

## CASE REPORT

# Conventional chondrosarcoma in the right hand with the invasion of the pisiform and the hamate bones – case report

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## Abstract

Chondrosarcoma is a malignant hyaline cartilage forming tumor. It is a rare pathology, with an estimated incidence of one in 200 000 individuals per year. It appears in two forms: primary, representing 90% of all cases and secondary emergence that develops on preexistent lesions. Primary chondrosarcomas are uncommon in the hand, with a frequency of only 1.5–3.2% of all chondrosarcomas. In conventional chondrosarcoma (cCS), the histological malignancy grading represents the main prognostic factor for surgical planning and prognosis. We present the case of a 60-year-old male, examined in the First Department of Orthopedics and Traumatology, "Dr. Pius Brânzeu" Clinical Hospital, Timișoara, Romania, with non-specific symptoms in the right hand. After clinical examination and imagistics, surgery and histopathological examination of the tumor were performed. This showed a conventional well differentiated – G1 chondrosarcoma, as suggested also by imagistic and clinical context.

**Keywords:** hand, chondrosarcoma, management.

## Introduction

Chondrosarcomas represent a heterogeneous group of malignant bone tumors that share in common the production of chondroid (cartilaginous) matrix. The estimated occurrence is one in 200 000 per year [1, 2]. Surgery remains the mainstay of treatment, as neither chemotherapy nor radiation therapy is effective in chondrosarcoma [2]. The prognostic factors of chondrosarcoma remain uncertain, as only a few large studies with long-term follow-up have been reported [3, 4]. Although it is more frequently found in phalanges and metacarpals, our case report intends to describe and to assess one of the more unusual localizations for this tumor.

## Case presentation

A 60-year-old male, former worker in a shoe factory for about 30 years, was admitted in February 2014 in the 1<sup>st</sup> Department of Orthopedics and Traumatology, "Dr. Pius Brânzeu" Clinical Hospital, Timișoara, Romania, with the following symptoms at the right hand: swelling of the 4<sup>th</sup> and 5<sup>th</sup> fingers right hand, paresthesia in the ulnar nerve's innervation territory and partial functional impairment. These complaints started two weeks ago without recalling any traumatic history or significant personal or family history and aggravated in the past three days.

The magnetic resonance imaging (MRI) investigation highlighted three structures.

The first structure, of 16/14/10 mm (Figures 1–4), with a predominantly fluid signal, irregular contour, well defined and apparently septate, was located on the dorsal face of the distal radial epiphysis. There was no evidence of bone signal changes adjacent to this septate structure.

Second structure, with the following dimensions of 31/36/26 mm, in hypersignal T2 and short *tau* inversion recovery (STIR), and iso-/hypo-signal T1 (compared to the muscle) – soft tissue, slightly inhomogeneous, septate, with a well defined contour, was found internally to the carp and the radio-carpal joint, between the base of 5<sup>th</sup> metacarpus and the distal ulnar epiphysis (Figure 5).

The third structure, with fluid signal in the triquetrum bone, occupies 2/3 of its surface and is well defined to the bone substance, with uncertain view of the internal cortical.

"Mirrored"-type erosions, bone edema in the articular surfaces of the capitate and hamate bone, as well as an intra-articular fluid line in the dorsal aspect of the radio-ulnar joint, can also be observed.

The X-ray series emphasizes an extensive tumoral mass, located at metacarpal level, on the external border, developed mainly in the soft parts, with the invasion of the pisiform and hamate bones, also encapsulating the external collateral ligament.

The lesion presents T2 hypersignal: the formation measures in its maximum diameter 43/30/38 mm. The lesion presents T1 hyposignal and picks up contrast typical aspect for a cartilaginous tumor, osteochondroma, osteosarcoma or chondrosarcoma. A similar image was located radio-scapoid, with maximum diameters of 155 mm.

T1 sequence after contrast, in coronal plane presents an intense contrast intake in the tumoral formation, also a global intake at the level of the whole tumoral mass can be observed (Figure 6, a and b).

T2 sequence in sagittal plane revealed the dislocating, deforming aspect of the tumoral mass, with inhomogeneous structure, solid cartilaginous and fluid components, a structure profound inhomogeneous (Figure 7, a and b).

The surgical intervention was planned. The surgery is described as follows: through a 7 cm centered approach on the tumoral formation followed by its uplifting, hemostasis and sampling bone and soft tissues for biopsy. The resection and curettage of the recognizable tumoral remains were practiced.

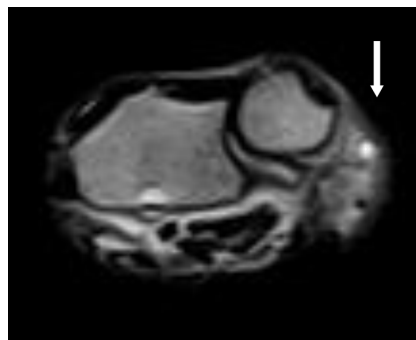
The pathology findings of the biopsy samples are relevant for a conventional well differentiated – G1 chondrosarcoma. A fibroadipose tissues, includes nodular tumoral formations, of variable dimensions, individualized or grouped, some lobulated, others defined by a conjunctive pseudocapsule and constituted mainly from the hyaline cartilaginous matrix, with limited areas of myxoid type and variable cellularity (Figure 8).

We also could observe mature lamellar bone, mainly cortical type, fragmented, accompanied by lobules, tumoral cartilage with permeation of the Haversian canals and spongy type lamellar bone, through the cartilaginous matrix (with architectural features similar to those of the tumoral soft tissue) (Figure 9). The periarticular tissue contains synovial tissue with metaplastic cartilaginous areas, small metaplasial bone islands and rare small tumoral cartilaginous nodules.

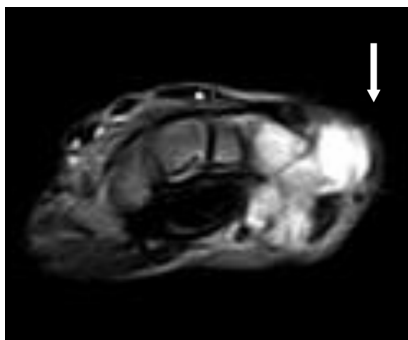
Some fragments are relatively limited by lamellar bone tissue, of cortical type, with focal aspects of Haversian canals permeation and few small cartilaginous nodules in the periosteal soft tissues; spongy bone had unequal lamellas surrounded by tumoral cartilaginous tissue (disconnected bone lamellas, with intertrabecular stroma almost completely replaced by tumoral tissue) (Figure 10). By architectural point of view, the morphological aspects are similar to those of the tumoral soft tissue, but with less expressed cellularity and a more diffuse growth pattern, without obvious nuclear pleomorphism (slight focal atypia), with frequent binucleated cells, small myxoid areas and often eosinophilic nuclei, suggestive for chondroid necrosis; on one of the sections, aspects of variable intrication of the adipose intertrabecular stroma with cartilaginous tumoral tissue and its continuation with cartilaginous nodules in the periarticular soft tissues, are identified, without cortical bone or remaining articular cartilage in the continuity areas (Figures 11 and 12).

Distinct hypercellular areas (mostly common at the periphery of the lobes, grouped focal, vaguely nodular-synovial chondromatosis-like pattern; they are intricated with normally, or less common hypocellular, with slight focal nuclear atypia frequent binucleated cells; blood vessels, nerves and focal striated muscle bundles were present in the biopsied material (Figure 13).

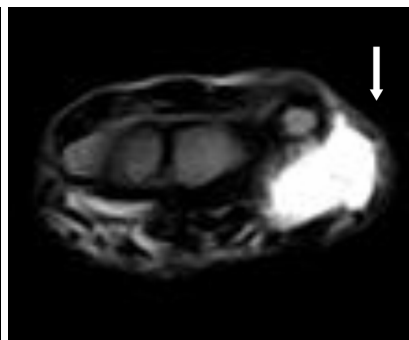
After six months, another MRI examination was performed. One year after the surgery, the patient was doing well with no evidence of tumor recurrence. The patient's follow-up showed no evidence of tumor recurrence.



**Figure 1 – T2 sequence in axial plane: tumoral mass with a solid component and the invasion of bone structures.**



**Figure 2 – T2 sequence in axial plane: fluid component of the multilobular radiocarpal mass.**



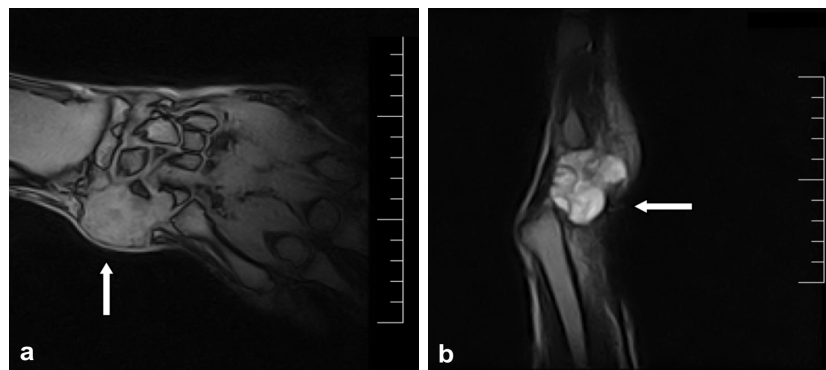
**Figure 3 – T2 sequence in axial plane: the fluid component of the tumoral mass; irregular multi-lobular shape, with radiocarpal deformity.**



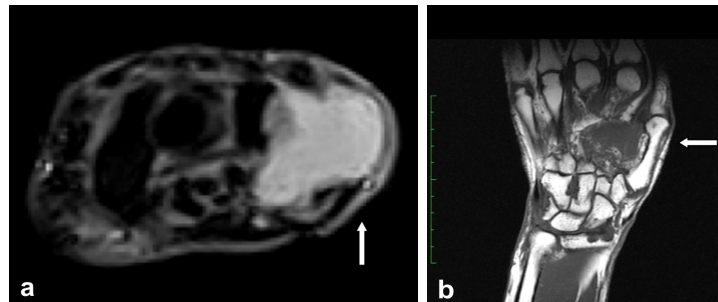
**Figure 4 – T2 sequence in axial plane: tumoral mass with inhomogeneous structure of mixed type, predominantly fluid, associating islands in isosignal with the cartilage structure.**



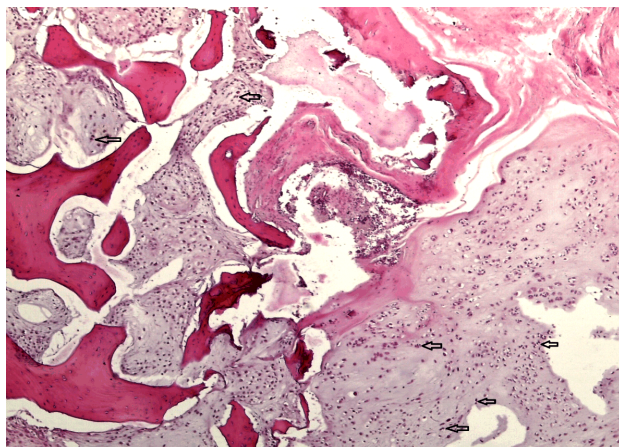
**Figure 5 – T2 sequence in axial plane: invading radiocarpal tumoral mass, with the disturbance of the adjacent bone architecture and deforming the aspect of the joint.**



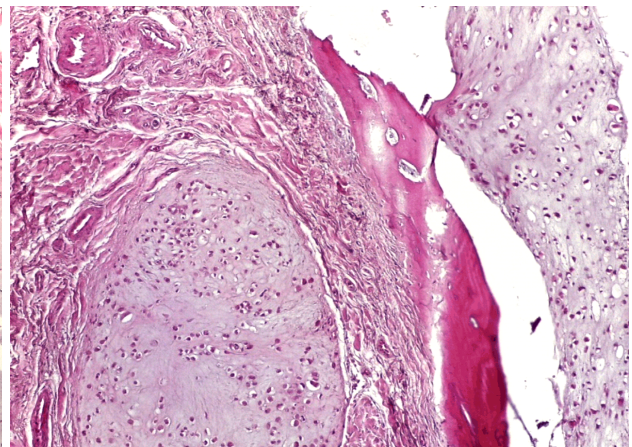
**Figure 6 – (a and b) T1 sequence in coronal plane: intense contrast intake in the tumoral formation.**



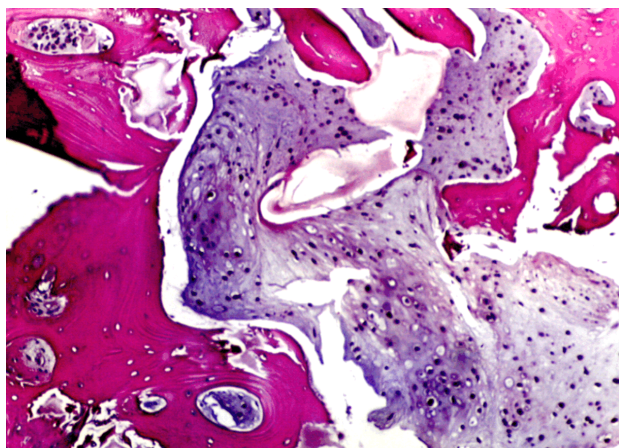
**Figure 7 – (a and b) T2 sequence in sagittal plane: deforming aspect of tumoral mass.**



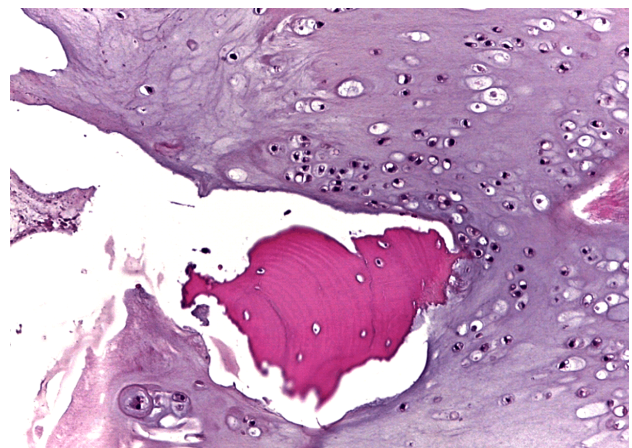
**Figure 8 – Cortical bone destruction and soft tissue tumor extension [Hematoxylin–Eosin (HE) staining, ×100].**



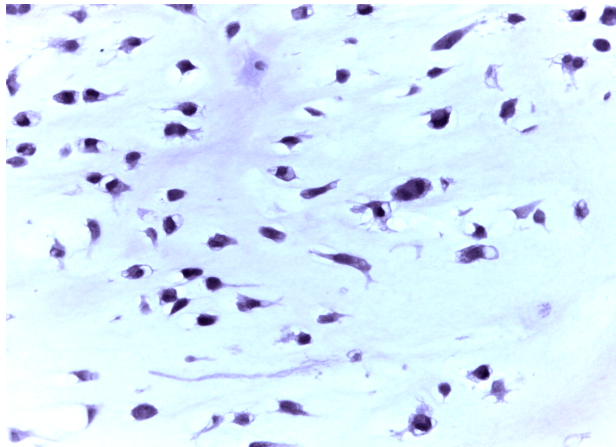
**Figure 9 – Island tumor in the proximity of the bone soft tissue and permeation of small Haversian canals (HE staining, ×100).**



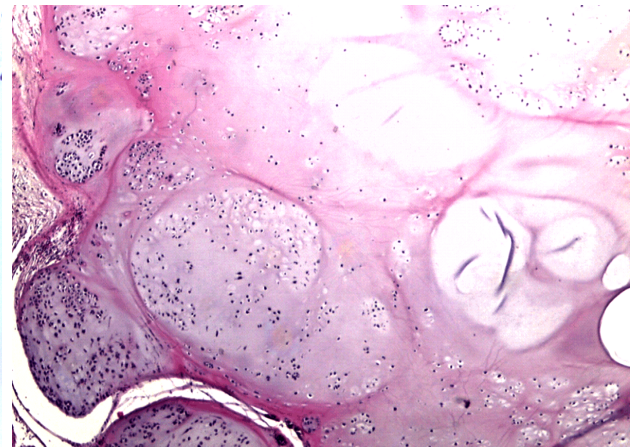
**Figure 10 – Permeation by tumor Haversian canals (HE staining, ×100).**



**Figure 11 – Image detail: discrete pleomorphism, few binucleated chondrocytes and inclusion of the remaining lamella by the bone tumor mass (HE staining, ×100).**



**Figure 12** – Moderate cellularity, slightly atypical cytonuclear aspect, several binuclear cells [May–Grünwald–Giemsa (MGG) staining,  $\times 400$ ].



**Figure 13** – Synovial-like pattern chondromatosis (HE staining,  $\times 100$ ).

## Discussion

The skeletal chondrosarcoma family is a heterogeneous group of malignant mesenchymal bone tumors represented by the production of a chondroid matrix [1]. Classically, they have a slightly higher incidence in the male population [5]. They are the third most common primary malignancy of bone (representing around 9% of all primary bone malignancies) after myeloma and osteosarcoma, the second most common primary bone sarcoma after osteosarcoma, with a prevalence of 11% to 22% and the most frequent one in adults. Three main subtypes are described: conventional, mesenchymal, and clear-cell [1, 5, 6].

With the most common sites being the pelvis followed by the proximal femur, proximal humerus, distal femur and ribs, it can rarely occur in the hand, with a frequency of only 1.5–3.2% of all chondrosarcomas [1, 5–7]. Phalangeal bones are affected in 60% of these cases and the metacarpals in about 40% [1, 5–7].

In conventional chondrosarcoma, the histological malignancy grading represents the main prognostic factor in surgical planning and prognosis [8]. Surgery is the mainstay of treatment regarding the limited cCS. While curettage is acceptable for grade 1 cCS, for higher grade cCS, wide excision is usually required. Skull base cCS is the exception, which may be treated with radiotherapy. Hadrons, in particular, can play an important role in the management of skull base cCS, very good outcomes being reported [9].

Periosteal chondrosarcomas are rare and represent only 1% to 2% of all chondrosarcomas [10]. Ulnar-sided wrist pain is a common complaint that presents a diagnostic challenge. The differential diagnosis of ulnar-sided wrist pain can be divided into six elements: osseous, ligamentous, tendinous, vascular, neurological, and miscellaneous [11]. The bones consist of the distal ulna, lunate, triquetrum, capitate, pisiform, and hamate. Osteoid osteoma of the hamate can produce ulnar-sided wrist pain in the dorsal or volar aspect of the wrist, depending on the location of the tumor in the bone [10, 11].

Chondrosarcomas are slow evolving, and symptoms can be found for years before the patient seeks medical attention. Pain in the absence of a pathological fracture

can be an important feature differentiating an enchondroma from a low-grade chondrosarcoma. Similar to enchondroma, it is a lesion arising in the medullary cavity with irregular matrix calcification, but chondrosarcomas have a more aggressive appearance with bone destruction, cortical erosions, periosteal reaction, and rarely a soft tissue mass [12]. Pain is more common with chondrosarcoma than with the benign enchondroma.

Histologically, chondrosarcomas are composed of malignant cells with abundant cartilaginous matrix, hypercellularity, plump nuclei, binucleated cells, a permeative pattern, and entrapment of bony trabeculae [13]. Tumoral markers and immunohistochemistry using vascular endothelial growth factor (VEGF), platelet-derived growth factor (PDGF), transforming growth factor-beta 2 (TGF $\beta$ 2), Ki67 or p53 are currently relevant for tumoral detection, improved diagnosis and recurrence follow-up [14].

The constant growth of the musculoskeletal tumoral pathology is determined by the increase in the average life expectancy on one side and by the constant exposure to various aggressive agents as a result of technological and industrial development, on the other side [15].

The main differential diagnosis of the low-grade chondrosarcoma is the enchondroma. Because they share a set of common morphological features, the differential diagnosis between these two entities is one of the hardest to establish in the bone pathology, often requiring the aid of patient's clinical and imaging data. Microscopic aspects favoring the malignancy diagnosis are: permeation of the trabecular bone, overcoming the bony cortical and the tumor's presence in the nearby soft tissues. Ollier disease is defined by multiple enchondromas with a disproportional distribution. The association of multiple enchondromas with multiple soft tissue hemangiomas or lymphangiomas is called the Maffucci syndrome. The malignant tendency is a common feature for both conditions. They often appear in the first decade of life. It remains uncertain if a single gene defect or a combination of mutations are involved in the occurrence of Ollier disease and Maffucci syndrome [16, 17]. In the latter, the heterozygous somatic mutations in the isocitrate dehydrogenase 1 and 2 (IDH1/IDH2) genes are suspected for their involvement [17].

Another differential diagnosis is the chondroblastic

osteosarcoma, more frequently found in teenagers and young adults. From a morphological point of view, this diagnosis implies the existence, at least focal, of the tumoral osteoid.

## ✉ Conclusions

Our study reviews the management of chondrosarcoma of the extremities, but also highlights the uncommonness of this in the metacarpal bones. Since the chondrosarcoma has a wide age distribution, its accurate diagnostic and treatment gives the patient the ability to continue his daily activities and be a fully social reinstated person, reducing time and costs sustained by the healthcare system.

## Conflict of interests

The authors declare that they have no conflict of interests.

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