Case Report

ID: JCDR/2012/3895:0043

Pathology Section

Polymorphous Low Grade Adenocarcinoma Presenting as Recurrent Oral Ulcerations

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ABSTRACT

Polymorphous Low Grade Adenocarcinoma (PLGA) is difficult to diagnose both clinically and histopathologically due to its indolent presentation and its morphological diversity, that includes several microscopic patterns. PLGA is a relatively newer entity under the sub-classification of adenocarcinoma, which arises from the minor salivary glands. It is an uncommon malignant neoplasm with low aggressiveness. We are reporting this case of PLGA which arose from the minor salivary gland over the right buccal mucosa with a nodular ulcerated swelling which was clinically suspected as pyogenic granuloma.

Key Words: Polymorphous Low Grade Adenocarcinoma (PLGA), Minor salivary glands tumour, Pleomorphic adenoma, Adenoid cystic carcinoma.

INTRODUCTION

Polymorphous low grade adenocarcinoma (PLGA) is a malignant neoplasm with a low aggressiveness, that occurs almost exclusively in the minor salivary glands, 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 [5]. Surgery is the treatment of choice and the survival in most of these cases is good. Recurrence has been reported in 9%-17 % of the cases and regional metastasis has been reported in 9%-15% of the cases [5].

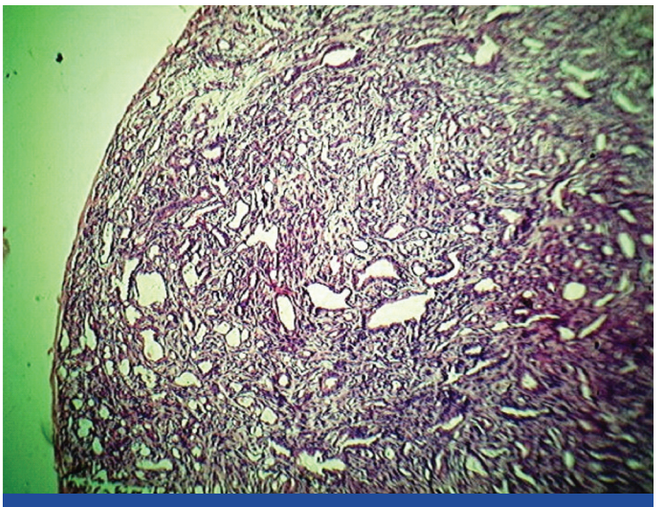
We are reporting here a case of PLGA which presented as a recurrent oral ulceration with a clinical suspicion of pyogenic granuloma.

CASE REPORT

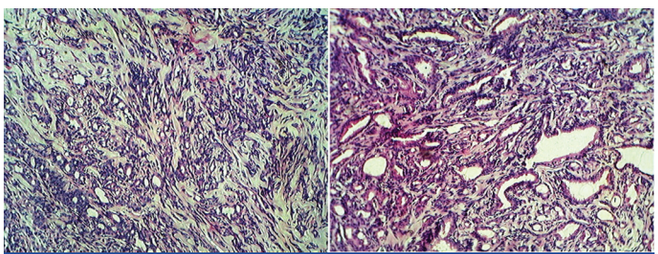
A 71-years old male patient presented to the surgical OPD with the chief complaint of recurrent, tiny, oral ulcerations since one and a half years. The patient was treated by local practitioners for symptomatic relief. The symptoms were however not relieved and the patient came to the surgery OPD in our hospital. On oral examination, an ulcer with a pink nodular swelling at its base was noted in the right buccal mucosa region. The ulcer measured 1.1x1 cm and the swelling measured 1.2x0.8 cm. Externally, the swelling was reddish in colour, with an ulcer which was covered by slough. The cervical lymph nodes were not palpable. The ear, nose and throat examination was within normal limits. On systemic examination, no significant diseases were noted. On the basis of the above findings, a clinical diagnosis of a chronic non-healing ulcer-pyogenic granuloma was made. Local excision of the nodular swelling was carried out and the specimen was sent for histopathological examination.

On gross examination, a well circumscribed lesion which measured 1.5x1 cm, with surface ulceration, was noted. Its external surface was reddish pink and its cut surface showed a gray-white nodular appearance.

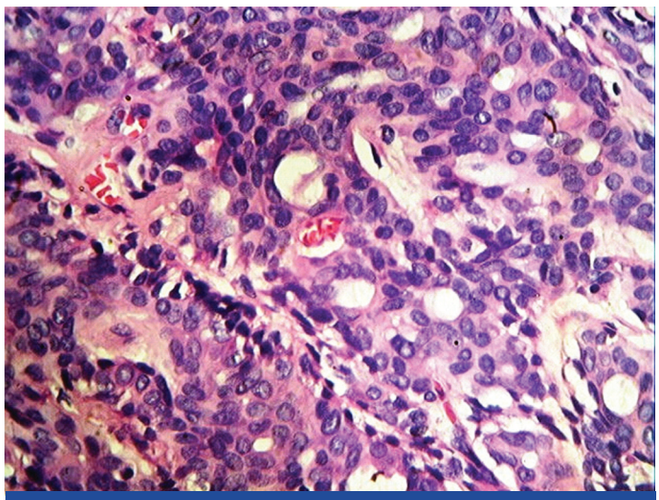
The microscopic examination – revealed a circumscribed tumour with focal areas of infiltrative growth at the periphery [Table/Fig 1].



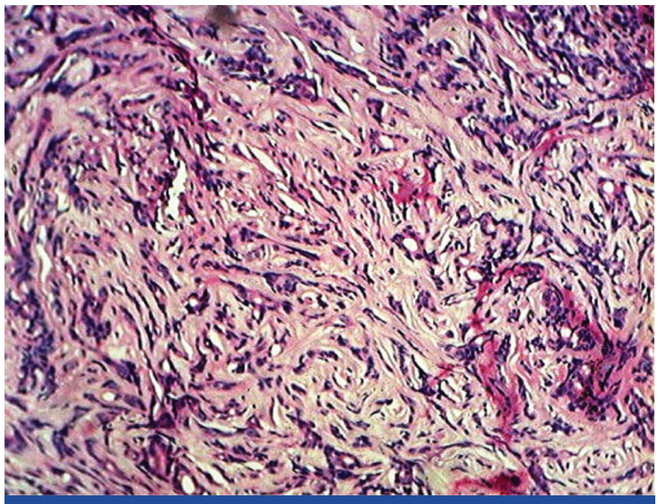
[Table/Fig-1]: Photomicrograph of well circumscribed salivary gland tumor having glandular and stromal components. (H & E stain, x100).



[Table/Fig-2 & 3]: Photomicrograph of tumor with variable architecture of neoplastic cells forming glandular, nodular, lobular and in cords appearance. (H & E stain, x100).



[Table/Fig-4]: High power view of tumor cells which are uniform round to oval, having bland nuclei with inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm.(H & E stain, x400).



[Table/Fig-5]: Photomicrograph showing Indian file or linear arrangement of tumor cells in hyalinised stroma.(H & E stain, x400).

The tumour was composed of cells which were arranged in the glandular, nodular, cystic, lobular and the whorls pattern [Table/Fig 2] and [Table/Fig 3]. The neoplastic cells were monomorphic and round to oval, having bland nuclear chromatin with occasional inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm [Table/Fig 4]

At places, hyalinized stroma with entrapped tumour cells was seen. The peripheral portion of the tumour showed an Indian file pattern of arrangement of the cells [Table/Fig 5]. The surface epithelium showed ulceration beneath in which the infiltration of many congested blood vessels and inflammatory cells was seen. The final histopathological diagnosis was given as PLGA which arose from the minor salivary gland at the right buccal mucosa.

DISCUSSION

PLGA has been recognized as a distinct salivary gland tumour that has a predilection to occur in the 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 0.4cms 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 patterns [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 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 [6].

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 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. Immunohistochemistry has as such no apparent diagnostic value in identifying this tumour [1]. But the tumour cells may be immunoreactive for cytokeratin, vimentin and S100, with variable results [6].

The treatment of PLGA consists of local excision with wide removal of the surgical margins. This procedure seems to be acceptable after full evaluation of the surgical margins and these are followed up by radiation therapy [9]. There is no evidence that indicates any benefit from postoperative radiation or adjuvant chemotherapy, although both the modalities have been used [10].

In our case, the patient presented with a history of a recurrent buccal mucosa ulceration with a nodular swelling and a clinical suspicion of pyogenic granuloma. On excision of the swelling 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.

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FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Submission: **Jan 02, 2012**
Date of Peer Review: **Jan 03, 2012**
Date of Acceptance: **Jan 04, 2012**
Date of Publishing: **May 31, 2012**