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Paediatric intraoral mucoepidermoid carcinoma mimicking a bone lesion

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Background. Mucoepidermoid carcinoma of the salivary glands occurs most frequently in adults during the fifth and sixth decades of life. Although uncommon, it is the main malignant salivary gland tumour in children, particularly adolescents.
Case report. A 14-year-old girl presented with a mass in her palate that had had a duration of one year. On panoramic X-ray, a well-delimited radiolucent area was observed on the left maxillary sinus region. The initial clinical diagnostic hypothesis was that this was a central giant cell granuloma. An incisional biopsy was performed and the final microscopic diagnosis was mucoepidermoid carcinoma. The tumour was removed by a wide transoral resection and the patient has been asymptomatic for 4 years.
Conclusion. Mucoepidermoid carcinoma in paediatric patients is uncommon, but it must be considered in differential diagnoses of intraoral submucosal mass/nodules in children, particularly in the palate.

Introduction

Salivary gland neoplasms are infrequent in the adult population and rarely affect children[1]. About 5% of all salivary gland tumours occur in children and adolescents, representing less than 10% of all paediatric head and neck tumours[2]. As in adults, the most common benign and malignant salivary gland tumours which occur in the paediatric population are pleomorphic adenomas and mucoepidermoid carcinoma (MECs). The majority of MECs have been histologically classified as having a low or intermediate grade of malignancy, favouring a good prognosis[1,3]. Jones and Franklin[4] analysed all cases of oral and maxillofacial pathology in children over a 30-year period, identifying 4406 cases, of which only 31 related to malignancies. Of these, six cases were salivary gland carcinomas: three MECs, two acinic cell carcinomas and one epithelial myoepithelial carcinoma.

Few series or case reports describing salivary gland tumours in the paediatric population have been published[1,3,5]. This report describes an additional case of a low-grade MEC affecting the palate of a 14-year-old girl that caused extensive bone destruction. The clinical, radiographic and histological features of this case are discussed, as well as the main differential diagnoses.

Case report

A 14-year-old girl was referred to the Dental Section of the Haematology and Hemotherapy Centre, State University of Campinas, Campinas, São Paulo, Brazil, complaining of a growing mass in her palate that had been present for one year. On clinical examination, a firm, painless, submucosal swelling of 5 cm in diameter was observed on the left side of the palate. The mucosa involved was smooth, normal in colour and without ulceration (Fig. 1). On panoramic X-ray, a well-delimited radiolucent area on the left maxillary sinus region was observed, causing displacement of the ipsilateral upper premolars (Fig. 2). Both premolars were clinically normal with pulp vitality. Initially, the lesion was aspirated, but no material was collected. After a clinical diagnosis of central giant cell granuloma, an incisional biopsy was carried out under local anaesthesia. During the surgical procedure, viscous material

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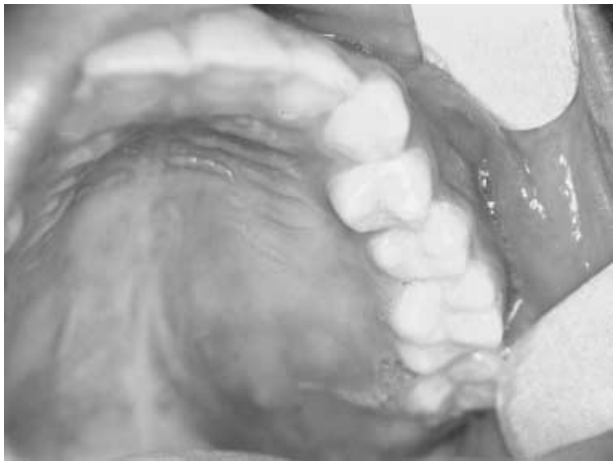

Fig. 1. Clinical view of a submucosal swelling on the left side of the palate of a 14-year-old girl that was diagnosed as mucoepidermoid carcinoma.

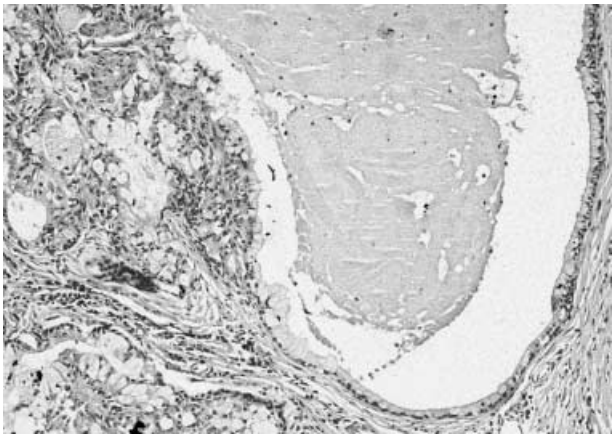

Fig. 3. Microscopic aspects of a low-grade mucoepidermoid carcinoma showing microcysts lined by mucous and epidermoid cells (H&E, × 500).

was produced, similar to mucous, suggesting a salivary gland alteration.

Microscopically, the lesion revealed mucin-filled macrocysts lined by mucous and epidermoid cells. Perineural invasion, mitotic figures, necrosis and cellular pleomorphisms were absent (Fig. 3). The diagnosis was MEC, low-grade subtype, according to Ellis and Auclair[6]. A clinical examination and chest X-ray were negative for neck and lung metastases. The tumour

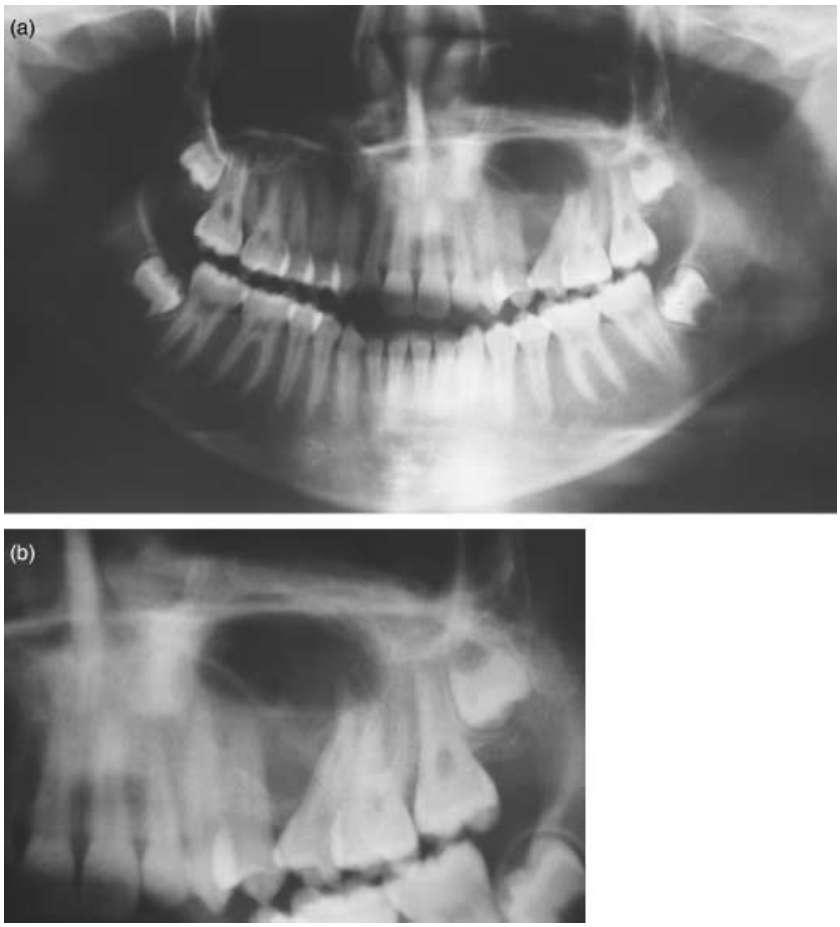

Fig. 2. Panoramic X-rays showing: (a) a well-delimited radiolucent area on the left maxillary region; and (b) displacement of the upper premolars roots caused by a mucoepidermoid carcinoma.

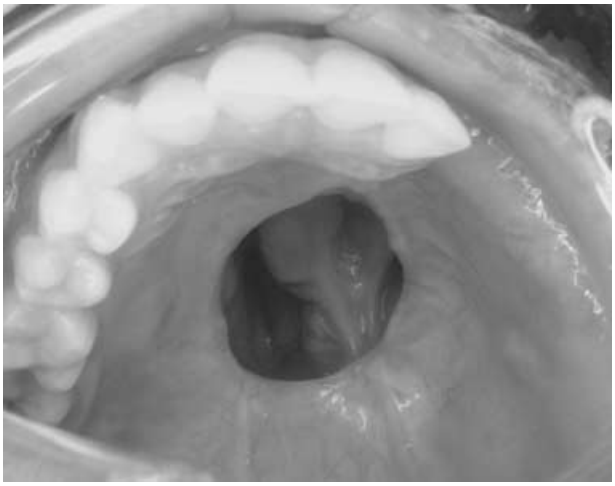

Fig. 4. Orosinonasal communication after surgical treatment of a low-grade mucoepidermoid carcinoma involving the palate of a 14-year-old girl.

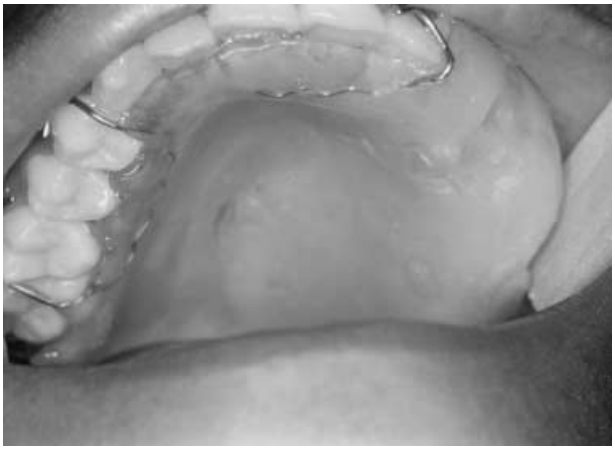

Fig. 5. Clinical view of the patient with an obturator prosthesis in place.

was removed by an ample transoral resection involving the hard palate and left maxilla. During the surgical approach, displacement of the sinus inferior wall was observed, but there was no tumour invasion of the sinus. Nevertheless, surgical management caused an oronasal/antral communication (Fig. 4), which was closed by an obturator prosthesis (Fig. 5). The surgical specimen measured $3.5 \times 3.0 \times 1.3$ cm and all margins were tumour-free. Currently, there are no signs of tumour recurrence after 4 years of treatment.

Discussion

Mucoepidermoid carcinoma is the most common malignant tumour of both the major and minor salivary glands, representing about 15% of all intraoral minor salivary gland tumours[6]. It is the most prevalent malignant salivary gland neoplasm in adults, as well as children and adolescents[3].

In the oral cavity, MEC is most common in the palate, presenting as a painless and well-circumscribed fluctuant nodule with a bluish or reddish-purple aspect, usually covered by a smooth and normal mucosa. Curiously, in this case, the tumour was firm to palpation and no liquid was aspirated, but microscopically, it was shown to be formed by various cystic cavities. Ulceration, bone resorption and numbness of adjacent teeth are associated with more aggressive tumours[5]. Although there was extensive bone involvement and tooth displacement in the current case, the tumour was classified histologically as having a low-grade of malignancy and no recurrence was observed after 4 years of treatment. Incisional biopsy is essential to establish a definitive diagnosis, assign a histological grade and prescribe adequate therapy. Before treatment, clinical examination and chest X-ray should be performed to rule out eventual metastases[7]. Differential diagnoses of palatal tumours in children include other minor salivary gland neoplasms, mainly pleomorphic adenoma, as well as benign and malignant mesenchymal tumours. Mucocele is the main reactive salivary gland lesion to be considered, but it is uncommon on the palate and sialometaplasia usually shows ulceration[4,5]. In paediatric patients with bone involvement, central giant cell granuloma, and odontogenic tumours such as ameloblastic fibroma, central odontogenic fibroma and adenomatoid odontogenic tumour should also be considered. Although rare in this age group and most frequent in the mandible, unicystic and conventional ameloblastoma must be also considered[8,9]. In this case, bone resorption was extensive, and the radiolucent image with displacement of adjacent teeth induced the authors to consider central giant cell granuloma as the first clinical and radiographic diagnosis. However, they observed the presence of mucus during the incisional biopsy and changing their clinical impression to that of a salivary gland tumour.

Most MECs in the paediatric group are classified microscopically as having a low or inter-

mediate grade of malignancy[1,3]. The favourable prognosis of MEC in young patients is considered to be associated with the low-grade characteristics of these tumours. However, it must be considered that 7.3% of low-grade MECs show recurrence and 2.4% regional metastasis[6].

All salivary gland neoplasms tend to recur after incomplete removal, and therefore, successful primary treatment is essential. A wide local excision with adequate free margins is the usual operative procedure for low-grade MECs. For high-grade MEC, combined surgery and radiotherapy treatment is recommended[10]. However, the value of adjuvant radiotherapy for MEC is controversial, particularly in young people. One must have strong indications to use radiotherapy in children with oral MEC because of serious side-effects such as trismus, growth disturbance and radiation-induced tumours[7]. Besides high-grade tumours, the indications for postoperative radiation therapy include close or positive margins, perineural invasion, deep invasion into muscle, and bone and lymph node metastasis[7,11]. Since this case was a low-grade MEC and none of the above features were observed, adjuvant radiation therapy was not performed.

Five-year survival correlates closely with histological grade, with values of about 92% and 63% for low- and intermediate-grade MEC, respectively. Survival for patients with high-grade MEC is poor[11]. A survival rate of 100 for low- and intermediate-grade MEC has been described by some authors[7,10]. Recurrence after 20 years of initial treatment has been reported, and thus, long-term follow-up is recommended[11]. This patient has been submitted to periodic follow-up, and currently, after 4 years, is alive without disease.

In summary, intraoral MEC in paediatric patients is uncommon and usually takes the form of a low-grade tumour. Mucoepidermoid carcinoma must be considered in the differential diagnoses of intraoral submucosal masses/nodules in children, particularly if the palate is involved. Complete surgical excision is the treatment of choice, resulting in excellent local control and survival.

What this paper adds

- This paper describes an uncommon case of salivary carcinoma in an adolescent. It also shows the clinical and radiographic features of the neoplasm.
- Differential diagnoses of bone lesions in children are discussed, particularly those in which the palate is involved.

Why this paper is important to paediatric dentists

- Salivary gland tumour must be considered in differential diagnoses of intraoral submucosal masses/nodules in children, especially in the palate.

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