Right ring finger Left great toe
Phalen et al.[3]	2	Not reported	Palmar aspect of the right thumb Dorsal aspect of the left middle finger
Phalen et al.[3]	3	Not reported	On the same finger Dorsum of the distal phalanx of the finger Palmar aspect of the middle phalanx
Phalen et al.[3]	4	Not reported	Dorsal and palmar aspect of a same finger
Jones et al.[2]	5	Not reported	Not described
Park[13]	6, 7, 8	Not reported	Not described
Ushijima et al.[10]	9	Male/46	Interphalangeal joints of the thumbs
Özalp et al.[15]	10, 11, 12, 13, 14	Not reported	On the hand
Hitora et al.[12]	15	Male/33	Flexor pollicus longus tendon sheath of the palmar aspect of the left thumb
Dalal and Bostock[11]	16	Male/28	Palmar aspect of the metacarpophalangeal joint Distal interphalangeal joint of the right little finger
Our case	17	Male/26	Distal interphalangeal level of the right little finger Palmar aspect of the level of $^1/_4$ distal right forearm

Klinert and Bennett[20] for pulley reconstruction in our case except that the FDS graft was not used because of the invasion of the mass into the tendon. Instead of the extensor retinaculum, a synovial graft "TCL", which is being divided during carpal tunnels surgeries was used in addition to the strip that had been harvested from the antebrachial fascia parallel to the wrist crease. The TCL was not divided completely and its pulley function was restored as much as possible. The graft that had been harvested as a strip was passed through the remnant of the pulley extensor mechanism.

On the basis of our case and the literature reviews, a multifocality of the neoplasm without any signs and symptoms rather than invasiveness or metastasis should be taken into consideration before planning any radiological examinations, during local surgical treatment modalities and the follow-up of GCTTS.

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