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Outcome of primary leiomyosarcoma of bone – Current evidence and a report of 21 cases from two musculoskeletal tumor centers

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Introduction: Primary leiomysarcoma of bone is an extremly rare musculoskeletal tumor. Therefore evidence regarding clinical characteristics, treatment strategies and surgical outcome is limited depending on single case reports and case series. The aims of the present study were to evaluate the (1) clinical characteristics, (2) the surgical treatment and, (3) the outcome of primary LMS of bone. of pooled data for this seldom musculoskeletal tumor.

Methods: A systemic search for the MeSH terms "leiomyosarcoma" AND "bone" with use of the online Databases MEDLINE, Embase, CINAHL and Google Scholar was conducted. The search was not limited to any language. Case reports and case series of primary LMS of extragnathic bone were included. The tumor had to be intraosseous, with other primary sites of origin clinically excluded (i.e. patients with previous history of LMS of the uterus). We evaluated demographic, pathological and therapeutic variables. Descriptive summary statistics included means and frequencies. Kaplan-Meier analysis with 95% confidence intervals (CI) was performed to estimate survival.

Results: Eighty-eight studies with a total amount of 197 primary LMS of bone have been found in the literature. Additionally 21 new cases treated at two musculoskeletal oncologic institutions were added, resulting in a total amount of 218 cases included in analysis. Mean age at diagnosis was 49 years (range 9 - 87). 50,5% of patients were female and 49,5% were male, respectively. Mean follow-up was 35,1 months (range 0 - 220,4). The most common site of appearance was the distal femur (36,1%) followed by the proximal tibia (22,0%). Amputation was performed in 21,5% of patients. In 27,0% limb salvage was achieved with endoprosthetic reconstruction and 5,0% had biological reconstrcution. 6,9% of patients presented distant metastasis at diagnosis and 29,8% of patients developed metastasis during follow up. The local recurrence rate was 10,1%. The mean overall survival was 125,7 months (range 2,2 - 220,4).

Conclusion: Pooled data analysis reveals clinical characteristics and outcome for rare tumor entities. This study analyzed all reported primary LMS of bone to date. About the knee (distal femur, proximal tibia) is the most common site of LMS occurence. Survival is comparable to other primary malignant sarcomas of bone.