Leiomyomatous hamartoma of the incisive papilla

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A case of unusual hamartoma in a six-year-old otherwise healthy Brazilian girl is reported, with emphasis on histological and immunohistochemical features. A mass observed in the incisive papilla was detected whose appearance was similar to congenital epulis or fibroma. Histological findings showed interlacing fascicles of large spindle cells resembling smooth muscle cells. Immunohistochemical staining for desmin and for smooth-muscle actin was positive. The histological diagnosis was leiomyomatous hamartoma, based on clinical and microscopic observations.
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INTRODUCTION

Hamartomas are defined by Willis[1] as "tumor-like, but primarily non-neoplasic, malformations or inborn errors of tissue development, characterized by abnormal mixture of tissues indigenous to the part with excess of one or more of these". Lymphangiomas and hemangiomas are examples of lesions that can be considered hamartomas or choristomas, although they are usually classified as benign neoplasms. Frequently, differences between neoplasic and hamartomatous processes are restricted to academic definitions, since clinical and morphological criteria are insufficient for exact classification. With the exception of the vascular types, hamartomas are uncommon in the head and neck region. A case of leiomyomatous hamartoma in the incisive papilla is presented, emphasizing the tissue composition revealed by immunohistochemical reactions. In addition the classification criteria of these lesions is discussed.

CASE REPORT

A six-year-old otherwise healthy girl was referred to the Oral Diagnosis Clinic of the University of São Paulo for investigation and treatment of a gingival swelling. The lesion was asymptomatic, and according to the mother was present at birth, and no alteration was noted since then. On the examination a penduculated nodule with 5 mm in diameter located in the midline of maxillary buccal gingiva was observed (Figure 1A). The nodule was fibrous upon palpation and covered by normal mucosa. There were no palpable lymph nodes in the region. On radiographic examination a

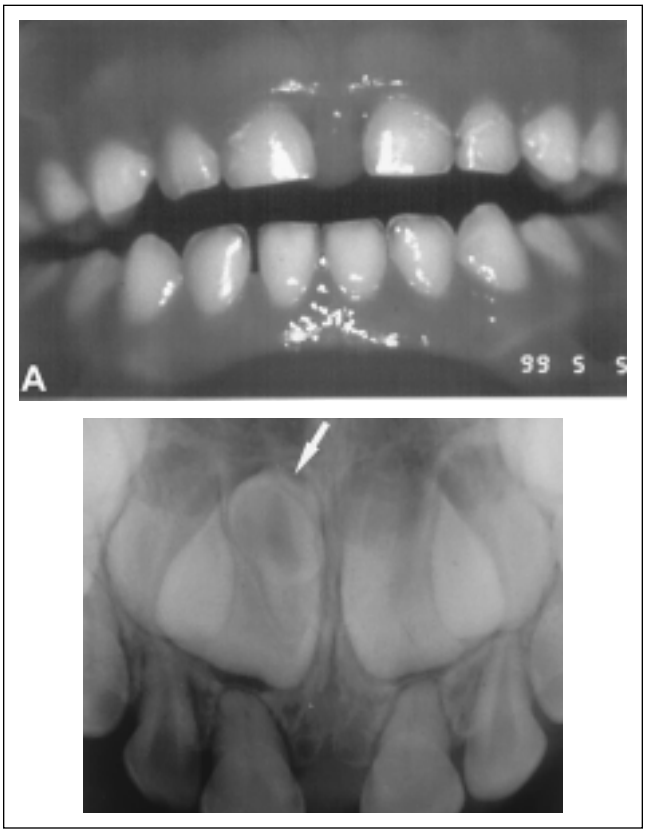

Figure 1 A. Presence of a penduculated nodule covered by normal mucosa located in the incisive papilla. B: Periapical radiographic showing a radiopacity compatible with a supernumerary tooth (arrow) adjacent to the tumor.

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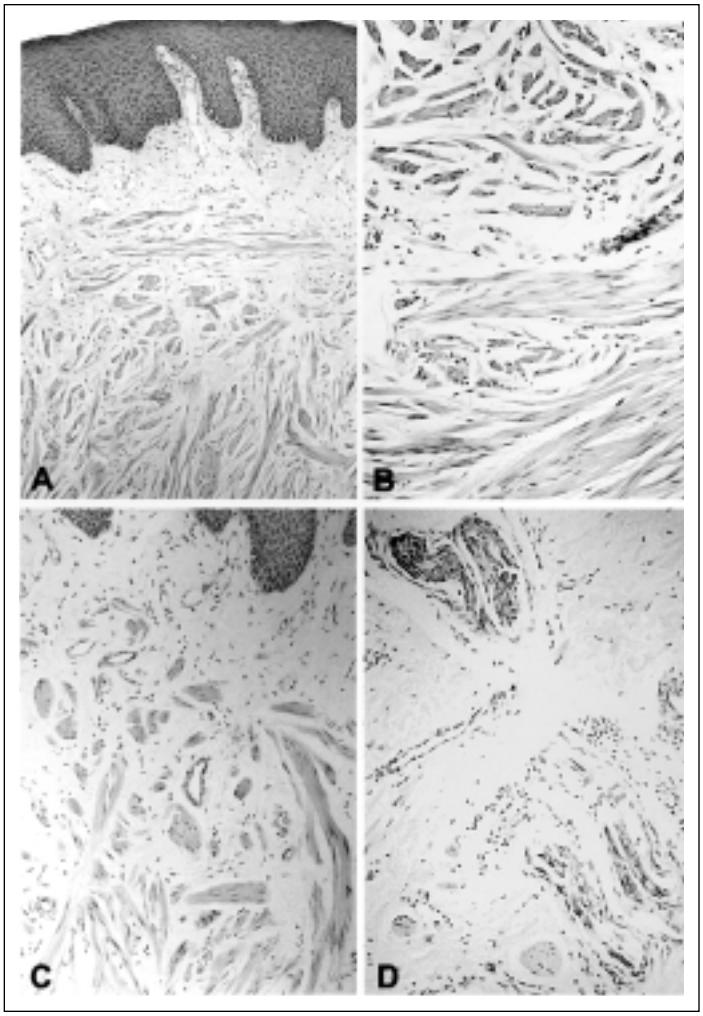

Figure 2 A. Subepithelial non-encapsulated ill-defined mass, composed of randomly orientated bundles of large spindle cells (Hematoxylin-eosin stain, original magnification X40). B: Detail of tumor cells, which resemble mature smooth muscle cells, with no evidence of atypia (Hematoxylin-eosin stain, original magnification X250). C: Positive staining for smooth muscle actin in smooth-muscle bundles (Original magnification, X100). D: Staining for S100 protein was positive in some areas of the tumor, confirming the presence of peripheral nerves scattered throughout the lesion (Original magnification, X125).

radiopacity compatible with a supernumerary tooth was present in the midline of the maxilla (Figure 1B).

An excisional biopsy was performed, and the specimen examined at the Oral Pathology Department of the University of São Paulo.

Microscopic examination showed a subepithelial non-encapsulated ill-defined mass, composed of randomly orientated bundles of large spindle cells with flattened nuclei (Figure 2, A and B). These cells were similar to mature smooth muscle cells. Lesional stroma was composed of loose connective tissue with increased number of thin-walled vessels, and conspicuous presence of peripheral nerves. In the deep aspect of the lesion an area of mixoid tissue was present. No evidence of atypical cells or adjacent tissues invasion was found.

Immunohistochemical staining was performed, and revealed intense positivity of spindle cells for smooth muscle actin (Figure 2C) and desmin. Immunostaining for S-100 protein demonstrated that peripheral nerves were present in large amounts and were scattered throughout the lesion (Figure 2D).

DISCUSSION

Hamartomas are tumor-like malformations not always easily distinct from benign neoplasms. Accurate analysis of the history, clinical findings, and microscopic aspects allow a diagnosis to be established.[2] In the present case several aspects favored a diagnosis of leiomyomatous hamartoma.

Firstly, the mother reported the presence of the lesion at birth, and no growth was observed since.

Secondly, the lesion was located in the midline portion of the maxilla (incisive papilla), a site where other hamartomatous lesions are common, such as developmental cysts and fibrous, neural, muscle and epithelial tumors.[3]

Thirdly, the histological features showed a proliferating

mass of mature spindle cells without invasive behavior, atypical cells or pleomorphism. This indicates that the existence of a tumor-like lesion, but not true neoplasic process.

Fourthly, the peripheral nerves were observed between bundles of spindle cells, as well as vessels, demonstrating the presence of different tissues in the mass with predominance of one of them.

Finally, no recurrence was noted after two years. In addition to the aspects related to the lesion itself, it is important to notice the presence of a supernumerary tooth, another developmental disturbance, located in the maxillary midline as well.

Positive results for desmin and smooth-muscle actin in the immunohistochemical reactions indicated that the spindle cells were smooth-muscle cells. With this finding, the differential diagnosis established was leiomyoma. Vascular leiomyoma, a sub-typing of oral leiomyomas, was considered in this case. However, in vascular leiomyomas the presence of peripheral nerves participating in the main mass is uncommon[3] and thick-walled vessels would be expected. Additionally, these lesions are observed in a very wide age range, but they are generally tumors of adults.[4] Clinically, congenital epulis was considered as differential diagnosis, but microscopically there was no evidence of granular cells.

We believe that only seven hamartomas of the smooth-muscle cells have been described in the literature involving either hard palate or gingiva. All of them are located in the midline portion of the maxilla. Furthermore, six of these cases were observed in Japanese population.[5-9,3] In most of them, the patients were females with ages between two months and four years old. One case was reported in a Caucasian patient, and the histological findings were similar to those observed by Japanese authors.[2] The case reported by Ng *et al.*[9] was diagnosed as leiomyoma. Discussions about this diagnosis have already been made by Napier *et al.*[2] and Semba *et al.*,[3] who believed this lesion was a true leiomyomatous hamartoma. We also believe this to be true, based on the fact that the lesion was located in the midline of the maxilla (incisive papilla), and presented histological features identical to the hamartoma of smooth-muscle cells reported in the literature. In addition, the age of the patient (two months old), and the fact that Ng *et al.*[9] did not perform immunohistochemical staining to confirm the existence of various cell types in the tumor mass, indicate that this lesion probably constitutes a hamartoma. Despite the discussion about classification criteria for these tumors, this case is an example that highlights the overlap between hamartomas and neoplasms.[2]

Goldsmith *et al.*[10] reported one case of a healthy sixteen-month-old boy with leiomyomatous hamartoma in the posterior tongue. Histological examination showed a circumscribed mass composed of randomly orientated fascicles of smooth-muscle cells in a fibrous tissue. Minor salivary glands were observed as well. There is another report of a case in the tongue in the literature, also in a Japanese patient.[11] The reasons for the preponderance of cases in the Japanese population remain unknown.

Hamartomas with other tissue components, such as minor salivary glands,[12] vascular structures,[13] and odontogenic epithelia,[14] are reported in the literature, but are rare. Most of them present a variety of these tissues simultaneously, and are detected in young age. On the other hand, some vascular hamartomatous lesions in the head and neck, as hemangiomas and lymphangiomas, are common. Willis[1] discussed some cases of benign angiomas and angiomatoses that do not constitute real neoplasms. The fact that these lesions can be congenital or appear soon after birth, and that they do not grow disproportionately and indefinitely, as true neoplasms, gives support to these arguments. Furthermore, other tissues (for example, muscular and nerve fibers) were observed in the main mass of these tumors, which are uncommon in neoplasic the process. With this explanation, it becomes clear that there is no sharp line of distinction between neoplasm and hamartoma concepts.

In spite of the rarity of leiomyomatous hamartoma in the upper respiratory-digestive tract, more investigations are needed in order to clarify the histogenesis of these lesions, and to establish the differences between neoplasic and hamartomatous processes.

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