Primary Xanthoma of the Mandible—A Case Report

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Xanthomas are common cutaneous and subcutaneous lesions that occur due to altered metabolic or endocrinal function. They are found on skin and around tendon sheaths in individuals with dyslipidemias. In extremely rare cases, they can present as isolated intrabony lesions in otherwise healthy individuals. The isolated intrabony lesions are referred to as primary xanthomas. This report describes a case of an incidentally found primary xanthoma, its management, and follow-up in an otherwise healthy patient.

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Xanthomas are commonly found on skin and around tendon sheaths in individuals with systemic diseases of altered endocrine or metabolic states, such as diabetes and dyslipidemias. In otherwise healthy individuals, xanthomas are exceedingly rare, especially as isolated lesions within bone, where they are referred to as primary intrabony xanthomas.

The vast majority of noninfective intrabony oral and maxillofacial pathologies present as incidental nonspecific radiolucencies or radiopacities at routine dental radiography. These present diagnostic difficulties because of the overlapping radiologic appearance of many of these lesions. Probabilities can be assigned based on several clinical and radiologic features but ultimately these lesions require biopsy examination for definitive diagnosis.

Xanthomas can range from completely radiolucent to mostly radiopaque and usually have ill-defined margins. This variability in radiographic appearance represents further diagnostic difficulties because it can mimic a wide range of pathologies.

Report of Case

A 30-year-old man was referred to the oral and maxillofacial department of a public hospital for removal of his third molars. The patient's medical history was relevant for 12 pack-years of smoking. An orthopantomogram (OPG) displayed an incidental ill-defined radiolucency in the anterior mandible (Fig 1). Further imaging with cone-beam computed tomography showed an ill-defined lesion extending from teeth 32 to 43 with loss of trabecular bone, erosion of the buccal cortical plate, but no cortical expansion (Fig 2).

An incisional biopsy was performed under local anesthetic. With a buccal mucoperiosteal approach, the bony window was enlarged and yellow soft tissue material was debrided (Fig 3). Histopathology showed that much of the marrow space within the woven bone was largely replaced by xanthoma type cells. These are pale with small uniform nuclei and abundant pale foamy cytoplasm. No bone necrosis or inflammation was seen (Figs 4, 5).

After discussion with the pathologist, a complete excision was performed under general anesthesia because xanthomatous cells can be present in other more sinister pathologies. It was noted intraoperatively that the central portion had changed to a white hard friable tissue, consistent with newly laid woven bone. Histopathology showed sections of viable woven bone with prominent reversal lines, areas of stromal fibrosis and fatty marrow, and a small amount of dystrophic calcification indicating healing. No further xanthoma or other pathology was noted. Follow-up OPGs at 6 and 12 months showed no

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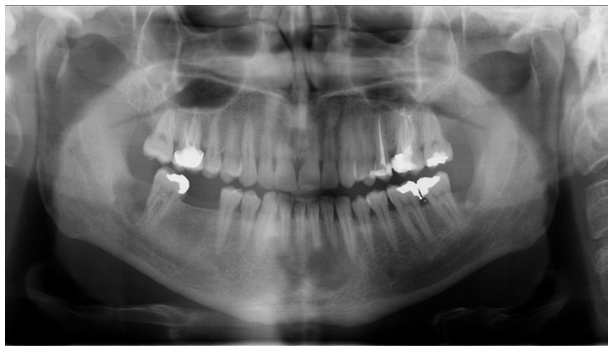

FIGURE 1. Orthopantomogram obtained 6 weeks after removal of third molars showing a well-defined radiolucent lesion in the anterior mandible.

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signs of recurrence and the lesion was filling up with new bone.

Discussion

The gross appearance of a yellow intraosseous substance can resemble several physiologic and pathologic processes. This appearance can represent age-related fatty marrow replacement, a benign healing process after previous treatment, an intraosseous lipoma, or osteolipoma. The etiology of central xanthomas is poorly understood and controversy exists on whether this is a benign neoplastic or reactive process. All mandibular xanthomas reported in the literature have arisen in patients who were otherwise healthy with no history of trauma or infection, giving evidence toward it being a slow neoplastic process. In addition, there is no evidence that they spontaneously regress; rather, they progress slowly and have been known to infiltrate surrounding bone.[1] Conversely, the presence of inflammatory cells, hemorrhage, reactive bone, and cholesterol clefts and associated giant cells suggest it might be a reactive process instead.

Gnathic bone xanthomas are mostly asymptomatic and are incidental findings at routine dental radiography. They have a wide age distribution (range, 11 to 72 yr), with most presenting in the second and third decades of life.[2] The mandible is more commonly affected than the maxilla (ratio, 9:1).[2] Pain and tooth mobility have been reported for xanthomas of the maxilla[2]; however, mandibular xanthomas are largely asymptomatic with the exception of 2 cases presenting with pain and paresthesia of the lower lip.[2,3] There seems to be no gender predilection for gnathic xanthomas, but they are twice as common in men when considering all intraosseous xanthomas.[2,4] They do not tend to cause soft tissue changes intraorally, but mild swelling can be noted.

There is high variability in their radiographic appearance. In the mandible, xanthomas are usually small (range, 1 to 4 cm), although there has been 1 case of a xanthoma extending from the sigmoid notch to the mandibular midline.[5] Internal features have varied from totally radiolucent to a mixed appearance with "honey-combing." The peripheral appearance of these lesions also varies, with descriptors of corticated, scalloped, ill-defined, and slightly sclerotic

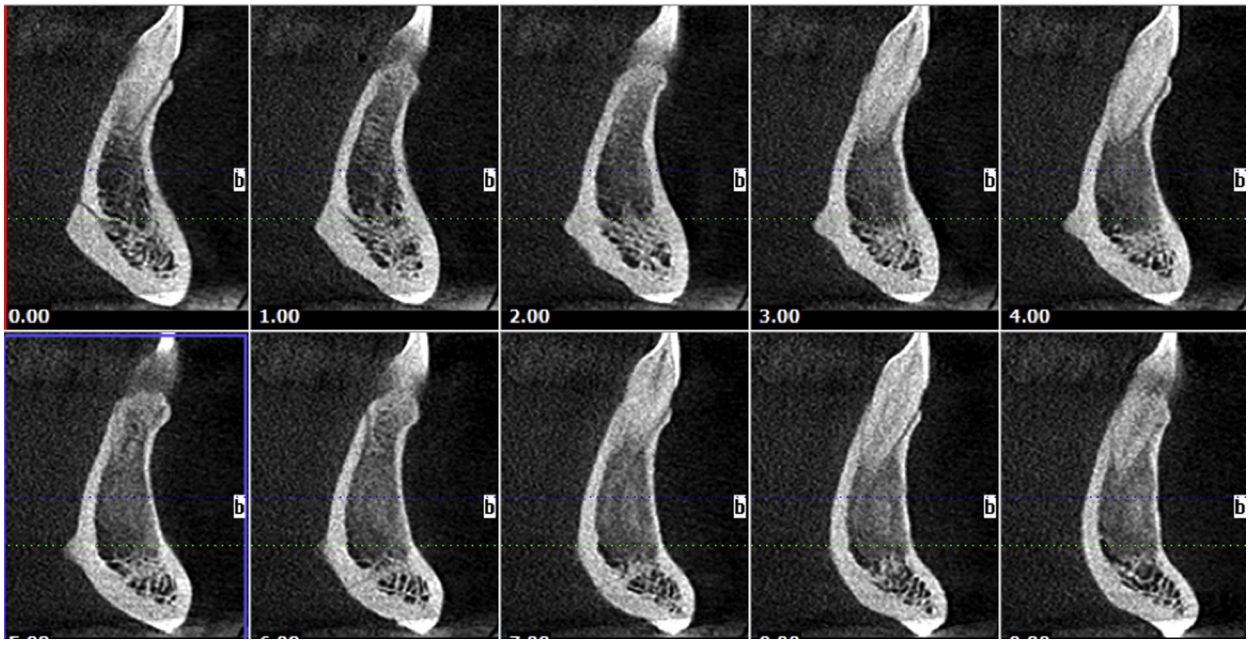

FIGURE 2. Cone-beam computer tomograms of anterior mandible showing replacement of trabecular bone with ground-glass appearance.

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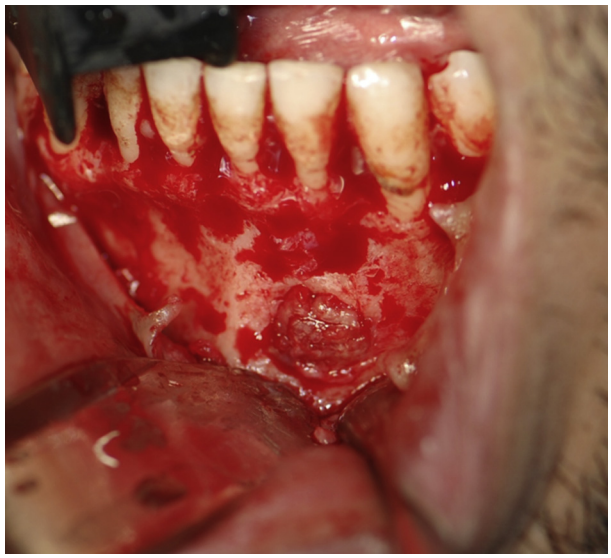

FIGURE 3. Intraoperative clinical photograph showing the lesion perforating the anterior plate of the mandible.

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margins being used previously. Root resorption and displacement of the inferior alveolar canal has been reported in a small number of cases, but largely they do not influence surrounding structures.[2]

The classic histopathologic appearance has been described to be dominated by foam cells with a variable fibrous component. Fibrous cortical defects, nonossifying fibromas (NOFs), and benign fibrous histiocytosis (BFH) have often been confused with xanthomas and some researchers have argued that xanthomas are on a spectrum of disease that includes these conditions. Histologically they are indistinguishable from each other and must be separated based on clinical and radiologic features. Fibrous cortical defects and NOFs are usually small and self-limiting and found as incidental findings confined to the metaphysis of long bones.[6,7] More recent reports have elaborated the biological behavior of xanthomas and currently it is considered a separate pathologic entity. NOFs more often occur in the pediatric population and can regress spontaneously. Xanthomas occur more commonly in an older group, although this can be variable. There have been no cases of spontaneous regression.[3] Compared with BFH, which is known to recur after treatment, xanthomas do not exhibit this behavior even when partial curettage is performed and therefore require shorter follow-up.[3] Histopathologically, BHP differs from xanthomas with a greater abundance of fibrous tissue that form a storiform pattern. Therefore, these clinical and pathologic differences suggest that gnathic xanthomas are an entity separate from NOF or BFH.

There are a few other conditions that result in xanthomatous cells. Rosai-Dorfman disease (RDD) results

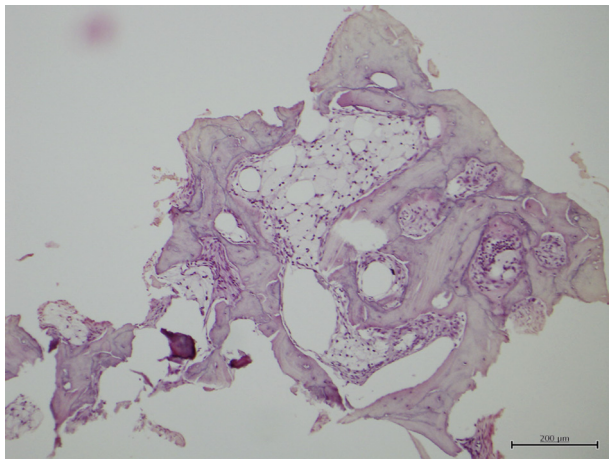

FIGURE 4. Bony lesion exhibiting collections of foamy macrophages (hematoxylin and eosin stain; magnification, ×100).

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from an overproduction of non-Langerhans sinus histiocytes. This is a non-neoplastic disease that results in massive cervical lymphadenopathy. Oral RDD is present in only 8% of affected patients and almost always in conjunction with nodal or other extranodal disease. Only 3 cases of isolated intraosseous lesions have been noted.[8-11] RDD is usually a self-limiting condition treated with complete enucleation and close long-term follow-up. Erdheim-Chester disease (ECD) is another rare non-Langerhans cells histiocytosis. There has been only a single reported case of involvement of the gnathic bones; however, it was associated with widespread skeletal involvement.[12] ECD carries a 1- and 5-year survival rate of 96 and 68%, respectively.[5] It is much more common to find xanthomatous cells secondary to hyperlipidemia

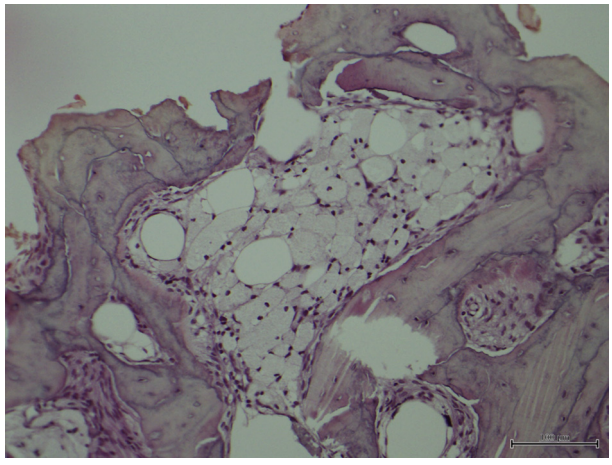

FIGURE 5. Higher power of collection of foamy macrophages (hematoxylin and eosin stain; magnification, ×200).

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types II and III, aneurysmal bone cyst, fibrous dysplasia, simple bone cyst, osteomyelitis, and histiocytosis X.[13] Clear cell carcinoma also can take on the appearance of xanthomatous cells.[14]

The variable radiographic appearance and similarities in histopathology to other conditions present diagnostic difficulties and management dilemmas. An initial biopsy examination might be insufficient to confirm gnathic xanthoma, and repeat tissue sampling might be required to reach a definitive diagnosis, as in the present case. Systemic health and extragnathic signs and symptoms should be investigated to exclude mimicking entities. Because of the limited number of reports and unknown underlying pathologic process, there also is a lack of consensus about the appropriate treatment. Nevertheless, long-term follow-up with simple enucleation with or without curettage has shown that these lesions tend not to recur.[2] Even subtotal excision has been shown to achieve a satisfactory prognosis.[15] The present case showed evidence of this, because the second biopsy, performed within 1 month of the initial biopsy, showed formation of new bone. Recurrence has been noted only with xanthomas secondary to systemic hyperlipidemia.[16]

This report describes a rare occurrence of primary bony xanthoma of the mandible, which was adequately treated. This case highlights the need for similar cases to be collated to better understand this rare pathology and possible systemic ramifications.

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