

## SECONDARY CHONDROSARCOMA FOUR CASES AND REVIEW OF THE LITERATURE

E. C. R. MERCHAN, S. SANCHEZ-HERRERA, J. M. GONZALEZ

The authors present four new cases of chondrosarcoma secondary to multiple osteochondromatosis (MOC). As MOC is a familiar and hereditary condition, the patients should be evaluated throughout their lives, from childhood to adulthood. Because of the risk of malignant transformation in this condition, any adult patient complaining of pain and excessive increase in tumor size should be investigated for sarcomatous degeneration. Wide resection, when possible, must be the elected treatment in this type of patient, in order to avoid the risk of local recurrence.

**Keywords :** multiple osteochondromatosis ; secondary chondrosarcoma.

**Mots-clés :** chondromatose multiple ; chondrosarcome secondaire.

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Cartilaginous tissue forming tumors can sometimes show benign behavior but at other times can transform into a chondrosarcoma. These benign tumors have been classified as chondromas, osteochondromas, chondroblastomas and chondromixoid fibromas. On the other hand, these malignant cartilaginous tumors can arise in both skeletal or extraskkeletal locations (1-6).

Skeletally-located chondrosarcomas can be divided into primary and secondary types ; the primary tumors have been classified as central, juxtacortical, mesenchymal, clear cell and dedifferentiated. On the other hand, the secondary skeletal cartilaginous tumors can be divided according to central and peripheral location (1, 2, 4, 6, 7).

These central tumors can be secondary to chondroma, to multiple enchondromatosis (Ollier's

disease), or to intravenous injection of Thorium dioxide (8-10), and they may occur after radiation of benign cartilaginous tumors (11). Furthermore, some central skeletal tumors may be associated with bone infarction and unicameral bone cysts (12) ; sometimes they can present as complications of fibrous dysplasia (13, 14) and Paget's disease (15, 16). The peripheral tumors may be secondary to osteochondroma, MOC and juxtacortical chondroma. Finally, the extraskkeletal tumors may be synovial, mesenchymal, or mixoid (1, 2, 4, 6, 7).

The purpose of this paper is to study four secondary chondrosarcomas which have been reviewed from an overall of 47 chondrosarcomas treated in a 24-year period.

### MATERIALS AND METHODS

From 1968 to 1991, forty-seven chondrosarcomas were treated in the Bone Tumor Unit of our centre.

The location of these tumors was as follows : mandible 3, scapula 5, humeral diaphysis 5, ribs 5, lumbar spine 4, pelvis 10, femoral diaphysis 10, knee joint 1 (synovial), fibula 1, tibia 1, and 2 were located in the feet. The clinicopathological variety of this series is summarized in table I. The mean age of the patients was 48.7 years (range : 15 to 85 years) ; 24 were male and 23 were female. In this series, four were secondary chondrosarcomas ; all were secondary to MOC. The main clinical characteristics of these four cases are now presented.

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Table I. — Clinicopathological diagnosis of 47 chondrosarcomas (24-year period)

1. Primary		
	Well differentiated	17
Central	Moderately differentiated	11
	Poorly differentiated	4
	Dedifferentiated	3
	Juxtacortical	5
	Clear cell	1
	Mesenchymal	1
2. Secondary		
	Multiple osteochondromatosis	4
3. Extrasketal		
	Synovium of the knee	1

#### Case 1

A 39-year-old female suffering from MOC presented in June 1978 with a tumor located in her right scapula. The lesion was considered to be stage IB according to Enneking (17); two weeks later a resection-biopsy was carried out and the diagnosis of secondary chondrosarcoma was confirmed. In July 1986 a first recurrence was found, which required a marginal scapulectomy. Unfortunately, in November 1990 a second recurrence (fig. 1) required a wide total resection. The last assessment performed in April 1991 did not show any recurrence.

#### Case 2

A 25-year-old male presented with a secondary malignancy in a previous MOC four years previously (fig. 2). The condition was located in the upper third of his right femur. The tumor was stage IB according to Enneking (17). A wide resection and substitution with a megaprosthesis Lima-PGR was carried out. The last assessment, performed in April 1991, did not show evidence of recurrence.

#### Case 3

A 19-year-old female suffering from MOC presented with a tumor in the right iliac in November 1988 (fig. 3). She was assessed as stage IB of Enneking (17). In November 1989 a marginal resection was carried out, followed by a partial but wide resection in January 1990. In April 1991 she did not show evidence of recurrence.

#### Case 4

A 47-year-old female presented in March 1990 with a tumor of the right scapula secondary to MOC. The lesion was stage IB according to Enneking (17). In October 1990 a marginal resection was carried out (fig. 4). The last assessment in April 1991 did not show signs of recurrence.

### DISCUSSION

The origin of secondary chondrosarcoma is a preexisting benign cartilaginous tumor, and it is most frequently associated with MOC or enchondromatosis. This condition precedes 25 to 30% of chondrosarcomas (18-20).

Our series of four secondary chondrosarcomas from a total of 47 chondrosarcomas over a 24-year period (8.4% rate) is slightly less than that in the literature. The explanation of this may be that our centre is not a national one, in contrast to the other centres studying this condition and reporting higher rates of occurrence. Clinical findings usually include pain, increase in tumor size, neurovascular complications or even pathological fracture (4, 6).

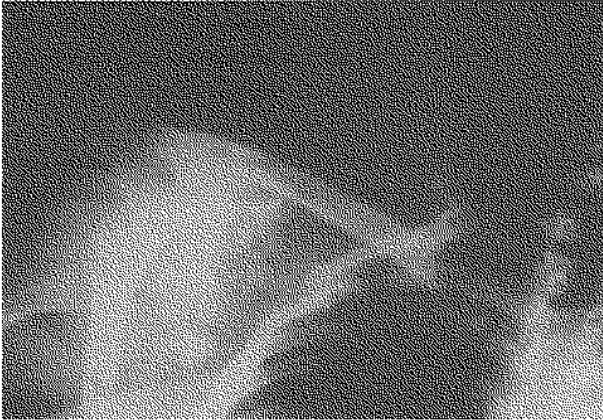
Our patients did not complain of symptoms of neurovascular complications or fractures, although pain and increase in tumor size were characteristic symptoms in this series.

The common location of this condition is the shoulder girdle or pelvis when associated with MOC, in contrast to the long bone metaphysis when related to enchondromas. All our cases were located in the pelvis or shoulder girdle.

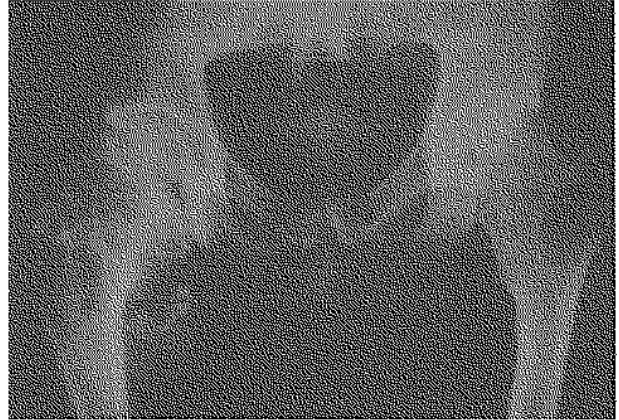
The pathological assessment of these tumors must be similar to the one utilized in primary chondrosarcomas. Furthermore, they have a low grade of malignancy, a better prognosis, and usually they are included in stages IA or IB of Enneking (17).

It is commonly accepted that standard radiographs, CT scan, MRI, arteriography and bone scintigraphy yield specific findings adequate to formulate the diagnosis.

Main radiographic features are popcorn, ring-like, or punctate intraosseous densities, focal lucencies greater than 2 cm, indistinct lesional mar-



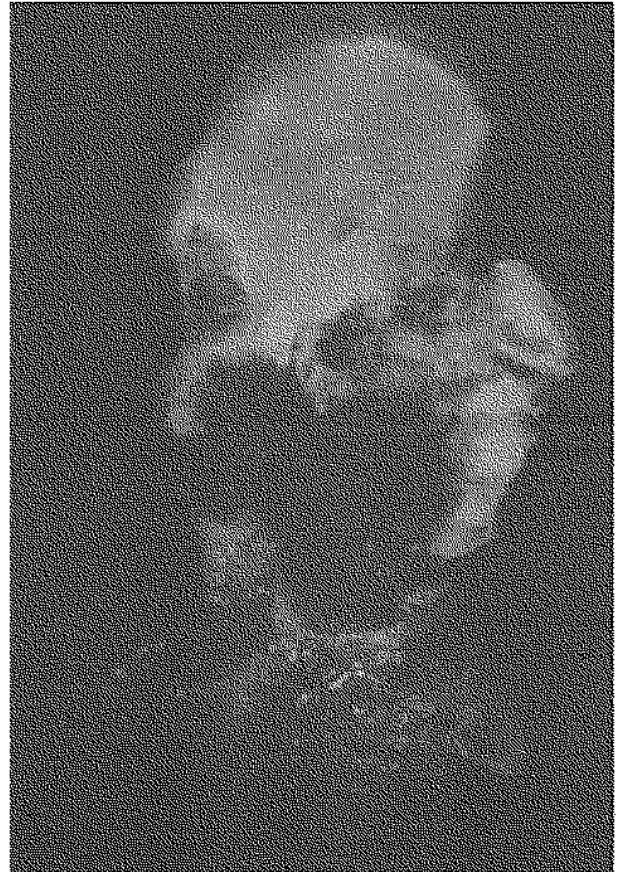
*Fig. 1.* — Case 1: radiographs at the time of the second local recurrence, 12 years after the beginning of the disease.



*Fig. 2.* — Case 2: initial radiographic examination showing the lesion in the upper third of the right femur.



*Fig. 3.* — Case 3: initial radiograph showing a tumor in the right iliac bone.



*Fig. 4.* — Case 4: the specimen obtained during surgery shows the marginal resection that was carried out.

gins and bone expansion (4, 6). Our data completely agree with these radiographic features referred to in the literature. When a preexisting osteochondroma shows a cartilage cap from 1 to 3 cm it is highly suspicious of sarcomatous degeneration. It has been reported that in Mafucci's syndrome a malignant transformation rate ranging from 15 to 56% can be expected (21, 22).

Malignant transformation of a solitary osteochondroma occurs in 1 to 16%, while MOC has shown sarcomatous degeneration rates ranging from 10 to 38% (3, 4, 6).

Our transformation rate of 8.5% is slightly lower than in the literature. Furthermore, four secondary chondrosarcomas were related to a preexisting MOC in this study. On the other hand, degeneration of a solitary chondrosarcoma is very rare. In fact, important series reported did not find cases of this condition. However, multiple enchondromatosis (Ollier's disease) formerly presented high rates of risk of malignant transformation ranging from 22 to 50% (6, 18, 19).

Degeneration after radiotherapy has been rarely reported; four out of 23 sarcomatous degenerations, excluding osteosarcomas have been reported by Huvos *et al.* (11), 3 out of 70 in Dahlin's series (2), and even 1 extraskeletal among 53 cases reported by Mankin (4).

A dose greater than 3000 cGy associated with a high degree of pathologic malignancy seems to be significantly related to this condition. The interval between radiotherapy and malignant transformation has been reported to be 3 years by Huvos *et al.* (11), in comparison with 8 to 10 years in Mankin's series (4).

Regarding chondrosarcoma presenting as a complication of fibrous dysplasia, the first case was reported by Jaffe in 1958; a rate of 0.5 to 1% transformation has been published for this condition (13, 14).

Chondrosarcoma as a complication of Paget's disease has been found in 3% of cases (6). Other authors have reported rates ranging from 5 to 10%, and the mean survival period has been found to be 6 to 12 months (15, 16). The appearance of a chondrosarcoma after intravenous injection of Thorium dioxide is a rare condition but it has been previously cited in the literature (8-10).

On the basis of this small series of four secondary chondrosarcomas related to MOC, we can state that any patient suffering from a disease known to pose a risk of developing sarcomatous degeneration, who is between 20 and 40 years of age and presents with pain and a rapid increase in tumor size, is highly likely to have secondary chondrosarcoma.

The rarity of this condition in the absence of MOC has been confirmed by this study. In fact, only 4 out of 47 chondrosarcomas were secondary, all of them being related to MOC.

Although 2 of our cases have not been followed for a sufficient length of time, it appears that a complete local resection of this tumor is most important for treating this rare condition, in order to avoid local recurrence.

#### Acknowledgements

We wish to thank Carol Rowan for her help with the manuscript.

This article has been performed with a partial help of Fundacion Caja de Madrid.

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## SAMENVATTING

*E. C. R. MERCHAN, S. SANCHEZ-HERRERA en J. M. GONZALEZ. Secundair chondrosaroom. Beschrijving van 4 gevallen en overzicht van de literatuur.*

De auteurs beschrijven 4 gevallen van chondrosaroom, secundair aan multipele osteochondromatose. Deze laatste aandoening is erfelijk en familiaal ; de patiënten kunnen gevolgd worden vanaf de geboorte tot op volwassen leeftijd. Rekening houdend met de mogelijkheid van een maligne degeneratie, moet elke klacht of volumetoename van de tumor verder onderzocht worden, om een sarcomateuze evolutie uit te sluiten. Een brede resectie, indien mogelijk, is de eerste keuzebehandeling ter voorkoming van het risico van locale recidieven.

## RÉSUMÉ

*E. C. R. MERCHAN, S. SANCHEZ-HERRERA et J. M. GONZALEZ. Chondrosarcome secondaire. Présentation de 4 cas et revue de la littérature.*

Les auteurs décrivent 4 cas de chondrosarcome secondaire à une ostéochondromatose multiple. Cette dernière affection est familiale et héréditaire ; les malades peuvent être suivis de la naissance jusqu'à l'âge adulte. Vu le danger de dégénérescence maligne, des plaintes ou toute augmentation de volume des masses chondromateuses, justifient des examens complémentaires pour éliminer une évolution sarcomateuse. Le traitement de choix est la résection large, évitant le risque de récidence locale.