Myofibroma of the Mandible: A Case Report

Chiu-Kwan Poon* and Po-Cheung Kwan**

* Chairman, Division of Oral and Maxillofacial Surgery, Department of Dentistry, Taichung Veterans General Hospital
** Attending Physician, Department of Pathology, Taichung Veterans General Hospital

Abstract

Myofibroma is an uncommon neoplasm characterized by myofibroblastic proliferation. The tumor may present as single or multiple nodules of soft tissue, bone, or internal organs. When encountered in the jaws, the lesions exhibit clinical and radiographic features suggestive of several odontogenic and non-odontogenic neoplasms. Myofibromas of the jaws are usually well-demarcated, but sometimes can be poorly delineated or infiltrated that may result in misdiagnosis and mistreatment. In this article, a case of myofibroma of the mandible of a 10-year-old Taiwanese girl is reported. The patient received local wide excision of the lesion and remained disease free with regular follow-up for 6 years. Details of the clinical features, histopathologic diagnosis, and management of the case are provided to augment the paucity of literature available to clinicians managing the rare disease.

Keywords: Myofibroma, Mandible.

Introduction

Myofibroma and myofibromatosis are rare, benign proliferation of myofibroblastic tissue. The lesions typically occur in early infancy and involve various kinds of tissues and internal organs.1-9 Series of reports have demonstrated the myofibroma, particularly in their solitary form, affect a broad range of ages.4,10-17 The similarity in both the clinical and histopathologic features of the “infantile” and “adult” lesions support the proposal that myofibroma is a more accurate and acceptable term for solitary myofibroblastic lesions of the oral cavity.10 The tumors demonstrate a predilection for the head and neck, especially the oral or perioral structures.9,18-19 In descending order, tumors involved mandible, tongue, lip, cheek, or buccal area, maxilla or palate, and floor of mouth. Intraosseous lesions are relatively common in childhood but uncommon in adults. The lesion is considered to be completely benign but there is the potential for it being confused with more aggressive spindle cell tumors.

Report of Case

A 10-year-old girl presented in May 1995 with a painful mass measuring 3.5 × 3.0 × 2.0 cm on the right retromolar area (Fig. 1,2). The lesion was rubbery firm, with an ulcerated surface. Extrinsically, there was a facial asymmetry due to
bulging of the right cheek. No lymph node enlargement in the right submandibular and cervical regions could be detected. Panoramic radiograph showed an ill-defined crestal bone loss and spacing between the developing right lower second and third molars. (Fig. 3) Computed tomography (CT) scan disclosed osteolysis of the posterior portion of the right mandibular body and angle areas that were replaced by soft tissue mass (Fig. 4). A $^{99}$Tc-MDP whole body bone scan revealed an area of increased radioactivity of the mandible at the site of tumor growth. Chest radiograph and liver sonogram failed to disclose any abnormal finding. All the laboratory data were within normal limits. An incisional biopsy was performed and a diagnosis of “smooth muscle tumor, probably malignant” was returned. Radical operations including wide excision of the tumor, en bloc resection of the mandible, and radical neck dissection were suggested. However, patient’s relatives only consented to the more conservative surgery. Wide excision of the tumor was performed under general anesthesia. In order to preserve her facial appearance, an intraoral approach was used to perform excision from the right lower first molar to the angle regions. The permanent right lower premolars and the right inferior alveolar nerve were preserved. During operation, it was found that the tumor had perforated the buccal and lingual cortical bone and infiltrated the adjacent soft tissue. Thus, the surrounding soft tissue was resected. Multiple frozen sections were performed until the margins were reported as tumor-free. The patient remained disease free with regular follow-up for 6 years (Fig. 5, 6).

Microscopically, the unencapsulated lesion consisted of interlacing bundles and whorls of spindle cells with blunt-ended nuclei, prominent nucleoli, and slightly eosinophilic cytoplasm (Fig. 7, 8). The interlacing fascicular arrangement of myofibroblast closely resembling smooth muscle tumor. In some areas a hemangiopericytotic cellular pattern with multiple slit-like vascular spaces was noted. Focal bone invasion and necrosis were present. A few mitotic figures were observed (Fig. 9), which ranged from 1 to 3 per 10 high-power fields. However, no atypical mitoses could be found. The supporting stroma was focally myxoid in appearance, with interspersed areas of hyalinization. In immunohistochemical studies, the tumor cells were positive for tests of actin and negative for desmin. Electron microscopy examination revealed spindle cells resembling fibroblasts rather than smooth muscle cells or Schwann cells.

Discussion

Solitary myofibroma and myofibromatosis are nonaggressive, benign myofibroblastic proliferations that are relatively common in the head and neck. The clinical appearance is nonspecific, such as enlarging mass that may or may not be painful; and secondary ulceration of the mucosal surface. The lesions should be differentiated from leiomyoma, nodular fasciitis, neurofibroma and some other spindle cell tumors. The differentiation of myofibroma from leiomyoma generally has little clinical significance, although myofibromas may be multifocal. Both tumors were found to occur in the mandibular posterior region of young patients, with cortical involvement and a slight male gender preference. Microscopically, leiomyomas exhibit an orderly proliferation of fascicles that intersect at right angles. The tumor cells are elongated and demonstrate cigar-shaped or blunt-ended nuclei with occasional perinuclear vacuolization. Jordan and Regezi reported that 2 of the 8 cases of leiomyoma were reclassified as
Fig. 1. (A) Frontal and (B) three quarter lateral views of the patient with swollen right buccal cheek before surgery.

Fig. 2. Photograph showing a firm, ulcerative lesion (arrow) of the right retromolar and buccal areas.
Fig. 3. Panoramic radiograph showing an ill-defined crestal bone loss (arrow) and spacing between the developing right lower second and third molars.

Fig. 4. Axial view of the CT scan demonstrates a poorly-defined radiolucency of the right mandibular region.
Fig. 5. (A) Frontal and (B) lateral views of the patient 6 years after surgical treatment. There was no evidence of facial swelling or asymmetry.

Fig. 6. The patient remained disease free with regular follow-up for 6 years. (A) Intraoral view showed normal mucosa of the surgical site. The patient has not yet had denture reconstruction of the mandible. (B) Panoramic radiograph demonstrated normal bone regeneration.
Fig. 7. Photomicrograph showing interlacing fascicular arrangement of myofibroblast. Invasion of the tumor elements between bone trabeculae was evident. (Hematoxylin and eosin. Original magnification, x100)

Fig. 8. The myofibroma exhibited typical pale-staining spindle cells with an eosinophilic cytoplasm and tapered nuclei arranged in a fascicular pattern (Hematoxylin and eosin. Original magnification, x200)
myofibroma on the basis of the positive expression of smooth muscle actin but negative expression of desmin. By contrast, leiomyoma expressed both markers. Nodular fasciitis (NF) is well documented in various body locations with rapid growth rate and is occasionally painful. Histologically, NF is characterized by haphazardly arranged fascicles set in a prominent myxoid stroma. The individual cells resemble fibroblasts with vesicular nuclei and small nucleoli. Extravasated erythrocytes and scattered lymphocytes are common features. Neural tumors, such as neurofibroma, usually stain for S-100 protein and not for desmin and actin. Therefore, immunohistochemistry always plays an important role in the differential diagnosis for oral myofibroma.

The present case has been misinterpreted as malignant or locally aggressive lesions prior to tumor excision. The clinical course did not parallel the radiographic findings and histological appearance. Imaging examination including panoramic radiograph which showed ill-defined crestal bone loss; and computed tomography scan disclosed multilocular radiolucency of the destructed mandible. Incisional biopsy revealed high cellularity and mitotic figures among the specimen, which was suggestive of “smooth muscle tumor, probably malignant”. Wide excision of the lesion and radical neck dissection seemed to be indicated. Fortunately, the patient’s relatives refused any radical surgery and strongly requested to preserve her facial appearance as possible. Histopathologic examination of the surgical specimen proved that the lesion was not leiomyosarcoma but myofibroma. She responded well to conservative surgical excision, and has shown no signs of recurrence.
years after surgery. This also affirmed the non-aggressiveness of the disease and the final diagnosis of myofibroma as well. As to prevent misdiagnosis of malignancy, it is important to understand and familiar with the distinctive histologic features of myofibromas.21,26,27

Myofibromas pursue a benign clinical course and may regress spontaneously.24,27 In most cases, conservative surgical excision is curative and only rare recurrences have been described. Intraosseous myofibromas of jaw bones are also biologically indolent and rarely recur after surgical excision. However, the decision in regard to conservative or radical surgical excision should be based on the aggressiveness of tumor growth, tumor location, esthetics, and functional considerations. In addition, the ability to obtain adequate surgical margins by intraoral conservative measures may be limited. The patient and their relatives should be informed of the working diagnosis, histopathologic findings, and the possibility of local recurrence after the “compromised” conservative treatment. Issues of local control may supersede the importance of biologic potential.28 Routine postoperative follow-up is mandatory.

References


下顎肌纖維瘤—病例報告

潘超群* 關寶祥**
*台中榮民總醫院牙科部口腔顴面外科主任
**台中榮民總醫院病理部主治醫師

摘 要
肌纖維瘤是一種非尋常的肌纖維母細胞增生性腫瘤。可呈現單一或多個腫瘤結節於軟組織、骨質、及內臟器官等。顎骨病灶的臨床及影像特徵與許多齒源性和非齒源性腫瘤相似。顎骨肌纖維瘤多為界限清晰，亦偶有浸潤破壞性質致誤診及不當治療。本文報告一例發生於十歲女童的下顎肌纖維瘤。病患僅接受局部廣泛性切除手術，術後六年未曾復發。由於相關文獻資料貧乏，故報告本病例詳細臨床徵象，組織病理發現與治療過程以供參考。

關鍵語：肌纖維瘤，下顎。