Primary Mandibular Xanthoma: Case Report and Review of the Literature

Primer Mandibuler Ksantoma: Olgu Sunumu ve Literatür Derlemesi

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Geliş Tarihi/*Received:* 26.04.2015
Kabul Tarihi/*Accepted:* 14.09.2015

This study was presented as poster presentation at 17th International Congress on Oral Pathology and Medicine, 25-30 May 2014, İstanbul, Turkey

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doi: 10.5336/dentalcase.2015-45878

ABSTRACT Xanthomatosis is yelow pigment accumulation which occurs as a result of alterations in lipid and cholesterol metabolisms. Primary xanthomas are idiopatic forms of xanthomas and not linked with lipid and cholesterol metabolism. Primary xanthomas are generally asymptomatic and mostly localized on mandible. They are observed as well-defined, sclerotic unilocular or multilocular lesions on panoramic radiography. However, aggressive forms with ill-defined borders and marked expansion are presented in the literature. In this study, a rare case of primary mandibular xanthoma was presented with comprehensive review of the literature.

Key Words:: Xanthomatosis; foam cells; mandible

ÖZET Ksantomatozis lipid ve kolesterol metabolizması bozuklukları nedeniyle ortaya çıkan, vücut dokularında sarı pigment birikimidir. Primer ksantoma, lipid veya kolesterol metabolizmasında herhangi bir sorun olmadan nedeni belli olmayan bir şekilde ortaya çıkan ksantomaya verilen isimdir ve çok nadir görülmektedir. Klinik olarak genellikle asemptomatik olan primer ksantoma, radyolojik olarak genellikle alt çenede sklerotik sınırlı, uniloküler veya multiloküler kistik görünümdedir. Ancak sınırları belirgin olmayan ve ekspansiyon gösteren formları da görülebilmektedir. Bu çalışmada nadir bir primer mandibuler ksantoma olgusu sunulmuş ve kapsamlı literatür araştırması yapılmıştır.

Anahtar Kelimeler Ksantomatoz; köpük hücreleri; mandibula

Turkiye Klinikleri J Dental Sci Cases 2015;1(4):229-31

Xanthomatosis is an accumulation of yellow pigment in the skin and internal organs in the form of a solitary tumor, presenting with alterations in the metabolism of lipids and cholesterol.[1,2] Xanthomatosis is rarely seen in bone and often associated with a secondary lipid metabolism altering disease such as hyperlipoproteinemia or hypercholesterolemia.[2]

Xanthomatosis is classified as primary xanthoma in the absence of secondary manifestations.[3] Primary xanthomas are extremely rare and mostly localized on mandible.[4-6] This study presents a rare case of mandibuler primary xanthoma with detailed review of the previously reported cases.

CASE REPORT

Twenty years old male patient referred to our clinic for routine dental examination. A panoramic radiograph was obtained and a radiolucent cystic lesion was observed in left mandibuler molar region (Figure 1). Patient did not have any complaints regarding the lesion and there were no signs of expansion or inflammation in the clinical examination.

Patient's consent was obtained and the lesion was removed with local curettage under local anesthesia with a provisional diagnosis of apical granuloma. The macroscopic appearance of the lesion was bulky and yellowish. Specimen was sent for histopathologic examination.

Microscopic examination revealed mononuclear inflammatory cell infiltration dispersed in collagenized fibrous connective tissue stroma. Focal cell clusters with pale cytoplasm also could be seen in the connective tissue stroma (Figure 2). These cells were foamy histiocytes which had lipid deposition with vesicular nuclei and vacuolar cytoplasms (Figure 3, 4). The case was diagnosed as mandibuler xanthoma. After histopathological diagnosis, patient was referred for further investigation to reveal any other metabolic disorder such as lipidemia or hypercholesterolemia. The lipid and cholesterol metabolism of the patient was in normal levels. Thus, the case was diagnosed as primary mandibuler xanthoma. Recovery of the surgical site was uneventful on postoperative 7th day.

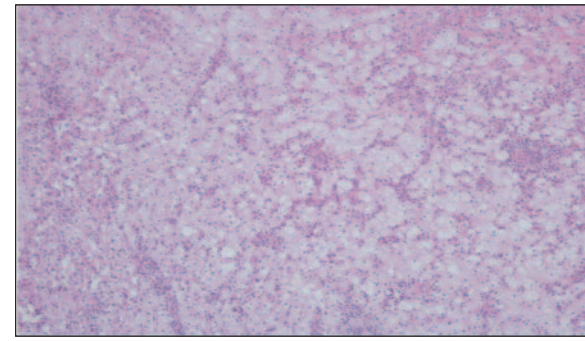

FIGURE 2: Histiocytic cells with pale cytoplasm can be seen among inflammatory cells in fibrous connective tissue (HE, x100).

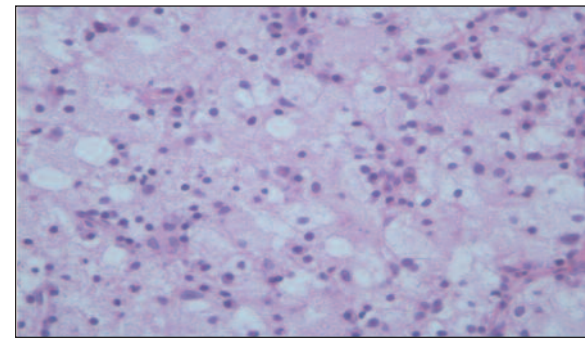

FIGURE 3: Inflammatory cells comprises mainly lymphocytes and histiocytic cells have vacuolar cytoplasm (HE, x200).

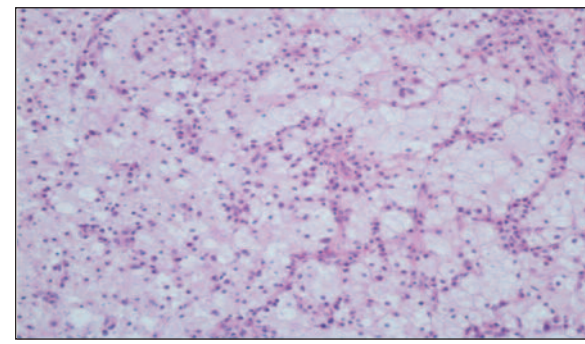

FIGURE 4: Histiocytic cells are filled with lipid and do not show nuclear atypia (HE, x400).

DISCUSSION

Bone xanthomas are usually seen with cutaneous manifestations and endocrine or metabolic disorders such as hyperlipoproteinemia, hyperlipidemia and diabetes mellitus. They are most commonly localized on hand and long bones, rib cranium and vertebra.[7-10]

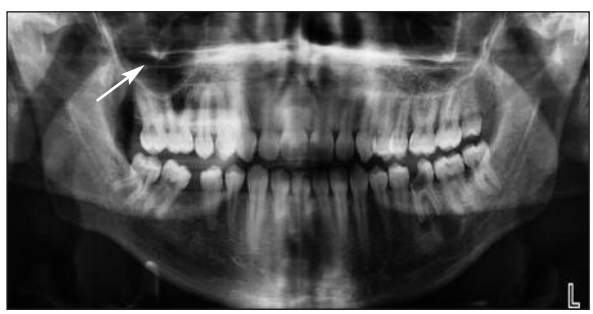

FIGURE 1: Radiographic appearance of the lesion.

Primary jaw bone xanthomas are extremely rare and occur exclusively on mandible. The previously reported primary mandibuler lesions which are diagnosed as xanthoma or xanthomatous lesions are shown in Table 1. Radiographic appearance is

TABLE1: Reported cases of primary mandibuler xanthoma.

Authors	Number of Cases	Publication Year	Histologic Features
Mosby et al.	1	1983	Histiocytes or foamy cells surrounded by fibrous connective tissue, secondary to pre-existing simple bone cyst
Harsanyi and Larson	7	1988	Sheets of histiocytes with foam cells, fibroblasts and fibrous tissue, and inflammatory cells as well as cells in transition with characteristics of both histiocytes and fibroblasts
Slootweg et al.	1	1993	Abundant foam cells combined with extensive reactive bone formation
Marques Mateo et al.	1	2004	Presence of an accumulation of histocytarian cells with a foamy cytoplasm with typical small round nucleus surrounded by immature fibrous tissue and adipocytes
Moraes Ramos Perez et al.	1	2011	Xanthomatous macrophages with foamy and granular cytoplasm and central small, round nuclei surrounded by scarce fibrous connective tissue

usually diffuse and ill-defined. Well-defined forms with sclerotic margins were also reported.[5] In cases with ill-defined borders, primary or metastatic bone malignant tumors should be considered in differential diagnosis.

Harsanyi and Larsson suggested that primary mandibuler xanthomas should be defined as histiocytic lesions.[1] Foam cells are a form of histiocytes and comprise droplets of fat which gives the cells a foamy aspect.[5] Primary xanthomas also include fibrous connective tissue, inflammatory cells and transitional cells which shows characteristics of both fibroblasts and histiocytes.[6]

The differential diagnosis of primary xanthoma includes a broad range of lytic jaw bone lesions such as radicular and residuel cysts, apical paradontitis and lateral periodontal cysts.[2,4] Generally, primary xanthomas have diffuse and irregular borders in radiography. Therefore, many authors include primary or metastatic malignant neoplasm in the differential diagnosis.[4]

The case in this study was pre-diagnosed as apical granuloma due to the close relation to the decayed left first molar tooth. However, patient was lost to follow-up after sutures were removed. Total removal of the lesion with surgical curettage is adequate for the treatment of primary bone xanthoma and recurrence was not reported. However, lipid or metabolic disorder associated xanthomas tend to recur after surgical removal.[1]

In this study, a rare case of primary mandibuler xanthoma was presented with prominent histologic features. Comprehensive literature search revealed eleven cases of primary mandibuler xanthoma which have been reported in detail. Primary mandibuler xanthoma is a benign lesion, however, it may be confused with agressive or malignant lesions of the jaws.

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