Comparison between MDM2-amplified sarcoma without well-differentiated sarcoma component and dedifferentiated liposarcoma

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INTRODUCTION: Dedifferentiated liposarcoma (DDLS) is a soft tissue sarcoma harboring the MDM2 gene amplification. It histologically possesses both a differentiated component and a well-differentiated component. However, a consensus is lacking on how to diagnose DDLS when a well-differentiated liposarcoma component is absent. In our study, we characterized MDM2-amplified soft tissue peripheral sarcoma (MSPS) as a soft-tissue sarcoma arose in extremity and superficial trunk while having MDM2 gene amplification, does not have a well-differentiated liposarcoma component. We compared the clinicopathologic and radiologic characters between DDLS and MSPS. Our primary focus was on the clinicopathologic and radiologic distinctions between DDLS and MSPS, and we subsequently compared their clinical outcome.

METHODS: We conducted an examination of the clinicopathological characteristics (including age at resection, sex, tumor size, surgical margin, and prognosis) and radiological features of DDLS (n=23) and MSPS (n=16). All cases underwent wide resection at our hospital starting from 2011. For all MSPS cases, MDM2 amplification was confirmed through FISH, and there was no presence of a well-differentiated liposarcoma component in any of the split specimens. The radiological characteristics were assessed using MRI T2-weighted or Gd-enhanced T1-weighted images to detect any signs of tumor invasion. We used the Wilcoxon test for quantitative variables and chi-squared test to compare pairs of continuous and categorical variables, respectively. Survival curves were calculated using the Kaplan‐Meier method and the differences were evaluated by the log-rank test. Statistical significance was defined as $p < 0.05$. Data analysis was performed using JMP statistical software package (version JMP version 14.0.0; SAS Institute Inc., Cary, NC). This study was approved by our hospital's institutional review board (2017-336).

RESULTS SECTION: On DDLS and MSPS, average ages at surgery were 66 and 61 years, respectively ($p>0.05$). The male representation was 74% for DDLS and 81% for MSPS, and the mean tumor sizes were 12 cm for DDLS and 10 cm for MSPS ($p>0.05$). Radiological tumor invasion was 4% for DDLS and 100% for MSPS, respectively ($p<0.0001$). The incidence of positive surgical margins was significantly different, with 27% for DDLS and 73% for MSPS, respectively ($p=0.0022$). The overall survival rates for DDLS and MSPS were 91% and 100% at 2 years, and 91% and 100% at 5 years, respectively ($p>0.05$). The progression-free survival rates were 78% for DDLS and 79% for MSPS at 2 years, and 65% for DDLS and 59% for MSPS at 5 years ($p>0.05$).

DISCUSSION: Previous studies have indicated that the clinical course of MSPS is like that of DDLS when compared to MDM2-negative pleomorphic sarcoma. In our current study, the clinical courses of DDLS and MSPS were also found to be similar. However, from a radiological perspective, MSPS distinctly differs from DDLS, exhibiting a high degree of invasiveness. This invasiveness is attributed to positive margins. Furthermore, genomic and epigenomic analyses are in the pipeline to compare MSPS and DDLS, aiming to establish a novel disease concept for MSPS.

SIGNIFICANCE/CLINICAL RELEVANCE: (1-2 sentences): The radiological characteristics of MSPS and DDLS were different. However, their clinical courses were similar.

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