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ABSTRACT

Introduction: Extraskeletal myxoid chondrosarcoma (EMC) is a rare neoplasm that affects soft tissue and is independent from normal cartilage, bone and periosteum. Due to its low frequency, there is no clear consensus on its treatment.
Case Report: We present the case of a 39-year-old male with pain in plantar region of right foot of several months of evolution. After a Tru-Cut biopsy, the patient was diagnosed with EMC of the right foot and was performed an amputation below-knee.
Conclusion: EMC located on the foot is an extremely infrequent condition that has rarely been published. This article reviews the published literature to reach a consensus on the best approach for its treatment.

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Keywords: Chondrosarcoma, Extraskeletal, Extraskeletal myxoid chondrosarcoma (EMC), Foot

Carlos Cano Gala[1], Germán Borobio León[2], Roberto González Alconada[2], Laura Alonso Guardo[3], Diego A. Rendón Díaz[1], Francisco J. García García[1], Juan F. Blanco Blanco[4]

Affiliations: [1]MD, Resident, Department of Trauma and Orthopaedic Surgery, Health Center Complex, Salamanca, Spain; [2]MD, Physician, Department of Trauma and Orthopaedic Surgery, Health Center Complex, Salamanca, Spain; [3]MD, Physician, Department of Anesthesiology and Perioperative Medicine, Health Center Complex, Salamanca, Spain; [4]PhD, Chief of Department, Department of Trauma and Orthopaedic Surgery, Health Center Complex, Salamanca, Spain.

Corresponding Author: Carlos Cano Gala, C/ VilarFormoso 2-12, 33, Salamanca, Spain37008; Tel: +34653611309, Fax: +34923291100; Email: ccanogala@gmail.com

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INTRODUCTION

In the year 1972, Enzinger and Shiraki [1] defined the myxoid variant of extraskeletal chondrosarcoma, based on the multinodular growth of primitive chondroblast-like cells in an abundant myxoid matrix. The age at presentation of this neoplasm ranges from 4 to 92 years, and it mainly affects middle-aged patients. There is no sexual or racial predilection.

Histogenesis of EMC is controversial. In any case, chondroblast differentiation is a proven fact in several histochemical and ultrastructural studies.

In spite of the fact that, according to the reviewed literature, most cases appear on the lower limbs, location on the foot is very rare. In this article, we present the case of EMC of the foot and we review the cases published over the last years.

CASE REPORT

We present a case of a 39-year-old male who is admitted in our department after being referred from Primary Care, with pain on the plantar region of the right foot of several months of evolution. In the physical examination, the patient presents a hard immobile tumor of approximately 3–4 cm on the base of the fourth metatarsal bone. The rest of the physical examination is normal. An MRI is performed, and it reveals a solid lesion of 3.4x5.3x3 cm

in size, with necrotic areas inside, located on the plantar region and infiltrating different musculotendinous and bone structures in the area (Figure 1).

Further bone scan, chest X-ray and blood tests are normal.

A Tru-Cut biopsy is performed on the safest and most accessible area (dorsum of the base of the fourth metatarsal bone). The histopathological report describes a chondral origin tissue, with low cellularity and atypia, surrounded by abundant myxoid matrix. The specimen was unencapsulated. After a review of literature and confirmation that there is no clear consensus on an optimum treatment, an amputation below-knee is performed because it is the most functional choice with non-affected borders. The patient was not undergone to chemotherapy.

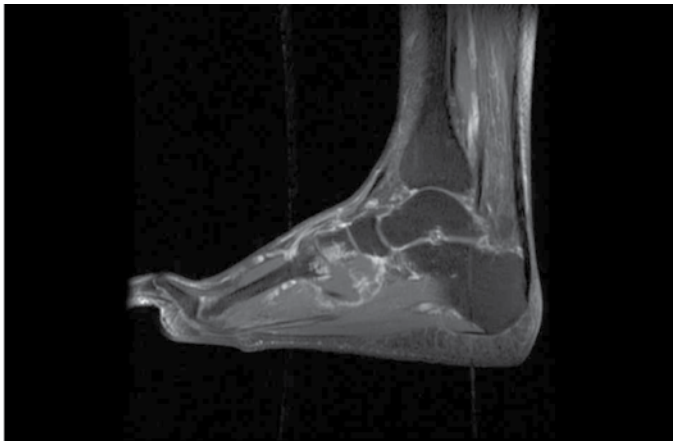

Figure 1: Magnetic resonance imaging scan showing the chondrosarcoma involving the base of the fourth metacarpal bone.

Table 1: Summary of cases of chondrosarcoma of the foot published in literature

Case	Sex	Age	Staging at diagnosis	Size (cm)	Cellularity	Treatment	Follow-up
1 [3]	M	50	NO	5	High	Local resection + extended resection + amputation	Disease-free/4 years
2 [6]	M	41	Lung metastasis	7	Low	Chemotherapy	Lung metastasis/14 years
3 [8]	W	78	NO	5	Low	Amputation	Disease-free/5 years
4 [8]	M	48	NO	9	Low	Amputation	Disease-free/6 years
5 [8]	M	51	NO	6.5	Low	Amputation	Disease-free/2 years
6 [7]	M	49	NO	6	Low	Extended resection	Chest wall metastasis/ Alive after 14 years
7 [7]	W	76	NO	8	Low	Extended resection	Disease-free/7 months
8 [7]	W	61	NO	6	Low	Amputation	Disease-free/4 months
9 [7]	W	43	NO	3	Low	Extended resection	Disease-free/8 years
10 [7]	M	32	NO	4	Low	Amputation	Disease-free/4 years
11 (Cano et al.)	M	39	NO	5	Low	Amputation	Disease-free/4 months

The postoperative evolution was favorable. Five months after surgery the patient clinical state is good and there is no evidence of distance metastasis in the monthly controls that are performed.

DISCUSSION

Extraskeletal myxoid chondrosarcoma is a rare entity that can be located both on the upper and the lower limbs, and is more commonly found on the thigh and the knee [2]. The foot is an extremely rare location for this pathology, and it receives different treatments depending on reviewed literature [3].

The age at presentation ranges from 4 to 92 years, and it is most common in midlife. There is no sexual or racial predilection.

Usually, patients present a clinical history of pain on the affected region and the appearance of a growing mass of months of evolution. The tumor grows very slowly and it is sometimes a casual finding, which can delay diagnosis significantly [4, 5].

Plain X-rays usually show a soft tissue mass that sometimes penetrates the underlying bone. The

diagnostic study is completed with MRI and an adequate staging study that includes a bone scan and an analysis with tumor markers. Diagnostic certainty is obtained with the anatomopathological study of a sample obtained either via biopsy or complete removal of the tumor.

There is no consensus in reviewed literature with regard to the optimum treatment. Table 1 gives an analysis of the cases of EMC of the foot found in the literature. As we can see, in most of them (63%), including our case, the patients underwent amputation (amputation level is not specified in the studies). In view of the data, both the patients who underwent amputation and those in whom the tumor was resected with free margins are alive and disease-free in the follow-up control studies that were carried out (with the exception of one case in which the patient maintains the lung metastasis of the initial diagnosis [6] and another case in which the patient developed a metastasis in the chest wall 36 months after diagnosis and who is still alive 14 years after the initial treatment [7].

The reviewed literature shows a relatively good prognosis of the tumor, with prolonged survival. This characteristic is not only applicable to cases of the foot; most studies report a good prognosis regardless of the location. Saleh et al., in a study from 1992 on 10 patients with EMC of different locations (one of them of the foot) question this good prognosis. The authors present a series in which, in spite of a long survival, 100% of the patients developed distant metastasis or local recurrence (many of them more than 10 years later). The only case of their series in which the location of the tumor was on the foot presented with lung metastasis at diagnosis, and the patient received chemotherapy as a single treatment (without response). Fourteen years later, the patient is still alive and the lung metastases are still present [6].

Our analysis reveals that most of the patients show long-term survival, even in the presence of metastasis. The low number of cases with location on the foot makes it difficult to discern clear prognostic factors. Oliveira et al. [8] refer to prognostic factors such as the size of the tumor, cellularity, mitotic activity, or the presence of different immunohistochemical markers, regardless of the location.

CONCLUSION

Extraskeletal myxoid chondrosarcoma (EMC) of the foot is an extremely rare condition with a relatively good prognosis even in the presence of distant metastasis, and which in most cases shows good results with amputation, which should be as functional as possible, as a treatment of choice. Larger case series are required to establish clear criteria with regard to prognostic factors.

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Author Contributions

Carlos Cano Gala – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
German Borobio Leon – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Roberto González Alconada – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Laura Alonso Guardo – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Diego A. Rendóndiaz – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Francisco J. García García – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Juan F. Blanco Blanco – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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