Giant Cell Tumor of the Tendon Sheath with Medullary Extension: a case report

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Giant cell tumor of the tendon sheath with medullary extension in the foot has rarely been reported before. We report a 28-year-old man who had a painless swollen lesion over the proximal phalanx of the second toe of the left foot for several years. Conventional radiographs showed a lobulated lucency in the bone marrow of the proximal phalanx surrounded by a soft-tissue density over the second toe. MRI revealed a 1.2-cm tumor mass at the volar side of the proximal phalanx with medullary extension. Histological findings were compatible with giant cell tumor of the tendon sheath with tumor extension to the marrow spaces.

Key words: Giant cell tumor; Magnetic resonance imaging (MRI); Tendon

CASE REPORT

A 28-year-old man suffered from pain, erythema and swelling of the metatarsophalangeal joint of his left great toe and second toe for a week. He had a history of gouty arthritis under regular medical treatment for several years. The patient also had another painless and swollen lesion at the proximal phalanx of the left second toe for several years. Neither tenderness nor erythema over the proximal phalanx was noted.

Conventional radiographs showed slightly increased soft-tissue density with stippled calcification (Fig. 1a), which was presumed to be a tophaceous gout, around the proximal end of the proximal phalanx of the left second toe. There was another soft-tissue mass surrounding the distal end of the proximal phalanx and lucency in the adjacent bone marrow of the second toe. No calcifications of the tumor matrix or periosteal reaction was noted. Bone scan of both early and delayed phases revealed hot-spots over the great toe and the second toe of the left foot.

MRI (0.5-T, Vectra, General Electric, Wis.) showed a soft-tissue tumor, measuring 1.2 cm, on the...
volar side of the distal end of the proximal phalanx of the second toe of the left foot (Fig. 1b, 1c, 1d). The mass (in relation to the flexor tendon) was clearly delineated on the axial plane, showing low signal intensity on T1-weighted images and higher signal intensity on proton-density images. The medullary extension of the mass was well identified on coronal short tau inversion-recovery (STIR) images. The soft-tissue lesion with calcifications in the second toe, which was presumed to be a tophaceous gout on radiographs, showed high signal intensity on T1-weighted images and low signal intensity on STIR images.

Figure 1. a. Frontal radiograph shows a well-defined lobulated lucent lesion (thick arrow) in the distal end of the proximal phalanx of the second toe. Note that there is periarticular soft tissue with stippled calcifications (arrows) around the proximal end of the proximal phalanx of the second toe, consistent with tophaceous gout. b. Coronal T1-weighted (TR/TE = 300/25 ms) and c. STIR (TR/TE/IR = 4000/25/100 ms) images show a multilobulated tumor (curved arrow) with bone marrow (thick arrow) involvement in the distal end of the proximal phalanx of the second toe. The signal changes of the juxtaosteal soft tissue (arrow) around the proximal end of the proximal phalanx and the metatarso-phalangeal joint of the second toe can be due to surface relaxation mechanism of the tophi (see text). Focal bone marrow change due to gouty arthritis in the adjacent distal metatarsal bone are also seen. d. On proton-density image (TR/TE = 2500/22 ms), mass of isointensity (arrows) eccentrically located on the volar aspect of the flexor tendon (open arrow). Note some stippled low-signal-intensity foci within the tumor, indicative of hemosiderin deposition. e. There are some destroyed bony strips (arrow) and the marrow spaces are completely effaced by mildly hyalinized stroma of the giant cell tumor of tendon sheath. (H&E, ×200)
Three days later, the tumor was excised with bone grafted to the volar side of the proximal phalanx. Grossly, the proximal phalangeal bone of the second toe was penetrated by the soft-tissue mass on its volar side. Two fragments of yellowish white masses with intramedullary extension were excised. Bone graft was then applied. On histology, the tumor mass was composed of fibroblast-like cells, histiocyte-like cells, hemosiderin-containing foamy cells, and osteoclast-like multinucleated giant cells with destroyed bony strips in the marrow spaces (Fig. 1e). Neither obvious nuclear atypia nor prominent mitotic figures were identified.

The patient had favorable outcome after operation without functional impairment of the left second toe.

**DISCUSSION**

Previous reports [1, 2] have shown that 3 to 17% of giant cell tumors of the tendon sheath occur in ankles and feet. Among those affecting the feet, the great toe is most commonly involved. Symptoms of mass effect produced by the lesion account for 93% of patients [1]. Monaghan et al. [1] reported that a provisional clinical diagnosis of giant cell tumor of the tendon sheath being made in only 3 of 71 cases. With adjuvant images, Kotwal et al. [3] made an accurate preoperative diagnosis of the tumor in 42 of 48 patients.

Conventional radiographs commonly show a soft-tissue mass on the volar aspect of a digit or phalanx. Some of them may present with pressure erosions. Rarely, intramedullary radiolucency occurs, indicating medullary extension of giant cell tumor of the tendon sheath.

On MRI, giant cell tumor of the tendon sheath is typically isointense to those of skeletal muscle, or between those of muscle and fat on both conventional T1- and T2-weighted images [4]. Most lesions are inhomogeneous, represent deposition of hemosiderin within the tumor. Postcontrast spin-echo T1-weighted images may show marked enhancement throughout the lesion, with tiny low-signal-intensity foci of hemosiderin. Differential diagnoses of giant cell tumor of the tendon sheath include superficial and musculoaponeurotic fibromatosis, synovial amyloid, hemophilic arthropathy, mineralized lesions, and foreign bodies.

As in our case, giant cell tumor of the tendon sheath can produce predominantly high signal intensity on STIR images that can be difficult to distinguish from the images of other soft-tissue tumors. Presence of low-signal-intensity foci within the tumor, compatible with hemosiderin deposition, can be a useful diagnostic clue for giant cell tumor of the tendon sheath. The demonstration of the tumor in relation to the flexor tendon is also an important clue for the diagnosis.

In a study of 133 cases with giant cell tumor of the tendon sheath [5], 23% showed signs of pressure erosion of an adjacent bone or joint without cortical perforation on radiographs, and 11% of these cases had signs of cortical perforation. In another study of 91 cases of giant cell tumor of the tendon sheath in the digits [6], 12% had erosion of bones adjacent to the soft tissue mass, and none of these cases had cortical perforation by the tumors. Moore et al. [7] did not encounter any bony invasion in their series of 115 cases of giant cell tumor of the tendon sheath. A similar observation was noted by Phalen et al. [8]. Kitagawa et al. [4] reported that MRI failed to detect medullary extension of the tumor in none of the five lesions. In our case, both the radiographs and MRI images were able to demonstrate the medullary extension, which were documented on histological examination.

The incidence of malignant variant of giant cell tumor of the tendon sheath is extremely rare, carrying poor prognosis. Bertoni et al. [9] reported eight cases of malignant variant. Four of these cases had intraosseous involvement or bony erosion by the tumors. Of these eight patients, only four were alive at 3.5 to 5 years’ follow-up.

The local recurrence rate of giant cell tumor of the tendon sheath ranges from 4 to 45% [1, 10]. Degenerative joint disease is one of the risk factors for local recurrence [10]. Jones et al. [2] reported that 45% of patients with giant cell tumor of the tendon sheath had degenerative joint disease in adjacent joints. Bone marrow extension by the tumor is a known risk factor for local recurrence [10].

Marginal excision of the soft-tissue mass is a treatment of choice. In case of intramedullary involvement, the intramedullary tumor should be completely enucleated at surgery, followed by curettage and irrigation of the residual cavity. Bone graft can be applied when it is indicated. Adjuvant radiotherapy is recommended in patients at risk of a higher recurrence rate [3].

In our case, the unusual signal changes of the tophaceous gout in the second toe can be explained by a surface relaxation mechanism of calcium deposits [11]. Calcium can occasionally reduce T1 relaxation times by such mechanism. For concentrations of calcium particulate of up to 30% by weight, the signal intensity on standard T1-weighted images increases
but subsequently decreases [11].

This report has some limitations. First, the erosion of the cortex, in relation to the medullary extension of the giant cell tumor of the tendon sheath, was not clearly shown on MR images. This is probably due to suboptimal spatial resolution by using a 0.5-T magnet. Second, there was no gadolinium-enhanced MRI available in our case, which could be helpful in differentiating giant cell tumor of the tendon sheath from other solid soft-tissue tumors of the foot.

In summary, we present a case of a giant cell tumor of the tendon sheath with medullary extension of the second toe of left foot. The MRI findings are well correlated with the histological findings.

REFERENCES

肌腱鞘巨大細胞癌合併骨髄侵犯：病例報告

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腳趾肌腱鞘巨大細胞癌侵犯骨髄少有報告，我們報告一例28歲男性因左腳第二趾無痛性腫脹數年，一般X光攝影顯示第二趾骨髄出現葉狀透光區合併周圍軟組織密度增加，磁振造影發現腳掌側有一軟組織腫瘤（約12公分）侵犯至前趾端的骨髄內，並由病理組織確實診斷是肌腱鞘巨大細胞癌侵犯骨髄。

關鍵詞：巨大細胞癌；磁振造影；肌腱