Coexistence of giant cell tumor of tendon sheath and enchondroma in the middle phalanx of the little finger mimicking a malignant tumor: A case report

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Abstract. Giant cell tumor of the tendon sheath is a type of slow-growing benign soft tissue tumor that typically arises from the synovium of the tendon sheath. Enchondroma is a benign bone tumor comprising of mature hyaline cartilage that centrally develops within the tubular bone. While giant cell tumor of the tendon sheath or enchondroma are common benign soft tissue and bone tumors, respectively the simultaneous occurrence of these tumors in the same region of the hand is exceedingly rare, and it can mimic a malignant tumor, thereby making the diagnosis more challenging. Herein, we report an unusual imaging presentation of the coexistence of these tumors in the middle phalanx of the little finger, which to the best of our knowledge has not been previously reported, and this initially present as a single intrinsic osseous lesion mimicking malignancy. The coexistence of these tumor types must be considered in the differential diagnosis of an intramedullary lytic lesion with a poor margin associated with a soft tissue mass of the fingers, and a meticulous preoperative magnetic resonance imaging investigation was required.

Introduction

The giant cell tumor of the tendon sheath (GCTTS) is a type of slow-growing benign soft tissue tumor that typically arises from the synovium of the tendon sheath. The disease was characterized by the proliferation of synovial-like mononuclear cells mingled with dispersed multinucleate giant cells, siderophages, and inflammatory cells (1,2). In terms of its growth pattern according to the World Health Organization classification, GCCTS can be divided into a localized type that mainly occurs in the digits and a diffuse type associated with a more aggressive growth and high recurrence rate that predominantly occurs in large joints (1).

A solitary enchondroma is a benign bone tumor comprising mature hyaline cartilage that centrally develops within the tubular bone. It is typically asymptomatic and accidentally found because of a deformity, fracture, or a more frequent imaging [e.g., radiographs and magnetic resonance imaging (MRI)] (3).

GCTTS and enchondroma are categorized as one of the most common benign soft tissue and bone tumors of the hand, respectively, with the finger being the most common site among all locations (4-7). However, the coexistence of both these tumors in the finger, one in the phalangeal region, is exceedingly rare and may mimic a malignant tumor, which makes the diagnosis more challenging. Herein, we report an unusual case of the simultaneous existence of GCTTS and enchondroma, which was initially considered on the imaging results as a single primary or secondary malignant bone tumor.

Case report

A 79-year-old female, right hand dominant, presented to our hospital with a 3-month history of a painless palpable growing mass in the left little finger. Clinically, the mass was on the volar aspect of the middle phalanx with the discoloration of the overlying skin, measuring 12x9 mm, with a firm consistency and was not tender. She had a past medical history of breast cancer, which had been treated with a multidisciplinary approach (surgical resection, chemotherapy, and radiation therapy) approximately 8 years prior. The patient was regularly followed-up by clinical examination, additional imaging (mammography, ultrasound, computed tomography,

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Abbreviations: GCTTS, giant cell tumor of the tendon sheath; MRI, magnetic resonance imaging; CEA, carcinoembryonic antigen; CA, cancer antigen; Gd, gadolinium

Key words: coexistence, giant cell tumor of tendon sheath, enchondroma

and positron emission tomography), and laboratory and biomarker tests [e.g., carcinoembryonic antigen (CEA) and cancer antigen (CA) 15-3] and showed no signs of recurrence, and no metastases were detected. The patient denied any history of preceding trauma, discharging sinuses, or constitutional symptoms. General examination did not reveal any abnormality.

Radiographs of the middle phalanx in the little finger revealed an ill-defined radiolucent lesion containing a partially sclerotic rim and internal septations with a thinned distal half of the anterior cortex. A soft tissue mass was anteriorly and laterally identified. No calcification of the tumor matrix, joint involvement, or periosteal reaction was identified (Fig. 1). MRI was subsequently performed to further evaluate the mass on the little finger. The study demonstrated a 14x6x6-mm lesion within the fifth middle phalangeal bone. The lesion extraosseously extended into the adjacent soft tissue. A T1-weighted MRI revealed a lesion with a homogenous low-signal intensity on the entire lesions with an H-shaped lesion partially enveloping the tendon sheath (Fig. 2A and B). A T2-weighted image showed an area of homogenous high-signal intensity on the proximal half intraosseous region and low-signal intensity on the distal half intraosseous, as well as the extraosseous extension (Fig. 2C). The lesions exhibited contrast enhancement on the T1-weighted image after gadolinium (Gd) contrast administration (Fig. 2D).

In the absence of antecedent injury and infection, an ill-defined border and extraosseous extension was found on the imaging evaluation, suggestive of a malignant tumor. Preoperative differential diagnoses of the primary chondrosarcoma of the bone invading the surrounding soft tissues or secondary breast cancer in the bone was considered. Two distinct types of tumor lesions were found during the open biopsy: 1) A soft tissue extraosseous lesion with medullary invasion in the distal half of the phalangeal bone and 2) a cartilaginous intraosseous lesion at the proximal half of the bone without an extraosseous extension. Excisional biopsy of the extraosseous lesion and curettage with an artificial bone graft of the intraosseous lesion were subsequently performed after an intraoperative pathology consultation.

Two distinct characteristics of the gross specimen were examined. One gross specimen, the soft tissue extraosseous mass, was a circumscribed yellowish brown piece of tissue measuring 12x8x6 mm. Microscopically, this tissue lesion comprised an uneven and sparse distribution of osteoclast-like multinucleated giant and mononuclear cells. The mononuclear cells varied from oval histiocytoid or epithelioid cells to plump spindle cells. Both had an eosinophilic cytoplasm and central nuclei that closely resembled the nuclei within adjacent osteoclast-like cells. The histiocytoid cells frequently exhibited indented or folded nuclei. The spindle mononuclear cells were arranged in storiform patterns and were associated with collagen production. The cartilaginous intraosseous lesion was a white cartilaginous tumor measuring approximately 5 mm^3 in the aggregate. Microscopically, the tumor comprised mature cartilage lobules surrounded by mature bone with chondrocytes and displayed no obvious atypia (Fig. 3). In both specimens, no malignant features were observed (e.g., high number of mitoses and pleomorphic nuclei). The soft tissue extraosseous mass was considered a benign soft tissue tumor, GCTTS, with intramedullary invasion into the distal half of the phalangeal bone, whereas the proximal cartilaginous intraosseous lesion was considered a primary benign bone tumor (enchondroma). A final histopathological diagnosis of concurrent giant cell tumor of tendon sheath and an enchondroma of the middle phalanx was established. During the most recent follow-up visit at 21 months postoperatively, no evidence of recurrence was observed on MRI. We obtained written informed consent from the patient for publication of this case report.

Discussion

GCTTS is one of the most common tumors involving the hand and accounts for 74.2% of all benign soft tissue tumors in this region (4). GCTTS has also presented as localized nodular tenosynovitis, pigmented villonodular synovitis (8), and fibrous xanthoma (9). In a study of 207 GCTTS cases, the finger was the most common site (75.8%) (10), with a predominant involvement of the distal joint (5,9). For localized GCTTS, radiographic features typically display a soft tissue mass with or without bone changes, including bone pressure erosion, osseous invasion, cystic change, degenerative changes, periosteal reaction, and calcification (2,5,11-13). GCTTS with phalangeal bone involvement in the hand region has also been reported (12,14,15). In the form of an intraosseous lytic lesion on radiography, GCTTS may mimic a primary bone tumor, as observed in our present case (11).

Enchondroma is the most common benign bone tumor of the hand, accounting for 35-65% of cases (6). In the digit distribution meta-analysis of 327 cases conducted by Gaulke *et al*, the little finger was the most common site (7). This tumor can usually be diagnosed with radiographs, which show a well-defined central osteolytic lesion with or without calcification. In the present case, the patient presented with a mass in the little finger with a radiographic feature of central lucency with mild endosteal scalloping but lacking the typical calcification at the base of the middle phalanx, which intraoperatively corresponded to the enchondroma lesion site.

GCTTS with an intramedullary bone invasion, which may mimic primary bone tumor, is considered rare (11). The concurrent presence of the intraosseous extension of GCTTS and enchondroma, a primary bone tumor, in the same phalangeal bone is extremely rare. Our literature search revealed that GCTTS has not been previously described in association with enchondroma. The coincidence of these two entities can mimic malignancy due to its intramedullary accompanying lesion with soft tissue mass involvement, making the diagnosis more challenging. In the present case, the lesions were primarily centered in the phalangeal bone, which involved the entire intraosseous region, with mixed signal intensity and diffuse contrast enhancement associated with an extensive soft tissue mass. Considering the patient's age, history of previous cancer, and these imaging findings, a malignancy including primary bone tumor (chondrosarcoma) and bone metastases from breast cancer was initially considered.

Chondrosarcoma is the most important condition to be differentiated. This tumor, located at the phalangeal bone, is locally aggressive and exhibits minimal metastatic potential (16). Moreover, the distribution of the tumor site

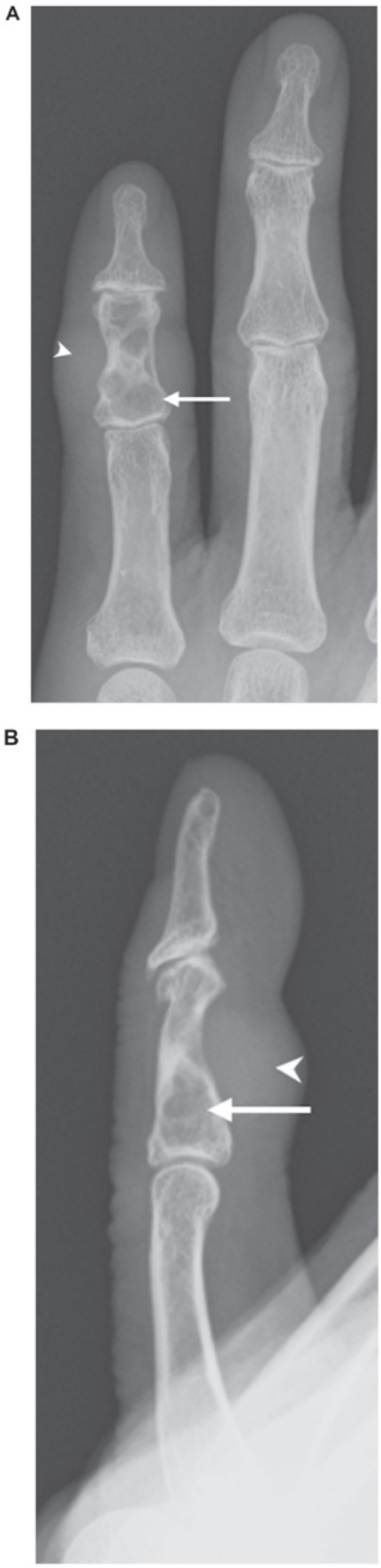

Figure 1. Frontal (A) and oblique (B) radiographs reveal a well-defined margin lucent lesion (arrow) with a partially sclerotic rim and thinned distal half of anterior cortex. A soft tissue mass was identified (arrowhead). No calcification of the tumor matrix, joint involvement, or periosteal reaction were identified.

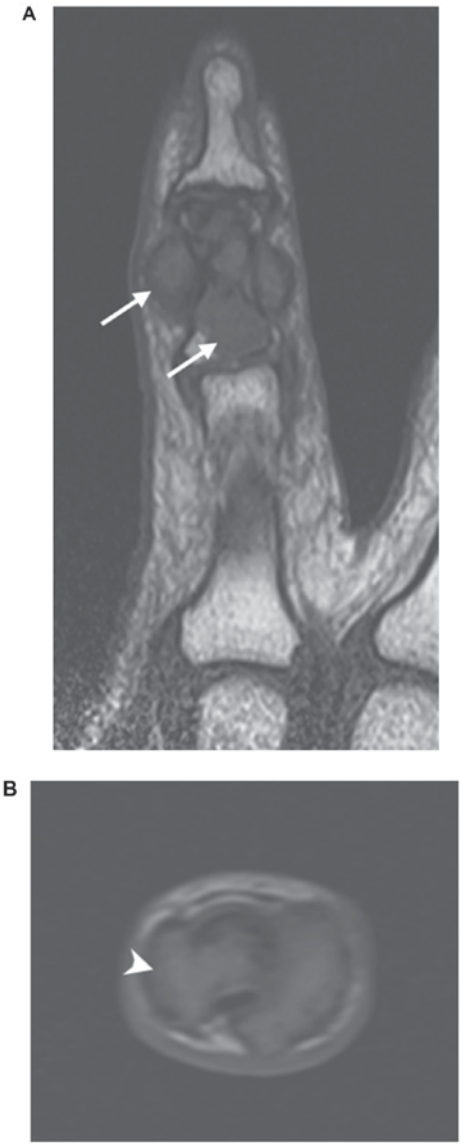

Figure 2. MRI T1 coronal image (A) demonstrating that signal intensities of the lesion were homogenous hypointense (arrows). MRI T1 axial image (B) demonstrating an H-shaped lesion partially covering the tendon sheath (arrowhead). MRI, magnetic resonance imaging

in the hand is similar between chondrosarcoma and enchondroma (17). The type of aggressive chondrosarcoma (high grade) generally displays an intraosseous ill-defined lytic area with a mouth-eaten or permeative pattern, periosteal reaction, and large soft tissue invasion through cortical destruction (18). However, in this case, an ill-defined intraosseous lesion was found on imaging findings due to synchronous double tumors, which was not characteristic of a high-grade pattern. Typical features of high-grade chondrosarcoma, including a lobulated high T2 signal and a ring-and-arc enhancement pattern, were not identified. The evidence that there were two distinct lesions, with the isolated cartilaginous intraosseous lesion being unrelated to the surrounding soft tissue mass (GCTTS), and no breach of the cortex on this lesion site was intraoperatively identified, which suggested that no soft tissue invasion arose from the intraosseous cartilaginous lesion. Additionally, histological examination of intraosseous and extraosseous lesions revealed no cytological atypia featuring

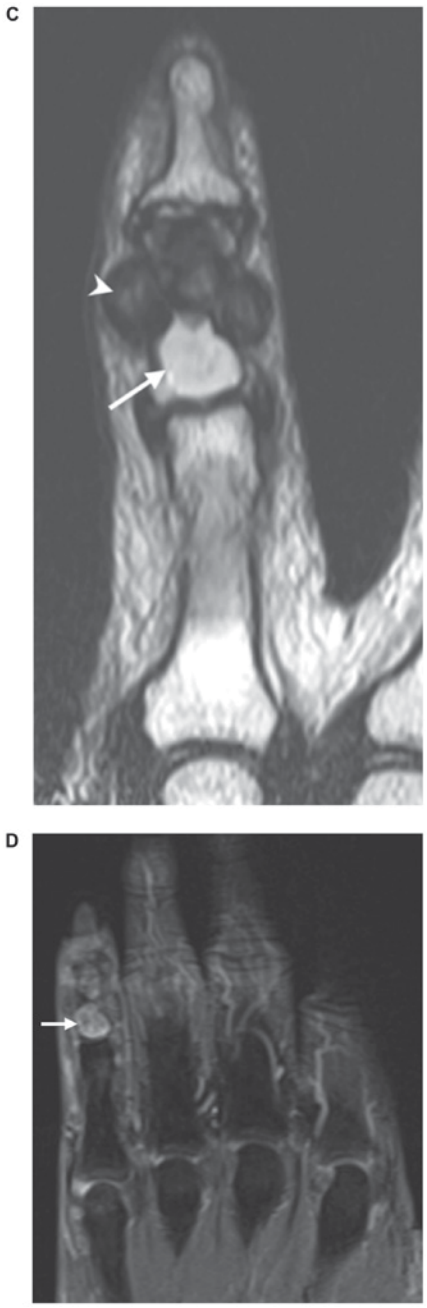

Figure 2. Continued. MRI T2 coronal image (C) demonstrating that signal intensities of the lesion were hyperintense on the proximal half intraosseous region (arrow) and hypointense on the distal half intraosseous as well as the extraosseous extension (arrowhead). MRI post-Gd T1 coronal image (D) showing contrast enhancement of the lesions (arrow). MRI, magnetic resonance imaging; Gd, gadolinium.

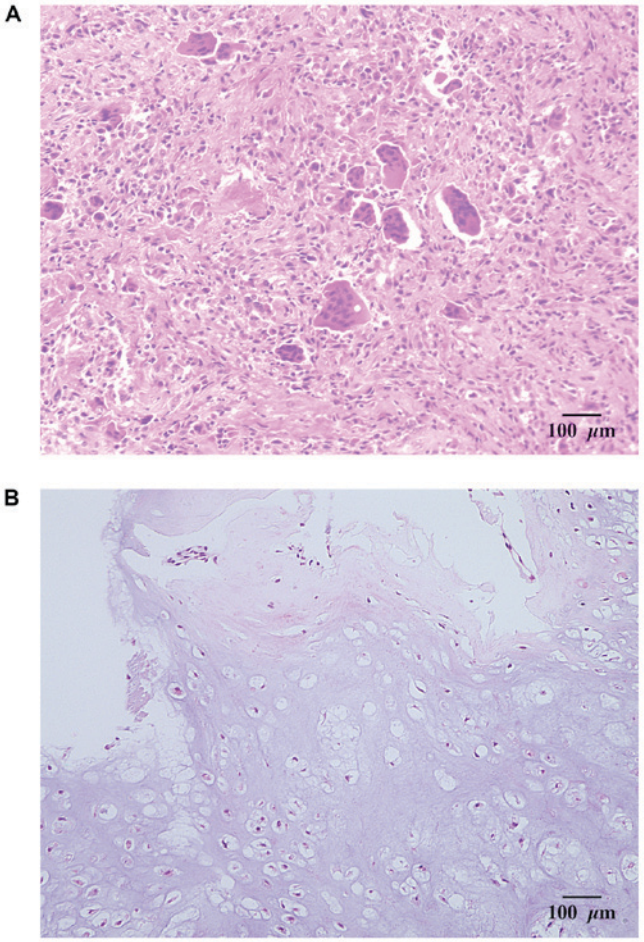

Figure 3. Microscopic findings of two distinct forms of benign tumor. GCTTS (A) shows scattered multinucleated giant and mononuclear cells in hyalinized collagen bundles. H&E, original magnification 200x. The enchondroma (B) showing a lobulated hyaline cartilage tumor surrounded by mature bone and no obvious atypical chondrocytes. H&E, original magnification x200. No features of malignancy (e.g., a high number of mitosis or pleomorphic nuclei) were found in either specimen. GCTTS, giant cell tumor of the tendon sheath; H&E, Hematoxylin and eosin.

high-grade chondrosarcoma. Since the type of low-grade chondrosarcoma (grade I) exhibits only minimal histological atypia, it is difficult to histologically distinguish it from enchondroma (18,19). Radiographically, cortical thickening or disruption may be evident if soft tissue is involved (20). In our case, because no cortical thickening or destruction was observed on imaging and intraoperative findings in the cartilaginous lesion site, a diagnosis of low-grade chondrosarcoma could not be made.

Metastases to the hand from a primary tumor elsewhere are very rare, with an incidence of 0.007-0.3% (21). In a literature review of 163 cases conducted by Kerin *et al*, lung carcinoma was the most common primary malignancy to metastasize (42%), followed by the kidney (13%) and breast (11%) (22). According to their radiological features, bone metastases are classified as either osteoblastic or osteolytic (bone-destruction). The latter can be radiographically observed only if at least 50% of the bone material has been destroyed (23). In the present case, from a clinical point of view, based on the patient's advanced age and particularly, the history of preceding breast cancer, the development of a bone metastasis in the ill-defined osteolytic finger lesion may be considered, despite its rarity. Bone metastases can appear in any pattern on radiographic findings using X-rays (24). Osseous metastases typically display T1 low-signal intensity, T2 high-signal intensity, and gadolinium enhancement, as were found in our case (25). However, regular follow-up care that showed no evidence of recurrence and distant metastases warranted a confirmatory biopsy, which was consistent with dual benign primary tumors.

In conclusion, the simultaneous presentation of GCTTS with intramedullary invasion and an enchondroma on the

phalangeal bone has not been previously reported and can initially present as a single intrinsic osseous lesion mimicking malignancy on imaging findings. As a result, their coexistence must be considered in the differential diagnosis of a poorly margined intramedullary lytic lesion associated with a soft tissue mass in the fingers, and a meticulous preoperative MRI investigation is required.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Author's contributions

MPJ, TK, TF, TS and NA were responsible for the study concepts. Data acquisition, analysis, and interpretation was undertaken by MPJ, TK, TF, TS and NA. Drafting of the manuscript was the responsibility of MPJ, TK, TF, TS and NA. MPJ, TK, TF, TS and NA gave final approval of the manuscript to be published, and MPJ, TK, TF, TS and NA are in agreement to be accountable for all aspects of the work.

Ethics approval and consent to participate

Not applicable.

Patient consent for publication

Written informed consent was obtained from the patient for publication of this case report.

Competing interests

The authors declare they have no competing interests.

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