CASE REPORT

Primary leiomyosarcoma of the mandible

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ABSTRACT
Leiomyosarcoma (LMS) is a malignant soft tissue tumor that exhibits smoothmuscle differentiation. Its occurrence in the oral cavity is exceedingly rare. This article presents a 67-year-old woman with 9-month history of a painful tumoral lesion on the right side of mandible. Tumor was composed of fascicles of spindle-shaped cells with cigar-shaped nuclei and eosinophilic cytoplasm. Immunohistochemistry showed that tumor cells were positive for smooth muscle actin (SMA) and desmin. The findings were consistent with the diagnosis of LMS.
Key words: Leiomyosarcoma, mandible, oral cavity, soft tissue tumor

INTRODUCTION

Leiomyosarcoma (LMS) is a malignant smooth muscle tumor, which is usually seen in the uterus and gastrointestinal tract.[1] The proportion of malignancies among smooth muscle tumors of the oral cavity is disproportionately high in comparison with the low ratio of malignant to benign smooth muscle tumors in the female genital tract.[2] The sex incidence is approximately equal and patients of all ages in a range of 10 months to 90-years-old may be affected.[3]

The purpose of this manuscript is to describe a case of primary LMS in the mandible with immunohistochemical staining of tumor cells, which was useful for diagnosis.

CASE REPORT

A 67-year-old woman was admitted for evaluation of a painful swelling in the right side of mandible with a 9-month history [Figures 1 and 2]. On panoramic radiographic examination, a large ill-defined radiolucent lesion extending from condyle to symphysis was seen [Figure 3]. Computed tomography (CT) sections presented a destructive soft tissue lesion involving the posterior region of mandible and ramus [Figure 4]. There was no regional lymphadenopathy and CT scan of abdomen, pelvic sonography and chest X-ray were normal.

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Based on clinical presentation and radiographic features, a malignant odontogenic tumor-like ameloblastic carcinoma and other odontogenic carcinomas were considered as the first differential diagnoses. Ameloblastic carcinoma is an aggressive tumor with a mean age of 33-years and frequently arises from a previous recurrent ameloblastoma.[4] However, this patient was much older with no previous history of recurrent ameloblastoma.

Primary bone malignancies like as osteosarcoma and chondrosarcoma were the next differential diagnoses. Osteosarcoma of the jaw is an uncommon malignancy and presents as a painful swelling, as in this case. However, the mean age of patients is about 33 years and it is more common in males. In addition, the radiographic feature of osteosarcoma contains radiopaque areas in many cases, but the massive radiolucent lesion in this case did not show any calcification or radiopaque areas. Chondrosarcoma is the other primary bone malignancy that occurs in old patients. Nevertheless, chondrosarcoma is not painful in most patients.[4]

The other malignancy that affects the jaw bones is multiple myeloma. However, it is multicentric in bones and more common in men. Moreover, it is associated with some systemic signs and symptoms.

Central mucoepidermoid carcinoma and primary intraosseous carcinoma are also considered in differential diagnoses. Radiographically, central mucoepidermoid carcinoma usually reveals either a unilocular or multilocular radiolucency with well-defined borders[4] in contrast to the ill-defined borders of this case.

Soft tissue sarcomas such as malignant peripheral nerve sheath tumor (MPNST) and LMS are exceedingly rare in the jaw

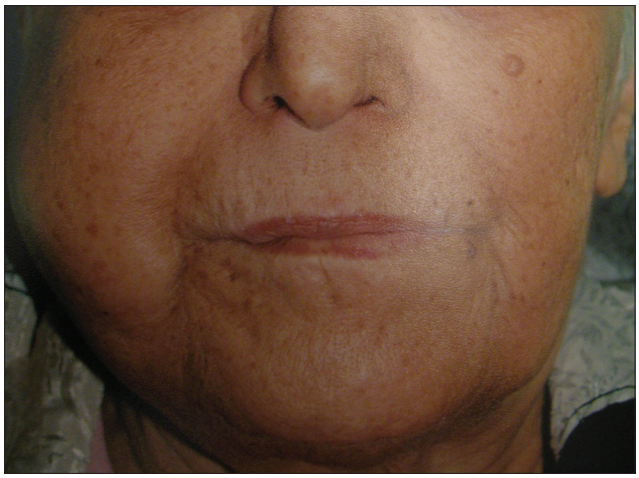

Figure 1: Photograph shows massive swelling in the right side of mandible

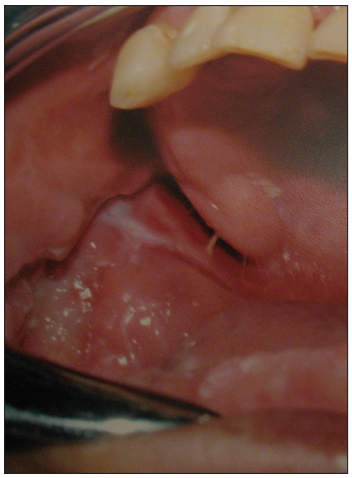

Figure 2: Photograph shows intraoral view of the swelling in the right side of mandible

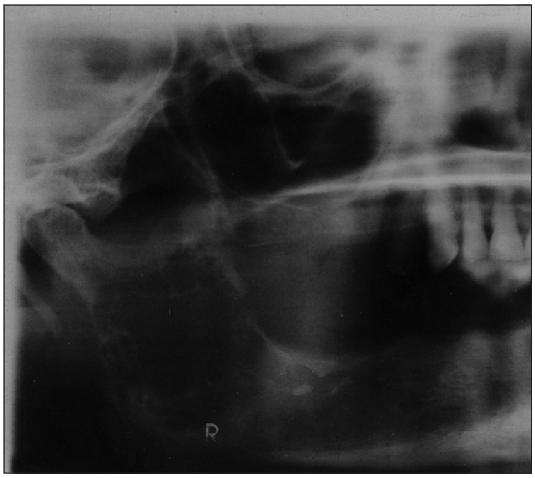

Figure 3: Panoramic radiograph shows a large ill-defined radiolucent lesion extending from condyle to symphysis

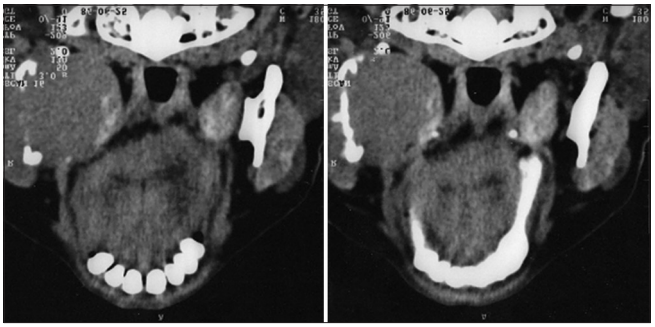

Figure 4: Computed tomography section presents a destructive soft tissue lesion in the posterior region of mandible and ramus

but they should also be considered. Also, metastatic tumors are a major consideration in patients with previous history of malignancy. Post irradiation sarcoma was also excluded, as the patient had no previous history of irradiation.

Hemimandibulectomy was performed under general anesthesia. Gross examination of the resected specimen was a creamy firm tumoral mass, measuring 8 × 5 × 4 cm, extending from condyle to symphysis [Figure 5]. Microscopic examination revealed a malignant mesenchymal tumor that was composed of spindle cell proliferation forming rough bundles and fascicles with interlacing pattern [Figure 6]. Tumor cells frequently had features of cytologic atypia as pleomorphism, hyperchromatism and scattered mitotic figures (five per 10 high power fields). No tumoral necrosis was observed. Spindle cells had blunt-ended, oval, centrally located nuclei and abundant eosinophilic cytoplasm [Figures 7 and 8].

Due to these histopathological features, differential diagnoses of a malignant spindle cell tumor, including spindle cell carcinoma, melanoma, LMS, adult spindle cell rhabdomyosarcoma, myofibroma and MPNST were considered.

Spindle cell carcinoma is a high-grade malignancy arising from superficial epithelial layer. This diagnosis was excluded by the absence of any dysplasia or carcinoma *insitu* of overlying epithelium. As well, malignant melanoma was excluded, as the tumor was not a mucosal lesion and had an origin within mandible. In addition, myofibroma of the jaw has some distinguished clinical and histopathologic features. It is more common in the first four decades of life and has biphasic appearance in histopathology. Adult rhabdomyosarcoma should also be mentioned, as it is a spindle cell tumor. Nevertheless, it has a more myxoid stroma. As well, MPNST shows fascicles of atypical spindle-shaped cells. However, the nuclei are wavy despite the oval blunt-ended morphology in this case.

Therefore, immunohistochemical analysis for desmin, SMA (smooth muscle actin), S100, myogenin and CK(cytokeratin) was performed. The cytoplasm of tumor cells was positive only for SMA [Figure 9] and desmin [Figure 10]. Negative immunoreactivity for S100 was against the neural origin of tumor cells. Spindle cell carcinoma

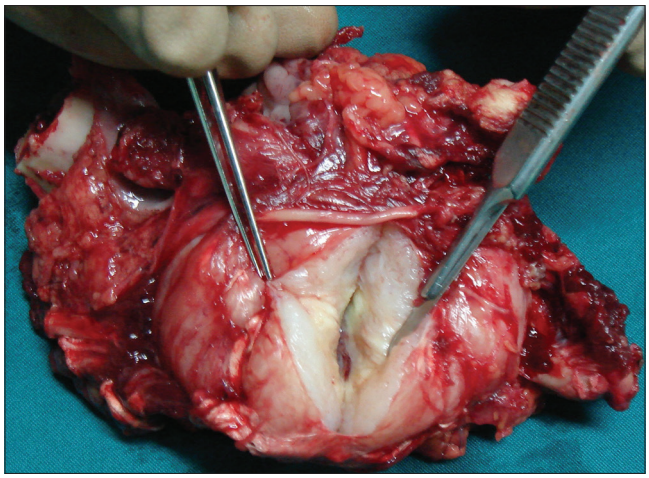

Figure 5: Gross appearance of resected specimen reveals a creamy firm tumoral mass

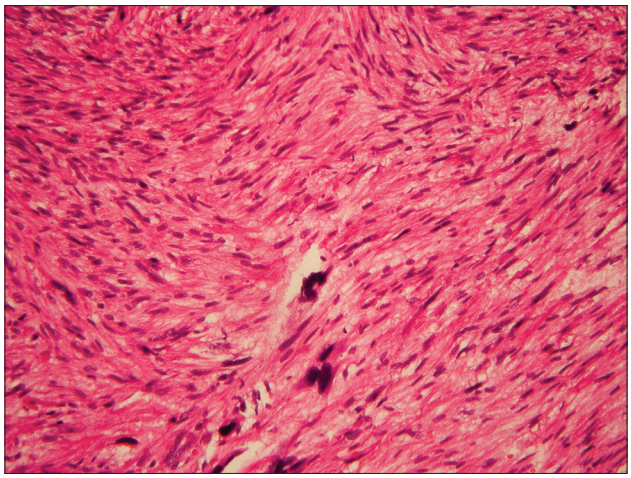

Figure 6: Photomicrograph shows a malignant neoplasm composed of fascicles of spindle cells (H&E stain, ×100)

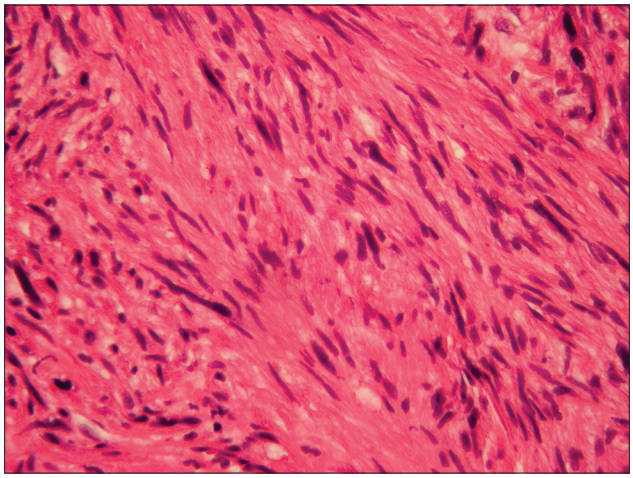

Figure 7: Photomicrograph shows fascicles of spindle cells with eosinophilic cytoplasm (H&E stain, ×400)

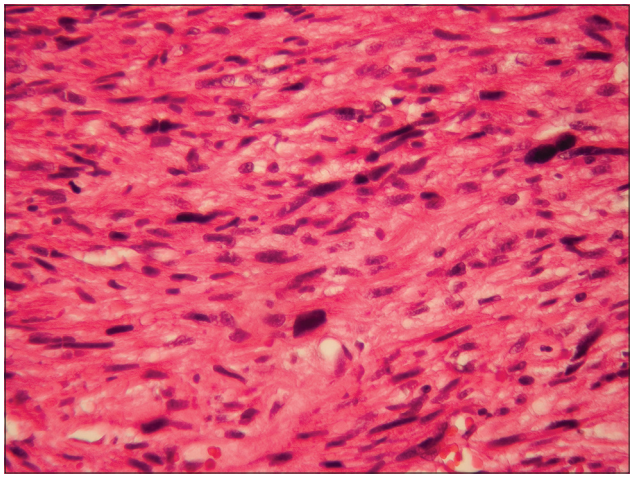

Figure 8: Spindle cells show pleomorphism and hyperchromatism (H&E stain, ×400)

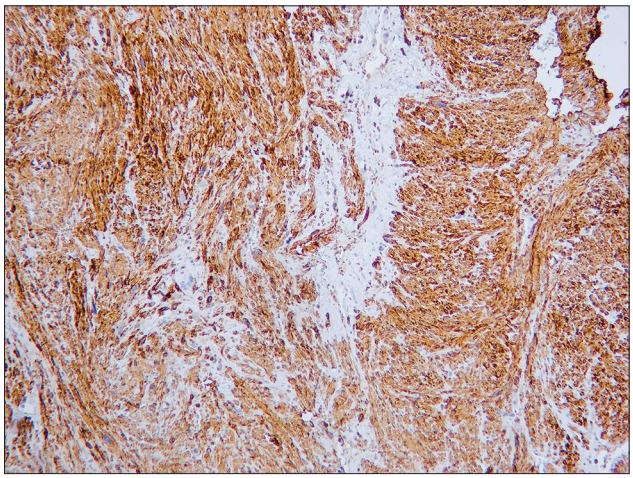

Figure 9: Photomicrograph illustrates considerable SMA expression by neoplastic cells (IHC stain, ×100)

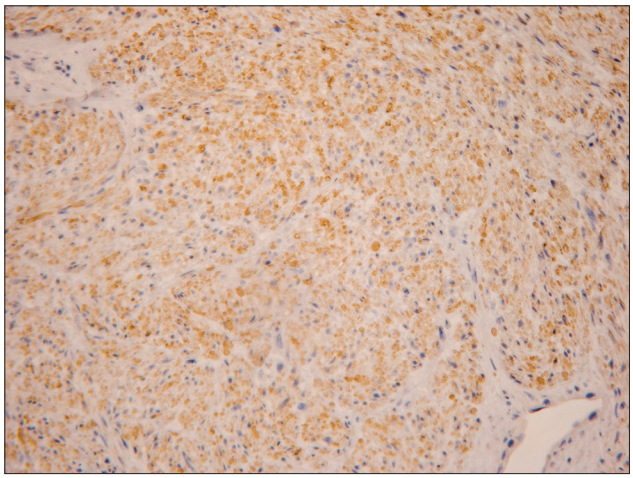

Figure 10: Photomicrograph illustrates considerable desmin expression by neoplastic cells (IHC stain, ×100)

was also excluded, as CK was not expressed in lesional cells. Also, myogenin negativity of tumor cells was against the diagnosis of rhabdomyosarcoma.

These findings, including the morphology of tumor cells and their nuclei with the results of immunohistochemical studies were consistent with the diagnosis of LMS. Unfortunately,

patient died because of myocardial infarction 10 days after surgery.

DISCUSSION

Intraoral LMS is an extremely uncommon neoplasm, with about 80 cases reported in the literature. The intraoral locations of these tumors are as follows: cheek, mandible, gingiva, maxilla, floor of the mouth, tongue, soft and hard palate mucosa.[1] Jaw bones appear to be a site of predilection for oral LMS and approximately 50% of these tumors arise in the jaws, predominantly mandible.[3] Mandibular canal may also be the site of origin in some cases of mandibular involvement.[5] Recently, Rege *et al*., have reported a case of high-grade primaryLMSin the mandible.[6]

Possibility of a metastatic LMS to the oral cavity from other primary sites should always be considered.[2] Oral LMS tends to metastasize to cervical nodes, lung and liver, unlike the LMS in other soft tissues, which rarely has nodal involvement.[7]

Primary and metastatic LMS of the parotid and submandibular glands has also been reported.[8] Metastatic LMS may originate from several potential primary sites and the lung is the most common target tissue for metastatic deposits.[1] Difficulty in the microscopic diagnosis of LMS, especially in its differentiation from leiomyoma, has been widely recognized.[7] Although histologic features such as atypia, cellularity and necrosis are correlated with malignancy, the number of mitoses per high-power field (HPF) seems to represent the most reliable criterion of malignancy. However, the criteria of malignancy differs according to the anatomic location.[2]

These neoplasms may easily be mistaken for other more common tumors including spindle cell carcinoma, spindle cell myoepithelioma and melanoma.[7] Periodic acid–Schiff (PAS) and Masson trichrome stain can be helpful for diagnosis. Longitudinal striations may be seen with phosphotungstic acid hematoxylin (PTAH) stain. Immunohistochemical analysis usually reveals the presence of desmin, muscle-specific actin and SMA.[1] In addition, P53 expression has been associated with higher recurrence rate and shorter survival.[2]

The prognosis of LMS is usually poor.[9] Wide surgical excision with radical neck dissection for lymph node metastases is the mainstay of treatment. Because it has high rate of recurrence and metastasis, long term regular follow up is necessary.[10]

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