

Extraskelatal Myxoid Chondrosarcoma: A Case Report

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Abstract

Extraskelatal 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: Extraskelatal myxoid chondrosarcoma; Local recurrences; Lung metastases; Bone metastases

Introduction

Extraskelatal 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. The dermatological examination at admission found a polylobed painless subcutaneous tumor of the right forearm, sitting on the scar of the old excision, measuring 5 cm with superficial telangiectasia (Figure 1A). It was hard and fixed to neighboring muscle tissues but respects the deep plane. A similar lesion was noted at the forehead measuring 1.5 cm in diameter (Figure 1B). The rest of the clinical examination showed no other abnormalities.

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 an abundant basophilic and hypovascularized chondromyxoid substance. Tumor cells arranged in small bays, cords or islands (Figure 1).



Figure 1: (A) Polylobed subcutaneous tumor of the right forearm, sitting on the scar of the old excision. (B) Nodular lesion in the forehead measuring 1.5 cm in diameter.

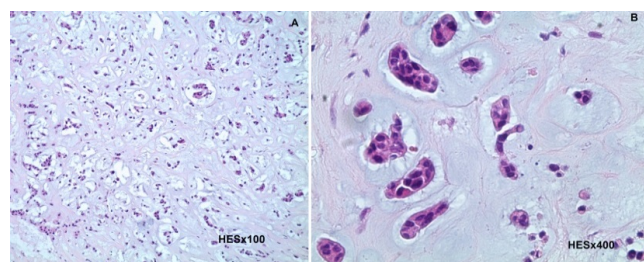


Figure 2: Histological examination (A) HES Coloration x100 magnification, proliferation of cuboid cells in spans and islets within an abundant chondroid substance. (B) HES Coloration x400 magnification. Proliferation of cuboid cells in spans and islets within an abundant chondroid substance with sometimes pseudoacinar structures.

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)

(q22;q11), thus the diagnosis of a high-grade extraskelletal myxoid chondrosarcoma of FNCLCC was retained (Figure 3).

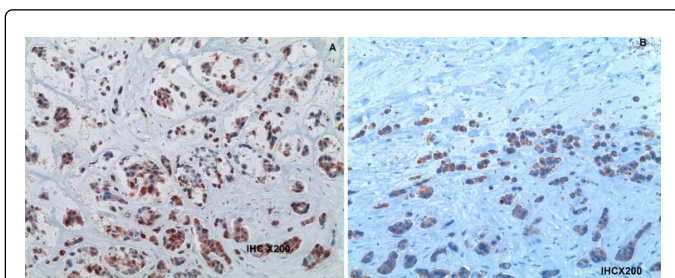


Figure 3: Immunohistochemical examination, x200 magnification. (A) Expression of protein S100. (B) Expression of Cytokeratine CK (AE1, AE3).

The staging imaging included chest, abdomen and pelvic computed tomography (CT) as well as a bone scintigraphy. It showed pulmonary and bone metastases at the humeral head, scapula and iliac bone. Axillary ultrasound found suspicious lymphnodes.

Medical record was discussed in a multidisciplinary consultation meeting; palliative chemotherapy was indicated. The patient was died two months after the initiation of treatment.

Discussion

Extraskelletal 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 septas and sometimes necrotic degeneration can guide the presumption of histological diagnosis. A circumscribed lobulated hyperintense mass with hypointense intralobular fibrous septas on T2MRI may give a hint of EMCs.

Up until now, wide local excision remains the standard of care for EMC. 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 plasmocytoid 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 NR4A3 [8].

The histological differential diagnosis includes: soft tissue chondroma, muciparemetastatic carcinoma, soft tissue myoepithelioma (myxofibro) high grade sarcoma/malignant fibrous histiocytoma, chor-dome, myxoid liposarcoma, extrarenal rhabdoid tumor. Detection of a rearrangement of NR4A3 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].

EMC 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

Extraskelletal 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.

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