Extraskeletal Myxoid Chondrosarcoma: A Case Report

Hafsae Bounniyt¹, Majda Askour¹, Mouna Rimani², Laila Benzekri¹, Nadia Ismaili¹, Karima Senouci¹ and Badredine Hassam¹

¹Department of Dermatology Venerology, Faculty of Medicine and Pharmacy, University Mohammed V Souissi, Rabat, Morocco
²Department of Anatomical Pathology Hassan, Rabat, Morocco

Corresponding author: Hafsae Bounniyt, Department of Dermatology Venerology, Faculty of Medicine and Pharmacy, University Mohammed V Souissi, Rabat, Morocco, Tel: +212662395350; E-mail: b.hafsae@hotmail.fr

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Abstract

Extraskeletal myxoid chondrosarcoma (EMCS) is a rare malignant soft tissue tumor that usually develops in the deep parts of the proximal extremities and limb girdles in middle-aged adults. Its biologic behavior and pathogenesis are poorly understood. EMCS has a prolonged and indolent clinical course, with a high rate of local recurrences and distant metastases before tumor-related death. The diagnosis and management of this tumor must be early and multidisciplinary.

Keywords: Extraskeletal myxoid chondrosarcoma; Local recurrences; Lung metastases; Bone metastases

Introduction

Extraskeletal myxoid chondrosarcoma is a distinctive soft tissue sarcoma; it accounts for approximately 2.5% of soft tissue sarcomas, commonly occurs in middle-aged adults, and arises mainly in the proximal extremities and limb girdles. We report a case of a patient with local and metastatic recurrences of fatal evolution [1].

Case Report

A 62 years old female patient, with no particular history, who complained in 2003 from a painless nodule at the right forearm progressively increasing in size. Excision was performed but no histological study was done. Two years later, the nodule developed again and was resected by a traumatologist. The histological study this time was in favor of an epithelioid sarcoma. The patient was lost of sight and revisited the doctor two months later with a second tumor recurrence at the same site. She was addressed to our department for medical care.

In order to confirm the diagnosis of cutaneous sarcoma, a cutaneous biopsy with an immunochemical study was carried out, with re-reading of the old block of the second excision. Histological examination showed a tumor proliferation of lobules within a hypovascularized chondromyxoid substance. Numerous mitoses have been noted, as well as extensive areas of necrosis. In the immunochemical staining on deparaffinated sections showed positivity to PS100, cytokeratin and EMA antibodies (Figure 2). Moreover, tumor cells were negative to CD34 antibody. We performed a cytogenetic examination, the karyotype was t(9;17).
Extraskeletal myxoid chondrosarcoma is a soft tissue sarcoma that was first described by Stout and Verner in 1953 [1] and formally defined as a distinctive entity by Enzinger and Shiraki in 1972 [2]. It is a very rare entity representing less than 3% of all soft tissue sarcomas [3]. It mostly affects adult patients with a median age of 50 years while it is considered extremely rare in childhood and adolescence where approximately 20 cases have been described in the literature [4].

Clinically, most patients present with a palpable frequently painful mass in the extremities or the trunk. Most tumors develop in the soft tissues, and only a minority in the subcutaneous tissues. According to a recent Chinese study of 40 patients, 60% of the tumors arose within the deep soft tissues of the lower limbs followed by the upper limbs (20%) then the trunk (10%) [3].

On imaging, magnetic resonance imaging is the gold standard for local extension assessment in the preoperative setting [5]. Although the radiological characteristics are not specific in this tumor because of the presence of myxoid component, presence of spetsas and sometimes necrotic degenerescence can guide the presumption of histological diagnosis. A circumscribed lobulated hyperintense mass with hypointense intralesional fibrous septas on T2MRI may give a hint of EMCS.

Up until now, wide local excision remains the standard of care for EMCS. After excision, the tumor is generally well delineated and ovoid with several septa-ting a multinodular appearance. There are often cavities containing mucous material, hemorrhagic foci and necrosis. The size is variable (median 7 cm) [6].

Histologically, classic EMCs are characterized by hypocellular myxoid nodules separated by fibrous septas of variable thickness. Tumor cells are elongated polygonal and of intermediate size with a dark pink cytoplasm. The nucleus is somewhat eccentric and contains a small nucleus. Some mitoses are visible on examination. There is sometimes a prospective cytoplasmic enlightenment giving a rhabdoid or vaguely plasmacytoid aspect. The tumor cells are arranged in coronas, sometimes interanastomosed or in more or less dense plaques, in a poor myxoid matrix. There is no cartilage matrix of the hyalin type. The morphology is rather characteristic and the diagnosis is generally easy to do if one knows the entity. Some cases deviate from this stereotyped description, with a high cellularity, more pleomorphism and more mitoses. Immunohistochemistry is not very useful in diagnosis. Some labeling with S100, CD117, NSE and synaptophysine is described, but keratins are generally negative or very weakly and focally positive and S-100 positivity is found in approximately 40% of myxoid liposarcomas, and 50% of EMCs. Thus, S-100 does not help distinguish myxoid liposarcomas from EMC [7]. Therefore a cytogenetic study is performed. EMCS is a distinct sarcoma characterized by recurrent chromosomal translocations, typically t(9;22) (q22;q12.2), fusing EWSR1 to NTRK3 [8].

The histological differential diagnosis includes: soft tissue chondroma, muciparemetyastic carcinoma, soft tissue myoepithelioma (myxofibro) high grade sarcoma/malignant fibrous histiocytoma, chor-dome, myxoid liposarcoma, extrarenal rhabdoid tumor. Detection of a rearrangement of NTRK3 by FISH court-PCR is useful in cases where the morphological diagnosis is unclear [9].

To date, surgery remains the mainstay of treatment for localized disease. Wide local excision is the treatment of choice. Neo-adjuvant radiation therapy may be indicated to reduce the tumor size and enable surgery.

Generally, adjuvant chemotherapy after complete resection is not indicated, although anthracycline-based chemotherapy was shown to be active in one report [10].

EMCS is known for its high recurrent and metastatic potential. According to the Chinese series, the local recurrence rate ranged from 37% to 48% with a median time of 3.3 to 3.5 years. 42.5% of patients had a single recurrence, whereas 57.5% of patients had repeated local recurrences (range from 2 to 7) over as long as 10 years. The metastatic rate ranged from 26% to 46% with a median time of 2.5 to 3.2 years. Metastatic disease could be diagnosed up to 14 years after first diagnosis. Approximately 31% of patients presented first with revealing metastases before the primary tumor were identified. The most common metastatic site was the lung, followed by lymph nodes, bones, soft tissues, and brain. Rare sites of metastasis such as liver, pancreas, and testis have been recently reported. The overall survival at 5, 10, and 15 years was 82% to 91%, 65% to 78%, and 58% to 78%, respectively [3].

Recently, a case of spontaneous regression of metastatic EMCS has been reported in a 25-year-old woman who presented with multiple bilateral pulmonary metastases [11].

It has been shown that clinical parameters such as large tumor size (N 10 cm), proximal location, and presence of metastatic disease are strongly adverse prognostic factors [3].

Conclusion

Extraskeletal myxoid chondrosarcoma belongs to soft tissue sarcomas. It is known for long clinical course and high rates of local recurrence and metastatic spread mainly to the lung and bones. Surgery remains the gold standard of care for local tumors.
**Conflict of Interest**

None.

**References**