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Primary leiomyosarcoma presenting as a rapidly enlarging gingival mass of the mandible

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ABSTRACT

Leiomyosarcoma of the oral cavity is a very rare tumor that is associated with aggressive clinical behavior and low survival. In this paper, we report a case of leiomyosarcoma presenting with a gingival exophytic mass that rapidly grew, causing facial asymmetry within 16 days, in a 9-year-old boy. After an excisional biopsy, microscopy revealed a spindle cell neoplasm that, on immunohistochemistry analysis, demonstrated reactivity for SMA. This established the diagnosis of leiomyosarcoma; subsequently, a marginal mandibulectomy and supraomohyoid neck dissection were performed. ***(Korean J Oral Maxillofac Radiol 2006; 36 : 227-31)***

KEY WORDS : Leiomyosarcoma; Mandible; Oral Cavity; Immunohistochemistry

Leiomyosarcoma of the oral and maxillofacial region is rare; relatively few cases of this malignancy have been reported.[1-5] According to Farman's review of 7,748 smooth muscle tumors, only 0.064% appeared in the oral cavity.[6] Oral leiomyosarcomas have been reported to occur over a wide age range with most patients in their sixth or seventh decade of life.[7-9] They are uncommon in childhood,[10] and only a few cases were reported.[5,11,12]

The leiomyosarcoma occurring in the jawbones and oral tissues has been postulated to arise from the tunica media of blood vessels,[13] arrectores pilorum,[14] circumvallate papillae,[15] myoepithelial cells,[16] or pluripotential, undifferentiated, mesenchymal cells.[3,17-19] The most common complaints among patients were swelling (38.5%) and pain (30.8%).[8] Other signs and symptoms were tenderness, rapid growth, loose teeth, bleeding gums and numbness.[8] There is no distinct clinical feature that might suggest the presence of oral leiomyosarcoma. Leiomyosarcoma on histologic examination is characterized by proliferation of eosinophilic spindle cells with blunt-ended nuclei in interlacing fascicles. The presence of frequent and atypical mitotic figures and necrotic foci indicates aggressive behavior.[20] Leiomyosarcoma shows positivity in immunuhistochemical reaction with antibodies against muscle specific actin (MSA) and smooth muscle actin (SMA).[7] The mitotic count, the presence of nuclear atypia, and the size of the tumor are important criteria in the designation of malignancy. Early, wide surgical resection is considered the primary treatment modality for relatively accessible leiomyosarcomas.[2] It has been suggested that the prognosis of soft tissue leiomyosarcomas is mainly dependent on their anatomic location in that peripheral leiomyosarcomas behave in a much more favorable fashion than tumors located in deep tissues.[21] Despite their relatively peripheral location, oral leiomyosarcomas are very aggressive tumors associated with a high propensity for recurrence, local and distal metastasis and a low survival rate.[3,5,7-9]

We report a case of primary leiomyosarcoma presenting

Fig. 1. Clinical photograph at initial visit shows exophytic gingival mass arising from distal and lingual aspect of mandibular left first molar.

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Fig. 2. Panoramic radiograph reveals ill-defined radiolucency surrounding the mandibular left first molar. Note loss of the lamina dura of the first molar and the distal displacement of the developing second molar.
Fig. 3. Occlusal radiograph demonstrates ill-defined radiolucency of lingual cortical bone.

with a gingival exophytic mass of a 9-year-old boy which rapidly grew, causing facial asymmetry within 16 days.

Case report

A 9-year-old boy with a chief complaint of mandibular gingival mass was referred by his general dentist on September 7, 2004. The patient had first noticed the mass 10 days earlier. There was no pain, neurosensory disturbance or lymphadenopathy. He reported no weight loss or febrile episodes. His medical history was negative for any systemic disease. An intraoral examination showed a firm and exophytic gingival mass distal and lingual to the mandibular left first molar (Fig. 1). The lesion measured 3 × 4 cm in its greatest dimension. The overlying mucosa was smooth, pinkish red, and hemorrhaged easily on palpation. A panoramic radiograph revealed an ill-defined radiolucency surrounding the mandibular left first molar with an enlarged periodontal space and loss of lamina dura. The developing permanent second molar was displaced distally by the lesion, whereas the follicle of the third molar was developing normally (Fig. 2). An occlusal view of the left posterior mandible showed cortical bone loss lingually (Fig. 3).

On the patient's first visit, an incisional biopsy of the gingival mass was performed under local anesthesia. Microscopic examination revealed acute and chronic inflammation. Surgery was scheduled for September 23, 2004. While waiting for surgery over the course of 16 days, the lesion rapidly increased in size (Fig. 4). A surgical resection of the entire gingival mass was then performed under general anesthesia

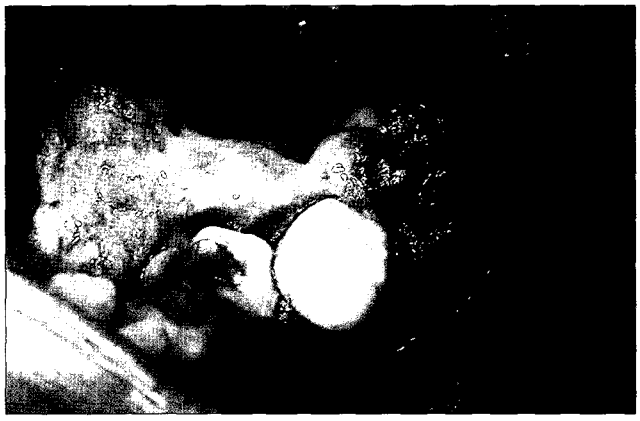

Fig. 4. Mass increased dramatically over the course of 16 days from the time of initial visit.

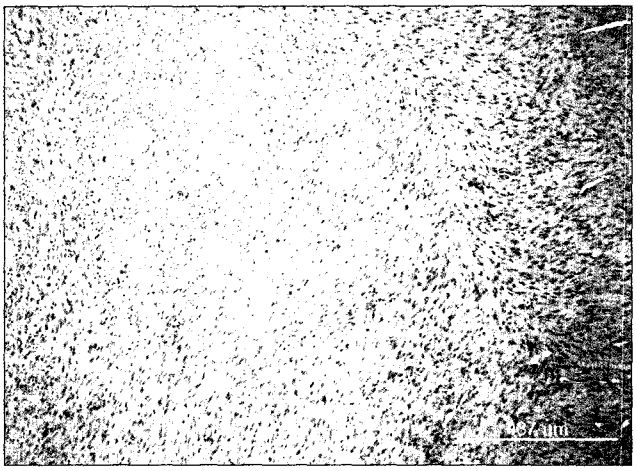

Fig. 5. Photomicrograph shows interlacing fascicular arrangement of spindle cells with cigar-shaped nuclei (H & E, × 100).

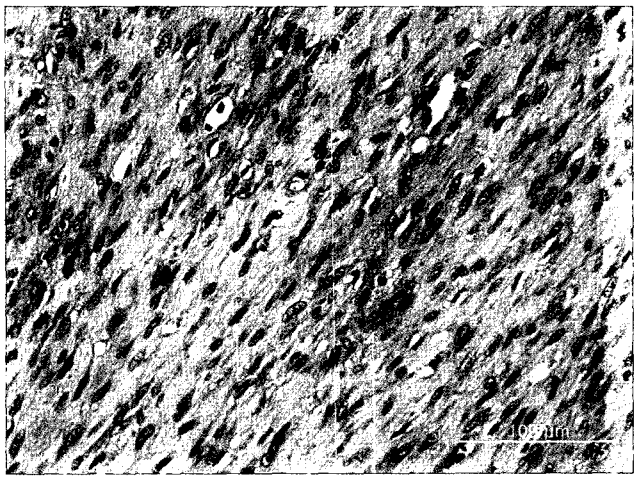

Fig. 6. Frequent and atypical mitotic figures are observed (H & E, ×400).

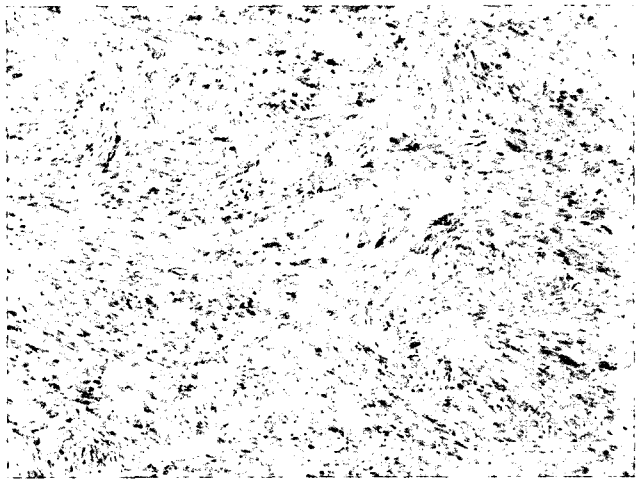

Fig. 7. Photomicrograph demonstrates strong immunoreactivity for SMA in the majority of the tumor cells (immunohistochemical staining for SMA, ×200).

and the frozen section diagnosis was spindle cell neoplasm. Histologically, the specimen consisted of interlacing fascicles of spindle-shaped cells characterized by eosinophilic cytoplasm and blunt-ended nuclei (Fig. 5). The tumor cells showed mild to moderate cellular atypia, 10 mitotic figures/10 HPFs, and atypical mitoses (Fig. 6). In immunohistochemical stain, the neoplastic cells were positively stained for antibody against SMA (monoclonal, clone 1A4, 1 : 200, DAKO, USA) (Fig. 7); they were negative for desmin (monoclonal, 1 : 200, DAKO, USA), CD34 (monoclonal, 1 : 200, DAKO, USA), CD68 (1 : 200, DAKO, USA), and S-100 protein (polyclonal, 1 : 200, DAKO, USA). The final diagnosis was leiomyosarcoma of the left mandibular gingiva.

After the final histologic diagnosis, left supraomohyoid neck dissection and segmental mandibular resection were additionally performed on October 25, 2004. There was no associated lymph node metastasis. The patient has been followed up for eighteen months and no evidence of local recurrence has been observed (Fig. 8).

Discussion

This case involves a rapidly growing lesion of the left mandibular gingiva in an otherwise healthy, young boy. In our initial assessment, the typical screening categories of infectious, reactive, and neoplastic processes were duly considered. An infectious process is unlikely in this case because the mass has attained considerable size without constitutional symptoms such as malaise or fever. Reactive lesions could be ruled out because of the presence of rapidly growing mass with cortical bone destruction. Eliminating those categories, the differential diagnosis points to neoplastic lesion.

When considering neoplasms, one must distinguish between

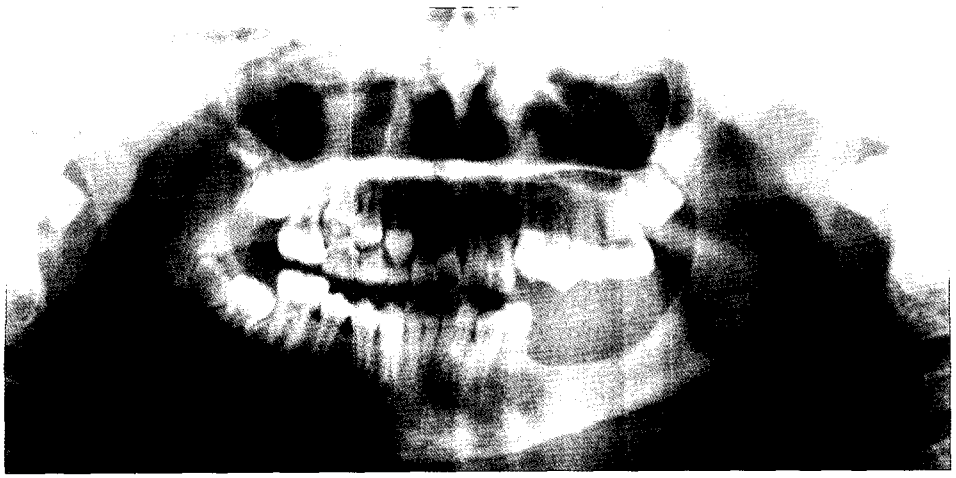

Fig. 8. Panoramic radiograph taken eighteen months postoperatively shows no evidence of recurrence.

malignant and benign processes. We favor a malignant over a benign process in the current case because of the rapid growth and ill-defined radiolucency. However, an aggressive benign lesion must be included as a possibility in the differential diagnosis. Fibromatosis of the craniofacial area presents as a firm mass of either rapid or slow onset.[22] It manifests a behavior between that of a benign lesion and that of a fibrosarcoma.[23] It most commonly occurs in children or young adults.[24,25] Fibromatosis may erode or destroy adjacent bone, as in the current case.[25,26]

Desmoplastic fibroma, the osseous counterpart of soft tissue fibromatosis, would be considered. Most of desmoplastic fibromas are discovered in patients younger than 30 years of age. A painless swelling of the affected area is the most common initial complaint. Radiographically, the lesion appears as a unilocular or occasionally multilocular radiolucent area. If the lesion erodes through the cortex, an accompanying soft tissue mass will be present.[23]

Myofibroma is another benign possibility. It is a relatively common soft tissue tumors of the maxillofacial region, and has have been misinterpreted as malignant or aggressive lesions.[27] A number of myofibromas of the oral region, in both children and adults, have been described previously.[28-30] The most common oral location is the mandible, followed by the lips, cheek, and tongue. The tumor is typically a painless mass that sometimes exhibits rapid enlargement. Intrabony tumors create radiolucent defects that tend to be poorly defined.[23,27]

One must consider a rapidly growing tumor resorbing bone to be malignant. Sarcomas would be at the top of our list of possible malignancies. They are a relatively common pediatric malignancy and can present very variably. Rhabdomyosarcoma (RMS) should be considered. There was a predilection for occurrence in the first two decades.[31] The most common sites of this tumor in children are the head and the neck. RMS of the oral cavity accounts for 10-12% of all head and neck RMS,[32] and the tongue, palate and cheek are the most common sites in the oral cavity.[33,34] Rhabdomyosarcoma is consistent with the current case except for the location of the lesion. Fibrosarcoma is also a strong consideration. It usually presents as a firm mass, more commonly in children and young adults.[23,35] Radiographically, fibrosarcoma appears in most cases as a lytic lesion with an ill-defined border.[36] The possibility of the current lesion being a leiomyosarcoma is unlikely, because leiomyosarcoma of the oral cavity is rare and most common in the sixth or seventh decade.[7-9] Another sarcoma that can be considered is osteosarcoma. Jaw lesion occurrences typically peak in the fourth decade. Although in some patients, juxtacortical osteogenic sarcoma occurs as exophytic hard nodules on the attached gingiva, it is uncommon in children.[37]

A malignant lesion of hematologic source to be considered is Burkitt's lymphoma. The first radiographic sign is a patchy infiltrative process beneath the alveolus, which causes resorption of the lamina dura.[23] However, Burkitt's lymphoma is endemic in central Africa, where it is the commonest childhood malignancy and is strongly associated with Epstein-Barr virus infection. Non-endemic Burkitt's lymphoma is rare and occurs primarily in North America.

Malignant peripheral nerve sheath tumors are also considered. They are most common in young adults. The tumor is an enlarging mass that sometimes exhibits rapid growth. The lesion manifests itself most commonly on the proximal portion of the extremities and the trunk; it is rare in the head and neck. In contrast to the current case, associated pain or a nerve deficit is common.[23]

Although we could not entirely exclude fibromatosis which might exhibit rapid growth, we favor a malignant process over a benign because of the growth rate and ill-defined radiolucent nature. Sarcomas would be at the top of our list of malignancies, as they generally occur in younger patients and can have a very variable presentation.

In the present case, a firm exophytic mass exhibiting rapid growth warranted consideration of malignancy. The ill-defined radiolucency with erosive lingual cortex also supported that impression. The incisional biopsy on the first visit revealed inflammatory tissue only because of inadequate sampling. The diagnosis from the frozen section examination was spindle cell neoplasm, probably malignant, and the final diagnosis was leiomyosarcoma. The final diagnosis of leiomyosarcoma was based on the recognition of typical cytoplasmic, nuclear features and immunohistochemical evidence.

We reported a case of leiomyosarcoma presenting with a gingival exophytic mass that rapidly grew in a 9-year-old boy. Leiomyosarcoma of the oral cavity is a very rare tumor that is associated with aggressive clinical behavior and a low survival rate. Because of the potential for recurrence, close follow-up is recommended.

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