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Primary and secondary leiomyosarcoma of the oral and perioral region--clinicopathological and immunohistochemical analysis of a rare entity with a review of the literature.

Abstract:
Leiomyosarcoma (LMS) rarely occurs in the head and neck region. These tumors present with a wide range of clinical features, so the diagnosis is predicated on conventional microscopic findings coupled with immunohistochemical analysis. Clinical and histologic data of 7 patients with LMS of the head and neck were recorded retrospectively. In addition to routine immunohistochemistry, staining for cell cycle regulator proteins p16 and p21 was performed. Five LMSs (4 intraoral, 1 dermal cheek) occurred primarily in the oral and perioral region. Two LMSs (parietal and sinonasal) were diagnosed as metastases originating from the uterus and pelvis. Treatment of the primary LMSs consisted of radical tumor resection with clear margins. Distant metastases from LMSs were irradiated or excised as palliative treatment. Three of 5 patients (60%) with primarily excised LMS developed recurrence after an average of 7 months, with lung metastases occurring after 17 months. In 1 patient, cervical lymph node metastases were detected after 10 months. Of all patients, 5 died after an average survival period of 2.4 years. The mean survival period of the 5 patients with primary LMS of the head and neck was 3.3 years. All tumors were positive for vimentin and
-smooth muscle actin, with 57% of tumors showing positive nuclear expression of p16 and 71% of p21. Lack of p16 nuclear expression was associated with a shorter mean survival time (1.3 vs 4.3 yr for p16 positivity). Lung and cervical lymph node metastases often occur in LMS of the head and neck. Presurgical staging, including gynecologic examination, whole-body computed tomography, and sometimes positron-emission or computed tomography, to rule out LMS metastasis is mandatory. Surgical resection of the tumor should be given top priority. Lack of p16 reactivity may have a prognostic value for LMS because it was related to a trend toward poorer survival.