Xanthomatosis is yellow pigment accumulation which occurs as a result of alterations in lipid and cholesterol metabolisms. Primary xanthomas are idiopathic 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.

Xanthomatosis; foam cells; mandible


Anahtar Kelimeler Ksantomatoz; köpük hücreleri; mandibula

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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. Xanthomatosis is rarely seen in bone and often associated with a secondary lipid metabolism altering disease such as hyperlipoproteinemia or hypercholesterolemia.

Xanthomatosis is classified as primary xanthoma in the absence of secondary manifestations. Primary xanthomas are extremely rare and mostly localized on mandible. This study presents a rare case of mandibular 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 mandibular 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 mandibular 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 mandibular xanthoma. Recovery of the surgical site was uneventful on postoperative 7th day.

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

Primary jaw bone xanthomas are extremely rare and occur exclusively on mandible. The previously reported primary mandibular lesions which are diagnosed as xanthoma or xanthomatous lesions are shown in Table 1. Radiographic appearance is
usually diffuse and ill-defined. Well-defined forms with sclerotic margins were also reported. In cases with ill-defined borders, primary or metastatic bone malignant tumors should be considered in differential diagnosis.

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

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

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.

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

<table>
<thead>
<tr>
<th>Authors</th>
<th>Number of Cases</th>
<th>Publication Year</th>
<th>Histologic Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mosby et al.</td>
<td>1</td>
<td>1983</td>
<td>Histiocytes or foamy cells surrounded by fibrous connective tissue secondary to pre-existing simple bone cyst</td>
</tr>
<tr>
<td>Harsanyi and Larson</td>
<td>7</td>
<td>1988</td>
<td>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</td>
</tr>
<tr>
<td>Slootweg et al.</td>
<td>1</td>
<td>1993</td>
<td>Abundant foam cells combined with extensive reactive bone formation</td>
</tr>
<tr>
<td>Marques Mateo et al.</td>
<td>1</td>
<td>2004</td>
<td>Presence of an accumulation of histocytarian cells with a foamy cytoplasm with typical small round nucleus surrounded by immature fibrous tissue and adipocytes</td>
</tr>
<tr>
<td>Moraes Ramos Perez et al.</td>
<td>1</td>
<td>2011</td>
<td>Xanthomatous macrophages with foamy and granular cytoplasm and central small round nucleus surrounded by scarce fibrous connective tissue</td>
</tr>
</tbody>
</table>

**REFERENCES**