Median maxillary alveolar osteolytic lesion in a 50-year-old female

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CLINICAL PRESENTATION

A 50-year-old Middle Eastern female was referred for evaluation of an asymptomatic median maxillary alveolar lesion. She denied the presence of symptoms, including pain, bleeding, swelling, or altered sensation associated with the area, but admitted "looseness" of her anterior teeth that had not been noted before the presentation of the lesion. The patient also denied a history of surgery or trauma to the area and use of alcohol, tobacco, or recreational drugs. Apart from the current symptoms, the patient's past medical history was significant for hypothyroidism and osteoarthritis. Her current medical regiment included levothyroxine as well as naproxen, as needed, for periodic arthritic pain.

Extraoral examination revealed no facial swelling or asymmetry. Regional lymphadenopathy was not noted. Intraoral examination revealed fair overall oral hygiene, no appreciable soft tissue swelling in the median maxillary alveolar area (labial or palatal), class I mobility of her maxillary lateral incisors, class II mobility of her central incisors, and reproducible, atraumatic occlusion that was free of incisor contact in centric occlusion. Each of the maxillary lateral and central incisors tested vital to cold testing and electric pulp testing. Sensation over the distributions of the nasopalatine and anterior superior alveolar nerves were normal.

Panoramic radiograph (Figure 1), periapical radiograph (Figure 2), and cone beam computed tomography (CBCT) (Figures 3-5) imaging revealed a 9 × 9 mm, round, well-defined, corticated, low-density area in the region of the incisive canal. Vertically, the lesion extended from the area close to the periapical aspects of the maxillary central incisors superiorly to a region just inferior to the anterior nasal spine. The lesion appeared to cause thinning of both the labial and palatal cortices, but preferential palatal cortical erosion was noted, raising the possibility of nasopalatine nerve or canal involvement. The lesion also appeared to cause enlargement and a mild, uniform expansion of the inferior aspect of the nasopalatine foramen. Consequently, the roots of the bilateral maxillary lateral and central incisors were notably shortened. Needle aspiration of the lesion was found to be negative for any type of fluid. Surgical exposure of the area during biopsy yielded a solid, doughy, but friable mass of tissue which yielded no signs of foreign bodies within or around the lesion.

DIFFERENTIAL DIAGNOSIS

In cases of anterior intrabony maxillary midline lesions with cortical erosion, entities such as nasopalatine duct cyst, keratocystic odontogenic tumor, periapical inflammatory disease, focal cemento-osseous dysplasia, Langerhans cell histiocytosis, and sinonasal schwannoma should be considered. Malignant lesions are unlikely here, as malignancies originating in the vicinity of the nasopalatine duct are rare,[1] possibly because of lack of actively mitotic areas. Limited size, well-defined borders, and the asymptomatic nature of the presentation also indicate a benign etiology.

A nasopalatine duct cyst (NPDC) is a nonaggressive cyst of oronasal duct epithelium that has been trapped within the incisive canal.[2] It is considered the most common nonodontogenic cyst of the oral cavity, with approximately 73% of all oral nonodontogenic lesions having a squamous, nasopalatal etiology.[3] Like our patient's lesion, NPDCs typically present as a median round or ovoid radiolucency overlapping the nasopalatine duct and a peak occurrence in middle age. Radiographic presentation is also generally well circumscribed and unilocular, with minimal involvement of the nearby bony trabeculae.[4] The relatively asymptomatic nature of NPDC also correlates well with the presentation in this case in that the lesion had not been detected earlier, although NPDCs more often induce palatal swelling without cortical erosion.[4] Overall, NPDCs differ significantly from the lesion in the present case in that they are more strongly associated with the Caucasian race[5] and the male gender, as well as a slightly larger size, with diameters typically ranging from 1.2 to 3.2 cm.[3] Palatal perforation as visualized in our case is unusual

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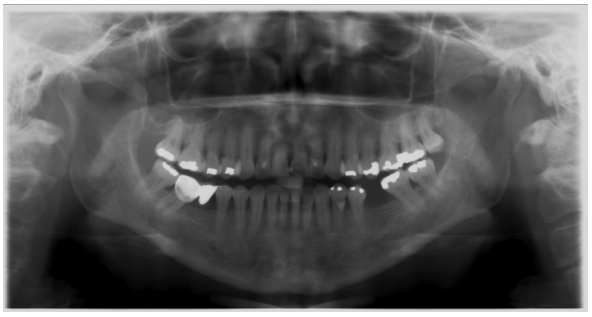

Fig. 1. Panoramic radiograph revealing median maxillary alveolar radiolucent lesion, along with shortened apices of the bilateral maxillary lateral and central incisors.

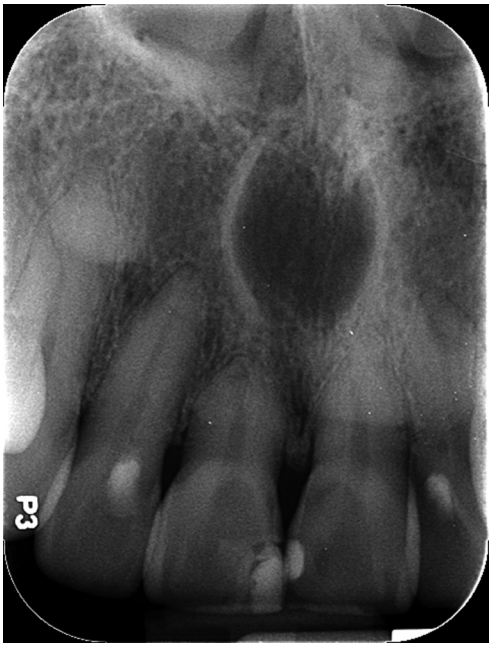

Fig. 2. Periapical radiograph demonstrating a well demarcated 9 × 9 mm midline maxillary radiolucency and shortened apices of the bilateral maxillary lateral and central incisors.

for NPDCs, which seldom form intraoral communications despite their expanded volumes.[5]

In NPDCs, in addition to the squamous epithelium, the nasopalatine duct is also known to contain cells of neuronal and inflammatory lineages.[3,4] In particular, sinonasal schwannoma, which is a benign neoplasm of Schwann cells, may arise from the various nerve-containing ductal structures of the maxilla and the face. The head and neck variants make up approximately 25% to 45% of all schwannoma presentations, whereas only about 4% of those arise within the sinonasal cavity.[6] There is no age, race, or gender predilection in the case of sinonasal schwannomas. Although typically asymptomatic, schwannomas may

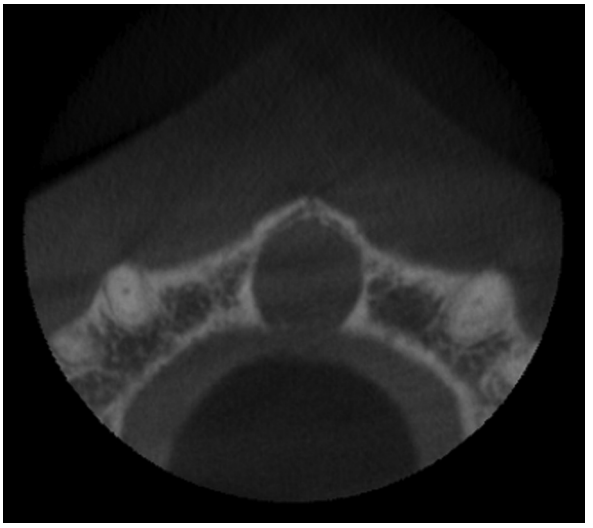

Fig. 3. Cone beam computed tomography axial image demonstrating midline maxillary 9 × 9 mm well demarcated unilocular radiolucency, labial bone thinning, and palatal bone thinning or perforation.

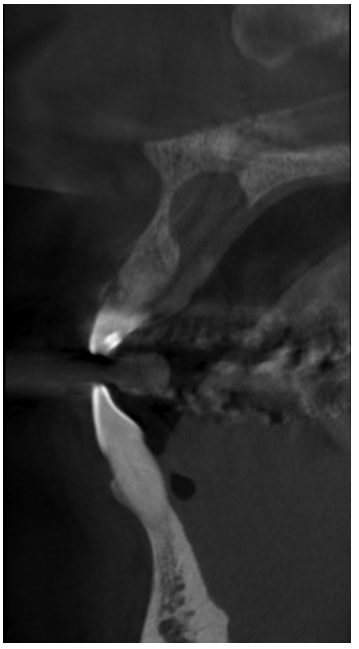

Fig. 4. Cone beam computed tomography sagittal image demonstrating labial bone thinning and palatal bone thinning or perforation in the area of the nasopalatine canal.

grow large enough to exert a mass effect on surrounding structures and present with symptoms, such as nasal obstruction, discharge, and anosmia.[7] Schwannomas of the sinonasal region are especially rare; they are rarely present without notable clinical symptoms because of their close association with neurosensory structures.

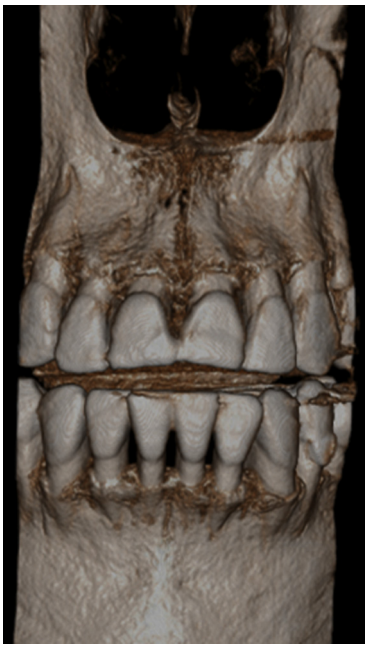

Fig. 5. Cone beam computed tomography three-dimensional image demonstrating a small midline periradicular area labial bone perforation just inferior to the anterior nasal spine.

In contrast, Langerhans cell histiocytosis (LCH) is an abnormal proliferation of inflammatory components—dendritic cells and macrophages—that may manifest in the anterior maxilla as an isolated disease of a single bone or as a systemic disease of multiple organs.[8,9] Its radiographic presentation often features a unilocular radiolucency of a flat bone that may or may not exhibit clear demarcation. Interestingly, LCH retains some of the characteristics associated with hematologic malignancies, such as the capacity to erode through calcified structures.[9] It is also associated with an unusually high rate of recurrence, approximately 60%, which often necessitates the use of chemotherapy and radiation.[9,10] Although LCHs show a general predilection for bony locales, including the nasopalatal area, they are several times more likely to arise within the mandible than within the maxilla.[8] Furthermore, studies have found that over 80% of LCH cases were seen concurrently with inflammatory, systemic presentations, such as skin lesions, hepatosplenomegaly, and prolonged fever, which were not evident in our patient.[8,9]

The current case may represent an odontogenic lesion as well. Two prominent odontogenic lesions with an established presence in the medial, anterior maxilla are periapical inflammatory disease (PID) and keratocystic odontogenic tumor (KCOT), previously known as odontogenic keratocyst (OKC). Of these, PID presents with the most common odontogenic lesion, with a wide spectrum of presentations that vary from simply inflammatory to cystic to granulomatous.[2] It is considered a natural sequelae of the bacterial invasion of the pulp that occurs as a result of decay or trauma.[11] PID lesions are decidedly radiolucent with well-circumscribed borders and may yet cause significant dissolution of the surrounding bone.[2,12] These radiolucent lesions have been known to produce swelling and cortical erosion, although both symptoms lack a directional preference and appear over a more generalized section of the face instead of resulting in a single discontinuity of the palatal cortex.[11] Not surprisingly, undeniable pain and discomfort also precede the physical destruction of oral structures as a result of the expansile, inflammatory mass,[2] as well as the physiologic stressors triggered by the presence of bacterial inflammation. PID is not possible in this patient's case, since all of the teeth in the area had vital, healthy pulps.

Compared with PID, KCOT/OKC is characterized by uncertain radiographic presentations and a greater potential for bony destruction. In the maxilla, it most commonly manifests as a lesion of the canine eminence, which often occurs in the vicinity of tooth roots with or without apparent association with an apex.[2] KCOT/OKC is notable for being one of the few pathologies that may cross the oral midline, although it typically originates in the posterior mandible and attains a midline presence by proliferating to such an extent as to resemble a borderline malignant lesion.[2,13] Interestingly, investigators have shown that maxillary midline presentations are predominantly found in men (72.2%) and in those over 60 years of age (88.9%).[13] Further analyses of KCOT/OKCs also have revealed their propensity for multilocularity, as is common with most rapidly growing lesions.[9]

DIAGNOSIS AND MANAGEMENT

At the time of biopsy, the lesion was removed in its entirety via a palatal full-thickness flap accessed through a sulcular incision. A peripheral ostectomy of the bony crypt was performed to ensure that all lesional soft tissues were removed. A mineralized allograft was placed into the bony crypt to provide bony support for the adjacent central incisors.

The biopsy specimen was evaluated by an oral and maxillofacial pathologist. Microscopy revealed a thick capsule of moderately cellular, dense, fibrous connective tissue surrounding the cholesterol clefts associated with a giant-cell reaction, with no evidence of epithelial cystic lining (Figure 6). A thick collagenous capsule surrounded aggregations of pale fusiform cholesterol clefts, many of which were partially surrounded by multinucleated giant cells. White, spindle-shaped cholesterol clefts, some of which were partially surrounded by multinucleated giant cells, showed dark

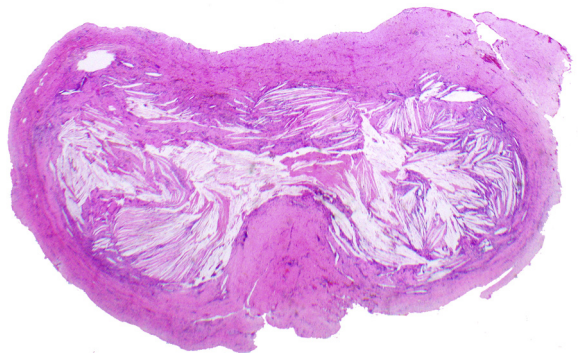

Fig. 6. A thick capsule of moderately cellular, dense, fibrous connective tissue surrounds cholesterol clefts associated with a giant cell reaction. No evidence of epithelial cystic lining was observed (H&E, magnification ×12.5). A high-resolution version of this slide for use with the Virtual Microscope is available as eSlide: VM02638.

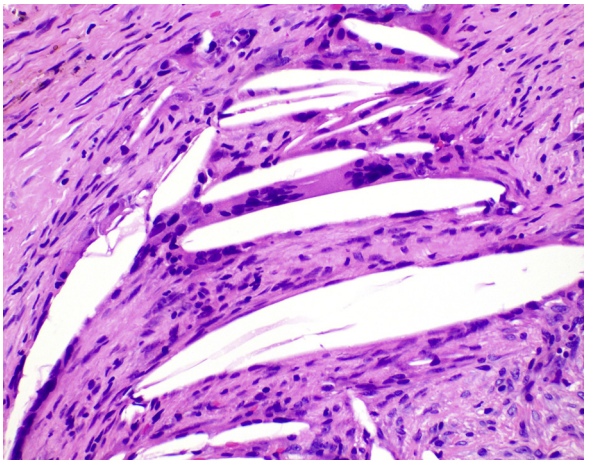

Fig. 7. White spindle-shaped cholesterol clefts, some of which are partially surrounded by multinucleated giant cells showing dark nuclei and abundant dense amphophilic cytoplasm (H&E, magnification ×400).

nuclei and abundant dense amphophilic cytoplasm (Figure 7). The absence of any foreign bodies within the specimen was also noted.

Clinical and histologic features led to a diagnosis of cholesterol granuloma. In light of the findings, no further surgical intervention was indicated. Following excisional biopsy and bone graft surgery, the patient recovered quickly. At 24 months following the procedure, she remains symptom free and pathology free, and all four of her maxillary incisors have maintained vitality with no more than class I mobility.

DISCUSSION

Cholesterol granulomas are histologic entities that present as fatty depositions within bony structures.[14] The lesions are typically identified histologically by visualizing collections of thin cholesterol crystals and fibrous tissue within a granular mass accompanied by foreign body giant cells and macrophages.[15] Cholesterol granulations may or may not be accompanied by epithelial elements surrounding the fatty deposits with a cystlike appearance in radiographic examination.[16,17] Although their exact etiology remains unclear, cholesterol granulomas are thought to occur as a result of poor ventilation of the lymph and/or air. They remain rare entities in the mediofacial region (including the mouth) but show a strong predilection for the aerated regions of the head, such as the middle ear and the mastoid air cell complex.[18,19] Although maxillary cholesterol granulomas are slightly more common, only 43 or fewer cases have been documented in the English literature by 2010.[17,19]

The most commonly accepted model of pathogenesis suggests that trauma and bleeding in enclosed spaces lead to entrapment of erythrocytes, which rupture upon death to release cholesterol crystals and membrane lipids. The precipitating cholesterol particles are perceived as foreign bodies and taken up by macrophages, which, in turn, (1) transform into engorged histiocytes because of their inability to properly disintegrate cholesterol and (2) release inflammatory mediators that initiate bone resorption and granulation.[20] Despite the absence of pneumatized structures within the alveolar bones, intrabony cavities, such as ducts, theoretically provide an environment in which trauma to the surrounding vasculature could cause leakage into an enclosed space without proper drainage.[19] Traditional cholesterol granulomas typically occur in young or middle-aged men,[17,21] and when it occurs in the nasal or paranasal sinus area, it may cause allergy-like symptoms, including nasal obstruction and discharge.[15] In certain cases, the lesion may present as a nasal polyp, seen as partial opacification on radiographs.[22] The inflammatory destruction of nearby structures as well as the risk of facial pain necessitates their removal (prophylactic and otherwise) via surgical means, which, in the case of nasal presentations, may be conducted endoscopically.[22,23]

In their study of periapical biopsies, Slutzky-Goldberg et al.[14] explored the rare and destructive presentations of cholesterol within periapical cysts in adolescents and older adults and found that the rate of cholesterol granulation increased with age. Additionally, the authors indicated that osteolysis from cholesterol deposition within the craniofacial region poses another threat to those with hypercholesterolemia.

Although an oral presentation is not expected, cholesterol granuloma should be considered in the differential diagnosis of mediofacial oral lesions,

especially in light of the increasing incidence of obesity and hyperlipidemia in the developing nations. The benign features and the low recurrence rate of cholesterol granuloma indicate that early detection can eliminate virtually all unfavorable sequelae.[17]

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