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CASE REPORT

A Case of Odontogenic Myxoma with Unusual Histological Features Mimicking a Fibro-Osseous Process

Yi-Ling Lin · John R. Basile

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Abstract Odontogenic myxoma is a rare benign but locally aggressive odontogenic tumor. This report describes a case of odontogenic myxoma producing diffusely dispersed calcified products in a pattern reminiscent of a fibro-osseous lesion of the jaw. Differential diagnoses for myxoid lesions of the jaws also are discussed. This paper highlights how an odontogenic myxoma can produce a large amount of calcified products to mimic a fibro-osseous process.

Introduction

Myxoma occurs both in bone and soft tissue [1]. Although intraosseous myxoma has been reported in various anatomical sites [2, 3], the majority of these tumors occur in the mandible, followed by the maxilla [4]. Hence the term "odontogenic myxoma" is often applied when the tumor occurs in the jaws to reflect its odontogenic origin. Microscopically, it consists of a proliferation of stellate or spindle cells similar to the stellate reticulum of a developing tooth bud, proliferating in a myxoid stroma. Islands of inactive odontogenic epithelium also may be present, but they are usually minimal and not required for the diagnosis. This resemblance to the mesenchyme of a developing tooth or the periodontal ligament has led investigators to believe that these tissues give rise to the tumor [5]. However, some studies have found that the cells in the matrix of the odontogenic myxoma are different from the ectomesenchymal tissues of a developing tooth [6, 7]. In addition, though it is rare, this tumor does occur in extragnathic bones [2, 3, 8]. These findings have prompted some pathologists to consider that the central myxoma may actually have an osteogenic origin. At present, there is no universally accepted theory about its histogenesis.

The odontogenic myxoma is a rare benign tumor that represents about 3% of all odontogenic tumors, affecting mostly young patients in their second and third decade of life [4, 9]. Cortical expansion and perforation are common, although small lesions may be asymptomatic and are discovered only during a radiographic examination. The tumor appears as a unilocular or multilocular radiolucency and the borders may be well or poorly defined. It can be locally aggressive despite its slow growth, which is reflected in the fact that less than 1% of tumor cells are positive for the proliferation marker Ki-67 [10]. Although small myxomas are generally treated by curettage, larger lesions require extensive resection [5]. Since myxomas tend to recur, it is mandatory that the patients be carefully followed after the surgery. It is widely accepted that local infiltration accounts for its aggressive nature and high recurrence rate, resulting in sparsely scattered deposits of residual bone and dystrophic mineralization in the tumor stroma. Here we report a case of odontogenic myxoma exhibiting osteo-cementum-like material diffusely dispersed throughout the lesion, similar to that occurring in fibro-osseous lesions of the jaws.

Y.-L. Lin
Department of Diagnostic and Surgical Sciences, University of California at Los Angeles, School of Dentistry, 10833 Le Conte Ave., Los Angeles, CA 90095, USA

J. R. Basile
Department of Oncology and Diagnostic Sciences, University of Maryland Dental School, 650 W. Baltimore St., 7-North, Baltimore, MD 21201, USA

Y.-L. Lin (✉)
10833 Le Conte Avenue, Room 53-058B CHS, Los Angeles, CA 90095, USA
e-mail: ylin@dentistry.ucla.edu

Case Report

A 25-year-old Caucasian woman was referred to an oral surgeon by her general dentist for evaluation of a unilocular radiolucency between teeth #26 and #27 identified on a radiograph during a routine dental exam (Fig. 1a). The patient had no clinical symptoms and the lesion exhibited a sclerotic border and had displaced both tooth roots. The teeth were stable and vital. Upon clinical examination, the lesion did not cause obvious perforation of the mandible but had eroded a small area of cortical bone in the facial papilla region. The tumor was removed in two stages, with aggregate sizes of 15 mm × 15 mm × 7 mm and 6 mm × 4 mm × 3 mm, respectively. Although no radiopacity was noted in the radiograph (Fig. 1a), the tumor was described by the surgeon as solid. The patient did well postoperatively without evidence of recurrence after 2 years of observation.

Microscopically, the specimen exhibited key features of odontogenic myxoma. It contained a poorly circumscribed proliferation of stellate or spindle-shaped cells in an abundant, loose myxoid stroma (Fig. 1b, c). The tumor cells did not exhibit mitoses or pleomorphism. However, the histology of this case diverged from that of an ordinary odontogenic myxoma (Fig. 1d). Numerous calcified products were present throughout the specimen (Fig. 1c). The calcified products had a globular or trabecular pattern, producing a histological image reminiscent of fibro-osseous lesions of the jaws, except for the presence of the myxoid stroma. The calcified products are best described as osteo-cementum, as the bulk part of them are acellular, non-polarizable and often are surrounded by a rim of cellular osteoid that is birefringent under polarized light (Fig. 2). No areas containing woven bone were identified in the sections. The calcified material likely contributed to the solid texture of the tumor. There were no immunohistochemical studies carried out for this case.

Discussion

The clinical presentation of this case suggested an infiltrative tumor of the mandible. The surgeon described the gross specimen as solid with a granular appearance, despite the fact that the lesion was radiolucent. We speculated that the tumor was associated with a considerable amount of hard tissue, either because the tumor had calcified products or because the tumor contained residual bone. The differential diagnosis based upon the radiographic, clinical and surgical findings included ameloblastoma, odontogenic myxoma, central giant cell granuloma, ameloblastic fibroodontoma and odontogenic fibroma.

Microscopically, the current case exhibits many features of an odontogenic myxoma except for the presence of numerous calcified products. It is generally accepted that the bone associated with odontogenic myxomas represents residual bone left behind following tumor infiltration and subsequent bone destruction. However, it is unlikely that the calcified products associated with the current case are residual bone. Residual bone is typically trabecular and lamellar and usually present focally or at the periphery of

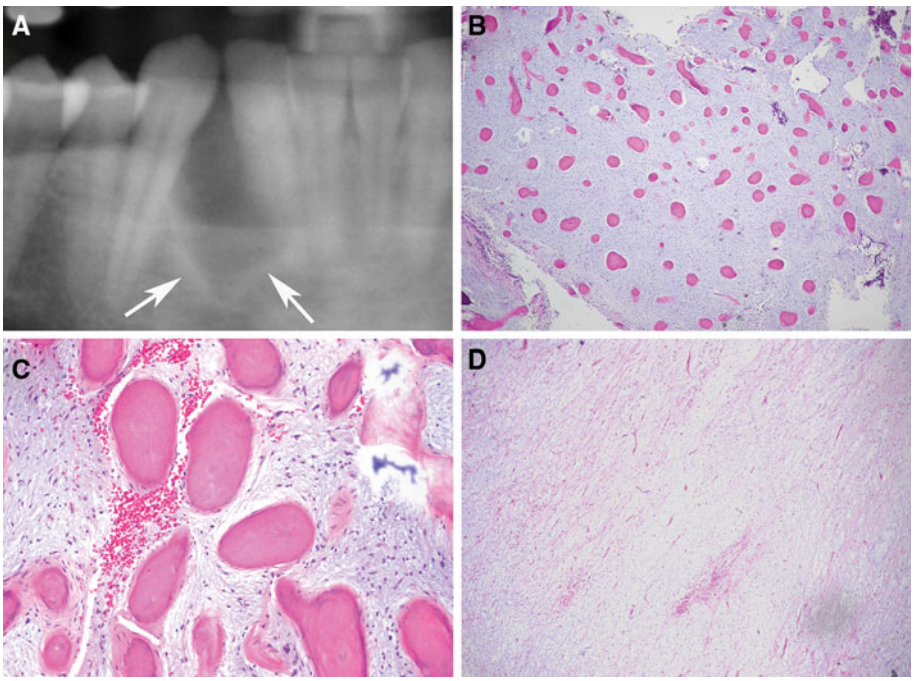

Fig. 1 **a** Radiograph showing a unilocular radiolucency (*arrows*) causing tooth displacement. **b** Photomicrograph showing stellate and spindle cells in a myxoid stroma containing diffusely dispersed calcified products (H&E stain, original magnification ×40). **c** Medium-power view of **b**, showing calcified products exhibiting a globular and trabecular configuration (H&E stain, original magnification ×200). **d** Photomicrograph of a classical odontogenic myxoma for comparison with **b** (H&E stain, original magnification ×40)

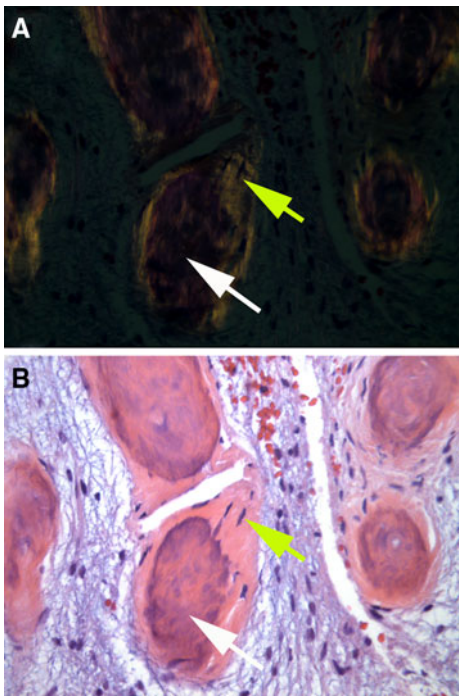

Fig. 2 The main body (*white arrow*) of the calcified products is non-polarizable (**a**), acellular (**b**) and is surrounded by a rim of cellular osteoid (*yellow arrow*) (**b**) that is birefringent under polarized light (**a**) (H&E stain, original magnification ×400)

the tumor. In this case, calcified products were distributed evenly throughout the lesion, creating a homogenous appearance. They did not exhibit properties of mature lamellar bone (Fig. 2). Therefore, these globular and trabecular calcifications are most likely osteo-cementum, having arisen by metaplasia, as they are not rimmed by plump osteoblasts or cementoblasts.

The current case has a strong resemblance to fibro-osseous lesions of the jaws, save for the myxoid stroma. Microscopically, fibro-osseous lesions of the jaws are characterized by newly formed mineralized products, including woven bone and cementum, in a cellular fibrous stroma. Although it cannot be completely ruled out, we consider that the possibility is low for the current case representing a fibro-osseous lesion whose fibrous stroma underwent significant myxomatous degeneration. Clinically, it is unusual for fibro-osseous lesions to present as an interproximal radiolucency and to erode bone. It is also unusual for a stroma to undergo such a complete myxomatous degeneration without a trace of original stroma. Therefore, it is more likely that the myxoid stroma is intrinsic to the current lesion. In addition, we strongly believe that a diagnosis should always be based on the biological nature of the disease. In this case, the myxoid stroma appears to be infiltrative and has eroded a small portion of cortical bone. Therefore, even if this were a fibro-osseous lesion initially, the infiltrative nature of the myxomatous area warrants a diagnosis of myxoma rather than a fibro-osseous lesion. A collision tumor with synchronous occurrence of an odontogenic myxoma and a focal fibro-osseous lesion also was considered. Nonetheless, we cannot find any area that was specific for either tumor. Colliding tumors almost always have distinct areas for individual lesions, and it is unusual for two lesions to be so homogenously intermingled as one lesion. In addition, it is unusual for a focal fibro-osseous lesion to present as an interproximal radiolucency. Therefore, the possibility of a collision tumor is also very low.

The other major differential diagnosis for the current case is odontogenic fibroma. Similar to odontognic myxoma, odongtognic fibroma is also a benign odontogenic tumor composed of stellate or spindle fibroblasts, and the associated stroma can range from collagenous to fibromyxoid. This tumor has been classified into simple and World Health Organization (WHO) types. For the simple type, the tumor may or may not have odontogenic epithelium and may have a small amount or no calcified material. Most pathologists nowadays regard the simple type as a spectrum of odontogenic myxoma. The World Health Organization (WHO) only recognizes the so-called WHO odontogenic fibroma, which requires the presence of odontogenic epithelium for the diagnosis. The choice between odontogenic fibroma and myxoma for the current case is not straightforward since odontogenic fibroma can have osteocementum and the stroma can be fibromyxoid. For pathologists that do not adopt the WHO criteria, the current case can be an example of odontogenic fibroma. However, we favor the diagnosis of odontogenic myxoma over fibroma since, as stated earlier, the myxoid stroma in the current case appears to be infiltrative, which warrants the lesion to be diagnosed as a myxoma. The current case also lacks odontogenic epithelium, which does not fit the WHO criteria for odontogenic fibromas. Since the current case is not typical for any known lesion, both histology and biological activity were taken into consideration for an appropriate diagnosis, and our final diagnosis for this case was odontogenic myxoma containing globular and trabecular osteo-cementum.

Chondromyxoid fibroma and chondrosarcoma should also be included in the differentials of myxoid lesions exhibiting calcified products. Though both lesions can have a myxoid stroma, they also exhibit areas of tumor with chondrocytes residing in lacunae of cartilagenous material, which was not present in this case.

Desmoplastic fibroma and neurofibroma also need to be considered in the differential diagnosis for conventional odontogenic myxoma. Histologically, the desmoplastic fibroma is composed of a proliferation of spindle cells in a fibromyxoid stroma, which superficially resembles an

odontogenic myxoma. However, the desmoplastic fibroma displays a greater degree of cellularity and more interstitial collagen than the odontogenic myxoma and its collagen fibers have a tendency to be arranged in fascicles. The neurofibroma also is composed of a proliferation of spindle cells in a myxoid stroma. The spindle cells in neurofibroma are slender and often exhibit wavy nuclei, in contrast to the plump spindle cells in an odontogenic myxoma. In addition, neurofibromas demonstrate a scattering of small axons.

There are many reports of odontogenic myxoma in the literature discussing their clinical behavior and surgical management. Most of these have a uniform histology and do not contain much hard tissue. Examples of odontogenic myxomas producing exuberant calcifications are exceedingly rare. In fact, there is only one published case of odontogenic myxoma in the English literature with similar histological findings as the current case [11]. However, since the paper did not show low-power micrographs of the tumor, it is difficult to assess whether the "osteo-cementum-like spheroid bodies" described by the authors are as extensive and evenly distributed as in the current case. More cases will be needed to draw any conclusion in terms of whether or not the ability of odontogenic myxoma to produce exuberant amounts of calcification has any impact on its biological behavior. In conclusion, it is important to recognize that odontogenic myxoma, similar to the WHO odontogenic fibroma, can produce a wide range of calcified products, and this possibility should not be excluded from the differential diagnosis when the lesion appears to be locally infiltrative and is associated with a significant amount of calcified products.

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