GSC Advanced Research and Reviews
eISSN: 2582-4597 CODEN (USA): GARRC2
Cross Ref DOI: 10.30574/gscarr
Journal homepage: https://gsconlinepress.com/journals/gscarr/

(CASE REPORT)

Extra skeletal myxoid chondrosarcoma of the ankle with pulmonary metastasis. Case report and a brief review of the literature

Anthony Bonilla [4], Nadine Akin [3], Amanda Canellas [4], Grace Perez [2], Sydni Rivera [2], Hadiyah Page [3], Marina Moussa [2], Jessica Jahoda [1, 2] and Mohamed Aziz [1, 5, *]

[1] *Research Writing and Publication (RWP), LLC, NY, USA.*
[2] *American University of the Caribbean School of Medicine, USA.*
[3] *Ross University School of Medicine, Barbados.*
[4] *St. George's University School of Medicine.*
[5] *Saint Vincent's Comprehensive Cancer Center, New York City, NY.*

GSC Advanced Research and Reviews, 2025, 24(01), 080-088

Publication history: Received on 21 May 2025; revised on 03 July 2025; accepted on 05 July 2025

Article DOI: https://doi.org/10.30574/gscarr.2025.24.1.0194

Abstract

Extra skeletal chondrosarcoma is a collective term for a group of malignant tumors that are not directly related to the bone or cartilage and is a diverse category. Primary chondrosarcomas typically arise in the skeleton, with a minority developing in an extra skeletal setting, which introduces challenges in diagnosis and classification. Of these, two entities are well-defined and characterized: extra skeletal myxoid chondrosarcoma (EMyC) and extra skeletal mesenchymal chondrosarcoma (EMC). While both are rare, they differ in their morphology and behavior, having little in common besides the myxoid matrix and soft tissue origin. EMyC has a wide differential diagnosis, including benign and malignant tumors. The rarity of EMyC poses a significant diagnostic challenge, both radiologically and pathologically.

We report the case of a 29-year-old healthy man who sought advice from his family doctor regarding a lump that had formed on his left ankle for almost a year. Imaging studies, tissue histomorphology, and immunohistochemical (IHC) studies supported the diagnosis of EMyC. The diagnosis was confirmed by the identification of the EWSR1-NR4A3 fusion gene through fluorescence in situ hybridization. The patient was treated with wide local excision and adjuvant radiotherapy. At 19 months postoperatively, three pulmonary metastases were found and effectively resected. At 26 months post metastasectomy, he remained asymptomatic with no recurrence. This case underscores the necessity of correlating radiologic, histologic, immunohistochemical, and molecular features for a definitive diagnosis of EMyC. It highlights the late metastatic potential of the tumor in the absence of metastases at presentation and after complete resection. Long-term follow-up is essential for monitoring patients with EMyC.

Keywords: Myxoid Chondrosarcoma; Extra Skeletal; Classification; Immunohistochemistry; Molecular; Metastasis

1. Introduction

MyC is a rare soft tissue neoplasm that has undergone substantial evolution in its classification and conceptual understanding since its original description. [1] [2] In the most recent WHO 2020 classification of chondrosarcoma, MyC is not classified as a cartilaginous tumor. Instead, it is categorized as a malignant soft tissue tumor and subcategorized as a tumor of uncertain differentiation. [2] EMyC is known as an uncommon malignant soft tissue tumor, contributing to remarkable difficulties for diagnosis and treatment in the primary health care setting. Unlike most chondrosarcomas, which originate from bony or cartilaginous tissues, EMyC originates from soft tissues and contains little to no true cartilaginous matrix, which makes EMyC paradoxically distinct from other cartilaginous tumors. [3] [5] This peculiar

* Corresponding author: Mohamed Aziz

histological feature, combined with its rather bland cytological appearance, occasionally leads to difficulties in differentiating it from either benign myxoid tumors or other soft tissue sarcomas. [4]

EMyC arises most commonly in the deep soft tissues of the extremities, with the thigh and leg being the most common sites; however, it may also occur at other sites, such as the trunk and retroperitoneum. The ankle area, as in our case, is not a common location of EMyC. [5] The tumor predominantly occurs in adults in the 4th-6th decades of life, and its occurrence in younger patients is uncommon. EMyC diagnosis is challenging due to morphologic diversity, and molecular confirmation is required by demonstrating a characteristic chromosomal translocation, specifically EWSR1-NR4A3 or TAF15-NR4A3 fusions. [6]

The treatment of EMyC is challenging due to its local recurrence and distant metastasis, which typically occur in the lung, as in our case. Wide surgical excision is the mainstay of the management, while adjuvant chemotherapy and radiotherapy are still evolving. Molecular diagnostics and targeted therapies have held promise for better outcome in patients in recent years. [7] [8]

We report a case of a 29-year-old male diagnosed with EMyC of the left ankle, illustrating the diagnostic dilemma and how new technologies can be applied to improve future diagnostic and treatment algorithms for this rare malignancy.

2. Case presentation

A 29-year-old male, never-smoker, was referred to the orthopedic clinic by his general practitioner due to a large mass over his left ankle. He had first noticed the mass approximately a year earlier and reported gradual growth over time. The mass had rapidly grown over the preceding months and had become more pronounced, leading to daily functional impairment. There was no history of trauma or constitutional symptoms, and his medical history was non-contributory. There was also no significant family history. Examination revealed a stiff, adherent, and slightly mobile mass of 6-7 cm in the deep soft tissues of the lateral aspect of the left ankle. No ulceration or discoloration was evident over the skin. The lesion was tender and well-demarcated, with no sensory or motor impairment.

The patient was referred to the surgery department for evaluation of the mass. The first radiographs showed an ill-defined soft tissue density. Further evaluation with computed tomography (CT) revealed a well-defined, lobulated soft tissue mass with areas of low attenuation and heterogeneous enhancement. These findings suggested a myxoid-rich component. MRI showed a multilocular, lobulated lesion with internal septae. The mass was isointense on T1-weighted sequences, with a few foci of hyperintensity due to hemorrhage. T2-weighted images demonstrated characteristically high signal intensity, reflecting the high-water content of the myxoid matrix. Following gadolinium administration, the lesion exhibited peripheral and septal enhancement. The radiological impression, particularly the T2 bright signal and multilobulated morphology, favored a diagnosis of EMyC.

Imaging differential diagnosis included synovial sarcoma, clear cell sarcoma, epithelioid sarcoma, and solitary fibrous tumor. Other less likely benign lesions included a large ganglion cyst, intramuscular myxoma, or a nerve sheath tumor with massive cystic degeneration. The mass's large size, deep location, lack of involvement with joint spaces or tendons, multilocular structure, and enhancement pattern all favored a diagnosis of EMyC. However, tissue confirmation was deemed essential. A core biopsy was taken. Histopathologic analysis from core needle biopsy exhibited a multinodular pattern of growth, separated by fine fibrous septa within an abundant myxoid matrix that contained mostly ovoid tumor cells. Tumor cells exhibited eosinophilic cytoplasm and displayed a polygonal to epithelioid morphology, forming cords, and scattered islands. Nuclei were bland and ovoid, with prominent nuclei and delicate chromatin. Many cells were plasmacytoid, with scattered multinucleated and spindled forms, focal hemorrhage, and occasional mitotic figures. Rare foci of necrosis were identified. (Figure 1 A, B, C)

IHC studies showed that the tumor cells were mostly positive for synaptophysin and showed focal reaction for NSE and S100. The Ki-67 proliferation index was high, with 30% nuclear staining. Stains for the epithelial markers, including cytokeratin AE1/AE3, CK19, EMA, CK5/6, and E-cadherin, were all negative. The immune profile was characteristic of EMyC, with overlapping features like those of other myxoid neoplasms, that required molecular confirmation. Molecular examination was carried out at the family's request. A FISH assay with EWSR1 and NR4A3 loci demonstrated the involvement of a t(9;22) (q22;q12) translocation, indicating the existence of EWSR1-NR4A3 gene fusion. Split signals for EWSR1 and NR4A3 were observed in 84% and 89% of nuclei, respectively, providing molecular confirmation of EMyC.

The staging workup was negative for metastasis or involvement of systemic organs. The patient's case was reviewed by a multidisciplinary sarcoma tumor board, which recommended wide local excision with wide negative margins and

adjuvant radiotherapy. The lesion was completely removed with 2 cm free margins. The mass measured 6.5 cm in size, with a lobulated appearance and a gelatinous, blue-gray hue, exhibiting foci of hemorrhage. Histological examination of the specimen confirmed the preoperative diagnosis. The patient finished adjuvant radiation therapy and was tracked via clinical follow-ups and surveillance imaging. Nineteen months postoperatively, imaging revealed three pulmonary nodules, the largest of which measured 3 cm in diameter. There was no evidence of disease at other sites. All metastatic nodules were resected. The patient declined chemotherapy and elected for observation. At 26 months following pulmonary metastasectomy, he remained clinically well and free of disease recurrence.

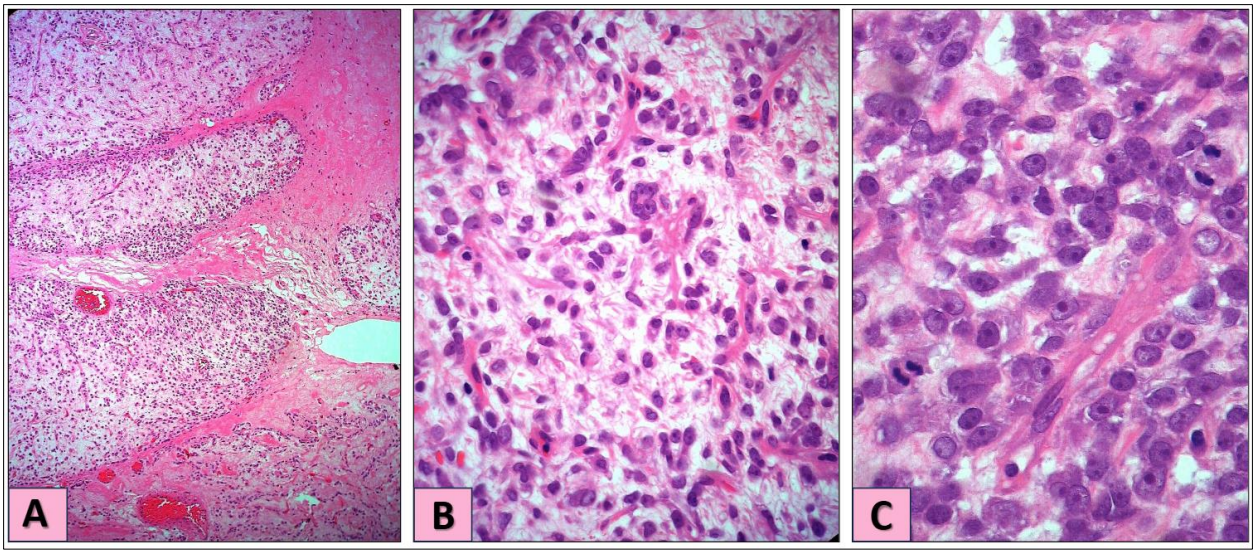

Figure 1 Histomorphology of the extra skeletal myxoid chondrosarcoma

- **1A** Low power view showing multinodular pattern of growth, separated by fine fibrous septa within an abundant myxoid matrix containing tumor cells (H&E stain X20)
- **1B:** Intermediate power view showing tumor cells with eosinophilic cytoplasm, polygonal, epithelioid, and plasmacytoid morphology, forming connecting cords (H&E stain X40)
- **1C:** High power view showing ovoid bland nuclei, with prominent nuclei, delicate chromatin, and occasional mitosis (H&E stain X60)

3. Discussion

3.1. History, epidemiology, and classification

EMyC is an uncommon soft tissue neoplasm that has been redefined in terms of its classification and relevant concepts since it was first reported. Enzinger and Shiraki first described EMyC in 1972. [3] EMyC was originally considered a myxoid soft tissue tumor characterized by cartilaginous differentiation, largely due to its myxoid stroma and infiltrating chondroblast-like tumor cells. [7] Nevertheless, the histogenesis of the tumor was a matter of controversy for many years [1] [2] [5], during which time it was uncertain whether it belonged to chondrosarcoma or a separate type of sarcoma with myxoid elements.

Historically, EMyC has been classified with general chondrosarcomas. The relationship of these tumors is frequently based on similar histologic morphology and nomenclature rather than similarity of molecular or developmental profiles. Nevertheless, there have been significant changes in this view due to developments in molecular genetics and immunophenotypic analyses. Unlike typical chondrosarcomas, which arise in clear cartilaginous sites and frequently contain IDH1/2 mutations, EMyC is defined by recurrent oncogenic chromosomal translocations, including the t(9;22)(q22;q12) creating an EWSR1-NR4A3 fusion gene. This individualistic therapeutic model and its characteristic clinical behavior eventually earned it a new designation. [6] [9]

The latest WHO classification of tumors of soft tissue and bone, 5th edition (2020), has incorporated this new knowledge. EMyC has been re-categorized under the tumors of uncertain differentiation in the soft tissue tumor category and is no longer reported with the cartilaginous tumors. [1] This shift in categorization is indicative that beyond its name, EMyC is not a genuine chondrogenic neoplasm and seems to function as a biologically related,

translocation-associated soft-tissue sarcoma. The reclassification also highlights the importance of molecular diagnostics in accurately diagnosing and managing these rare tumors. [1] [2]

From an epidemiological perspective, EMyC accounts for less than 3% of all soft tissue tumors. It commonly presents in adults aged 40–60 years and is more common in males, with a male-to-female ratio of 2–3:1. [5] EMyC can develop virtually anywhere in the body, but it prefers the deep soft tissues of the proximal extremities and limb girdles. More distal cases, for example, the ankle, were rarely reported [5], as was the case with our patient.

3.2. Spectrum of extra skeletal chondrosarcomas (EMyC)

EMyC is known to be the most frequent type of extra skeletal chondrosarcoma. It exhibits a multinodular pattern of growth, an abundant myxoid stroma, and centers of cords or free tumor cells, as well as hallmark gene fusions with NR4A3 (most commonly, the EWSR1-NR4A3 fusion). Even in the setting of slow growth, EMyC is well known to tend to local recurrence and hematogenous dissemination to distant organs, such as the lungs, often after a long time following the first treatment. [9] EMyC is the most frequent bona fide extra skeletal chondrosarcoma, unlike mesenchymal chondrosarcoma, which is rare in soft tissue. [10] Accurate diagnosis of these entities is largely based on a histopathologic and molecular correlation, especially in differentiating them from morphologic mimics.

Beyond EMyC and EMC, no other distinct primary extra skeletal chondrosarcoma subtypes are currently recognized in the established WHO classification. Rare conventional or clear cell chondrosarcomas may seem to be extraskeletal. However, these reported cases are quite exceptional and likely refer to direct extension from a primary skeletal location or may have been reclassified after more extensive molecular analysis. [11] Some tumors with chondroid differentiation, such as myxoid liposarcoma, chordoma, or even synovial sarcoma, can morphologically or radiologically mimic the EMyC, but they differ in histogenesis and molecular profile. Some reports have presented EMyC mimicking mucinous ovarian cancer (11), and others reported it clinically mimicking plantar fibromatosis. [10]

3.3. Clinical presentation, diagnostic workup, and differential diagnosis

In the presence of suspicion of EMyC, diagnosis is usually initiated by identifying its typical clinical presentation. EMyC usually presents in the deep soft tissues of the proximal limb and limb girdles, with the thigh and buttock as two of the most reported locations. Nonetheless, it can also occur in other sites, including deep soft tissue locations, like the upper extremities, trunk, and, least frequently, distal locations, such as the ankle, as seen in our patient. [12] [13]

The tumor is usually slow-growing and symptom-free, therefore most patients are asymptomatic for many years. As the lesion grows, regional pain, swelling, or limited motion may develop. Systemic symptoms are rare. The mass is typically firm, immobile, and deeply seated, with overlying intact skin and no neurovascular compromise unless the process is advanced. [9]

The diagnostic workup begins with imaging. Radiographs enable us to detect the presence of a soft tissue mass and whether it contains calcifications or not. CT scanning can be useful in defining the borders and internal architecture of the lesion, which typically shows a well-circumscribed, lobulated mass with regions of low attenuation representing the matrix component of the myxoid material. [3] MRI is the most informative modality. EMyC is usually lobulated and multilocular, with a high signal on T2-weighted images, a low to intermediate signal on T1-weighted images, and mild peripheral and septal enhancement after contrast administration. These imaging findings, particularly the high T2 signal, are suggestive but not pathognomonic. [14] PET-CT is not a standard procedure, but it could assist in staging or detecting metastasis. [15] The atypical ankle location of our patient's tumor, along with tumor behavior and imaging findings, provided the appropriate differential diagnosis. Precise diagnosis was, however, not possible without histopathological profiling and molecular validation.

Tissue sampling is essential, regardless of whether it is achieved through core needle biopsy or excisional biopsy. Macroscopically, EMyC appears as a lobulated, glistening, grey-blue tumor with occasional foci of hemorrhage or necrosis. Microscopically, the tumor is characterized by multinodularity throughout the field, with a morphology of an abundant myxoid matrix invested by tumor cells. The cells are usually small to medium-sized, round to polygonal or epithelioid, with cords or chains, and have bland, ovoid nuclei with finely dispersed chromatin and a low mitotic count. Plasmacytoid cells, spindled cells, and foci of hemorrhage may be seen. [12] These features were all appreciated in our case. IHC is useful to clarify the differential; however, it is not diagnostic. EMyC is usually reactive for synaptophysin diffusely and may stain for S100 and neuron-specific enolase (NSE) with variable results. It is negative for cytokeratins, EMA, and E-cadherin. The proliferation index (Ki-67) may vary but is commonly present in higher amounts in more aggressive tumors [3] [16], as in our case, which showed a Ki-67 index of 30%.

Molecular analysis is usually required for diagnosis confirmation. The most distinctive finding is the EWSR1-NR4A3 gene fusion, which is confirmed by fluorescence in situ hybridization (FISH) or reverse transcription-polymerase chain reaction (RT-PCR). There may be other rarer fusion partners; however, EWSR1-NR4A3 remains the most frequent and specific marker of EMyC. [6]. Table 1 summarizes the diagnostic features of EMyC and Selected Mimics.

Table 1 Diagnostic Features of Extra skeletal Myxoid Chondrosarcoma (EMyC) and Selected Mimics

Feature	**EMyC**	**Synovial Sarcoma**	**Myxoid Liposarcoma**	**Intramuscular Myxoma**	**Low-Grade MPNST**
Typical age	30–60 yrs	Adolescents and young adults	30–50 yrs	40–60 yrs	Any age
Common location	Deep soft tissue of proximal extremities	Para-articular regions	Thigh, retroperitoneum	Large muscle groups	Peripheral nerves
Growth	Slow, deep, painless mass	Variable, may be painful	Painless enlarging mass	Very slow growing	Variable
MRI T2 signal	Very high, lobulated	Intermediate-high	Very high, with septa	Uniformly high	Variable
Histology	Myxoid stroma, cords/nests of uniform cells	Biphasic or monophasic spindle cells	Myxoid background with lipoblasts	Hypocellular myxoid stroma	Spindle cells in fascicles
IHC	Synaptophysin+, S100±, NSE±, CK–	Cytokeratin+, TLE1+, EMA+	S100–, CD34–	S100–, CD34+	S100+, SOX10+
Molecular	EWSR1-NR4A3	SS18-SSX fusion	FUS-DDIT3 or EWSR1-DDIT3	No fusion	Often NF1 alterations
Distinguishing feature	NR4A3 fusion, indolent but metastasizing	TLE1+/SSX fusion	Lipoblasts, DDIT3 fusion	Benign, low cellularity	Nerve origin, S100+

3.4. Treatment of EMyC and the expected prognosis in localized versus metastatic disease?

After diagnosis, the treatment is predominantly surgical, and wide local excision with negative margins is essential. Adjuvant radiotherapy is widely used, particularly for larger tumors or those with narrow margins. There is limited efficacy of chemotherapy, and systemic therapy generally is withheld for unresectable or metastatic disease. Sustained follow-up is necessary due to the potential risk of late recurrences and distant metastases, primarily to the lungs. [13] [17]

EMyC has a prognosis that differs from that of other soft tissue sarcomas. Given this, it presents a paradoxically high rate of local recurrence and hematogenous metastasis, primarily to the lungs, as in our patient. [12] Therefore, the need for lifelong follow-up in all patients is important, regardless of the apparent success of the therapeutic strategy. [18] The prognosis is usually favorable when the tumor is localized at the time of diagnosis and when the tumor is completely excised with free margins. [17] The atypical ankle location of our patient's tumor, along with tumor behavior and imaging findings, provided the appropriate differential diagnosis. Precise diagnosis was, however, not possible without histopathological profiling and molecular validation. [12]

This case illustrates the role of multimodality therapy in rare sarcomas. In descriptive studies, effective diagnosis, treatment planning, and follow-up were achieved with the support of radiology, pathology, surgery, molecular diagnostics, and oncology. The patient in our case has sustained a durable remission from surgery alone after metastatectomy, highlighting the necessity for personalized risk-benefit assessment and patient-centered decision-making; for example, our patient declined post metastatectomy chemotherapy and elected observation. He remained tumor free for 29 months before he was lost to follow-up.

The occurrence of pulmonary metastases, frequently several years after primary therapy, represents a distinguishing feature of EMyC and accounts for its distinctive clinical behavior. Distant metastases are reported to develop in approximately 50% of cases, with the lungs being the most affected, followed by bones and, less frequently, soft tissues. [19] It is also important to recognize that the presence of pulmonary metastasis is not equivalent to a rapid prognosis

since many patients have a long-term stable disease even in the absence of systemic treatment. [19] However, in selected patients, even with metastasis, especially those with lone resectable lung metastasis or limited systemic spread, chemotherapy can achieve independent long survival and durable control. [17]. Table 2 summarizes the prognostic factors in EMyC.

Table 2 Prognostic Factors in Extra skeletal Myxoid Chondrosarcoma (EMyC)

Prognostic Factor	**Favorable**	**Unfavorable**
Tumor size	≤5 cm	>5–7 cm
Surgical margins	Negative (R0)	Positive (R1/R2)
Anatomic location	Extremities	Retroperitoneum, axial sites
Ki-67 index	Low (<10–15%)	High (>20–30%)
Histologic grade	Low cellularity, bland nuclei	Increased mitoses, pleomorphism
Molecular subtype	EWSR1-NR4A3	Rare/variant fusions
Response to metastasectomy	Isolated pulmonary nodules, complete resection	Diffuse metastatic spread
Follow-up interval	Long-term surveillance	No follow-up/early discharge

3.5. With advancing technology, what does the future hold for the diagnosis and treatment of EMyC?

Given the rapid pace at which diagnostic and therapeutic methods are evolving, the treatment of EMyC will likely become more personalized. One of the most recent advances is the integration of molecular diagnostics into daily practice as part of personalized therapy for sarcomas. In addition to confirming this definition of EMyC, the identification of EWSR1-NR4A3 and other unusual fusion transcripts has provided a target for study in the biology and oncology of this tumor. (6)

Additionally, in the future, NGS sequences and RNA fusion panels will also identify new or variant fusion partners that may correlate with certain genotypes and different prognoses, therapeutic responses, or patterns of metastasis. Furthermore, with a continued understanding of the molecular pathways mediated by NR4A3 fusions, inhibitors of these pathways, such as transcriptional modulating agents or targeted inhibitors, are potential therapeutic options for metastatic or non-resectable disease. (20)

On the therapeutic front, there is increasing excitement in profiling the immune and microenvironmental landscape of sarcomas, including EMyC, which may allow for the identification of a subset of patients who will derive benefit from immunotherapy or other novel biologics. (21) (22) The management of EMyC is likely to be more personalized in an era where the pace of diagnostic and therapeutic developments is rapid. One of the most significant recent developments is the widespread adoption of molecular diagnostics in personalized therapeutic approaches to sarcomas. (9) Although systemic therapy has generally played a minor role for EMyC patients, precision oncology trials could provide alternatives to cytotoxic therapy in the future, especially for those with molecularly defined susceptibilities. (23) There is also the ongoing development of functional imaging techniques, such as PET-MRI. As testing technologies continue to evolve, the diagnostic and therapeutic paradigms for EMyC are expected to progress toward increasingly molecular, precise, and preemptive strategies. (24)

3.6. What did we learn from this case?

Multiple teaching points can be gleaned from this case regarding the evaluation and management of EMyC. It has valuable educational messages to offer; it reminds us that we should not ignore rare tumors in the differential diagnosis, especially when we encounter deep, slow-growth, and painless soft-tissue masses in a young adult with no history of trauma or systemic disease. Our patient's atypical ankle site, in association with imaging features and behavior of the tumor, enabled us to narrow our differential diagnosis. The exact diagnosis, however, required histopathological profiling and molecular confirmation.

While EMyC is, in general, regarded as low-grade, it is not biologically silent. Late metachronous metastases, with a predominant lung localization, can occur and justify long-term follow-up and information to the patient about a possibly chronic disease. In presenting this case, we aim to increase understanding of EMyC clinicopathology and contribute to an ongoing discussion of the nuances of treating rare, molecularly defined malignancies globally.

4. Conclusion

Extra skeletal myxoid chondrosarcoma (EMyC) is an uncommon soft tissue sarcoma with distinctive histological and molecular features that can be diagnostically challenging. Its seemingly benign nature, despite its risk for local recurrence and late pulmonary metastatic tendencies, is a false impression. Our case illustrates the importance of multimodality imaging, detailed histopathologic and molecular characterization, and the central role of multidisciplinary collaboration in providing optimal patient care. With the emergence of molecular diagnosis and targeted therapy, the future is set to receive more accurate diagnosis and probably more effective systemic therapy for this rare tumor. This case highlights the need for lifelong surveillance among the EMyC population. It is illustrative that a personalized, evidence-based treatment can yield superior results even when metastasis appears late in the disease development.

Compliance with ethical standards

Acknowledgments

Special thanks to MD candidates Harrison Atanga and Sabrina Escobar for their assistance in reviewing the final manuscript.

Disclosure of conflict of interest

The following declarations are made by all authors.

Statement of ethical approval

Ethical review and approval were not required for this study involving human participants. The paper has been sufficiently anonymized to maintain the patient's confidentiality.

Statement of informed consent

The patient was lost to follow-up, and all attempts to reach the family members were unsuccessful. Therefore, the paper has been sufficiently anonymized to maintain patient confidentiality.

Payment/services information

All authors have declared that they received no financial support from any organization for the submitted work.

Financial relationships

All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might be interested in the submitted work.

Other relationships

All authors declare that no other relationships or activities could be perceived as influencing the submitted work.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript

References

[1] Murphey MD, Kransdorf MJ. Staging and classification of primary musculoskeletal bone and soft-tissue tumors according to the 2020 WHO update, from the AJR special series on cancer staging. American Journal of Roentgenology. 2021 Nov 14;217(5):1038-52.

[2] Sbaraglia M, Bellan E, Dei Tos AP. The 2020 WHO classification of soft tissue tumours: news and perspectives. Pathologica. 2020 Nov 3;113(2):70.

[3] Zhang L, Wang R, Xu R, Qin G, Yang L. Extraskeletal myxoid chondrosarcoma: a comparative study of imaging and pathology. BioMed research international. 2018;2018(1):9684268.

[4] Fice MP, Lee L, Kottamasu P, Almajnooni A, Cohn MR, Gusho CA, Gitelis S, Blank AT. Extraskeletal myxoid chondrosarcoma: a case series and review of the literature. Rare Tumors. 2022 Feb 11; 14:20363613221079754.

[5] Wagner MJ, Chau B, Loggers ET, Pollack SM, Kim TS, Kim EY, Thompson MJ, Harwood JL, Cranmer LD. Long-term outcomes for extraskeletal myxoid chondrosarcoma: a SEER database analysis. Cancer Epidemiology, Biomarkers and Prevention. 2020 Nov 1;29(11):2351-7.

[6] Paioli A, Stacchiotti S, Campanacci D, Palmerini E, Frezza AM, Longhi A, Radaelli S, Donati DM, Beltrami G, Bianchi G, Barisella M. Extraskeletal myxoid chondrosarcoma with molecularly confirmed diagnosis: a multicenter retrospective study within the Italian sarcoma group. Annals of surgical oncology. 2021 Feb; 28:1142-50.

[7] Hisaoka M, Hashimoto H. Extraskeletal myxoid chondrosarcoma: updated clinicopathological and molecular genetic characteristics. Pathology international. 2005 Aug;55(8):453-63.

[8] Stacchiotti S, Baldi GG, Morosi C, Gronchi A, Maestro R. Extraskeletal myxoid chondrosarcoma: state of the art and current research on biology and clinical management. Cancers. 2020 Sep 21;12(9):2703.

[9] Chiusole B, Le Cesne A, Rastrelli M, Maruzzo M, Lorenzi M, Cappellesso R, Del Fiore P, Imbevaro S, Sbaraglia M, Terrier P, Ruggieri P. Extraskeletal myxoid chondrosarcoma: clinical and molecular characteristics and outcomes of patients treated at two institutions. Frontiers in Oncology. 2020 Jun 16; 10:828.

[10] Saraggi D, Salmaso R, Montesco MC, Zamuner C, Tiengo C, Rugge M, Fassan M. Extraskeletal myxoid chondrosarcoma of the foot clinically mimicking plantar fibromatosis. The American Journal of Dermatopathology. 2017 Mar 1;39(3): e38-40.

[11] Güler Çekiç S, Atay ÖF, Ülker V, Akbayir Ö. Extraskeletal myxoid chondrosarcoma mimicking mucinous ovarian cancer: A case report. Journal of Obstetrics and Gynaecology Research. 2025 Feb;51(2):e16235.

[12] Gusho CA, King D, Ethun CG, Cardona K, Harrison Howard J, Tran TB, Poultsides G, Tseng J, Roggin KK, Fields RC, Cullinan DR. Extraskeletal myxoid chondrosarcoma: Clinicopathological features and outcomes from the United States sarcoma collaborative database. Journal of surgical oncology. 2022 Dec;126(8):1533-42.

[13] Brown JM, Rakoczy K, Pretell-Mazzini J. Extraskeletal myxoid chondrosarcoma: clinical features and overall survival. Cancer treatment and research communications. 2022 Jan 1; 31:100530.

[14] Tateishi U, Hasegawa T, Nojima T, Takegami T, Arai Y. MRI features of extraskeletal myxoid chondrosarcoma. Skeletal radiology. 2006 Jan; 35:27-33.

[15] Antonescu CR, Argani P, Erlandson RA, Healey JH, Ladanyi M, Huvos AG. Skeletal and extraskeletal myxoid chondrosarcoma: a comparative clinicopathologic, ultrastructural, and molecular study. Cancer: Interdisciplinary International Journal of the American Cancer Society. 1998 Oct 15;83(8):1504-21.

[16] Zhang L, Hashimoto H. Extraskeletal myxoid chondrosarcoma: updated clinicopathological and molecular genetic characteristics. Pathology international. 2005 Aug;55(8):453-63.

[17] Ulucaköy C, Atalay İB, Yapar A, Kaptan AY, Bingöl I, Doğan M, Ekşioğlu MF. Surgical outcomes of extraskeletal myxoid chondrosarcoma. Turkish Journal of Medical Sciences. 2022;52(4):1183-9.

[18] Masrouha K, Multani I, Bhatt O, Ghert M. Extraskeletal Myxoid Chondrosarcoma: Long-Term Survival in the Setting of Metastatic Disease. Case Reports in Orthopedics. 2020;2020(1):2684746.

[19] Paoluzzi L, Ghesani M. Extraskeletal myxoid chondrosarcoma with massive pulmonary metastases. Clinical Sarcoma Research. 2018 Dec; 8:1-6.

[20] Zou T, Sethi R, Wang J, Budak G, Chandran U, John I, Watters R, Weiss K. Whole genome sequencing for metastatic mutational burden in extraskeletal myxoid chondrosarcoma. Frontiers in Molecular Medicine. 2023 Jul 24; 3:1152550.

[21] Oiseth SJ, Aziz MS. Cancer immunotherapy: a brief review of the history, possibilities, and challenges ahead. Journal of cancer metastasis and treatment. 2017 Oct 31; 3:250-61.

[22] Ogata R, Soda H, Senju H, Fujioka M, Shimada M, Yamashita K, Irifune S, Tagawa R, Dotsu Y, Iwasaki K, Taniguchi H. Immunosuppressive tumor microenvironment in extraskeletal myxoid chondrosarcoma: A case of pleural metastases. Thoracic Cancer. 2022 Oct;13(19):2812-6.

[23] Rulten SL, Grose RP, Gatz SA, Jones JL, Cameron AJ. The future of precision oncology. International Journal of Molecular Sciences. 2023 Aug 9;24(16):12613.

[24] Yan Q, Yan X, Yang X, Li S, Song J. The use of PET/MRI in radiotherapy. Insights into Imaging. 2024 Feb 27;15(1):63.