Abstract #11041

Dedifferentiated Chondrosarcoma: Survival Analysis of 179 Cases from the SEER Database

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Background: Dedifferentiated chondrosarcoma is a rare malignancy with a poor prognosis. Although universally poor, survival rates are noted to range from 0%-29% at five years. Current literature consists of mostly small series and is therefore quite limited with regard to specific factors affecting survival.

Questions/Purposes: The aim of this investigation is to analyze a national sampling of cases of dedifferentiated chondrosarcoma encountered during an eleven year time period to examine survivorship characteristics of this disease with relation to patient and tumor characteristics, surgical treatment factors, and overall and disease-specific survival.

Patients and Methods: To identify our patient series we performed a retrospective review of the Surveillance, Epidemiology, and End Results (SEER) database. One hundred seventy nine patients were identified in the SEER database from 2001 to 2011. We excluded patients who had incomplete data with regard to tumor location, survival, and who were classified as both "dedifferentiated" and low grade (AJCC Stage 1) histology. Information regarding the demographic and clinical characteristics, the histological features and grade of the tumors, the location and size of the tumors, the surgical stage at the time of diagnosis, the use of surgery and radiation treatment, and survival were recorded. Kaplan Meier survival analysis was performed for overall disease and disease specific survival. Additionally, univariate and multivariate analysis was performed to identify prognostic factors.

Results: Overall and disease specific survival for our patient cohort was 18% and 28%, respectively, at 5 years. Individuals with appendicular tumors tended towards a worse prognosis than individuals with axial, chest wall, and soft-tissue tumors (Figure 1). Patients with a primary chest wall tumor were 0.27 (95% CI: 0.10 – 0.77) times as likely to die from disease as those who had a primary lower extremity tumor (p=.01). Individuals with grade III or IV AJCC were more than twice as likely to die from disease at any given time compared to individuals with grade II AJCC (p = <.01). Those with larger tumors (i.e., >8 cm) were 2.17 (95% CI: 1.11 – 4.27) times more likely to die from disease at any given time than were individuals with small tumors (i.e., <= 8cm). Individuals presenting with metastatic disease at diagnosis were 4.05 (95%CI; 2.25 – 7.29) times more likely to die from disease compared to those with localized disease (p < .001). Nineteen patients had no surgical intervention and these patients were found to have a significant increase in mortality when examining overall survival compared to the groups treated with surgery (p < .01). Our results are summarized in table 1.

Conclusions:
The overall prognosis of dedifferentiated chondrosarcoma remains poor with less than an third of patients surviving to five years from diagnosis. Tumor size >8cm and presence of metastases at diagnosis were significant unfavorable prognostic factors. Patients with a primary tumor located in their chest wall had a better prognosis. Patients treated surgically were approximately 50% less likely to die from disease compared to those treated without surgery.

Table 1: Time to disease specific death as a function of univariable demographics and comorbidities

	***HR* (95% CI)**	***p***
> 60 years (versus =<60 years)	1.30 (0.83 – 2.03)	.26
Primary Tumor Site		.09
Axial (versus lower)	0.75 (0.43 – 1.32)	.32
Chest Wall (versus lower)	0.22 (0.07 – 0.72)	.01
Upper (versus lower)	0.85 (0.50 – 1.44)	.54
AJCC Stage		.003
III or IV (versus II)	2.72 (1.49 – 4.97)	.001
Unknown (versus II)	1.20 (0.72 – 2.00)	.48
Surgical Treatment		.16
Amputation (versus No Surgery)	0.54 (0.24 – 1.19)	.13
Limb Salvage (versus No Surgery)	0.52 (0.26 – 1.02)	.06
Radiation Treatment (versus None)	0.86 (0.51 – 1.46)	.58
Primary Tumor Size >8cm (versus <=8cm)	2.17 (1.11 – 4.27)	.02
Metastatic disease at diagnosis		<.001
Present (versus None)	4.05 (2.25 – 7.29)	<.001
Unknown (versus None)	1.50 (0.91 – 2.45)	.11

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Figure 1: Kaplan Meier Curve for overall survival by primary location