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Hand Chondrosarcoma: Clinical Case Report

Condrosarcoma de mano: Reporte de caso clínico

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Abstract

Chondrosarcoma is a malignant cartilaginous tumor usually located in the pelvis, spine, and long bones. Only 1.5% of chondrosarcomas occur in the hand, frequently in the phalanges. They can be primary or secondary, depending on whether its origin is a normal bone or a pre-existing cartilaginous lesion, which is more frequent in the hand. We present a 70-year-old female patient, painter, and pianist who presented a 5-month-old increased volume and pain at a preexistent pinky tumor. Physical examination showed a volume increase of stony consistency affecting the base and the proximal third of the proximal phalanx with no mobility or neurovascular abnormalities. Radiological studies, complemented by magnetic resonance imaging, showed an insufflating lesion with cortical thinning and soft tissue calcification. The case was presented to the Oncology Committee, which studied the tumor dissemination and performed a wide excision. An anatomopathological study showed a chondrosarcoma with free margins. Three months after surgery, the patient resumed her musical activity with no evidence of recurrence or metastasis to date. Determining whether a chondroid tumor is benign or malignant is essential for prognosis and treatment. In addition, it is critical to combine clinical symptoms with imaging and histological findings. As this tumor is neither radiosensitive nor chemosensitive, wide excision with negative margins is a treatment option seeking to preserve life over function and aesthetics. The recurrence rate is variable, reaching up to 60%. Therefore, despite the low risk of malignancy, it is advisable to follow up on these lesions both clinically and radiologically.

Keywords

- malignant cartilage tumor
- chondrosarcoma
- hand chondrosarcoma

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Resumen

El condrosarcoma es un tumor cartilaginoso maligno localizado usualmente en pelvis, columna y huesos largos, encontrándose solo 1,5% en la mano, frecuentemente falanges. Este puede ser primario o secundario, según su origen sea un hueso normal o lesión cartilaginosa preexistente, esta última más frecuente en mano. Se presenta a una paciente de 70 años, pintora y pianista, que consultó por aumento de volumen y dolor de 5 meses de evolución en relación a un tumor de meñique antiguo. Al examen físico destacó aumento de volumen de consistencia pétrea que afectaba base y tercio proximal de falange proximal, sin alteración de movilidad ni neurovascular. Se realizaron estudios radiológicos que evidenciaron una lesión insuflante con adelgazamiento cortical y calcificaciones en partes blandas, complementados con resonancia magnética. Se presentó al Comité de Oncología, realizó estudio de diseminación y excisión amplia cuyo estudio anatomopatológico evidenció condrosarcoma, con márgenes libres. A los 3 meses postoperatorios la paciente retoma la actividad musical, sin evidencia de recidiva ni metástasis a la fecha. Determinar si un tumor condroideo es benigno o maligno es primordial para el pronóstico y tratamiento, siendo fundamental para ello conjugar la clínica, imagenología e histología. Como este tumor no es radio ni quimiosensible, la escisión amplia con márgenes negativos es una opción de tratamiento, buscando preservar la vida por sobre la función y estética. La tasa de recurrencia es variable, alcanzando hasta el 60%, por lo cual, pese al bajo riesgo de malignización, es recomendable seguir tanto clínica como radiológicamente estas lesiones.

Palabras Clave

- tumor cartilaginoso maligno
- condrosarcoma
- condrosarcoma de mano

Introduction

Chondrosarcomas are the second most common primary malignant bone tumors after osteosarcomas.[1] They are chondroid lesions and account for 9% to 10% of all primary malignant bone tumors.[2–4] Chondrosarcomas appear between 40 and 80 years old,[2] mostly around the 5th and 6th decade of life.[2–5] The most common locations are the pelvis, proximal femur, and proximal humerus in up to 70% to 75% of cases.[3,5,6] Of all chondrosarcomas, 0.5 to 3.2% occur in the hand,[2,3,5,6] and mostly are low-grade lesions. Hand chondrosarcoma dissemination is exceptional, even in lesions with a high histological grade.[2,6,7]

Although they can be primary tumors, up to 88% of hand chondrosarcomas are secondary to the degeneration of preexisting lesions such as enchondromas and osteochondromas or occur with multiple enchondromatoses.[1,2,5] They affect men in a 3:1 ratio[3] and the phalanges in up to 68% of cases.[2] The proximal phalanx is the most common site, and its involvement occurs in approximately 53% of patients.[2,6] In addition, there is a predominance of the little finger at 33%.[5,6]

Based on cellularity, matrix and nucleus features, and number of mitoses, there are three chondrosarcoma grades:[1,3,8] low (grade I), intermediate (grade II), and high (grade III). Low-grade lesions are the most frequent and characterized by moderate hypercellularity and atypia compared with the high cellularity and pleomorphism from high-grade lesions.[5]

For chondrosarcomas, the challenge lies in their differentiation from a benign condition (for instance, an enchondroma), especially if the lesion is low-grade.[1] As a result, emphasis is placed on a complete study, with clinical, imaging, and histology as the cornerstones to determine the malignant origin of the lesion and, then, choose the definitive treatment, ranging from curettage and bone graft to wide resection (amputation) of the compromised area.[6]

Clinical case

A 70-year-old woman, painter, and pianist, hypertensive under treatment, had a long-standing history of a tumor in the right little finger. She presented pain and increased volume at the right little finger for 5 months. She did not report any history of trauma. The physical examination revealed an increased proximal volume of a stony consistency affecting the base and proximal third of the proximal phalanx. The patient presented good mobility of adjacent joints and no neurovascular compromise. We requested a conventional radiological examination (►**Figure 1**) and, to rule out a potential pathological fracture, a magnetic resonance imaging (MRI) with contrast (►**Figure 2**).

Since the study highly suggested a malignant lesion, we requested a chest computed tomography (CT) scan, which showed no evidence of dissemination. After discussing the case with the Oncology Committee from our institution and the Mayo Clinic, we decided on a wide excision instead of an excisional biopsy. Since the involvement of the proximal phalanx was almost complete, an excisional biopsy would virtually be an amputation of the finger, leaving it dysfunctional and with no bone support. The biopsy result confirmed

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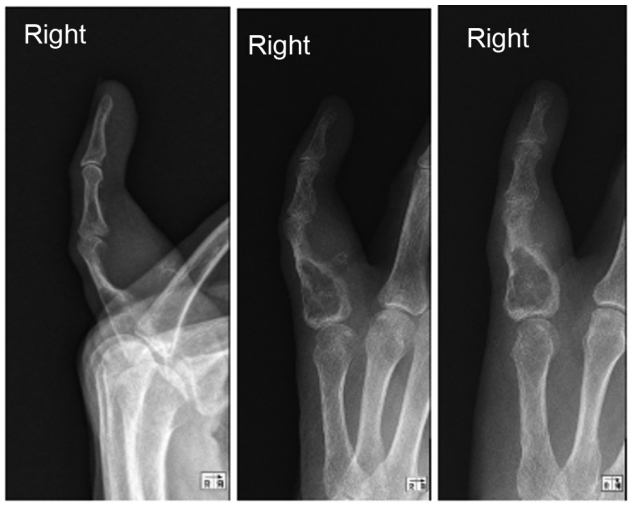

Fig. 1 Radiograph of the right little finger showing a radiolucent geographic bone lesion on the proximal half of the first phalanx, which inflates and thins the cortex, with endosteal scalloping, no suspicious periosteal reaction, and a non-aggressive appearance, suggesting an enchondroma. There was a 5-mm bone fragment displaced upward and increased volume of the regional soft tissue, possibly indicating a pathological bone fracture.

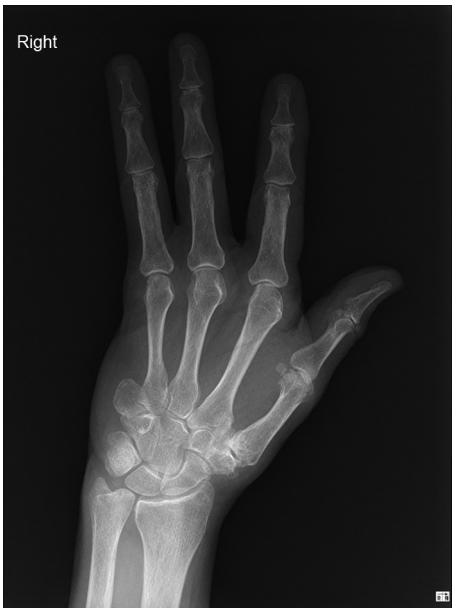

Fig. 3 Postoperative follow-up radiograph (1.5 years later). Amputation at the fifth metacarpal base level with no evidence of recurrence.

the diagnosis of a well-differentiated grade I chondrosarcoma and demonstrated the free margins.

Later, the patient presented an ulnar subluxation of the extensor tendon of the fourth finger with mild symptoms. Its treatment consisted of conservative therapy and the construction of a supportive orthosis. Three months after surgery, the patient resumed her musical activity. After 1.5 years, there was no evidence of metastases (►**Figure 3**).

Discussion

Differentiating chondrosarcoma from an enchondroma is a considerable challenge, even more so if the malignant tumor is low grade since they are similar in imaging and histological features.[5] As a result, combining clinical interpretation with imaging and histopathological studies is essential.[5,9] In this context, a histologically benign lesion could be considered malignant if the imaging suggested it, while an imaging-benign tumor would be interpreted as malignant if the clinical examination, histological findings, or both suggested it.[9,10] Making this difference is fundamental not only for diagnostic clarification but determining appropriate treatments and reducing recurrence rates.[5]

Clinically, malignancy-suggestive signs include a progressive volume increase and pain (a potential pathological fracture), and up to 10% of cases are asymptomatic.[2,6] For imaging, it is important to complement conventional radiology with CT, MRI, or both with contrast to characterize the lesion and its relationship with adjacent structures. In these cases, cortical destruction or thinning, soft tissue

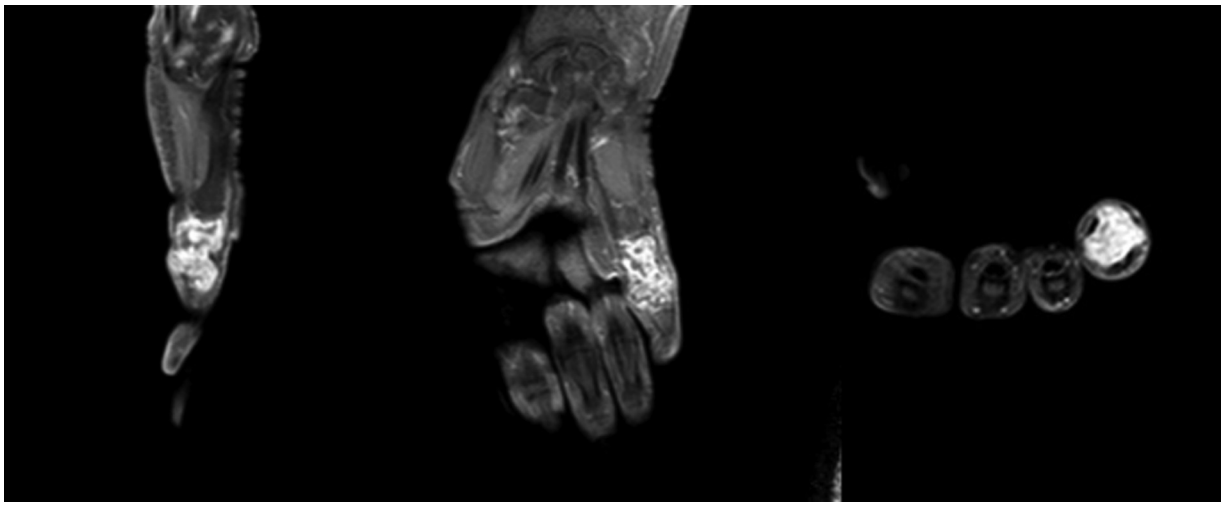

Fig. 2 Tumor lesion of the proximal phalanx of the little finger. Its characteristics and the previous radiographic study indicate a chondroid tumor with malignancy signs resulting from its extraosseous extension (chondrosarcoma) with no involvement of adjacent joints.

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involvement, periosteal reaction, and intralesional calcifications ("popcorn-like lesions") suggest malignancy.[2,3,6]

Regarding histology, it is worth mentioning that chondromas have high cellularity and more nuclear atypia than in other parts of the body.[2] As a result, the microscopic appearance alone does not confirm the diagnosis. However, a sample with abundant cartilaginous matrix, hypercellularity, bulging nuclei, binucleated cells, and bone trabeculae entrapment suggests chondrosarcoma.[3]

Most chondrosarcomas are reportedly resistant to chemotherapy and radiotherapy.[1,2,6,9] As such, their treatment of choice is surgery, and the type of surgical intervention is a matter of debate. Arguably, in a locally aggressive disease with exceptional dissemination potential,[5] intralesional curettage plus bone graft is an alternative in low-grade, central chondrosarcomas with no cortical involvement, sparing the body segment and its function (especially in thumb injuries). This therapeutic alternative has a higher recurrence rate,[1,2,6,11] ranging from 6% to 60%[1,6,11] compared with more aggressive management. On the other hand, to avoid the possibility of local recurrence and dissemination and as an alternative for definitive treatment, wide resection (amputation) is another option, as in this case, in which incisional biopsy can lead to diagnostic error and an excisional biopsy (with intralesional curettage) was not a good alternative since it would have left a dysfunctional finger with no bone support. For high-grade lesions (II or III), with severe function-altering deformity, extension to soft tissues, neurovascular compromise, and radiological and/or clinical signs suggestive of aggressiveness, wide excision is more acceptable, always trying to preserve the functionality and aesthetics.[1–3,5] The amputation must occur in the healthy tissue, sparing the lesion and removing the tissue from the biopsy site.[2] In addition, it is worth mentioning that adequate follow-up is always required to identify potential recurrence or dissemination, especially if treatment is more conservative.[3]

Conclusion

A hand chondrosarcoma is a rare condition with a low capacity for metastasis and high recurrence rates, especially with insufficient treatments. The definitive diagnosis is challenging, particularly when differentiating benign and low-grade malignant lesions. As a result, it is essential to contextualize the clinical picture with the imaging and histological findings. In some situations, intralesional curettage may be appropriate, but wide excision still has a role in local tumor control.

Conflict of Interest
None declared.

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