

# Indolent palatal swelling: Catch 22

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## ABSTRACT

We present an interesting but intriguing case of an indolent palatal swelling. The lesion was asymptomatic causing little discomfort to the patient and thus was an incidental clinical finding. Provisional diagnosis was a benign, minor salivary gland tumor. Clinical differential diagnoses included benign lymphoepithelial lesion or mucus extravasation phenomenon. Nevertheless, we also considered malignancies such as mucoepidermoid carcinoma, lymphoma, and neoplasm of the maxillary sinus. However, the histopathology revealed a rare clinicopathologic entity prompting immediate treatment of the lesion.

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## CASE PRESENTATION

A 65-year-old male patient visited the Department of Oral Medicine of the Institute for getting his missing teeth replaced. On intraoral clinical examination of the patient, a deep red-colored multinodular but small swelling (measuring 1.5 cm × 2 cm × 2 cm) was observed in the left posterior palatal region (missing 26, 27, 28) extending toward the midline [Figure 1]. The maxillary left first and second molars had been extracted a few months ago. As per the patient's information, the swelling was observed nearly 6 months back and was found to be gradually increasing. However, there was no history of pain in this mass. On extraoral examination, the swelling was not apparently visible. On palpation, the mass was firm and fixed to the underlying tissues. Neither loosening of teeth nor tenderness on percussion was noted. Orthopantomograph and occlusal radiograph did not reveal any significant bone changes, suggesting that the lesion was still superficial [Figure 2]. The patient was diabetic for the last 22 years and was on medication for the same. Family history was noncontributory.

## DIFFERENTIAL DIAGNOSIS

The solitary, firm, asymptomatic, and nonulcerated mass seen on the palate instantly suggested a benign salivary gland tumor. Clinical differential diagnoses included benign lymphoepithelial lesion or mucus extravasation phenomena. However, we also considered malignancies such as malignant salivary gland tumor, lymphoma, and neoplasm of the maxillary sinus as the possible diagnoses.

Incidentally, our patient did not complain of swelling or discomfort; in fact, the patient's visit to the hospital was aimed at getting his missing teeth replaced. Clinical observation revealed a solitary, firm nodule on the palate covered by epithelium. The most commonly observed benign salivary gland tumors on the palate include pleomorphic adenoma (PA) and basal cell adenoma.<sup>[1,2]</sup> PA, also known as mixed tumor, is the most common tumor of the major and minor salivary glands, more prevalent in elderly males.<sup>[3]</sup> Intraorally, palate is the

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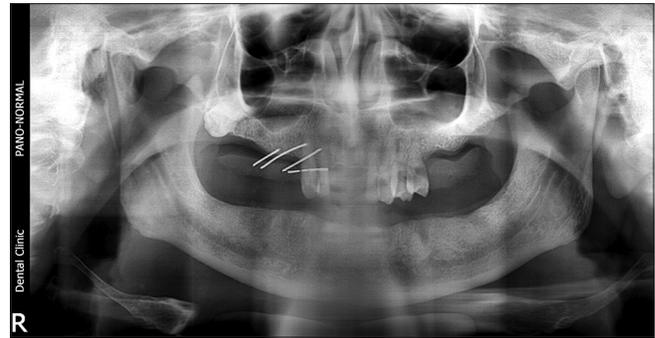
**Figure 1:** Small asymptomatic nodular swelling was observed on the palate

most common site where they appear as firm, painless swellings, and in the vast majority of cases, they do not cause ulceration of the overlying epithelium.<sup>[1]</sup> PA is typically lobulated and enclosed within a connective tissue pseudocapsule that varies in thickness within the major salivary glands, whereas in minor salivary glands, the capsule is poorly defined to absent in areas.<sup>[1,2]</sup>

Mucoepidermoid carcinoma represents the most common malignant tumor of salivary glands. They account for approximately 34% of parotid malignancies, 20% of submandibular gland malignancies, and 30% of minor salivary gland malignancies, with palate as the most common intraoral site.<sup>[1,4]</sup> Since there was no pain, paresthesia, or ulceration, the possibility of mucoepidermoid carcinoma was ruled out.

Lymphoma was considered plausible as the swelling was asymptomatic, showing spongy to firm tumescence; these features simulated our case. The clinical presentation of lymphomas of the oral region varies with their site of origin and tumor type, but most present as a mass or an ulcerated mass and resemble squamous cell carcinoma or salivary neoplasm.<sup>[1]</sup> Within the oral cavity, lymphoid tissue is chiefly represented in Waldeyer's ring; elsewhere within the oral cavity, it appears as unencapsulated lymphoid tissue within the base of the tongue and soft palate, as well as within the major and minor salivary glands. The tonsils are the most common oropharyngeal site, followed by the palate.<sup>[1]</sup>

Non-Hodgkin's lymphoma (NHL) is very often seen on the hard palate, primarily in elderly men and women with an average age of 70 years.<sup>[2]</sup> Oral lymphomas are relatively rare and often difficult to diagnose in a clinical setting, and most often mimic other pathological entities, such as dentoalveolar abscess, periodontal abscess, infected dental cyst, or benign jaw tumors.<sup>[5]</sup> NHL primarily appears in the head, neck, and jaw areas.



**Figure 2:** Orthopantomograph did not reveal bone changes

It typically is characterized by swollen and nonpainful enlargement of the lymph nodes, especially at the junction of hard and soft palates.<sup>[6,7]</sup> Although lymphoid lesions in the hard palate are very likely to be lymphomas, follicular lymphoid hyperplasia can present in this anatomic location; hence, a thorough histologic examination is of utmost significance. Histologic differentiation must be made among lymphoid hyperplasia, NHL, and benign lymphoepithelial lesion, when present in the hard palate.<sup>[1]</sup>

Since it was a nodular and nonulcerated swelling, trauma could have also caused the nodule formation because of mucus extravasation caused by the severance of salivary gland excretory duct.<sup>[1]</sup> Although palate is not the common site of mucus extravasation phenomenon, it usually presents as a relatively painless smooth-surfaced mass ranging in size from a few millimeters to 2 cm in size, consistent with our case. However, our patient reported that the mass was persistent for the last few months, which negated the possibility of a mucocele.

Last but not least, even neoplasm of maxillary sinus could be a reason for the palatal swelling. Occasional maxillary sinus cancers may present as a palatal ulcer and mass representing extension through the bone and soft tissue of the palate. However, sometimes, this swelling can appear without ulceration also.<sup>[1,2]</sup>

## DIAGNOSIS

Fine needle aspiration cytology of the lesion was done and the smear stained with Papanicolaou stain revealed mucin and keratin, which was nonspecific. Hematologic investigations revealed a normal blood picture. An excisional biopsy of the entire lesion was done. The specimen received was grayish white in color and soft in consistency. Three bits of soft-tissue specimen were subjected to routine tissue processing. Largest bit measured 1 cm × 1 cm × 0.7 cm in dimensions. Histopathology of the specimen revealed parakeratinized stratified squamous epithelium

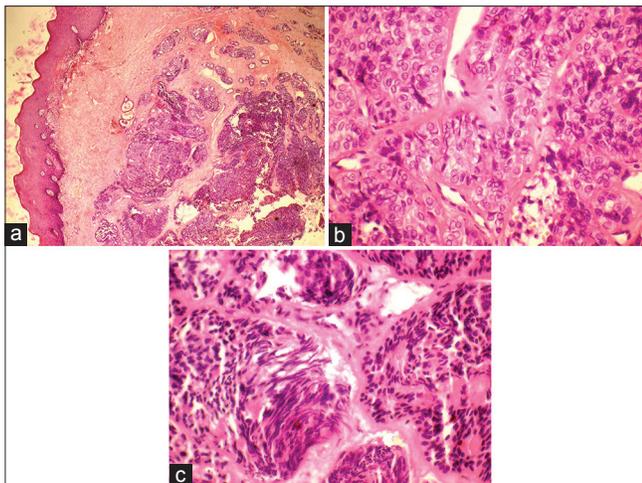
which was hyperplastic at areas with an underlying fibrocellular connective tissue stroma. The stroma showed unencapsulated, circumscribed tumor mass composed of homogeneous population of cuboidal to columnar isomorphic cells with prominent, bland, often vesicular ovoid to spindle-shaped nuclei and minimal eosinophilic cytoplasm. The tumor cells were arranged as solid nests, lobules, and glandular patterns extending till the entire depth of the received specimen [Figure 3a and b]. Perineural invasion was also evident at focal areas [Figure 3c]. Few areas of intraluminal mucin and hyalinization of stroma surrounding the tumor cells were evident. Mild lymphocytic infiltrate was also seen. On the basis of histopathologic findings, the report was signed out as polymorphous low-grade adenocarcinoma (PLGA).

## MANAGEMENT

Conservative surgical excision of the lesion was performed. The lesion healed uneventfully. Patient is on a regular follow-up and there is no recurrence reported even after 1 year of follow-up.

## DISCUSSION

Histopathologically, the most difficult neoplasms to differentiate from PLGA are benign mixed tumor and adenoid cystic carcinoma (ACC).<sup>[8]</sup> However, in contrast to PLGA, PAs are not infiltrative. Furthermore, neither benign mixed tumors do show neurotropism, nor they do demonstrate perivascular, osseous, or cartilaginous infiltration.<sup>[8,9]</sup>



**Figure 3:** (a) Low power magnification demonstrates parakeratinized stratified squamous epithelium and multiple lobules showing glandular and ductular patterns of tumor cells (H and E,  $\times 40$ ). (b) Tumor cells display prominent vesiculated appearance of the ovoid nuclei arranged in glandular patterns. Cellular atypia and multiple mitotic figures are also observed (H and E,  $\times 400$ ). (c) Perineural invasion by tumor cells is evident in focal areas ( $\times 400$ )

Possibly, ACC is the most difficult tumor to differentiate histopathologically from PLGA.<sup>[8,10]</sup> ACCs are most often found on the palate intraorally. They are also unencapsulated, infiltrating tumors that have a strong tendency for neural invasion. Some other features ACCs have in common with PLGAs include the histologic patterns such as cribriform, solid, and tubular. Nevertheless, the distinction between ACC and PLGA can be done based on histologic examination. For example, the nuclei of ACC are more hyperchromatic and more angular than those of PLGA. Cribriform areas with accumulation of basophilic glycosaminoglycans are observed frequently in ACC, not in PLGA. Moreover, the cytoplasmic staining of PLGA is eosinophilic to amphophilic, whereas that of ACC is very pale to clear staining.<sup>[8,10]</sup>

The distinction between ACC and PLGA is important because ACC is more aggressive and relentless clinically with multiple recurrences. On the other hand, PLGA is an indolent neoplasm where patients are free of the tumor after conservative wide surgical excision, with least requirement of postoperative radiotherapy and chemotherapy.<sup>[8,11]</sup>

Although considered to be a low-grade malignancy, a case of *de novo* PLGA was described which was observed in a minor salivary gland with a large radiographic extent. PLGA arising in a major salivary gland is considered extremely rare.<sup>[12,13]</sup> PLGA was earlier referred to as terminal duct carcinoma owing to its probable origin in the ductal system of the salivary glands.<sup>[14]</sup> PLGA is formed by luminal epithelial, myoepithelial, and basal epithelial cells simulating terminal duct carcinoma.<sup>[15,16]</sup> PLGA is a low-grade malignant neoplasm because of rare occurrence of regional lymph node metastasis, with even extremely rare manifestation of distant metastasis.<sup>[9]</sup> Microscopic or histopathologic observation of perineural invasion does not appear to affect the prognosis.<sup>[17,18]</sup>

This neoplasm manifests almost exclusively in minor salivary glands, with a high predilection for the palate.<sup>[9]</sup> However, some cases have been reported in major salivary glands, for example, in the parotid gland, where most cases have been diagnosed as part of a carcinoma ex PA, such as those in the case series reported by Kemp *et al.*, who found that only two out of twenty cases arose *de novo*, and the rest showed remnants of PA.<sup>[19]</sup> PLGAs arising in major salivary glands have clinicopathological and immunohistochemical features similar to those of PLGAs originating in minor salivary glands.<sup>[12]</sup>

The tumor, histopathologically, shows a uniform and bland cytological picture, a diverse yet characteristic growth pattern and a prominent neurotropism.<sup>[20]</sup> This clinicopathologic entity is, sometimes, a diagnostic

dilemma due to its multiple histomorphologic patterns and its cytologic uniformity. The histopathologic features simulate the picture observed in PA and ACC. Immunohistochemical studies comparing PA and PLGA have shown that glial fibrillary acidic protein is negative in PLGA and positive in PA.<sup>[21,22]</sup> However, PLGA differs from PA because it is characterized by infiltrative margins and an absence of chondromyxoid stroma.<sup>[1]</sup>

PLGA presenting as palatal lesion is not so rare and should be included in the differential diagnosis of palatal lesions, presenting as nonulcerated nodular swellings. Other salivary gland malignancies commonly seen on the palate, including mucoepidermoid carcinoma and ACC, should also be considered in the differential diagnosis. It is of prime importance to differentiate among these malignant lesions and reach a conclusive diagnosis as the management and prognosis varies for each of them. While ACC is associated with low long-term survival rates, PLGA is a low-grade malignancy, and its biologic behavior is apparently not influenced by the different morphologic and cell differentiation patterns that it may reveal. It is of prime importance for a pathologist to inform the predominant histologic pattern in PLGA, the presence of perineural invasion and vascular permeation, so as to affirm if these criteria are helpful to identify its biologic behavior.

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#### Conflicts of interest

There are no conflicts of interest.

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