POSTER 42

Extraskeletal Myxoid Chondrosarcoma: Retrospective Case Series Examining Prognostic Factors, Treatment Approaches, and Oncologic Outcomes

Casey Brodsky, PhD, *University of Michigan Medical School*, brodskca@med.umich.edu
Matthew Bucala, BA, *University of Michigan School of Public Health*, mbucala@umich.edu
Geoffrey Siegel, MD, *University of Michigan Department of Orthopedic Surgery*, gsiegel@med.umich.edu

Background: Extraskeletal myxoid chondrosarcoma (EMC) is an ultra-rare soft tissue sarcoma, and a limited number of studies are published concerning the clinical course of the disease and efficacy of treatment. As a result, there remain outstanding questions about how to best manage patients with EMC tumors.

Purpose: The goal of this retrospective case series of EMC patients seen at the University of Michigan Sarcoma Center is to characterize trends in treatment and outcomes to help inform future EMC management.

Patients and Methods: EMERSE database was used to identify all patients seen at the University of Michigan Sarcoma Center between 1998 and 2021 with the term "extraskeletal myxoid chondrosarcoma" in their medical record. A list of 91 patients was generated, and a retrospective chart review was performed to verify each patient's final pathologic diagnosis. Exclusion criteria included patients with a diagnosis other than EMC or inconclusive pathology. Chart review further identified demographics, tumor characteristics, treatments, and outcomes.

Results: A final list of 45 EMC patients were identified. Median follow-up from date of diagnosis was 48.9 months (range 1.2 – 340.1). Median age at diagnosis was 56.9 (range 25 – 79), 36 patients (80%) were male, and 9 patients (20%) were female. Tumor characteristics are summarized in Table 1. Twenty-six patients (58%) had metastatic disease; 10 had metastases at diagnosis and 16 recurred with or progressed to metastatic disease. Metastases were most common in the lungs (77%). Management approaches used are summarized in Table 2. Patients receiving curative-intent surgery were considered evaluable for recurrence if they had post-surgery follow-up greater than 6 months (n = 26, 79%). Sixteen of these patients (62%) experienced a recurrence, 4 local (25%) and 12 metastatic (75%), with median time to recurrence 24.2 months (range 2.2 – 163.7). Fisher's exact test was performed to determine prognostic significance of several factors on recurrence rate; tumor grade 1 vs. 2 was the only significant factor identified ($P = 0.0278$). Kaplan-Meier survival analysis was performed: 5-year overall survival was 80% for all patients, 86% for locoregional disease, and 58% for metastatic disease; for locoregional disease, 1-year disease-free and metastasis-free survival post curative-intent surgery were 71% and 69%, respectively; 1-year progression-free survival for metastatic disease from start of 1L systemic therapy was 35%.

Table 1: EMC tumor characteristics

Tumor characteristics (n = 45)	
Primary tumor site	
Upper extremity	11 (24%)
Lower extremity	26 (58%)
Axial body	6 (13%)
Unknown	2 (4%)
Primary tumor size in cm (n = 38)	7.9 (1.5, 27)
Histologic grade	
Grade 1	16 (36%)
Grade 2	9 (20%)
Grade 3	5 (11%)
Not reported	15 (33%)
Stage at diagnosis	
Localized	34 (76%)
Regional lymph node metastasis	1 (2%)
Distant metastasis	10 (22%)

Table 2: Management approaches for EMC patients

Locoregional disease management (n = 35)	
Surgery	33 (94%)
Local resection	30 (91%)
Amputation	3 (9%)
(Neo)adjuvant chemotherapy	3 (9%)
(Neo)adjuvant radiation therapy	20 (61%)
No surgery (radiation therapy only)	2 (6%)
Metastatic disease management (n =26)	
Non-curative intent surgery	13 (50%)
(Neo)adjuvant chemotherapy	2 (8%)
Radiation therapy	13 (50%)
(Neo)adjuvant radiation	5 (38%)
Standalone/palliative radiation	8 (62%)
1L systemic therapy	22 (85%)
2L systemic therapy	15 (58%)
3L systemic therapy	12 (46%)

Conclusions: EMC is not as indolent as has been historically thought; these data demonstrate a high recurrence rate, high propensity for metastasis, and high rate of progression of metastatic disease on systemic therapy. Work is ongoing to perform univariate and multivariate analysis of the prognostic significance of patient factors and treatment approaches on survival outcomes. Potential study limitations include small sample size and referral bias toward more advanced disease at a quaternary care center.