Case Report

Pleomorphic adenoma of the upper lip in a child

Mônica A. Lotufo¹), Celso A. L. Júnior²), Joyce P. de Mattos¹) and Cristiane M. França¹)

¹⁾Department of Oral Diagnosis, School of Dentistry, Ibirapuera University, São Paulo, Brazil ²⁾Department of Stomatology, School of Dentistry, University of São Paulo, São Paulo, Brazil

(Received 27 July 2007 and accepted 19 March 2008)

Abstract: This case report describes a rare and unusual lesion found in a 12-year-old boy, which was diagnosed as pleomorphic adenoma of the minor salivary glands in the upper lip. The tumor was a circumscribed, submucosal nodule, about 2.0 cm in diameter and characterized by slow growth and a rubbery consistency. Complete excision was performed and the histopathological analysis showed an epithelial salivary gland tumor with islands of plasmacytoid cells, duct-like structures, in a variable stroma with chondroid, fibrous and myxoid appearance. The tumor did not recur. A brief review of the relevant literature is also presented. (J. Oral Sci. 50, 225-228, 2008)

Keywords: pleomorphic adenoma; childhood tumors; salivary gland tumors; lip.

Introduction

Neoplasms of the salivary glands (SG) are mainly adult diseases, being rarely reported in children. Less than 5% of all pediatric head and neck tumors have their origin in the SG (1).

The most common salivary gland tumor is pleomorphic adenoma (PA), which accounts for 60-65% of such diseases. It mainly affects women in their fourth to sixth decade of life, and has a natural history of asymptomatic slow growth over a long period (2).

The etiology of PA is unknown. It is epithelial in origin, and clonal chromosome abnormalities with aberrations involving 8q12 and 12q15 have been described (3). Histologically, PA is characterized by a large variety of tissues consisting of epithelial cells arranged in cord-like cell pattern, together with areas of squamous differentiation or with plasmacytoid appearance. Myoepithelial cells are responsible for the production of abundant, extracellular matrix with chondroid, collagenous, mucoid and osseous stroma (4).

PA may occur in the first two decades of life although this is uncommon (5-9). The relative frequency of epithelial salivary gland tumors in children and adolescents ranges from 3.7 to 5.5%, with most cases arising in the major salivary glands, mainly the parotid gland, and only 10-15% occurring in the intra-oral minor salivary glands (5).

This paper describes the diagnosis and management of an asymptomatic, slowly growing, pleomorphic adenoma in the upper lip of a child. A brief review of the relevant literature is also presented. It is suggested that the practitioner has to be aware of such rare conditions.

Case Report

A 12-year-old schoolboy visited the Oral Diagnosis Center with a submucosal nodule in the maxillary right labial mucosa. According to the boy's parents, they had first noted the lesion about one year ago, after which it had gradually increased in size. The clinical development of the nodule was slow and asymptomatic. On examination, the nodule was circumscribed, mobile, sessile, rubbery in consistency and 1.5 to 2.0 cm in diameter. The overlying mucosa was smooth with a pinkish-yellow color showing evidence of superficial vascularity (Fig. 1). There was no pain or bleeding on palpation. Head and neck abnormalities were not noted on clinical examination. There was no evidence of the lesion on the external surface of the lip. The medical history was unremarkable, and no other abnormalities were found on clinical examination.

A dental etiology was ruled out based on clinical and radiographic examination. Thus, the clinical diagnosis

Correspondence to Dr. Mônica Andrade Lotufo, Department of Oral Diagnosis, School of Dentistry, Ibirapuera University, 1329 Interlargos Ave, São Paulo, Brazil Tel: +55-11-5694-7916 Fax: +55-11-5694-7915 E-mail: monicalotufo@uol.com.br

established was compatible with a benign minor salivary gland tumor or a lipoma.

The tumor was completely removed with an intra-oral biopsy. During the surgical procedure, the lesion was excised without difficulty with clinically normal margin because the specimen was fully encapsulated (Fig. 2). Subsequent follow-up after one year showed no signs of recurrence.

Histopathological analysis of the surgical specimen revealed a cellular mass well-encapsulated epithelial salivary gland tumor characterized by large islands of plasmacytoid cells, duct-like structures, in a variable stroma with chondroid, fibrous and myxoid appearance. There was no evidence of malignancy. The diagnosis was pleomorphic adenoma (Fig. 3).



Fig. 1 Submucosal nodule in the upper lip mucosa covered by normal mucosa with a slightly superficial, yellowish appearance, and teleangectasic vessels.



Fig. 2 Macroscopic view of the specimen, showing a fullyencapsulated nodule.

Discussion

Minor salivary gland tumors are rare in children, and are responsible for only five percent of all salivary gland tumors (1). The most likely salivary gland tumor found in children is a pleomorphic adenoma; its anatomical distribution follows that seen in adults, the palate being the most affected site (6,8). Jorge et al., provided an extensive review of cases of minor salivary gland PA in children, finding only 16 well-documented cases; they also reported five new cases of juvenile intra-oral PA, two of which were located in the upper lip (6). No other study has described pleomorphic adenoma arising from this location in a child.

A review of the cases shows that the incidence of PA during childhood usually occurs between 7 and 16 years of age, with an average of 10.5 years. The female to male ratio is 7:3, indicating a female preponderance. This tendency is similar to that seen in adults (8).



Fig. 3 Epithelial salivary gland tumor characterized by large islands of epithelial cells and duct-like structures (A), outer cells with a plasmacytoid appearance (B), in a variable stroma showing chondroid, fibrous and myxoid appearance (C) (H-E staining, ×100, ×400, ×400).

The most common symptom of a salivary gland tumor in children is a submucosal lump with few cases showing ulceration, pain or bleeding (7). Bentz et al. analyzed 324 cases of salivary gland masses in children, including major and minor glands. They found that 86.7% were vascular proliferations (59.2% hemangiomas and 27.5% lymphangiomas) and 13.3% were salivary gland tumors. Interestingly, major salivary gland tumors accounted for 79% of the lesions, while 7% were located in the palate, 5% each in the cheek and tongue and 2% each in the lip and gingiva. The authors did not specify the diagnosis of the lesion in the lip. Based on their findings, Bentz et al. proposed a diagnostic and treatment strategy for the clinician who encounters a child with a mass in the salivary gland region: the first step is to establish whether the mass is inflammatory or not (7).

Differential diagnoses of intra-oral, solid, asymptomatic nodules include minor salivary gland tumors, and benign and malignant mesenchymal lesions such as neurofibroma and rhabdomyosarcoma. For lesions affecting the buccal mucosa, lip and tongue, lipoma, neurofibroma and other benign mesenchymal tumors, should be considered. The encapsulation and mobility of the nodule are signs of probable benignity, although a biopsy must always be performed (6).

Some authors noted that pleomorphic adenomas in children can undergo rapid enlargement within a short period of time, but generally such tumors exhibit the same characteristic seen in adults, including slow growth (8,9).

The present case did not show local recurrence twelve months after surgery. However, taking into consideration the characteristics of salivary gland neoplasms, which may recur after many years, the patient will be followed up for at least five years. In a retrospective study, McGregor et al., showed that of 31 patients under 30 years of treatment for pleomorphic adenoma, 42% (n = 16) developed recurrence. All recurrences occurred in the parotid gland. The authors believed that the chances of recurrence are higher when the first incidence of PA appears before 30 years of age (9,10).

Pleomorphic adenomas are usually painless, slow growing tumors; however, some cases exhibiting rapid growth have been reported, especially in the palate. Pleomorphic adenoma appears to be encapsulated, but this capsule is often infiltrated by lateral extension of the tumor. Even though pleomorphic adenoma is benign, it has a high rate of implantability. Any rupture of the capsule or incomplete excision will leave residual tumor cells behind, resulting in recurrence (11).

A lesion referred to as salivary gland anlage tumor (SGAT) histologically resembles a pleomorphic adenoma

(12,13). The microscopic and ultrastructural pattern of this benign tumor shows epithelial and mesenchymal components, with tubular and cord-like structures composed of cells immunophenotypically compatible with myoepithelium, which express a broad spectrum of keratins and epithelial membrane antigens while the stromal component expresses vimentin and smooth muscle actin (14,15). Only 23 cases have been described in the literature so far, and studies comparing the nature of the epithelia, myoepithelial cells and stromal components of the SGAT with those of PA are lacking. It is known that the luminal cells of PA are positive for cytokeratin and S-100 protein and negative for vimentin, and that the myoepithelial cells markers are vimentin, alpha-smooth-muscle actin or calponin. This difference may be related to the cellular differentiation of both entities (16). However, the SGAT is seen as a nasal or nasopharyngeal obstruction in newborns, especially males, and there have been no reported recurrences. Thus, it has been proposed that it may have an hamartomatous nature albeit its histological resemblance with PA, which is a true benign salivary gland tumor.

This rare disease reported in a child serves to alert clinicians regarding the diagnosis of unusual cases of orofacial swellings. As these lesions are asymptomatic, the patient may not be aware of their existence or are discovered accidentally by a dentist during examination. The old rule is still valid: a nodule in the lower lip of a child is probably a mucocele; a nodule in a child's upper lip may be a salivary gland tumor.

References

- Ethunandan M, Ethunandan A, Macpherson D, Conroy B, Pratt C (2003) Parotid neoplasms in children: experience of diagnosis and management in a district general hospital. Int J Oral Maxillofac Surg 32, 373-377
- 2. Forty MJ, Wake MJC (2000) Pleomorphic salivary adenoma in an adolescent. Br Dent J 188, 545-546
- Farina A, Pelucchi S, Grandi E, Carinci F (1999) Histological subtypes of pleomorphic adenoma and age-frequency distribuition. Br J Oral Maxillofac Surg 37,154-155
- Seifert G, Brocheriou C, Cardesa A, Eveson JW (1990) WHO International histological classification of tumours. Tentative histological classification of salivary gland tumours. Pathol Res Pract 186, 555-581
- 5. Guzzo M, Ferrari A, Marcon I, Collini P, Gandola L, Pizzi N, Casanova M, Mattavelli F, Scaramellini G (2006) Salivary gland neoplasms in children: the experience of the Istituto Nazionale Tumori of

Milan. Pediatr Blood Cancer 47, 806-810

- Jorge J, Pires FR, Alves FA, Perez DEC, Kowalski LP, Lopes MA, Almeida OP (2002) Juvenile intraoral pleomorphic adenoma: report of five cases and review of the literature. Int J Oral Maxillofac Surg 31, 273-275
- Bentz BG, Hughes CA, Lüdemann JP, Maddalozzo J (2000) Masses of the salivary gland region in children. Arch Otolaryngol Head Neck Surg 126, 1435-1439
- Bradley P, McClelland L, Mehta D (2007) Paediatric salivary gland epithelial neoplasms. ORL J Otorhinolaryngol Relat Spec 69, 137-145
- 9. McGregor AD, Burgoyne M, Tan KC (1988) Recurrent pleomorphic salivary adenoma – the relevance of age at first presentation. Br J Plast Surg 41, 177-181
- Ellies M, Schaffranietz F, Arglebe C, Laskawi R (2006) Tumors of the salivary glands in childhood and adolescence. J Oral Maxillofac Surg 64, 1049-1058
- 11. Daniels JS, Ali I, Al Bakri IM, Sumangala B (2007) Pleomorphic adenoma of the palate in children and

adolescents: a report of 2 cases and review of the literature. J Oral Maxillofac Surg 65, 541-549

- 12. Har-El G, Zirkin HY, Tovi F, Sidi J (1985) Congenital pleomorphic adenoma of the nasopharynx (report of a case). J Laryngol Otol 99, 1281-1287
- Dehner LP, Valbuena L, Perez-Atayde A, Reddick RL, Askin FB, Rosai J (1994) Salivary gland anlage tumor ("congenital pleomorphic adenoma"). A clinicopathologic, immunohistochemical and ultrastructural study of nine cases. Am J Surg Pathol 18, 25-36
- 14. Herrmann BW, Dehner LP, Lieu JE (2005) Congenital salivary gland anlage tumor: a case series and review of the literature. Int J Pediatr Otorhinolaryngol 69, 149-156
- 15. Cohen EG, Yoder M, Thomas RM, Salerno D, Isaacson G (2003) Congenital salivary gland anlage tumor of the nasopharynx. Pediatrics 112, e66-e69
- Furuse C, Sousa SO, Nunes FD, Magalhães MH, Araújo VC (2005) Myoepithelial cell markers in salivary gland neoplasms. Int J Surg Pathol 13, 57-65