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# Atypical cartilaginous tumor imaging findings in the distal phalanx of the left thumb: case report and literature review

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**Background:** Atypical cartilaginous tumors (ACTs) usually occur in long bones rather than in the hands or feet. To date, detailed imaging features of ACTs in the hands or feet were reported in only a few case reports.

**Case presentation:** We report a case of an Asian woman in her early 80s who presented to our hospital with a painless mass in the distal phalanx of the left thumb. Radiography and computed tomography (CT) showed osteolytic destruction and cortex thickening in the distal phalanx with extension into soft tissue. Magnetic resonance imaging (MRI) demonstrated a local soft tissue signal mass with isosignal intensity in T1 and heterogeneous high-signal intensity in T2. Color Doppler ultrasound suggested that the tumor was hypovascularized. The patient underwent amputation, and histological analysis revealed an ACT. The patient's symptoms improved postoperatively, with no recurrence as of the 3-year follow-up.

**Conclusion:** Meanwhile, this study also reviewed the comparable diagnostic methods of ACT and chondrosarcoma. The analysis of previous similar cases showed that preoperative imaging diagnosis of ACT can be challenging and multimodal imaging appears to be beneficial in diagnosing ACTs and malignant chondrosarcoma grade II/III in the hands and feet.

## KEYWORDS

atypical cartilaginous tumor, chondrosarcoma, hands and feet, imaging, diagnosis

## Introduction

In the 5th Edition of the World Health Organization (WHO) tumor classification of bone and soft tissue tumors, published in 2020, atypical cartilaginous tumor/chondrosarcoma grade I (intermediate-locally aggressive) (ACT/CS1) in the 2013 version was divided into two diseases: ACT and CS1. The former was retained in the classification of intermediate-locally aggressive, and the latter was upgraded to malignancy.

The new version of the guideline differentiates between ACT and CS1 based on the location of the tumor: tumors located within the appendicular bone both in long and short tubular bones are called ACT, while those occurring in the axial skeleton (pelvis, scapula, and skull base flat bones) are called CS1 to reflect the poorer clinical outcome of these tumors at these sites. The most common sites affected by ACT are the femur, humerus, and tibia. ACTs of the hands and feet are not common. Because of the rarity of ACT in the hands and feet, the imaging literature on ACT is scarce and mainly comprises older or small case series, as well as case reports describing conventional radiographic features (1, 2). Preoperative imaging diagnosis of ACT can be challenging. We report a case of ACT in the distal phalanx of the left hand thumb and made a detailed report on its imaging performance. The imaging features of CS and ACT in hands and feet were summarized and reviewed.

## Case report

The authors have read the case report (CARE) Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016). An Asian woman in her early 80s presented with a palpable and painless mass in the distal phalanx of the left thumb. The patient noticed that mass 2 years ago, and the mass swelled progressively in the 2 years and its growth accelerated in the past 6 months, which prompted the woman to go to the hospital for medical assessment. Upon physical examination, the patient had a swollen left hand thumb, giving it a drumstick appearance (Figure 1). The patient claimed a history of type 2 diabetes and hypothyroidism with no known trauma or any masses elsewhere. Our patient did not have Ollier's disease, Maffucci syndrome, or hereditary exostosis and had no evidence of prior enchondroma. The results of routine blood tests and biochemical tests were normal. To further clarify the diagnosis, the patient received medical imaging examinations. The radiograph showed a mass in the distal phalanx of the left thumb with osteolytic bone destruction (Figures 2A, B). In computed tomography (CT) images, osteolytic destruction of the distal phalanx of the thumb with a local soft tissue density mass was observed (Figures 2C, D). Magnetic resonance imaging (MRI) also showed cortical destruction of the distal phalanx of the thumb and a local soft tissue signal mass of approximately 20 mm × 19 mm × 26 mm with isosignal intensity in the T1-weighted image (T1WI) and heterogeneously high signal intensity in the T2-weighted image (T2WI) (Figures 2E, F). The mass invaded the interphalangeal joint. In addition, the patient underwent ultrasound examination. Ultrasound imaging demonstrated a hypoechoic mass (25 mm × 18 mm × 22 mm) with patchy calcification in the distal phalanx of the left thumb (Figures 2G–I). The mass had a clear margin and irregular shape. The bone cortex around the mass is discontinuous and not smooth. Color Doppler showed a dot blood flow signal within the mass. Subsequent chest radiography and bone scans did not reveal any metastatic lesions.

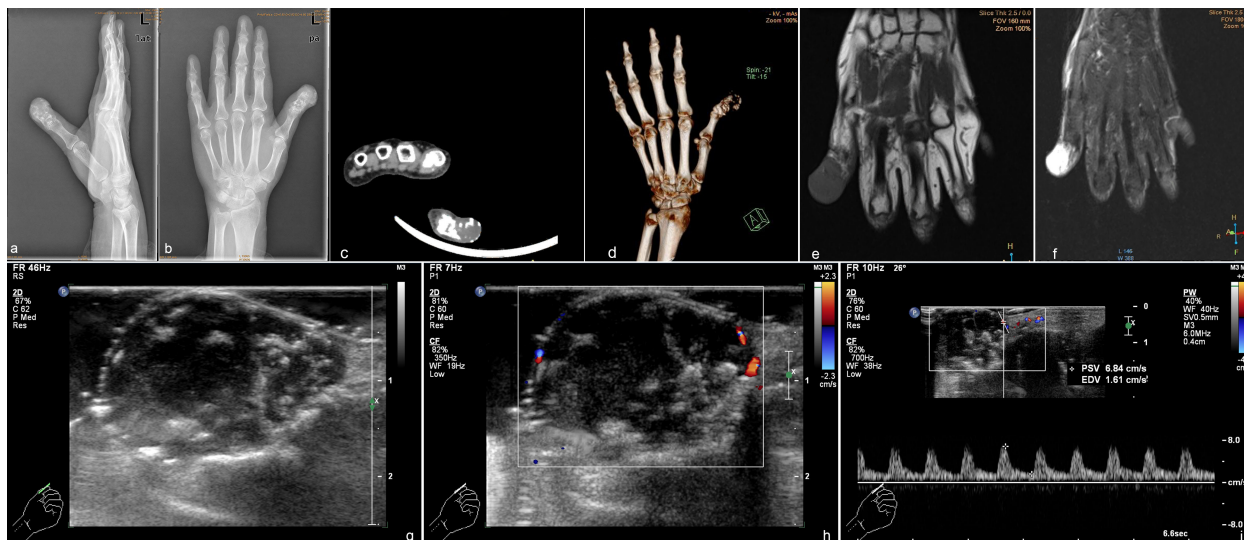
With the above imaging findings, the patient received amputation of the left thumb distal phalanx to achieve radical excision of the mass. The lesion tissues were stained with

hematoxylin-eosin (HE). Microscopically, blue-stained cartilaginous matrix was found, and chondrocytes showed obvious heteromorphism. The caryocinesia was observed, with invasive growth accompanied by a few bone trabeculae. Pathologic findings are consistent with a chondrogenic tumor, showing intermediate-locally aggressive behavior. Combined with tumor location (appendicular skeleton, not the axial skeleton), imaging findings (a mass with osteolytic bone destruction), and an aggressive clinical behavior (rapid growth in the last 6 months), the tumor was diagnosed as an ACT (Figures 3A–C). The postoperative x-ray examination showed a good surgical outcome (Figures 3D, E). After the surgery, the patient experienced an improvement in her symptoms and reported a better quality of life. During the 3-year follow-up, there was no evidence of recurrence detected by MRI or CT scans. Informed consent was obtained from the subject described in this report.

## Discussion

Although the 2013 version proposed the concept of ACT, it did not provide specific diagnostic criteria, and it was difficult to apply to clinicopathological diagnosis. The 2020 version clarified the diagnostic criteria for ACT: tumors occurring in the appendicular bones (long and short tubular bones) are called ACT, while those occurring in the axial bones (flat bones including the pelvis, scapula, and skull base) are called CS1. ACT and CS1 have the same histological characteristics, but the prognosis of appendicular





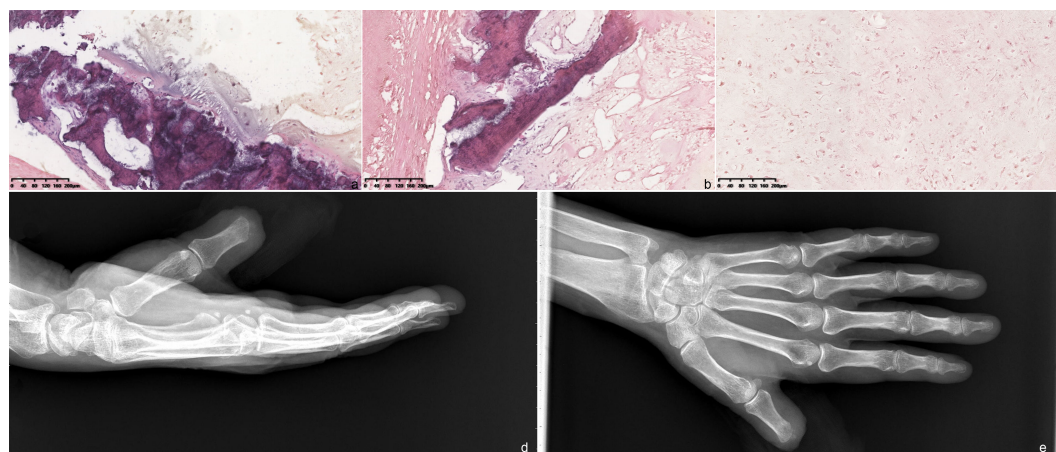
**FIGURE 2** Preoperative appearance of the patient’s hand shows a destructive cortex and osteolytic change with soft tissue masses. **(A)** Lateral radiograph; **(B)** radiograph. Computed tomography imaging **(C)** and 3D view **(D)** of atypical cartilaginous tumor of the hand shows osteolytic destruction of the distal phalanx of the thumb. Magnetic resonance imaging. **(E)** T1-weighted imaging shows a low-signal shadow of the mass; **(F)** T2-weighted imaging shows a heterogeneously high signal intensity. Ultrasound demonstrated a hypoechoic mass with calcification **(G)** and a dotted blood flow signal **(H)**. **(I)** Spectrum Doppler shows the arterial spectrum.

bone tumors is significantly better in axial bone (3). The new version of the guidelines divides ACT/CS1 into ACT and CS1 based on the anatomical location of the tumor, with the former retained as intermediate and the latter upgraded to malignant disease. ACT involving bones of the hands and feet is uncommon. Early diagnosis is helpful for the treatment and prognosis of the disease. Radiography, CT, and MRI are helpful for tumor biopsy and treatment.

In order to differentiate ACT and CS imaging characteristics of the hands or feet, we searched the following three databases: PubMed, Embase, and Cochrane Library, according to the

Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines (4). We also manually searched additional relevant studies using the references of the systematic reviews that were published previously. All of the searches were performed from inception to 22 August 2022, and imposed restrictions on the English language. The inclusion criteria specified studies and case reports describing patients with primary CS or ACT confirmed by pathologically involving the hand or the feet who received image examination.

The following search strategy was used for PubMed and was modified to suit Embase and Cochrane Library databases.



**FIGURE 3** Hematoxylin and eosin-stained section of the tumor (x10 magnification). **(A)** Microscopically, there are many slightly heterotopic chondrocytes with obvious heteromorphism and a Caryocinesia phase. **(B, C)** Most of the tumor cells show invasive growth, accompanied by a few bone trabeculae. Postoperative image of the patient. **(D)** Lateral radiograph; **(E)** radiograph.

#1 (chondrosarcoma [Title/Abstract]) OR (atypical cartilaginous tumor[Title/Abstract])

#2 (small bone [Title/Abstract] OR hand [Title/Abstract] OR foot [Title/Abstract])

#1 AND #2

Next, we screened the titles and abstracts of these articles, excluding articles in which the lesion did not involve the hands or feet, those not describing the imaging examination features, and those in which the tumors were secondary. Periosteal chondrosarcoma, clear cell chondrosarcoma, mesenchymal chondrosarcoma, and dedifferentiated chondrosarcoma were excluded because they have different biological behaviors (5). [Supplementary Figure 1](#) illustrates the study selection process.

As a result, 32 articles (6–37) discussing 92 cases were selected. The characteristics of the selected cases, including the present case, are listed in [Supplementary Table 1](#). All included cases were primary chondrosarcoma confirmed by pathology. There were 49 male and 43 female patients. All patients were adults, except for a 12-year-old male patient reported by Gupta et al. (28). Among all patients, 90 patients underwent radiographic examination, 8 patients underwent CT, 17 patients underwent MRI, 6 patients underwent CE-MRI, and 16 patients underwent bone scans. In [Supplementary Table 1](#), the radiography and CT imaging manifestations of chondrosarcoma and ACT in hands and feet were mainly seen as cortical destruction, soft tissue mass, and osteolytic change, which characteristically contained calcification. Additional features that may be present included endosteal scalloping and periosteal reaction. On MRI, a total of 9 cases provided detailed information on T1WI and T2WI. These lesions appeared as hyposignal or isosignal on T1WI images. On T2WI images, the above lesions were hypersignal compared with skeletal muscle. CE-MRI showed an enhanced area among all patients who underwent CE-MRI examination. In addition, Tos et al. (23) reported a case showing a rounded high-intensity image on coronal STIR and a low-intensity heterogeneous lesion on sagittal SET1. The bone scanning results of 16 patients all showed abnormal radioactive accumulation, one of which indicated that the tumor was hypervascular. Only the lesion in this case report was examined by ultrasound. Ultrasound imaging demonstrated cortical destruction and a hypochoic mass with patchy calcification. Color Doppler showed that the tumor was hypovascularized. Resection, amputation, and ray resection were commonly used in treatment.

Considering that in the previous classification versions, ACT and CS1 were classified into the same category (ACT/CS1), but in the new classification version published in 2020, the intermediate-locally aggressive chondrosarcoma grade I (CS1) occurring in the hand or foot is uniformly classified into ACT, in the following analysis of imaging findings, we excluded the data without clear pathological classification in [Supplementary Table 1](#). The data in [Supplementary Table 1](#) were reclassified as two categories: intermediate-locally aggressive ACT and high-malignant chondrosarcoma grade II/III (CS2/3), for comparison of imaging findings. After the reclassification, 10 cases of ACT and 35 cases of CS2/3 were retained for further analysis. Of the 10 cases of ACT, it is more

common in male than in female patients (female:male = 3:7). The age range was 29–87 years, with an average age of 52 years. In 35 cases of CS2/3, it is more common in female than in male patients (female:male = 20:15). The age range was 30–85 years, with a mean age of 63 years old. In terms of symptoms, the two categories may show pain, swelling, or both. In ACT, 20% of patients presented with pain (2/10), 20% also presented with swelling (2/10), and 30% had both (3/10). In CS2/3, 43% patients presented with pain (15/35), 17% presented with swelling (6/35), and 40% had both (14/35). Imaging findings showed that in ACT, 80% (8/10) of patients showed cortical destruction, 40% (4/10) displayed calcification, 70% (7/10) showed soft tissue mass, 40% (4/10) showed osteolytic destruction, and only 1 case (10%) showed periosteal reaction. In CS2/3, 83% (29/35) of patients showed cortical destruction, 80% (28/35) showed calcification, 74% (26/35) showed soft tissue mass, osteolytic destruction was observed in 11% (4/35), and periosteal reaction was observed in 2 cases (6%). In summary, both ACT and CS2/3 are common in middle-aged and elderly patients. ACT is more common in men, while CS2/3 is more common in women. In imaging findings, both of the two categories showed cortical destruction and soft tissue mass. Calcification was more likely to occur in malignant CS2/3. Therefore, there are certain difficulties in the imaging of the two categories, and pathological examination is still needed to distinguish them.

In clinical practice, radiography and CT remain the mainstay for the initial detection of chondrosarcoma or ACT of small bones and are helpful for characterization of the lesion (38). The main application of MRI is the preoperative assessment for staging the extent of disease (1, 2). However, there is a paucity of data on the use of PET/MRI in ACT or CS of small bones. However, some scholars (39) have noted that PET/MRI can provide additional functional information to supplement the morphologic mapping and histopathology of these tumors. It is expected that future research will highlight a potential role for PET/MRI in the management of CS or ACT of small bones. Ultrasound examination is noninvasive, painless, and economical. It can not only show small lesions of the bone cortex and observe the structural relationship between the tumor and surrounding blood vessels but also show different degrees of bone destruction, periosteal reaction, and soft tissue invasion (40). Color Doppler flow imaging can also provide hemodynamic information. These results also suggest that multimodal imaging is helpful to improve the diagnostic efficiency of CS or ACT.

This study also has some limitations. First, since only hands and feet with imaging findings were collected, the epidemiological data collected in this study are not comprehensive. Second, because the collected literature in [Supplementary Table 1](#) were published before the release of the new version in 2020, we failed to classify ACT and CS of the imaging information in some articles. In addition, for the analysis of imaging findings, fewer cases were included in this study, and no statistical analysis was made on the differences. The differential diagnosis of imaging features between ACT and malignant CS2/3 needs further study.

In conclusion, although ACT is very rare in hands and feet, the possibility of ACT should also be considered for soft tissue tumor occurring in middle-aged and elderly men whose clinical

manifestations are mainly pain and swelling, and the imaging findings are cortical destruction, soft tissue mass, and osteolytic change. Multimodal imaging may be helpful to improve the diagnostic efficiency of ACT.

## Data availability statement

The datasets presented in this study can be found in online repositories. The names of the repository/repository and accession number(s) can be found in the article/[Supplementary Material](#).

## Ethics statement

The studies involving humans were approved by Ethics Review Committee of Sichuan University. The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation in this study was provided by the participants' legal guardians/next of kin. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

## Author contributions

HW: Data curation, Methodology, Conceptualization, Formal analysis, Writing – original draft, Writing – review & editing, XT: Supervision, Writing – original draft. YW: Supervision, Project administration, Validation, Writing – original draft. XP: Validation, Investigation, Writing – original draft. YY: Conceptualization, Investigation, Supervision, Validation, Writing – review & editing.

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## Conflict of interest

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## Supplementary material

The Supplementary Material for this article can be found online at: <https://www.frontiersin.org/articles/10.3389/fonc.2025.1407012/full#supplementary-material>

### SUPPLEMENTARY FIGURE 1

A flowchart showing the literature search and analysis process.

### SUPPLEMENTARY TABLE 1

List of primary atypical cartilaginous tumor or chondrosarcoma cases receiving imaging examination. LF, left foot; RH, right hand; PP, proximal phalanx; PIP, proximal interphalangeal point; LH, left hand; MP, metacarpophalangeal; Gd, gadolinium-enhanced; MC, metacarpal.

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