CASE REPORT

Polymorphous low-grade adenocarcinoma: a case report

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We present a case of *de novo* polymorphous low-grade adenocarcinoma (PLGA) arising in a minor salivary gland with a relatively large radiographic extent compared with that of most of the PLGAs reported. This paper describes the radiographic extent of the lesion and the findings of CT imaging.

Dentomaxillofacial Radiology (2013) **42**, 14804843. doi: 10.1259/dmfr/14804843

Cite this article as: Potluri A, Prasad J, Levine S, Bastaki JM. Polymorphous low-grade adenocarcinoma: a case report. *Dentomaxillofac Radiol* 2013; **42**: 14804843.

Keywords: adenocarcinoma; salivary gland neoplasm

Case report

A 60-year-old African American female was referred to the University of Pittsburgh School of Dental Medicine's urgent care clinic with a history of persistent swelling on the left side of her face, which was said to have been present for approximately 2 years. A history of third molar extraction in the area was reported to have occurred at about the same time as the onset of facial swelling. The patient stated that the swelling had gradually enlarged since then. A review of the patient's past medical history, based on a health questionnaire, revealed a history of asthma and possible pulmonary sarcoidosis. In addition, she also indicated a history of cigarette smoking. The patient reported using a nebulizer and oxygen to relieve the symptoms of chronic obstructive pulmonary disease (COPD).

Clinical examination of the head and neck revealed a firm swelling of the left cheek area. Pain or paraesthesia was not reported. Ocular involvement was not observed. An intraoral exam revealed a firm lobular mass, measuring approximately 4 × 2.5 cm, extending from the left posterior maxillary vestibule and alveolar ridge and involving the superior third of the left buccal mucosa. Upon palpation, the lesion was noted to have discrete submucosal borders. The mucosa overlying the lesion was noted to be predominantly pink, with

erythematous foci and focal ulceration posteriorly (Figure 1).

A panoramic radiograph showed an edentulous maxilla and retained mandibular teeth exhibiting heavy calculus deposits and moderate marginal bone loss. The left maxillary sinus floor was difficult to trace. Bone loss in the posterior left maxilla in the area of previously extracted teeth 14, 15 and 16 was also noted (Figure 2). Based on the panoramic radiographic findings and clinical presentations, the working differential diagnosis was a malignant salivary gland neoplasm. A benign salivary gland tumour or soft tissue neoplasm was also included in the differential diagnosis. Although less likely, a haematological malignancy was also considered.

A CT scan revealed a large, heterogeneously enhancing soft tissue mass on the left side of the face measuring 7.0 × 4.0 cm in the coronal plane and 4.0 × 3.3 cm in the axial plane. The epicentre of the mass was determined to be located lateral to the left maxillary sinus and medial and anterior to the left mandible (Figures 3 and 4). Mass effect was present with smooth medial compression of the tongue and medial bowing of the lateral wall of the left maxilla and left maxillary sinus, with partial bone destruction and no pathological fracture (Figure 4). The mass extended into the left infratemporal fossa superiorly with inferior extension superior to the left parapharyngeal space, contiguous with the left lateral aspect of the tongue (Figure 5). Portions of the mass were noted to be contiguous with both the left lateral pterygoid muscle and the left

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Received 11 February 2011; revised 14 April 2011; accepted 22 April 2011

Figure 1 A firm lobular mass extending from the left posterior maxillary vestibule

medial pterygoid muscle (Figure 6). The posterolateral margin of the soft tissue mass was noted to abut against the anterior aspect of the ramus of the left mandible.

Histopathological review of microscopic slides showed a well-circumscribed, non-encapsulated tumour. The surface epithelium was found to be intact. The tumour was composed mostly of lobules of monotonous cells with scattered duct-like structures. The tumour cells were uniform in size, with round to ovoid nuclei and pale eosinophilic cytoplasm. The nuclei exhibited open, vesicular chromatin with a characteristic bland “salt and pepper” appearance and small nucleoli. Although one study showed p63 immunohistochemical stain to be positive in 100% of polymorphous low-grade adenocarcinomas (PLGAs),¹ p63 in this case was negative in tumour cells, while S100 staining was strongly and diffusely positive (Figures 7 and 8).

The patient was scheduled for a left inferior maxillectomy with transoral excision of the buccal space. An intraoperative biopsy was consistent with a diagnosis of PLGA. Following the maxillectomy, a buccal fat pad advancement flap was made in that area and a pre-fabricated palatal prosthesis was fitted into the defect and closed. At 1 year follow-up there were no signs of local recurrence or metastases.

Figure 2 Panoramic radiograph showing posterior maxillary bone destruction and missing sinus floor

Figure 3 CT axial section, soft tissue window, showing the epicentre of the lesion, enhancement and extent

Discussion

PLGA of the salivary glands was first described by Freedman et al² who named it lobular carcinoma owing to its resemblance to lobular carcinoma of the breast. The term PLGA was then introduced by Batsakis et al in 1984.³ This tumour is now a well-recognized entity arising almost exclusively in minor salivary glands, with a high predilection for the palate.⁴ PLGA arising in a

Figure 4 CT bone window showing partial bone destruction of the maxilla

Figure 5 CT coronal section, soft tissue window, showing the extension of the lesion superior to the left parapharyngeal space

major salivary gland is considered extremely rare.⁵ The tumour is characterized microscopically by having a uniform and bland cytological presentation, a diverse yet characteristic growth pattern and a low metastatic potential with prominent neurotropism.⁶ The mean age of patients presenting with this tumour is approximately 58 years and the male-to-female ratio is approximately 1:2.⁴ In a large series of 164 patients

Figure 6 CT axial section, soft tissue window, showing the mass to be contiguous with both the left lateral pterygoid and left medial pterygoid muscle

Figure 7 A 100 × photomicrograph (haematoxylin & eosin) showing sheets and lobules of tumour cells

with mean follow-up of nearly 10 years, 97.6% of all patients treated with surgery only were either alive or had died without evidence of recurrence.⁴ Of the 164 patients, 4 had evidence of disease at the last follow-up. Three patients had died with evidence of a tumour and one was alive with a tumour.

We described a case of PLGA arising in the minor salivary glands of the upper left buccal vestibule with an extensive growth involving the posterior maxillary left buccal vestibule, lateral to the left maxillary sinus and medial and anterior to the left mandible ramus. The mass extended into the left infratemporal fossa superiorly, with inferior extension superior to the left parapharyngeal space, contiguous with the left base of the tongue. Despite its large size, no evidence of metastatic disease was found.

PLGA, as the name implies, is considered to be a low grade malignancy almost exclusively arising from minor salivary glands. This tumour is generally considered to be a slow growing tumour in the soft tissue. Variable

Figure 8 A 400 × photomicrograph (haematoxylin & eosin) showing the characteristic (salt and pepper) nuclear feature

patterns of local recurrences and metastasis have previously been reported with some tumours.⁴ Very few cases have described the extent and radiographic features of PLGA. de Magalhaes *et al*⁷ described a mandibular lesion with unusual radiographic features similar to those of a benign tumour. Based on these findings, an ameloblastoma was the preferred diagnosis in that case. Sato *et al*⁸ also described an intraosseous PLGA of the maxilla in which panoramic radiographic features were more suggestive of a radicular cyst, whereas a CT scan showed irregular scalloping and perforation of the cortical border more suggestive of a malignant process.

Our case shows significant radiographic features that are characteristic of a slow growing but aggressive neoplasm. The panoramic radiographic findings included loss of left posterior maxillary bone and lack of visualization of the inferior border of the maxillary

sinus. CT showed irregular cortical density, thinning of the cortical plates, mass effect with bowing of the medial wall of the maxilla and maxillary sinus, localized perforation of cortical borders, hypervascularity and an irregular pattern of enhancement. The course of progression of the tumour, the location of the epicentre, partial bony destruction and radiographic features were more suggestive of a low grade neoplasm, possibly of minor salivary gland origin. Sometimes, this tumour could be difficult to diagnose because of its varied histomorphological patterns. As it was shown in previously reported cases and the present case, the tumour tends to have a pattern of radiological features that is consistent with a slow growing tumour, except for two cases that mimicked a radicular cyst and an ameloblastoma. Establishing a clear radiographic pattern for these histomorphologically diverse tumours will be helpful in early diagnosis of these lesions.

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