Case Report

Solitary Peripheral Osteoma at a Curious Site with an Ambiguous Etiopathogenesis: A Case Report and Review of Literature
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Abstract
Solitary peripheral osteoma of the mandible is singularly accounted and only a few cases not related to the Gardner’s syndrome is offered in the literature. Osteoma is a benign osteogenic neoplasm. The most familiar site of occurrence in the mandible is the posterior part and it is customarily asymptomatic. Osteoma is usually ascertained accidentally in case of mandible where it poses as an esthetic predicament. Peripheral osteoma has a distinctive clinical and radiographic appearance, but its exact pathogenesis is incomprehensible. Enumerated here is a patient with a peripheral osteoma of the mandible at an atypical site; which might have occurred as a result of a combination of trauma and muscle traction.

Keywords: Osteoma; Neoplasms; Connective Tissue; Bone Tissue; Mandible; Ambiguous; Muscle Traction; Pathological Conditions; Signs and Symptoms.


Introduction
Osteoma is a benign osteogenic lesion which according to Lichtenchstein, “is composed essentially of osteoblastic connective tissue forming abundant osteoid and new bone which may eventually become compact over a period of time”.

Osteomas may be grouped on the basis of their site of derivation as central, peripheral, or extra skeletal. Central osteomas surface from the endosteum, peripheral osteoma from the periosteum, and extra-skeletal soft tissue osteoma evolve within a muscle. Osteomas may also be categorized as compact and cancellous on the histopathological and clinical grounds.

A peripheral osteoma of the mandible is exceptional. Clinically, peripheral osteoma is a well circumscribed slow growing hard mass which is habitually asymptomatic. The osteoma goes undetected unless found incidentally on a routine radiographic survey or when it expands to the extent that it causes facial asymmetry or functional impairment. Multiple osteoma of the jaws are a frequent finding in Gardner’s syndrome. There are a few cases that are not related to Gardner’s syndrome. Radiographically, osteoma appears as a well-circumscribed radiopacity.

The current report relates to an interesting case of a peripheral osteoma at a remarkable site of occurrence and with a possible combination of etiological factors. The case is successfully managed surgically, and the healing is uneventful.

Case Report
A 25-year old male, reports to the department of oral medicine and radiology, for an evaluation of a painless knob on the right inner aspect of the lower jaw. The prominence was first noticed three years back at the right lingual mandible. It has emerged from an almond size to the present dimension. The patient denies pain or inability to masticate but desires removal of the lump being malignant. There are no records of any previous facial trauma or a contributory family or medical history.

Extraoral examinations bare no obvious facial or mandibular asymmetry. Intraoral examination reveals an ovoid, nodular, sessile projection at the antero-lingual aspect of the right mandible, which measures approximately 6 cm (antero-posteriorly) X 5 cm (bucco-lingually) in diameter extending from the lingual cortical plate overhanging the floor of the mouth (Figure 1a). The swelling anteriorly crosses the midline and is found in relation to distal aspect of 32, involving 31 to mesial aspect of 46 respectively. The lower anterior show imbrication with marked distolinguinl...
Inclination of tooth #42. The palpation of the protuberance confirms the inspection findings. The nodular protuberance shows bony hard consistency and appears to be in continuity with the underlying bone. The overlying skin appears slightly blanched and movable over it. Other findings on inspection include, mild local deposits of plaque and calculus with mild gingival inflammation.

Radiographic examination (Figure 1b & c) depict a well circumscribed ovoid radiopaque bulge associated with the right mandibular body seeming to arise from the lingual cortical plate crossing the midline to the contralateral incisors. The well demarcated protuberance appears to resemble bone of a cancellous quality. A written informed consent for the bone biopsy procedure was obtained (Figure 1d & e), and the same performed for the further evaluation of the swelling. The histopathological report (Figure 1f) depicts a vascular connective tissue stroma containing trabeculae of bone outlined by osteoblasts. Also, evident are areas of osteoclastic bone resorption. This was found to be consistent with that of a cancellous osteoma, which corroborates the clinical and radiographic picture.

Surgery of the peripheral osteoma was considered as the treatment of choice for the case. Surgical management involved excision of the osteoma under local anesthesia. Mucoperiosteal flap of the lingual gingiva was raised via blunt dissection by means of a periosteal elevator extending from 32 to 47 and extraction of 42 was performed. The initial cut was outlined using a surgical bur and the pedicle in conjunction with the nidus was resected through chisel and burs. Subsequently, the surgical site is smoothened with bone files and burs followed by wound debridement and finally closed by way of interrupted sutures. The surgical management of the patient was successful and was recalled a year later for a review follow-up.

Discussion
Osteoma is the proliferation of compact or cancellous bone characterized by a very slow, continuous growth. The peripheral osteoma is a tumor of mature bone of two varieties as per histopathological and clinical footings. Compact or “ivory” osteoma is histologically a normal-appearing dense bone with minimal marrow spaces, and occasional haversian canals. Clinically ivory osteoma has a sessile base which extends to a wide range of proportions. Alternatively, the cancellous osteoma histologically resembles the bone of origin, and contains trabeculae of bone and fibro-fatty marrow with osteoblasts. Clinically it is, in general, pedunculated with irregular or smooth surface. Conversely, the present case illustrates a sessile cancellous peripheral osteoma.

Peripheral osteomas occur more frequently in the paranasal sinuses such as the frontal, ethmoidal and maxillary sinuses. Additional documented locations in the craniofacial structures include external auditory canal, orbit, temporal bone, pterygoid processes, and infrequently, in or on the jaws. A solitary peripheral osteoma of the jaw bones is quite exceptional, involving the mandible more often than the maxilla. The most affected areas in the mandible are the posterior body, followed by the condyle, angle, ascending ramus, coronoid process, anterior border, and sigmoid notch. The article depicts peripheral osteoma at the right lingual part of the mandible crossing the midline.

Kaplan et al. reported the age at which the lesions occurred to be between 15 - 75 years, the majority being noticed after the age of 25. The duration of the lesions varies between one and 22 years. There is no sex predilection as noted by several investigators whereas Schneider et al have noted, previous reports favoring cancellous type in females and compact osteomas in males is not true as the case exemplifies.

The precise pathogenesis of peripheral osteoma is still unknown. Some investigators classify it as a reactive condition triggered by trauma, as peripheral osteomas is generally located on the lower border or buccal aspect of mandible which is the frequently traumatized area while others consider it as a true neoplasm. Peripheral osteoma is assumed to be benign in nature because in the preponderant number of cases its growth potential and rate deem to be limited. Twenty-four percent of cases of peripheral osteoma of mandible is associated with a history of trauma which may cause sub-periosteal bleeding or edema that simulate an osteogenic reaction. Trauma may be minor and may be unlikely to be remembered by the patient years later.
Bony hyperplasia associated with muscle traction is a documented phenomenon.\textsuperscript{4,6,10} It is suggested that a combination of trauma and muscle traction may play a role on its development. Either one or both might imitate an osteogenic reaction that could be perpetuated by continuous muscle traction in the area\textsuperscript{8,10} as might have transpired in our case.

\textbf{Figure 1:} The intraoral (a) and radiographic examination (b & c) reveals an ovoid swelling with sessile base depicting a well circumscribed radiopaque mass at the lingual aspect of the right mandible. The excisional biopsy procedure was carried out (d) and gross macroscopic examination revealed multiple bits of bony hard specimens (e). The histopathological examination shows trabeculae of bone scattered within vascular connective tissue (40x, H \& E stain)(f).

Also, another etiopathogenesis in the literature is that of an RFB osteoma virus (ectropic C-type retrovirus) and their clones which have shown to induce multiple
osteomas in mice and exhibit close relatedness to the Akv virus (endogenous, ecotropic murine leukemia virus). The data suggests that retroviruses directly affect osteoblast progenitor cells as primary target cells in the skeleton, resulting in the continuous formation of new bone. A number of molecularly cloned ecotropic, N-tropic murine leukemia viruses (MLVs) induce osteomas and osteopetrosis together with malignant lymphomas when the viruses are injected into newborn NMRI (Naval Medical Research Institute) mice.6,20 The virus-induced bone lesions are characterized by localized increased bone formation in the affected animals.19,20 In the present documentation the likelihood of a subclinical infection cannot be entirely ruled out.

Another interesting correlation of familial history or genetic transmission may be taken into account for osteoma, although a chance probability of an osteoma occurring in a mother and a child is 0.0016%, there may be a possibility of a dominantly inherited predisposition to osteoma formation in humans, or a recessive trait with a vertical transmission in inbred strains of mice.21 Furthermore, the discovery of an osteoma of the facial skeleton should raise the possibility of Gardener’s syndrome (GS).2,8,9 Patients with GS present with symptoms of rectal bleeding, diarrhea, and abdominal pain. The triad of colorectal polypsis, skeletal abnormalities, and multiple impacted or supernumerary teeth is consistent with this syndrome. Onset occurs in the second decade, with malignant transformation of the colorectal polyps approaching 100% by the age of 40. The skeletal involvement includes both peripheral and endosteal osteomas, which can occur in any bone but are found more frequently in the skull, ethmoid sinuses, mandible, and maxilla.5,14,15

Gardener’s syndrome is considered a variant of familial adenomatous polyposis (FAP) with certain extra-colonic manifestations (such as osteoma, gastric or duodenal polyposis and desmoid fibromatosis). It was reported that GS is caused by truncating mutations of the APC gene (codons 1403 and 1578) differing from classic FAP (codons 169-1600), attenuated FAP (amino terminal to codon 157), and congenital hypertrophy of the retinal pigmented epithelium (codons 463-1387).22,23 The APC protein is thought to mediate the stability of beta-catenin in the WNT signaling transduction pathway (‘wingless-type mouse mammary tumor virus integration site family member’) in normal colonic epithelial cells, thereby indirectly regulating the expression of WNT target genes such as the c-myc-oncogene. APC gene mutations cause the development of multiple adenomatous polyps in the colorectum, which strongly predisposes gene carriers to colorectal cancer along with other extracolonic manifestations like that of osteomas.24 However, there is evidence that patients with identical mutations may have different phenotypic expressions because of unclear reasons. The majority of GS patients may have a family history, but about 25% of GS patients can present with a new dominant mutation and are the first affected member of the family.23 Nonetheless, the exact etiopathological mechanism remains a blur so far, hence research is dictated towards establishing the etiopathogenesis of such a benign tumor, in humans, which may be syndrome associated with a possible cofactor for malignancy, especially the one with a viral etiology. However, no corroborating syndrome lesions were found in the present case report.

Peripheral osteoma is distinguished clinically and based on history from other growths occurring on the jaws like exostoses. Exostoses are bony excrescences that occur on the buccal aspect of alveolar bone. These lesions are of reactive or developmental origin and are not true neoplasms. They exhibit variable growth and stop growing by childhood.2,5 Osteoblastomas and osteoid osteomas, which perhaps ought to be considered, are more frequently painful and exhibit a more rapid rate of growth in comparison to osteomas. Osteomas may also be confused radiographically with sclerosing osteitis or odontomas. Sclerosing osteitis is differentiated based on its margin, which is ill-defined and is usually associated with some cause such as a retained root or an infected tooth.1,2 Odontomas on the other hand are usually surrounded by a radiolucent soft tissue capsule, the absence of which in osteoma is significant in its differentiation.1

With regard to the management of an osteoma, surgery is necessary for symptomatic ones that are growing continuously. Also, for the rationale of displeasing aesthetic aspect or through choice (oncophobia) or as in the present...
case which also includes speech impediment and limitation to maintain oral hygiene. The most controversial issue involves osteomas that are asymptomatic as neither they are recurrent, nor associated with a malignant change. The individualized assessment of the size and site of the tumor is essential for determining the possible complication derived from a wait and see approach and those derived from surgery. In case of surgical management, complete resection in critical areas is not necessary when the risk of surgical damage is high as the recurrence is truly rare. In the case of mandibular osteoma, the intraoral approach is preferable to the extraoral, as there is less surgical morbidity.

In the present case complete surgical removal was done as the risk was minimal and the prognosis was good. The surgical approach was carried out through intraoral incision. As is appropriate the patient was scheduled for periodic review and follow up after surgical removal of the osteoma.

Conclusion
Osteomas are benign tumors with unpredictable growth and with no probability of malignant transformation. Peripheral osteoma not associated with Gardner's syndrome is seldom documented in the literature especially solitary ones occurring at the lingual aspect of the mandible crossing the midline. Also, the case presents to involve multiple pathogenic factors. Hence, an astute clinician must not rule out the possibility of an osteoma at such a site which may consist of an indefinite etiopathogenesis.

In addition, research is dictated towards the conduction of conclusive human studies for establishing the accurate etiopathogenetic factor or factors, as in case of viral factor which may preclude malignancy leading to the speculation of the presence of an osteoma as a possible hallmark of cancer. Furthermore, when syndrome linked they establish an important maxillofacial clinical sign of premalignancy which then requires prompt diagnosis and management. The management of the asymptomatic osteoma is controversial. Each case should be assessed individually according to the size and location considering the risk of the surgery to that of a watch and wait strategy, in order to decide/determine the management options.

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References

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