Myofibroma of the mandible: A case report

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ABSTRACT

Myofibroma is a rare benign soft tissue tumor that in all ages usually occurs in the head and neck region, and at subcutaneous tissue, but rarely has bone origin within bone. Intraosseous lesions are more often found in childhood. Although intraosseous lesions are relatively common in mandible. Reports for mandible, reports on radiographic findings of myofibroma occurred on the mandible are uncommon. We describe the radiographic appearance on the conventional radiographs and CT of myofibroma of the mandible in a 9-year-old boy. This benign lesion closely resembles to odontogenic cyst or tumor on image. (***Korean J Oral Maxillofac Radiol 2006; 36 : 211-5***)

KEY WORDS : Myofibroma; Mandible; Odontogenic Cysts; Child

Myofibroma or myofibromatosis are nodular tumors of soft tissue, bone, or internal organs that affects all ages, but oral examples are less known, and jaw lesions are comparatively rare. The tumor may present as single or multiple nodules, myofibroma is usually solitary, but may occur as myofibromatosis. In 1954, Stout[1] first delineated the disease that newborns and infants presented with multiple nodular lesions of the skin, subcutaneous tissue, muscle, bone, and viscera as congenital generalized fibromatosis. Twenty-seven years later, Chung and Enzinger[2] reevaluated the clinicopathologic features of this entity and were the first to use the term infantile myofibromatosis to designate a condition in newborns or young children characterized by multiple (multilcentric) or singular (solitary) nodular tumors of myofibroblastic origin. They subdivided the disease into solitary and multiple types.

These lesions have been reported under various names, including infantile myofibromatosis, adult myofibroma, congenital fibromatosis, infantile myofibroma, and adult myofibroma. However, Lingen et al.[3] suggested that the term myofibroma, which has been proposed by Smith et al.[4] and others[5-8] was more appropriate for these solitary lesions of myofibroblastic origin. Therefore, as the tumor usually affects infants and younger patients, the condition was classified an "infantile" myofibroma. In young children multiple lesions are often present, in which case it is known as infantile myofibromatosis.[2] The solitary form is more common than the multiple form and tends to occur in adults.[1]

We report a case of myofibroma of the mandible in a 9-year-old boy and review the existing literature.

Case report

A 9-year-old Korean boy visited the Department of Pediatric Dentistry at Chosun University Dental Hospital with the chief complaint of a painless swelling over the right mandible premolar region of a few weeks duration and the lingual eruption of the right madibular lateral incisor. The child had no significant medical history. On examination some buccolingual expansion was noted in the lower right premolar region. Physical examination revealed a firm swollen region, which showed tenderness, pain at palpation. The right mandibular lateral incisor was erupted lingually that was sensitive to percussion. The right mandibular primary canine and two primary molars were non-vital. Gingival swellings extended from the right mandibular primary canine to the second primary molar. However, the patient had not experienced any symptoms.

On radiographic examination an extensive radiolucent lesion with sclerotic borders was revealed under the right mandibular primary canine and primary molars (Fig. 1). Root resorption of involved teeth was observed. Two right developing premolars were displaced to the inferior border of mandible. On cross-sectional view of occlusal projection expansion of buccal and lingual cortical bone of the mandible with partial resorption of lingual cortical bone was observed (Fig.

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Fig. 1. Panoramic radiograph demonstrates the extensive radiolucent lesion with sclerotic borders under the right mandibular primary canine and primary molars. Root resorption of the involved teeth is shown. The two right developing premolars are displaced to the inferior border of mandible.
Fig. 2. Cross-sectional view of occlusal projection demonstrates expansion of buccal and lingual cortical bone of the mandible with partial resorption of lingual cortical bone.

2). Based on these clinical and radiographic examinations radiographic impression was of an odontogenic cyst (including a radicular cyst, an odontogenic keratocyst), or an ameloblastoma.

Based on these findings, marsupialization of the lesion and extraction of the right mandibular first primary molar was performed under general anesthesia with enflurane+N_2O+O_2. During the procedure, the oral surgeon confirmed a hard mass without a cystic wall. An incisional biopsy of the lesion showed a tumor composed of interlacing bundles of spindle cells with tapered or blunt-ended nuclei mixed with foci of hemangiopericytoma-like appearance (Fig. 3). A diagnosis of myofibroma was made.

The surgeon then decided to resect the lesion. Conventional radiography and computed tomography (CT) were performed for re-evaluation purposes seven months later. The lesion was found to have extended to the mesial root of right mandibular first molar (Fig. 4), and more expansion to the buccal side than the original lesion (Fig. 5). CT revealed a well-demarcated, unilocular cystic appearance with partial resorption of the lingual cortical bone (Fig. 6). These radiographic features indicated an odontogenic cyst, although previous pathologic diagnosis was of myofibroma. Although myofibroma is characteristically slow-growing tumor, this case was shown rapid growth.

The patient was admitted for surgical resection. The lesion was easily excised under general anesthesia and the defect was filled with a corticocancellous bone graft. The posto-

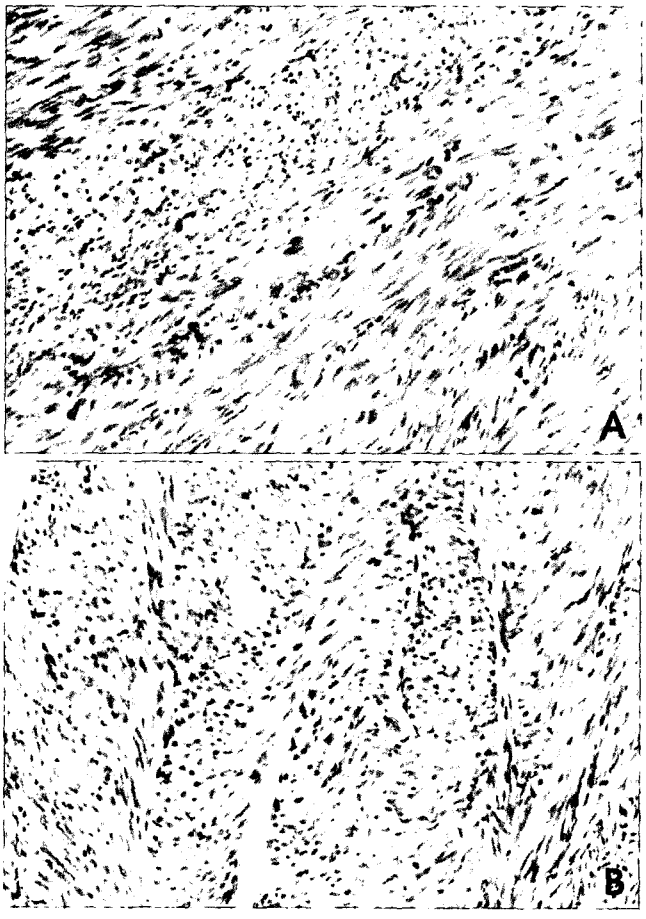

Fig. 3. Photomicrograph shows the tumor composed of interlacing bundles of spindle cells with tapered or blunt-ended nuclei (A) mixed with foci of a hemangiopericytoma-like appearance (B) (H-E, ×200).

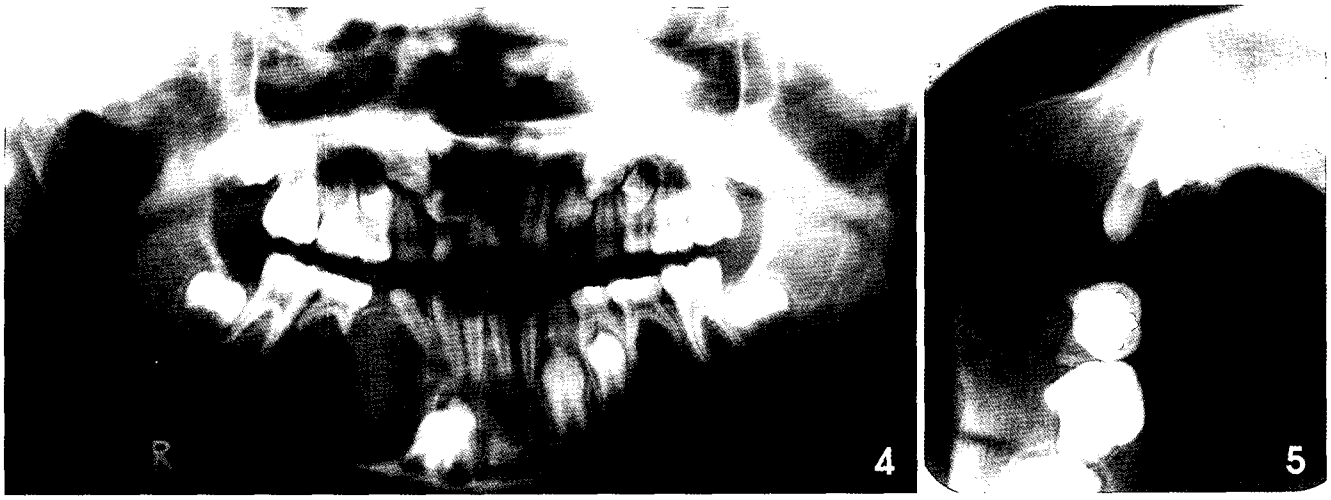

Fig. 4. At 7 months later, this panoramic radiograph demonstrates that the lesion had extended to the mesial root of the right mandibular first molar.
Fig. 5. At 7 months later, this cross-sectional view of the occlusal projection demonstrates that the lesion had extended more to the buccal side than the original lesion.

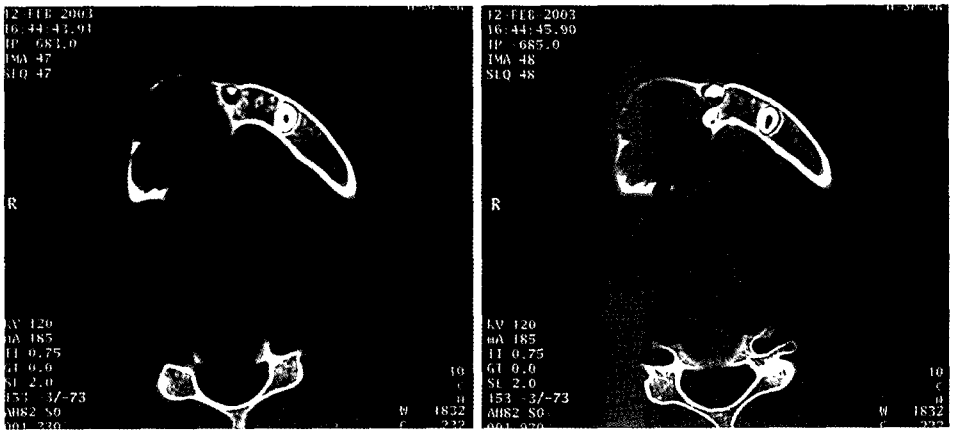

Fig. 6. Computed tomograph reveals a well-demarcated, unilocular cystic appearance with partial resorption of the lingual cortical bone.

perative course was uneventful and he was discharged 3 days later and followed periodically. No evidence of recurrence was noted 2 years after surgery.

Discussion

Solitary myofibroma is a benign lesion usually found in the superficial soft tissues and is viewed as a separate entity and distinct from the more aggressive deeper multiple lesions of infantile myofibromatosis.[9] Soft tissue lesions occur predominantly in the head and neck, including the mouth,[3,5,10-12] the skin of the trunk, and subcutaneous tissue.[2,9,13] Intraosseous lesions are much less common but have a marked predilection for the skull[13-16] and mandible[9,13,17-19] In the largest series of 79 myofibromas in the oral region, Foss and Ellis[20] reported that approximately one third of the tumors affected the jaw bones; 12 lesions were central and 15 involved the cortical or periosteal surface.

The cause of myofibroma is presently unknown. A number of authors have suggested that the tumors are inherited in an autosomal dominant[21] or alternatively in an autosomal recessive[22] trait. However, its low familial incidence suggests that there are probably factors other than genetics that play an important role in the cause of this disease.[5]

Clinically the lesions are more often seen in infants although the age range is wide.[3,8,9,13,20] According to these reports, all affected jaw bones occurred in children under 18 years of age. However, one report[19] revealed adult mandibular cases. Chung and Enzinger[2] found the solitary form to be more common (a three-to-one ratio) and to have a slight male predo-

minance, whereas the multicentric form was slightly more frequent in female patients. This finding is consistent with other reported cases[3,5,8,10,20] but Montgomery et al.[11] reported 4 men in their 9 cases. Clinical signs and symptoms are nonspecific, the presence of a painless mass or swelling is typical.[8,9,11,19,23,24] The tumors present as slow growing, nodular, firm submucosal swellings.

Radiographically the lesions appear benign and are often interpreted as cysts because they are sharply defined radiolucencies with a variably thick sclerotic rim.[9] Mandibular tumors have been described as well-defined either unilocular or multilocular radiolucencies with marginal sclerosis.[3,5,9,11] In addition, a cyst-like expansion and thinning of the cortical plate may occur; radiographs showed that lesions caused partial resorption of cortical bone and displacements of adjacent teeth or tooth germs.[5,8-10,23,24] Myofibroma was occurred on gingiva, radiographs showed ill-defined crestal bone loss and interproximal spacing between teeth.[5,9,12,25] Foss and Ellis,[20] reported radiographic features of 10 cases in a clinicopathologic analysis of 79 cases. In 8 of these, the radiolucent tumor defect was associated with erupting teeth. Tumors of the alveolus tended to form broad, shallow, lytic defects without distinct cortical margins. Central tumors, all mandibular, also were demonstrated only partial or poorly developed sclerotic borders around a radiolucent center. One tumor had a vague multilocular radiographic pattern. Cortically or periosteally based lesions caused an exophytic soft tissue shadow with a thin radiopaque shell overlying a non-corticated, lytic defect in the bone surface. Only one report described a solitary adult myofibroma of the mandible simulating an odontogenic cyst.[19] This also demonstrated expansive unilocular cystic mass in the left ramus extending from the distal aspect of the mandibular third molar to the mandibular foramen.

Histologically the lesions are characterized by being circumscribed, but not encapsulated, masses of spindle cells that sometimes show diverse features like hemangiopericytoma-like vascularity and necrosis, which may lead to their being confused diagnostically with more aggressive soft tissue spindle cell tumors.[3,8] The lesions are usually well circumscribed and display a biphasic pattern consisting of fascicles of spindle-shaped cells admixed with areas that demonstrate a hemangiopericytoma-like pattern.[3,18]

Lesions of the oral cavity present a wide differential diagnosis radiographically and histologically. As previously stated, because myofibroma may have a radiographic appearance similar to that of well-demarcated cyst, the diagnosis to make myofibroma is very difficult. The radiographic differential diagnosis may include an odontogenic keratocyst, an ameloblastic fibroma, and a cystic ameloblastoma. The diagnosis of myofibroma relies entirely on the histopathologic examination. Histologically, myofibroma must be differentiated from leiomyoma, neurofibroma, fibrosarcoma, leiomyosarcoma, low-grade myofibroblastic sarcoma, metastatic neuroblastoma, hemangiopericytoma, and desmoplastic fibroma of bone.[5,8,9,11] The presence of a zoning phenomenon specifically distinguishes myofibromatosis from leiomyoma, leiomyosarcoma, and other myofibroblastic lesions, regardless of the fact that cells in these other lesions also exhibit immunoreactivity for smooth muscle actin.

Surgical excision is the treatment of choice for myofibroma. Although myofibroma is nonencapsulated, previous studies have shown a low recurrence rate.[1,6,9] Most recurrences are associated with anatomic restraints on appropriate surgery.

In conclusion, myofibroma is a soft tissue tumor, which rarely causes intraosseous lesions in the mandible. Myofibroma in the mandible is described as having unilocular or multilocular radiolucencies with or without a sclerotic rim. Here, we describe the cystic manifestations of the solitary form of myofibroma, which may arise in the mandible. Care must be taken not to mistake these lesions for more aggressive or malignant lesions, which they may resemble.

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