

# Well-differentiated extraskeletal chondrosarcoma: about a new case

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## Key words

Extrasosseous chondrosarcoma • Well-differentiated • Soft tissue

## Summary

Extraskeletal chondrosarcoma is a rare malignant tumor. The well differentiated histological type, which is found primary in soft tissue, is extremely rare. This report describes the case of a 58-year-old woman presented with a large palpable mass in the right buttock. Imaging studies revealed a well-defined soft tissue

mass, with extensive calcification. A histological examination after surgical resection confirmed the diagnosis of well-differentiated extrasosseous chondrosarcoma. The outcome was favorable, without recurrence or metastasis.

## Introduction

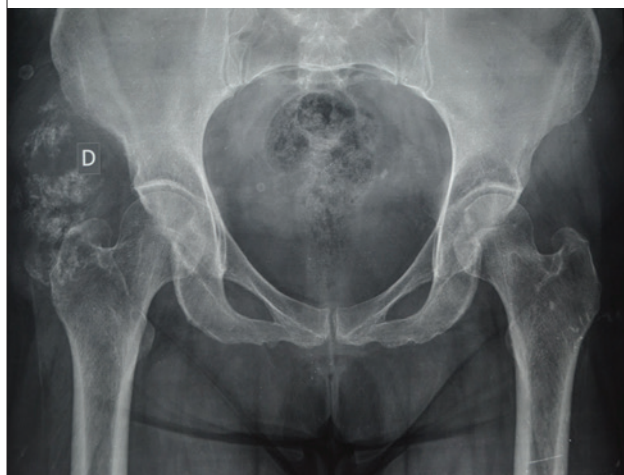
Chondrosarcoma, a malignant tumor that originates from cartilage cells, is the second most common malignant bone tumor, constituting 20-27% of all primary malignant osseous neoplasms<sup>1,2</sup>. Chondrosarcoma can also develop in other areas, such as soft tissue<sup>3-5</sup>, lung<sup>6</sup>, breast<sup>7</sup>, where cartilage is not normally found. The purpose of this report is to describe a rare example of a well-differentiated extrasosseous chondrosarcoma arising in soft tissue and discuss the differential diagnosis of cartilaginous lesions.

## Case report

A 58-year-old woman, with no significant past medical history, presented with a 6-months history of a painless lump in the right hip, without sensory or motor deficit or difficulty in mobilization in the physical examination. A radiograph revealed a soft tissue mass, with flaky calcification (Fig. 1). Magnetic resonance imaging (MRI) revealed a well-defined soft tissue mass in the right buttock. The mass measured 12 cm in diameter and showed isointense signal on the T1-wighted image and heterogeneous high signal intensity on the T2-wighted

image (Fig. 2). A surgical biopsy was recommended. The biopsy specimen featured numerous whitish cartilaginous fragments. A microscopical examination concluded to an atypical cartilaginous tumor. The presumptive diagnosis was an extraskeletal chondrosarcoma. Computed tomography (CT) confirmed the presence of a well-demarcated soft tissue mass, with arci-

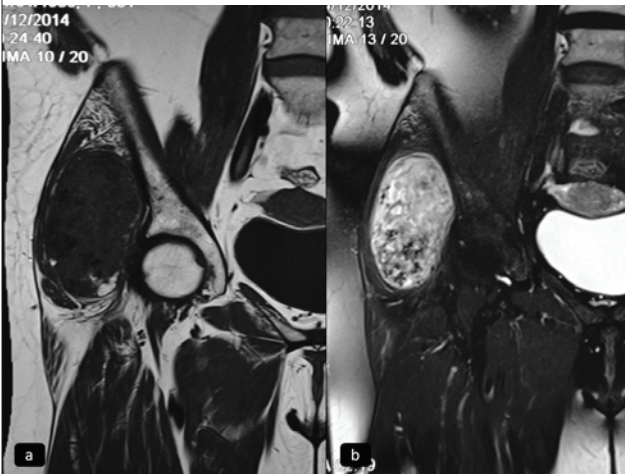
Fig. 1. Radiograph demonstrating a large soft tissue mass with calcification.



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**Fig. 2.** A well defined isosignal T1(a) soft tissue mass and heterogenous high signal intensity on T2 (b).



**Fig. 4.** On cut section, it is a circumscribed tumor measuring 18 cm at the greatest diameter, yellow-white with a cartilagenous appearance.

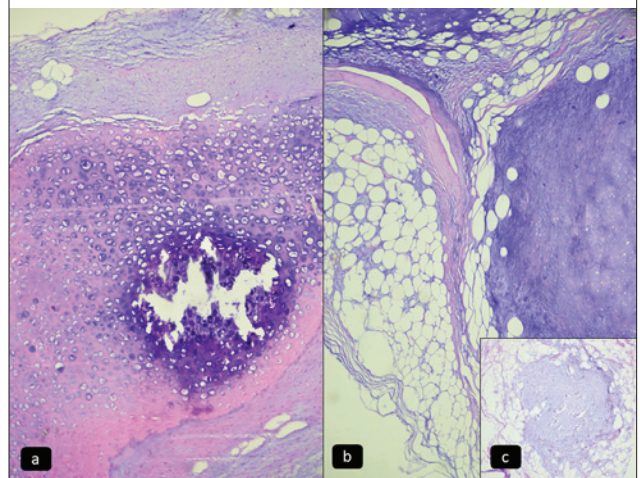


form calcification outside the right iliac bone, without obvious absorption or destruction (Fig. 3). The tumor was removed surgically, with wide excision. It consisted of a circumscribed solid mass measuring 18 × 17 cm with a yellow-white matrix and cartilaginous appearance (Fig. 4). Histologically, the tumor was composed of multilobulated, moderately to mildly cellular cartilaginous tissue with calcification and ossification. Chondrocytes contained occasional hyperchromatic nuclei, with binucleation and a mild atypical nucleus. No mitoses were noted. The cartilaginous lobules were separated by fibrous septa, which were continuous with a myxoid matrix. In addition, tumor proliferation infiltrated adjacent adipose tissue (Figs. 5, 6). Histology favored a diagnosis of a well-differentiated chondrosarcoma completely resected. The patient remained well, with no recurrence or metastasis at a 10-month follow up evaluation.

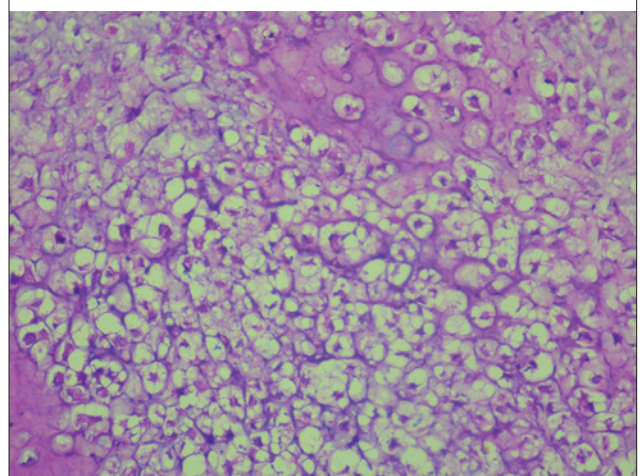
**Fig. 3.** CT shows that this mass is isodense to muscle and it is outside the right iliac bone.



**Fig. 5.** The histological examination reveals multilobulated hyaline cartilaginous tumor with calcification (a : HE×100), separated by myxoid matrix and infiltrate adipose tissue (b : HE×100, c : HE×200).



**Fig. 6.** Cartilaginous lobules are moderately cellular with mild atypical nucleus (HE×400).



**Tab. I.** Clinico-pathologic features of all well-differentiated extra-osseous chondrosarcoma.

Article	Year	Number of cases	Age/Sex	Location	Size (cm)	Tumor grade	Follow-up	Evolution
Wu et al. <sup>8</sup>	1980	5	NA	NA	NA	NA	NA	NA
Amir et al. <sup>4</sup>	1985	1	50/M	Foot soft tissue	NA	NA	NA	NA
Bailey et al. <sup>9</sup>	1990	1	70/F	pleural	8	Grade I	13,5 years	Alive with no recurrence or metastasis
Khalidi et al. <sup>5</sup>	2005	1	38/M	Foot soft tissue	7,5	Grade I	4,5 years	Alive with pulmonary metastasis
Shah et al. <sup>6</sup>	2007	1	60/M	Lung	14	Grade I	6 years	Alive with cutaneous and vertebral metastasis
Dianbo et al. <sup>3</sup>	2011	1	71/M	Inter costal soft tissue	9,1	Grade I	1 year	Alive with no recurrence or metastasis
Present case	2017	1	58/F	Buttock	18	Grade I	10 months	Alive with no recurrence or metastasis

NA: Not Available ; M: Male ; F: Female

## Discussion

Extrasosseous chondrosarcomas are infrequent malignant tumors, which account for less than 3% of all soft tissue sarcomas <sup>1,3</sup>. The most common histological types are myxoid and mesenchymal chondrosarcomas. Well-differentiated types are extremely rare, with only a few cases reported in the English literature <sup>3-6,8,9</sup> (Tab. I). A careful examination and radiological assessment should be performed to exclude metastasis. The present case was unusual because the tumor belonged to a well-differentiated subgroup of extrasosseous chondrosarcomas and originated in the soft tissue of the buttock.

The most common presentation of an extrasosseous chondrosarcoma is a mass. However, extrasosseous chondrosarcoma can reach an enormous size, as was found in the present case. CT revealed an oval-shaped mass, with a ring and arc pattern of chondroid matrix mineralization <sup>3</sup>. Histologically, the tumor was composed of multilobulated blue hyaline cartilage. These lobules of cartilage were not well defined. Previous studies reported increased cellularity and atypia, depending on the grade of the tumor <sup>3,5</sup>.

Well-differentiated extraskeletal chondrosarcoma generally have a good prognosis after wide excision, although cases of recurrences or metastases have been reported in the literature. For example, Khalidi et al. described a case of a well-differentiated extrasosseous chondrosarcoma of the foot, with pulmonary metastases <sup>5</sup>. In the present case, the differential diagnoses included a soft tissue chondroma and myxoid liposarcoma, with cartilaginous differentiation. Soft tissue chondromas are rare benign tumors, which occur mainly on hands and feet. They are well demarcated, small masses which rarely exceed 3 cm in diameter, and do not invade adjacent tissue <sup>10</sup>. A myxoid liposarcoma with cartilaginous differentiation presents as a well circumscribed, multinodular, intramuscular tumor, with a gelatinous cut surface <sup>11</sup>. Histologically, this tumor shows bland fusiform cells and lipoblasts in a myxoid background, with a prominent

capillary net work and focal cartilaginous differentiation <sup>11</sup>.

In conclusion, extraskeletal chondrosarcoma is a rare malignant tumor. We describe a recent case of well-differentiated extrasosseous chondrosarcoma. The current report is unique because of the rare histological type and because only a few cases of extraskeletal chondrosarcoma in the soft tissue have been reported. An extraskeletal chondrosarcoma should be considered as a possible diagnosis, especially when a soft tissue mass shows calcifications on imaging examinations.

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