DOI: 10.1111/j.1365-263X.2006.00785.x

Plasmacytoid myoepithelioma of the palate in a child

DANYEL ELIAS da CRUZ PEREZ[1], MÁRCIO AJUDARTE LOPES[2], OSLEI PAES de ALMEIDA[1], JACKS JORGE[1] & LUIZ PAULO KOWALSKI[3]

[1]Department of Oral Pathology and [2]Department of Semiology, School of Dentistry of Piracicaba, UNICAMP, Piracicaba, São Paulo, and [3]Department of Head and Neck Surgery and Otorhinolaryngology, Cancer Hospital, AC Camargo, São Paulo, Brazil

International Journal of Paediatric Dentistry 2007; 17: 223–227

Background. Myoepithelioma is a rare benign salivary gland tumour, localized most frequently in the parotid and in minor intraoral salivary glands. There have been only four cases of myoepithelioma in children and adolescents reported in the English-language literature, all of them involving the plasmacytoid variant.
Case Report. A 13-year-old boy, complained of a painless nodule of the palate. Incisional biopsy was performed and revealed large plasmacytoid cells with a round and eccentric nuclei. The diagnosis was consistent with myoepithelioma, plasmocytoid variant. Tumour cells were positive for cytokeratins, vimentin and S-100 protein. Surgical resection was performed and no evidences of tumour recurrence were observed after 6 years of the treatment.
Conclusion. Myoepithelioma is a very rare tumour in children and apparently presents a good prognosis, similar to occur in adult patients.

Introduction

Myoepitheliomas are benign tumours of the salivary glands that are predominantly or entirely composed of myoepithelial cells. Most authors affirm that these represent one extreme of the histological spectrum of pleomorphic adenoma, with either no ductal differentiation[1] or up to 5% of the tumour composed of ductal epithelium[2]. Myoepitheliomas account for less than 1% of all salivary gland tumours, and usually affect patients in the fourth and fifth decades of life, without gender predilection; the parotid gland is the most commonly affected site[2].

Microscopically, there are five cytological variants; however, the spindle-shaped and plasmacytoid cell subtypes are recognized as the two main ones[3]. The spindle-cell variant is the most common subtype, presenting a proliferation of spindle-shaped cells and eosinophilic cytoplasm[1], and forming a solid architectural pattern[3]. The plasmacytoid variant presents round cells with eccentric nuclei, and large and eosinophilic cytoplasm, also known as hyaline cells[1]. Diagnosis is based mainly on the histopathological features, although immunohistochemical analysis is useful to confirm myoepithelial origin of the tumour cells, which are positive for cytokeratin, vimentin, glial fibrilary acid protein, S-100 protein and muscle-specific actin to varying degrees[3,4].

This form of tumour is extremely rare in children and adolescents, and only four well-documented cases of myoepithelioma in patients ≤ 18 years of age have been reported in the English-language literature[4–7]. The aim of this paper was to describe a case of paediatric myoepithelioma, emphasizing the clinical, histopathological and immunohistochemical features.

Case report

A 13-year-old black boy was referred by a general dental practitioner to the Oral Diagnosis Clinic (Orocentro), School of Dentistry, State University of Campinas, Piracicaba, São Paulo, Brazil. He complained of a painless swelling of the palate of 2 years' duration. The patient reported that he did not regularly attend the dentist, but received oral hygiene instruction at his school. Oral examination revealed good oral hygiene and dental status. In addition, a well-circumscribed, firm and submucosal nodule,

Correspondence to:
Márcio Ajudarte Lopes DDS PhD, Faculdade de Odontologia de Piracicaba/UNICAMP, Área de Semiologia, Departamento de Diagnóstico Oral, Av. Limeira, 901. Areão. CEP: 13414-018, Piracicaba/SP, Brazil. E-mail: malopes@fop.unicamp.br

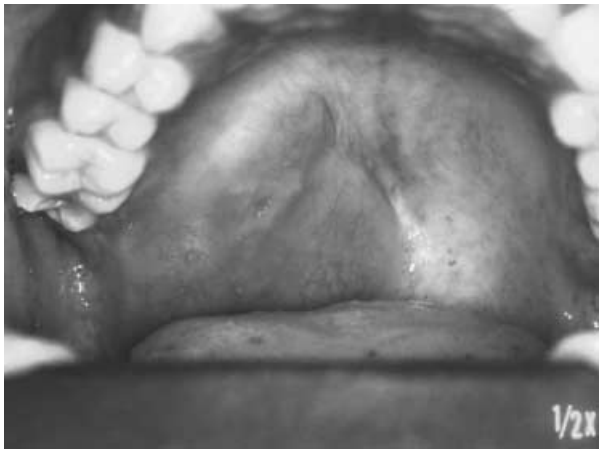

Fig. 1. Firm submucosal nodule with a central erythematous area at the junction of the hard and soft palate (diameter = 4.0 cm).

measuring 4 cm in diameter, was observed in the transition between the hard and soft palate (Fig. 1). The tumour presented a smooth and erythematous surface, and did not show radiographic evidence of bone involvement. Computed tomography revealed an increase in soft tissue in the soft palate, oropharynx and posterior wall of the nasopharynx, with a corresponding reduction in the size of the airways. No bone involvement was observed. The primary clinical hypothesis upon diagnosis was that this was a benign tumour of the salivary gland, and an incisional biopsy was performed under local anaesthesia.

Microscopic analysis showed large cells with a plasmacytoid aspect presenting round, eccentric nuclei, and an abundant and homogeneous eosinophilic cytoplasm, arranged in trabecules forming a solid pattern, surrounded by a loose stroma. Rare spindle-shaped cells were observed among the plasmacytoid cells. The histopathological diagnosis was compatible with myoepithelioma, plasmacytoid variant (Fig. 2). Immunohistochemical reactions were carried out using the avidin-biotin complex technique. Tumour cells showed strong and diffuse positivity for cytokeratins (AE1/AE3, Dako Cytomation, Glostrup, Denmark; dilution = 1:500), vimentin (VIM 3B4, Dako Cytomation; dilution = 1:400) (Fig. 3) and S-100 protein (Sigma-Aldrich Corporation, St. Louis, USA; dilution = 1:10 000), confirming the myoepithelial origin of the tumour cells. Actin muscle-specific (HHF35, Dako Cytomation; dilution = 1:800) and cytokeratin 14 (LL002, Novocastra Laboratories Ltd, Newcastle upon Tyne, UK; dilution = 1:200) immunostaining was negative. Thus, the patient was referred to the Department of Head and Neck Surgery and Otorhinolaryngology, Cancer Hospital AC Camargo, São Paulo, Brazil, for treatment. The patient underwent a complete surgical resection of the tumour under general anaesthesia. There were no postoperative complications. Histopathological examination of the surgical specimen found cells similar to those described above. Neither epithelioid nor clear cells, and neither ductal nor acinar

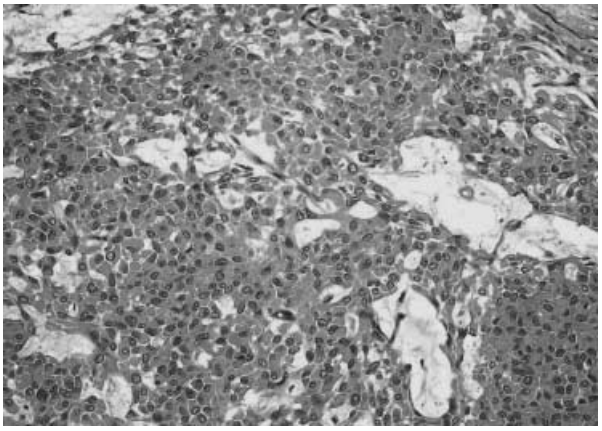

Fig. 2. Large cells with round eccentric nuclei surrounding homogeneous eosinophilic cytoplasm; these have a plasmacytoid aspect and present little pleomorphism (Haematoxylin and eosin, original magnification ×200).

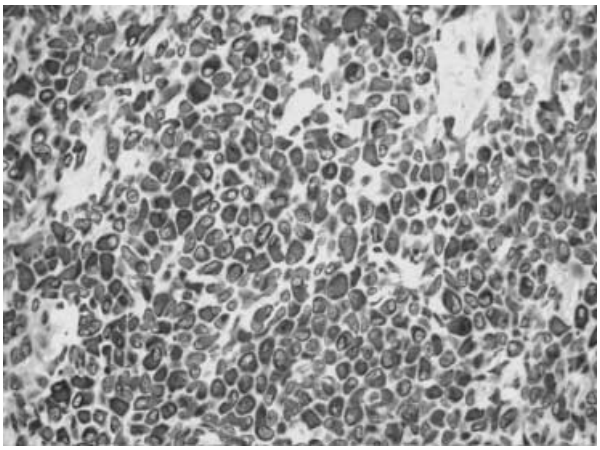

Fig. 3. Strong immunohistochemical expression of vimentin in neoplastic cells of myoepithelioma (Streptavidin-biotin-peroxidase method, ×400).

differentiation were noted. There was mild cellular atypia and mitotic figures were absent. Although the tumour was well circumscribed, no capsule or pseudocapsule was observed, and the surgical margins were free. Immunohistochemical reactions were run again, using the same antibodies, and the same positivity that was obtained in the biopsy specimen was observed. After complete scarring of the surgical wound, a palatal obturator prosthesis, which consists of a metal structure supported in the upper posterior teeth with a resin base covering the surgical defect, was attached. The patient has been followed-up for 6 years without showing any evidence of tumour recurrence.

Discussion

Myoepitheliomas represent about of 1% of all salivary gland neoplasms[4]. In the oral cavity, the palate is the most common site for these tumours, and the histological spindle-cell subtype is the most common; however, the plasmacytoid variant tends to occur more frequently in palatal tumours and in young patients[3,6]. All cases of myoepithelioma that have been reported in paediatric patients occurred in the palate and were of the plasmacytoid variant. Table 1 shows the clinical data of the present case and the previously reported paediatric cases. Although it is extremely rare, malignant myoepithelioma has also been described in paediatric patients[8].

The differential diagnosis of an intraoral submucosal nodule presenting in a child includes reactive and neoplastic lesions. Among the reactive lesions, abscess and deep mucocele should be considered. With regard to neoplastic lesions, salivary gland tumours are the main group, plemorphic adenoma and mucoepidermoid carcinoma being the most common forms[9,10]. Other benign and malignant neoplasms, such as neurofibroma, schwannoma and rhabdomyosarcoma, can also be included in the differential diagnosis of intraoral submucosal nodules in youngsters[11].

Clinically, intraoral myoepitheliomas present as painless firm submucosal nodular lesions[4–7], without involvement of bone or the soft tissue of adjacent structures[4–6], as in this case, where no bone involvement was observed during radiographic and computed tomography examinations, and as confirmed by surgery. The tumours in these patients group are relatively small in size, varying from 1.0 to 3.0 cm, with a mean of 1.9 cm[4–7]. This case presented with a tumour that was 4.0 cm in diameter, the largest ever reported in a paediatric patient.

Even if the myoepithelioma described above represents only one end of the spectrum of pleomorphic adenoma, it is important, from an academic point of view, to separate these two lesions. Indeed, because of their variable architectural pattern with diverse cell types, and frequently increased cellularity, myoepitheliomas can be misdiagnosed as a malignant epithelial or mesenchymal tumour[1,3,12]. For a diagnosis of myoepithelioma, as well as presenting tumour cells with the histopathological features of neoplastic myoepithelial cells, the lesion will have no ductal differentiation or up to 5% of the tumour will be composed of ductal epithelium. In this case, even after several histological cuts, the authors did not observe ductal formation, supporting their

Table 1. Clinical data for previously reported myoepithelioma in children and adolescents.

Reference	Age (years)	Gender	Site of lesion	Size (cm)	Length of presence of lesion (months)	Treatment	Length of follow-up (months)
Kahn & Schoub (1973)[5]	17	F	Hard palate	3.0	2	Surgery	No information
Stromeyer *et al*. (1975)[7]	14	M	Gingival mucosa of the maxilla	2.0	1	RT + Surgery	15*
Nesland *et al*. (1981)[6]	18	F	Soft palate	1.5	No information	Surgery	1*
Lins & Gnepp (1986)[4]	08	F	Soft palate	1.0	No information	Surgery	12*
Present case	13	M	Soft palate	4.0	24	Surgery	72*

RT, radiotherapy; *Alive without disease.

diagnosis of myoepithelioma. Microscopically, most cases in the paediatric group presented with cellular pleomorphism[5–7] and the absence of a well-defined capsule[4,6], although the tumours were circumscribed, as was seen in this case. However, these histopathological characteristics do not imply malignancy, since the tumours present a benign clinical behaviour and do not recur after local resection[2,6].

Although a diagnosis of myoepithelioma should be made mainly on the basis of histopathological features, particularly when these characteristics resemble neoplastic myoepithelial cells[3], immunohistochemical features can assist in establishing a final diagnosis. In normal myoepithelial cells, muscle-specific actin and cytokeratin 14 are the two most important immunoexpressed antigens[3]. Nevertheless, the tumour cells of myoepithelioma, particularly of the plasmacytoid variant, show little or no normal myoepithelial features[13]. Despite the fact that vimentin is not expressed in normal myoepithelial cells, it is a more important marker than muscle-specific actin or cytokeratin 14 for neoplastic myoepithelial cells[3,14]. S-100 protein is normally present in many salivary gland tumours and has also been identified as a marker of neoplastic myoepithelial cells[3,4], but it is not expressed in normal myoepithelial cells. In this case, the tumour cells showed strong and diffuse positivity for vimentin, S-100 protein and cytokeratins, but did not express muscle-specific actin or cytokeratin 14, confirming that, for myoepithelial neoplastic cells, vimentin and S-100 are more significant markers than muscle-specific actin or cytokeratin 14, for the reasons commented on above.

The treatment of these tumours consists of surgical excision with tumour-free margins[4]. Although the paediatric myoepitheliomas are rare, it is reasonable to assume that their biological behaviour is similar to tumours seen in adults, and that they will not recur after adequate treatment. Therefore, these patients must be maintained under periodic and long-term follow-up to ensure a good prognosis. The four cases of myoepithelioma in paediatric patients reported in the English-language literature all had short follow-ups after the treatment of no more than 15 months (Table 1). Following adequate surgical intervention, this patient did not present any signs of tumour recurrence after 6 years of treatment.

What this paper adds

- This paper reports an extremely rare case of intraoral plasmacytoid myoepithelioma in a child and reviews and discusses the clinical, histopathological, immunohistochemical features and outcome of previously published cases in the literature.

Why this paper is important to paediatric dentists

- It is fundamental that dental practioners, particularly the paediatric dentist, may recognize eventual oral lesions that occur in childhood and adolescence.
- Although oral salivary gland tumours are rare in children, particularly the myoepithelioma, they should be considered in the clinical differential diagnosis of oral submucous nodule in children.

References

1 Ellis GL, Auclair P. Tumors of the salivary glands. In: Ellis GL, Auclair P (eds). *Atlas of Tumor Pathology*, 3rd Series, Fascicle 17. Washington, DC: Armed Forces Institute of Pathology, 1996: 57–68.

2 Sciubba JJ, Brannon RB. Myoepithelioma of salivary glands: report of 23 cases. *Cancer* 1982; **49**: 562–572.

3 Dardick I. Myoepitheliomas: definitions and diagnostic criteria. *Ultrastruct Pathol* 1995; **19**: 335–345.

4 Lins JEW, Gnepp DR. Myoepithelioma of the palate in a child. *Int J Pediatr Otorhinolaryngol* 1986; **11**: 5–13.

5 Kahn LB, Schoub L. Myoepithelioma of the palate. Histochemical and ultrastructural observations. *Arch Pathol* 1973; **95**: 209–212.

6 Nesland JM, Olafsson J, Sobrinho-Simões M. Plasmacytoid myoepithelioma of the palate. A case report with ultrastructural findings and review of the literature. *J Oral Pathol* 1981; **10**: 14–21.

7 Stromeyer FW, Haggitt RC, Nelson JF, Hardman JM. Myoepithelioma of minor salivary gland origin. *Arch Pathol* 1975; **99**: 242–245.

8 Moriniere S, Robier A, Machet MC, Beutter P, Lescanne E. Massive infra-clinic invasion of the facial nerve by a myoepithelial carcinoma of the parotid. *Int J Pediatr Otorhinolaryngol* 2003; **67**: 663–667.

9 Flaitz CM. Mucoepidermoid carcinoma of the palate in a child. *Pediatr Dent* 2000; **22**: 292–293.

10 Fonseca I, Martins AG, Soares J. Epithelial salivary gland tumors of children and adolescents in southern Portugal. *Oral Surg Oral Med Oral Pathol* 1991; **72**: 696–701.

11 Jorge J, Pires FR, Alves FA, *et al*. Juvenile intraoral pleomorphic adenoma: report of five cases and review of the literature. *Int J Oral Maxillofac Surg* 2002; **31**: 273–275.

12 Takai Y, Dardick I, MacKay A, Leung R, Burford-Mason A, Mori M. Diagnostic criteria for neoplastic myoepithelial cells in pleomorphic adenomas and myoepitheliomas: immunocytochemical detection of muscle-specific actin, cytokeratin 14, vimentin and glial fibrilary protein. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1995; **79**: 330–341.

13 Jaeger RG, Oliveira PT, Jaeger MMM, Araújo VC. Expression of smooth-muscle actin in cultured cells from human plasmacytoid myoepithelioma. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1997; **84**: 663–667.

14 Takai Y, Mori M, Dardick I, *et al.* Myofilament localization and immunoelectron microscopic detection of muscle-specific actin in neoplastic myoepithelial cells in pleomorphic adenomas and myoepitheliomas. *Ultrastruct Pathol* 1994; **18**: 579–595.