Peripheral ameloblastoma (PA) is an uncommon subtype of ameloblastoma. In fact, it is the rarest variant, accounting for about 1 to 10% of all ameloblastomas.1 In contrast to the intraosseous locally aggressive ameloblastoma, PA is a soft tissue tumor with indolent biological behavior.2,3 It has been theorized that this tumor may arise from remnants of dental lamina beneath the oral mucosa or from the basal cells of the oral epithelium.4,5 PA tends to occur between the 5th and 7th decades of life with an average reported age of 52 years.2 Clinically, PA is usually a painless lesion that commonly affects the mandibular gingiva.6 Because PA has non-specific clinical presentation, it is mostly considered as pyogenic granuloma or fibroma.6 We report a case of PA that was observed in a 32-year-old man presenting with a gingival swelling in the lingual mandibular premolar area.

CASE REPORT

A 32-year-old Indian heavy-smoker man presented to the oral surgery clinic with painless gingival swelling on the left side of the mandible. The lesion was asymptomatic and had been noticed by the patient two years ago before the consultation. Intra-oral examination showed a sessile pink to red in color gingival mass and was located on the lingual gingiva between the left mandibular 2nd premolar and 1st molar (Fig. 1). The lesion was firm with granular surface. Orthopantomogram (OPG) was taken and showed no remarkable changes (Fig. 2). Clinically, the diagnosis was pyogenic granuloma based on the clinical presentation. Under local anesthesia, the lesion was surgically excised with 1 mm safe margin, formalin-fixed and sent for histopathologic examination. Grossly, the excised specimen was a whitish-brown, firm soft tissue mass measured 1.3 cm x 0.7 cm x 0.3 cm. Microscopically, the examination showed multicentric down-growth and

Figure 1: Intra-oral picture of gingival mass on the left mandibular premolar-molar area.
budding of the basal layer of the surface epithelium with some tumor cells were observed lying free in the connective tissue stroma (Fig. 3). Most of the proliferating islands exhibited peripheral rows of palisaded columnar cells that showed reverse polarization and hyperchromatism with infrequent mitosis. The major pattern of the proliferating nests is follicular with some areas showing acanthomatous and cystic changes (Fig. 4). In the lumen of some of the cystic areas neutrophil infiltrate was seen. No cell atypia or pleomorphism was noted. The connective tissue stroma was collagenous and mildly infiltrated by chronic inflammatory cells. Additional immunohistochemical studies for CK19, Ber-Ep4 and Ki-67 were ordered. The tumor islands were partially positive for CK19 with marked reaction in their luminal areas (Fig. 5), and negative for Ber-Ep4. Ki-67 was positive in 5% of the cells, which were found mainly in the basal/parabasal layers of the epithelial islands (Fig. 6). Based on the clinical, histopathology and immunohistochemistry results, the lesion was finally diagnosed as peripheral ameloblastoma.
 Odontogenic tumors are uncommon unique lesions of the jaw bones. They comprise a heterogeneous group of lesions that is derived from epithelium or ectomesenchym, or both.\(^7\) Though these lesions share the common origin from the tooth-forming apparatus, they possess diverse histopathologic forms and clinical behavior. Some of these lesions represent a true neoplastic while others are a tumor-like hamartomatous lesions.\(^8\)

PA is a rare soft tissue odontogenic tumor that was first described in the literature by Kuru in 1911.\(^9\) In 2005, the World Health Organization (WHO) classified PAs as one of the distinctive subtypes of ameloblastoma.\(^1\) This is because of its extraosseous location and less aggressive clinical behavior compared with the conventional ameloblastoma.\(^10\) Although PA is rare, it is the 2\(^{nd}\) most common peripheral odontogenic tumor (28\% of cases), preceded by odontogenic fibroma.\(^11\) Similar to the conventional ameloblastoma and other odontogenic tumors, the etiopathogenesis of PA is poorly understood and the origin is still unclear. Nevertheless, hypotheses have been suggested that it could originate from the epithelial rests of dental lamina or from the basal layer of the oral epithelium.\(^4,5\) In fact, many of the reported cases of PA (one of them is our case) were described to be in connection with surface epithelium suggesting PA might be derived from the surface epithelial layer.\(^12,13\) On the other hand, other cases were reported to be completely in the connective tissue without contact with surface epithelium suggesting that it might be to be derived from dental lamina residuals.\(^2,5,14\) Genetically, one study reported genetic aberration in chromosome 7 (trisomy) in PAs which may play a role in its tumorigenesis.\(^15\)

PA is typically presented as slowly growing painless mass with smooth surface. It is more in males than females by ratio of 1.9:1, and frequently occurs on the middle age of life with mean age of 52 years.\(^2\) In regard to the location, it is most commonly found on the mandibular premolar region with percentage of 33\%.\(^2\) Our reported case presented most of these clinical characteristics: A male patient with gingival swelling on the premolar-molar area. However, the patient here is 32 years old, which considered younger than the reported mean age of PA.

In PA, the step of detailed radiological investigation is essential to rule out any bone involvement and the misdiagnosis of central ameloblastoma. In the current case, radiological exam didn’t indicate any bone involvement, which directed the diagnosis toward peripheral lesions.

Although PA is rare extraosseous tumor, the clinical diagnosis of gingival swellings should involve PA as differential diagnosis since it resembles clinically other gingival swellings including pyogenic granuloma, traumatic fibroma, peripheral odontogenic fibroma, peripheral giant cell granuloma, and peripheral ossifying fibroma.\(^3\) Those lesions could be excluded based on the histopathological examination.

Under microscope, PA should be differentiated from peripheral squamous odontogenic tumor, peripheral odontogenic fibroma and oral basal cell carcinoma.\(^1,2,16\) The present case showed the typical features of ameloblastoma with islands exhibiting follicular and acanthomatous patterns. It has been shown that those two patterns are the most common in PA.\(^2\) Occurrence of calcifications in PA is not common finding, although it was reported recently in one case.\(^11\)

Currently, the role of immunohistochemistry is limited in diagnosing odontogenic tumors as these tumor types are largely identified by their morphologic features. However, some markers can be used to help in distinguishing the odontogenic tumors from non-odontogenic ones. For this purpose, it has been shown that CK19 and Ber-Ep4 can be used to differentiate between PA and basal cell carcinoma\(^16\). Our case showed diffuse positivity for CK19 and negativity for Ber-Ep4 confirming the diagnosis of PA. In addition, the positive reaction of PA neoplastic cells to CK19 indicates that PA is most likely to originate from remnants of odontogenic epithelium.\(^2\) The present case also showed low proliferating activity (Ki-67=5\%) which indicates low-level of aggressiveness of this tumor compared with intraosseous ameloblastoma that was shown to have greater expression of the proliferative markers.\(^5,17\) In fact, the labeling index of Ki-67 has been linked to the patient’s age, the gross appearance of the excised specimen (solid, mixed, cystic), general histological patterns and the cytological pattern of the outer layer cells of ameloblastoma.\(^17\)

Because of the indolent biological behavior of PA, it has been recommended that this tumor can be treated conservatively by local excision with small free margin.\(^2,10\) Up to date, there is no reported cases of malignant transformation of PA, although some cases have been published in regard to PA with malignancy.\(^2,18\) The recurrence rate is low, and close follow up is advised, especially for PA with atypical features.\(^10\)
CONCLUSION

We have reported a case of peripheral ameloblastoma in the gingival region because of its low incidence and the need to include this lesion on the differential diagnosis of swellings affecting the gingiva.

Author Contribution: DAQ collected the clinical and histopathological data, drafted, review and finalized the paper.

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REFERENCES


