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CASE REPORT
KAZUISTIKA

High-Grade Chondrosarcoma of the Proximal Phalanx: an Unusual Case of a Rare Entity

High-Grade chondrosarkom proximálního článku: neobvyklý případ vzácné entity

A. PAGNOTTA[1], V. M. FORMICA[1], S. TACCOGNA[2], G. SUMMA[3], C. ZOCCALI[4,5]

[1] Hand and Microsurgery Unit of the Jewish Hospital of Rome, Rome, Italy
[2] Pathology Unit of the Jewish Hospital of Rome, Rome, Italy
[3] Villa Stuart Eurosanità, Rome, Italy
[4] Department of Anatomical, Histological, Forensic Medicine and Orthopaedic Science, Sapienza University of Rome, Rome, Italy
[5] IRCCS Regina Elena National Cancer Institute, Rome, Italy

SUMMARY

Chondrosarcoma of the hand is a rare disease, but is one of the more common malignancies of the hand. Biopsies and imaging are a fundamental step in determining correct diagnosis, grading and selection for best treatment.

We describe the case of a 77-year-old male complaining of a painless swelling in the proximal phalanx of the third ray of left hand. A biopsy was performed and the histology revealed a G2 chondrosarcoma. The patient underwent III ray amputation with metacarpal bone disarticulation and sacrifice of the radial digit nerve of the fourth ray. Definitive histology revealed grade 3 CS. Eighteen months after surgery, the patient is apparently disease-free with a good functional and aesthetic outcome although with persistent paresthesia of the fourth ray.

Although there is no agreement in the literature for the treatment of low-grade chondrosarcomas, wide resection or amputation can be considered the mainstay treatment for high-grade tumors.

Key words: chondrosarcoma, proximal phalanx, ray amputation, surgical treatment, tumor hand.

INTRODUCTION

Chondrosarcoma (CS) is a malignant bone tumor producing cartilaginous tissue. It usually occurs in the fifth and sixth decades (3).

Excluding rare subtypes as clear cell CSs, mesenchymal CSs and juxtacortical CS, the conventional CSs are divided into three main groups based on their biological aggressiveness: low-grade CS or G1, grade 2 CS or G2, grade 3 Cs or G3; a fourth dedifferentiated form can also occur (3).

Biopsies are important for diagnosis, even though a correct image interpretation is fundamental for differentiating between low-grade CSs and benign chondromas (3).

Common localizations include bones of the pelvis, proximal femur, proximal humerus, distal femur and ribs (3).

Even though chondroma is the most common benign tumor of the hand, low-grade CS has to be considered quite rare in this site; otherwise, high-grade CS is exceptional.

In the context of a rare case, even though less than 10% of all chondrosarcomas occur in the hand, they are the most common primary malignant bone tumors of the hand representing about the 4% of all other hand malignancies (2, 6, 8).

If no consensus is present in literature about the correct treatment for low-grade tumors, whether intralesional or wide, radical surgery is recognized as the principal treatment in high-grade cases since neither chemotherapy nor radiation is effective (8).

CASE REPORT

A 77-year-old man at observation complained of a painless swelling in the proximal phalanx of the third ray of left hand which he had for 6 months.

He also noticed a progressive increase in size of the lesion and a progressive reduction of the range of motion. On physical examination, there was a swelling mass of taut-elastic consistency on the ulnar and volar side of the proximal phalanx restricting range of motion both in flexion and extension of the III metacarpophalangeal joint (Fig. 1A). There was no pain both during thumb-index pinch nor during digital pressure of the involved phalanx.

Radiographic examinations showed a geographic map osteolysis with calcified spots in the matrix indicating a slight erosion in the ulnar side, compatible with an aggressive chondroid tumor (Fig. 1B).

Subsequent MRI scans demonstrated an apparently cartilaginous bone tumor extending into the soft tissue involving the flexor tendon of the third finger and probably the radial nervous and vasal structures of the fourth finger (Fig. 1C).

Given the high level of suspicion of aggressive malignant tumors, a CT-guide trocar biopsy was performed and histopathologic findings initially revealed a grade 1 CS. Considering the aggressive aspect of the lesion, a second biopsy was performed with a related diagnosis more coherent with a grade 2 CS.

Based on the absence of distant metastases, the aggressive behavior demonstrated from the imaging scans and the histological tests, as well as the impossibility of

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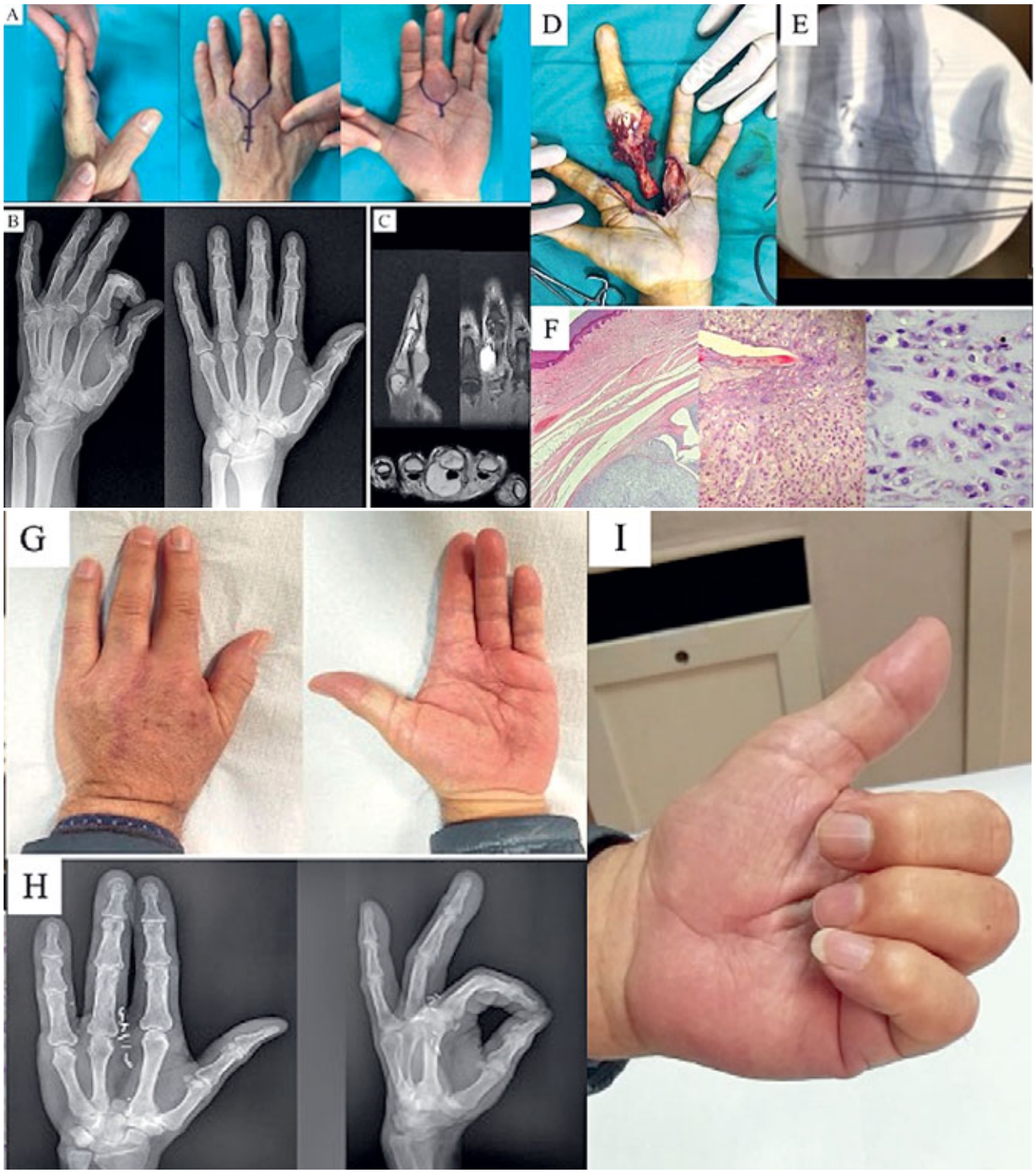

Fig. 1. A – clinical presentation of the swelling in the left hand in the lateral, dorsal and palmar view; B – X-ray showing a geographic map osteolysis with calcified spots in the matrix indicating a slight erosion in the ulnar side, compatible with an aggressive chondroid tumor; C – MRI demonstrating the erosion of the cortical bone and the involvement of the flexor tendon (lateral view: spin echo-T1; coronal view: gradient echo-STIR; axial view: turbo-spin echo-T2); D – intraoperative picture showing the amputated third ray; E – intraoperative X-ray showing Kirshner wires inserted to reconstruct the metacarpal arch; F – hematoxylin and eosin (HE) preparations; left square: the neoplasm infiltrates the subcutaneous. Note the skin on the bottom right (HEx100); central square: the tumor grows in lobules/nodules and shows cellular components embedded in a myxoid background stroma and shows hypercellular areas (HEx200); right square: the tumor has enlarged, binucleated and multinucleated atypical cells with an increased nuclear to cytoplasmic ratio and variable tumor necrosis. Cells are characterized by irregular nuclear chromatin, prominent nucleoli, mitotic figures (arrow) (HEx400); G – post-operative clinical presentation of left hand in dorsal and palmar view; H – post-operative X-ray at 18 months of follow-up; I – post-operative clinical presentation of the hand flexion.

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performing savage surgery, radical surgical treatment was indicated.

Considering better aesthetic and functional outcomes, a disarticulation of the third ray was performed instead of amputation of the finger (Fig. 1D). In observing the involvement of the radial digital nerve of the fourth ray during surgery, it was also sacrificed. Four Kirshner wires were inserted to reconstruct the metacarpal arch which was then removed after 45 days (Fig. 1E).

The operation was performed by surgeons who had wide expertise in muscular-skeletal oncology and reconstructive surgery.

The definitive histology exam revealed a grade 2 CS with focal areas of grade 3 (Fig. 1F).

The post-operative course was free of complications.

At 18 months of follow-up, the patient was apparently free of disease, completely satisfied with the aesthetic and functional results, although a paresthesia of the radial side of the fourth finger was present due to sacrifice of the related digital nerve (Fig. 1G).

The DASH-score was 25%.

The X-ray showed a good alignment of the metacarpal bones with a possible residual part of the basis of the fourth metacarpal bone (Fig. 1H) which corresponded to a satisfying related hand flexion (Fig. 1I).

The patient gave his consensus for publication of the present case; the publication of the case was also approved by the local ethical committee.

DISCUSSION

CS is the second most common primary bone malignancy after osteosarcoma. Chondrosarcomas of the hand account for just 6–10% of all reported cases but is the most common primary malignant bone tumor of the hand, accounting for about 4% of all hand malignancies (2, 3, 8).

If low-grade CS can be considered common, G3 CS is quite rare. Patil S. et al. carried out a review of literature which identified 23 cases of CS of the hand where one grade 1, 20 grade 2 and just two grade 3. Moreover, only few cases of dedifferentiated chondrosarcomas are reported in literature (2).

Due to the indolent behavior of CS, this tumor is often misdiagnosed. In literature, it has been reported that the time between first symptom appears and surgery is 2 months to 30 years (2, 8).

In our case, there was no pain present but swelling and a reduction in the range of motion which forced the patient to arrive at observation.

High quality imaging accuracy, both of CT scans and MRIs, is important in revealing clear lesions with the presence of possible calcifications and several probable patterns described as popcorn-like, punctate or comma-shaped spots/lesions, the cortical bone can be destroyed. Biopsies are important for diagnosis and grading. Nevertheless, the effectiveness of biopsies in determining the actual grade is quite low, thus imaging-pathology concordance and clinical behavior is crucial otherwise a more aggressive grade needs to be hypothesized.

Indeed, the final histology exam may differ from the first biopsy in terms of grade just like our case: the preoperative histology corresponded to a low-grade tumor but the images, the clinical aspects as well as an increase in diameter were more consistent with a higher-grade tumor, therefore a second biopsy was performed.

Treatment of CSs is an area of debated in the literature. CSs are quite radio and chemo-resistant, therefore surgery is the indicated treatment.

The difficulty in differentiating between the different grades above, as well as between chondroma and low-grade CS, are the main challenges faced when basing diagnosis on the preoperative grade.

Also, FDG-PET scans were less accurate in distinguishing between chondromas from low-grade CSs (1).

If wide surgery is recognized as mandatory for high-grade tumors independently from the site, no consensus has been reached for low-grade CS. Indeed, several authors sustain intralesional surgery for low-grade CS located in the limbs, based on the low-metastatic potential of that form.

Nevertheless, a recent review was critical towards this treatment sustaining that in several cases where curettage had been applied successfully, the diagnosis was rather questionable and the correct diagnosis was more likely to be chondroma (11).

Based on these factors, intralesional surgery should be considered questionable for treating CS. The actual tumor grade can be higher than that found at the preoperative histologic exam.

Nevertheless, it would seem that CS of the hand had the tendency to form local recurrence but not metastasis (7).

In 1999, Mankin et al. paper discussed the relatively "benign" behavior of the CSs located in the hand, and hypothesized that they were a different entity altogether naming them "primary non-metastasizing chondrosarcoma of the digits" (4).

In addition, in 1999 Bovèe et al. reported the results of 28 cases of hand CSs and performed a meta-analysis of 84 cases, they concluded that CS show high aggressive local behavior but low metastatic potential (2).

In 2014, Stomeo et al. also affirmed that chondrosarcomas of the hand are believed to be more benign in their behavior compared to those located elsewhere (10).

These observations could justify a less aggressive treatment for low grade CSs of the hand. The principal goal of surgery might be minimizing functional impairment which provides the rationale behind performing curettage, local adjuvant therapy and bone grafting.

Hickey et al. in 2011 were able to compare 190 patients from five separate studies affected by grade I chondrosarcoma, in which 112 patients underwent wide excision and 78 were treated with intralesional curettage (4). They concluded that the risk of recurrence or metastasis between the two groups was not significant and intralesional curettage was a valid alternative for chondrosarcomas of the hand.

Also in the hand, wide resection is considered the gold-standard treatment for high grade CS.

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High-grade bone chondrosarcoma of the hand is best treated surgically with wide en bloc excision or ray amputation to gain adequate tumor clearance and avoid the risk of local recurrence or metastasis (4, 5, 7, 10).

Considering the poor functional results associated with reconstruction through bone grafting, amputation plays a greater role than in other segments. In literature are proposed only few techniques to reconstruct the rays (except in the case of the thumb); in particular bone grafting can spere the ray with better aesthetic results but with poor functional outcome.

Ray amputation allows to reach radical excision and good functional outcomes (except in the case of the thumb) (9).

Puhaindran et al in 2010 published the results of 25 patients who underwent single ray amputation excluding the thumbs. They reported a good functional result and a high MSTS score at least 2 years after follow-up (9).

In our case a diagnosis of grade 1–2 CS was made at biopsy. Therefore, wide resection and reconstruction or ray amputation were the possible surgical indications. After accurate evaluation based on the very aggressive radiological aspects and clinical behavior of the tumor and having had discussed the case thoroughly with the patient, a third ray amputation was performed with III metacarpal bone disarticulation in order to provide better functional outcome and sacrifice of IV radial digital nerve due to the involvement of these structures. No flap was deemed necessary for the reconstruction of the II-IV commissure and for the complete closure of the gap. Patient had no complications after surgery and demonstrated to have good function in terms of strength of grip, extension and flexion movement of other digits and satisfactory aesthetical outcomes although he complained about paresthesia on the radial side of the fourth finger due to the sacrifice of the radial fourth digit nerve.

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Corresponding author:
Carmine Zoccali M.D. PhD.
Department of Anatomical, Histological,
Forensic Medicine and Orthopaedic Science
Sapienza University of Rome
Piazzale Aldo Moro 5
00185 Rome, Italy
E-mail: carminezoccali@libero.it