Polymorphous Low–Grade Adenocarcinoma of the Palate:Report of an Interesting Case & Review of Literature

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Abstract
Polymorphous low grade adenocarcinoma (PLGA) is a malignancy arising predominantly from minor salivary glands. PLGA is the most common intraoral malignant salivary gland tumor which accounts for 10% of all tumors and 25% of all malignancies. It has been frequently described as occurring in hard or soft palate minor salivary glands, approximately 60% of the cases involve palate; some cases being described in tongue and in minor salivary glands. We report a case of PLGA of the palate diagnosed on the basis of histopathology and treated by local excision. The review of literature concerning clinical, histological and immunohistochemical features, as well as the proper management concerning this tumor is described.

Introduction
Polymorphous low grade adenocarcinoma (PLGA) is a malignant neoplasm with a low aggressiveness that usually affects the minor salivary gland, primarily in those of the hard palate[1]. PLGA was first described simultaneously in 1983 by two groups of researchers under different names. Batsak et al.[2] described it as terminal duct carcinoma and Freedman et al.[3],named it as lobular carcinoma. Evans and Batsak[4] in 1984, eventually coined the term, PLGA. The age of the patients at the time of the diagnosis of PLGA ranges from 16 to 94 years, with a mean of 59 years and with a female predilection[5].The duration of PLGA varies from some months to several years and the lesion is usually asymptomatic. Recurrence has been reported in 9%-17 % of the cases and regional metastasis has been reported in 9%-15% of the cases. The clinicopathological features of PLGA overlap Pleomorphic Adenoma and Adenoid cystic carcinoma, that may result in a diagnostic pitfall. The malignancy of PLGA is usually low, and metastases to regional lymph nodes and distant areas are uncommon. Local recurrence rates of PLGA range from 10% to 20% and regional metastases are up to 10% in literature. (Gnepp et al., 1988; Vincent et al., 1994). Nevertheless, three histologically confirmed cases of distant metastases from PLGA have been reported (Tanaka et al., 1995; Thomas et al., 1995; Hannen et al., 2000)[9] .Surgical excision is the treatment of choice and the survival in most of these cases is good.

Case Report
A 45-year-old male patient reported to the deptt of oral & maxillofacial surgery,santosh dental college & hospital with a complaint of growth in the palate since last one year. The growth developed one year back and was static in size initially, but a sudden increase in the size of growth was noticed since last three months. Patient gave history of smoking a few cigarettes daily from last twenty years.

A well circumscribed exophytic proliferative mass, measuring approximately 2cm x 1.5cm in diameter on the left side of the hard palate, not crossing the midline,

Fig. 1: Frontal profile photograph of the patient

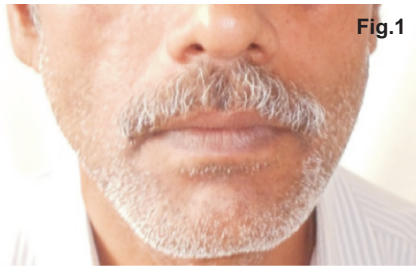


Fig. 2: Intraoral photograph showing a well-circumscribed mass on the left side of the hard palate

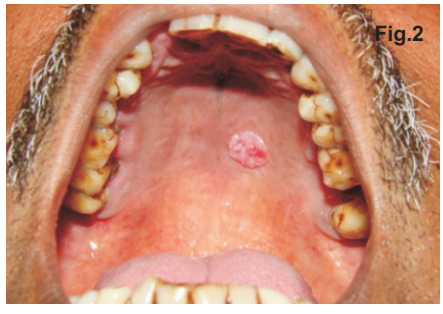
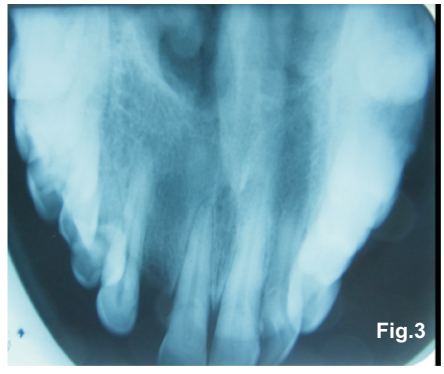
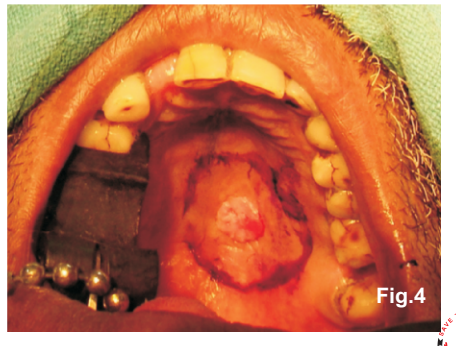


Fig. 3: Occlusal radiograph taken for hard palate revealed a radiolucent lesion involving the palate



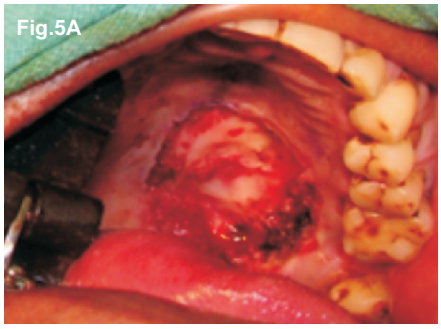
was present. It was firm in consistency and nontender. No bleeding on manipulation was noticed. Lymph nodes were nonpalpable. Hematological investigations revealed slightly raised ESR . The serum chemistry of the patient was normal. The patient was tested



for Hepatitis B and HIV I and II which were negative.

On the basis of the above findings, a clinical diagnosis of minor salivary gland pathology was made. Local excision of the growth was carried out with wide removal of the surgical margins and the specimen was sent for histopathological examination.

Fig. 4: Intraoperative photograph showing surgical markings,with the wide margins taken into consideration before resection of the mass



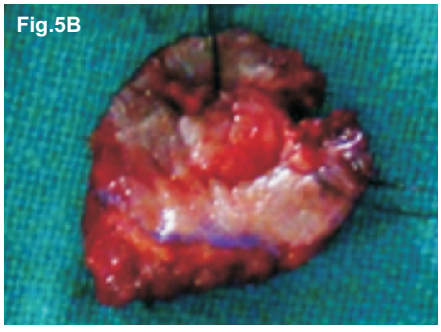


Fig. 5A and 5B: Palatal tissues after resection and resected mass

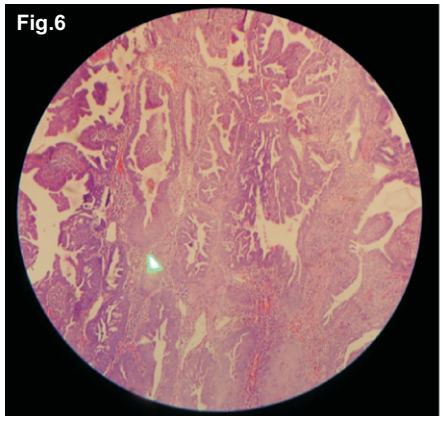


Fig. 6:Pictomicrograph showing the tumor cells in the form of large alveolar masses,glands lined by cuboidal epithelial cells and cells in whorls pattern

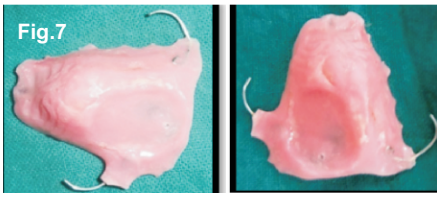


Fig. 7:Obturator for reconstruction of palatal defect

Discussion

Polymorphous low-grade adenocarcinoma (PLGA) is a distinctive salivary gland neoplasm with a propensity to arise almost exclusive from minor salivary glands and is associated with slow growth and an indolent biology. Previously, 'lobular carcinoma' and 'terminal duct carcinoma' were used as the terminologies for PLGA[2,3]. It is the second most common intraoral malignant salivary gland tumour, accounting for about 2% of all the salivary gland tumours[6,7] .PLGA occurs over a wide age range (16-94 years) with a mean age of 59 years, but it has not been found to occur in the first and second decades of life[8]. The tumour usually occurs in the palate (about 2/3rd), in the lip, buccal mucosa, alveolar ridge and the base of the tongue (in the remaining cases). The tumour ranges in size from 0.4 cms to 6 cms in the greatest dimensions, with a mean of 2 cms[4,8]. PLGA is a rare, malignant, salivary gland tumour, which is found almost exclusively in the minor salivary gland and it is very rarely seen in the major salivary glands. It is difficult to diagnose both clinically and histopathologically due to its indolent course and morphological diversity, which includes several microscopic pattern[6]. This tumour may display a mixture of growth patterns within a single tumour, including solid islands, glandular profiles, tubules, trabeculae, cribriform nests and linear or single file arrangements[6]. The tubular areas are lined one or two cell layers

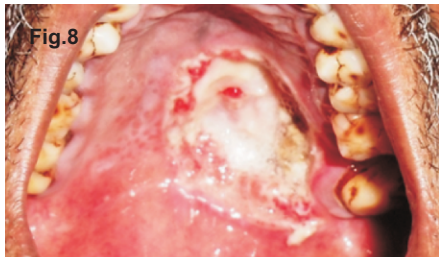


Fig. 8: One week postoperative photograph

of cuboidal to columnar cells. The tumour cells are uniformly round to polygonal, of small to medium size, with round to oval nuclei, with abundant pale to eosinophilic cytoplasm. Its nuclear pleomorphism is negligible and there are occasional mitotic figures. A characteristic slate gray stroma is frequently seen[7].

Because of its morphological variations, PLGA has often been misdiagnosed as pleomorphic adenoma or adenoid cystic carcinoma (ACC). However, PLGA differs from pleomorphic adenoma in the presence of infiltrative margins and an absence of chondromyxoid stroma. The differential diagnosis with adenoid cystic carcinoma, basal cell adenoma, and other salivary gland tumors is important because it does have an overall excellent prognosis.The primary difference between PLGA and ACC is based on both the cytological and the histological characteristics. The cell cytoplasm in PLGA is eosinophilic with rounded nuclear borders, while the cells in ACC are more basaloid with angled and hyperchromatic nuclei. It is important to distinguish ACC from PLGA because the former is associated with low long term survival rates. PLGA is a low grade malignancy and its biological behaviour is apparently not influenced by the different morphological and cell differentiation patterns that it may exist. Immuno-histochemistry has as such no apparent diagnostic value in identifying this tumour[1].

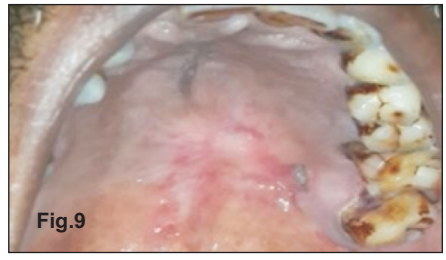


Fig. 9: Six months postoperative photograph showing complete healing and no clinical signs of recurrence

The treatment of PLGA consists of complete surgical excision with wide removal of the surgical margins. This procedure seems to be acceptable after full evaluation of the surgical margins, although it is not uncommon to have an incisional biopsy as the initial diagnostic procedure. This procedure is usually followed by an excisional biopsy or a wide local excision. There is no evidence that indicates any benefit from postoperative radiation or adjuvant chemotherapy, although both the modalities have been used[10]. Tumors localized to the hard palate are significantly more likely to be associated with tumor recurrence. Recurrence or persistence is present in about 10% of cases, emerging up to 14 years after initial presentation (mean, 7 years)[8].

In our case, the patient presented with a history of a mucosal ulceration with a nodular swelling on the palate and a clinical suspicion of minor salivary gland pathology. On excision of the mass with wide surgical margins, it revealed the features of PLGA and the patient is on regular follow up. To conclude, PLGA is an unusual tumour with low grade aggressiveness. Morphological pleomorphism is the hallmark of PLGA and so, histopathological examination is an important factor which helps in its diagnosis. IHC is not of particular help, but the differentiation of PLGA on light microscopy from pleomorphic adenoma and adenoid cystic carcinoma is necessary to yield an excellent long term clinical outcome with a conservative but complete surgical excision.

References

References are available on request at editor@healtalkht.com