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Neurovascular hamartoma: a case report of a tongue lesion

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Abstract

Hamartoma is a proliferation of normal cells and tissues, which are considered endogenous to the site of injury. Oral neurovascular hamartoma (NVH) presents as exophytic lesions, usually with a smooth surface, and less than 10 mm in diameter. Hamartomas arising in the oral cavity are uncommon and may show a variety of clinical presentations and histological and growth patterns. We present a case of oral neurovascular hamartoma lesion in the tongue of a 47-year-old individual, consisting of nerve bundles and blood vessels. Although rare, the neurovascular hamartoma should be considered in the differential diagnosis of tongue lesions.

Keywords: Congenital; Hamartoma; Oral cavity.

Introduction

Hamartoma consists of the proliferation of normal cells and tissues, which are considered endogenous to the injury site and can be found in different areas of the body. In hamartoma, tissues appear disorganized and ill-defined, merging within the normal surrounding tissues [1].

Hamartomas that occur in the oral cavity are uncommon and can present a variety of clinical and histological manifestations and growth patterns. The biological behavior is benign and the reasons for development are unclear [1-4].

Although epithelial and mesenchymal hamartomas that severely affect the oral cavity are rare, it is undeniable that the presence of nervous

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or neurovascular components in hamartomas is even infrequent. Within the tongue, the endogenous components that can cause hamartoma include blood vessels, nerves, lymphatic vessels, skeletal muscles, adipose tissues, and salivary gland components [2, 3, 5].

Oral NVH presents as exophytic lesions usually with a smooth surface and less than 1 cm in diameter. Their color may range from pink to yellow, according to its predominant histological constitution, and in general, they are painless but can cause general discomfort to patients [2].

Although oral NVH are considered uncommon and under-reported lesions, with few studies in the literature, they should be considered in the differential diagnosis of oral lesions, so it is important to understand their clinical characteristics, histopathological findings, and treatment [2]. Thus, the objective of this work is to characterize the neurovascular hamartoma through the description of a clinical case with a brief literature review (Table 1).

Case report

Male (ASF), 47 years old, melanoderm, without systemic alterations, shows bilateral nodular injury on the dorsum of the tongue with an undetermined evolution period. The lesions present with well-defined limits, with approximately 10mm of diameter, surface and coloration similar to lingual mucosa, and firm consistency (Figure 1).

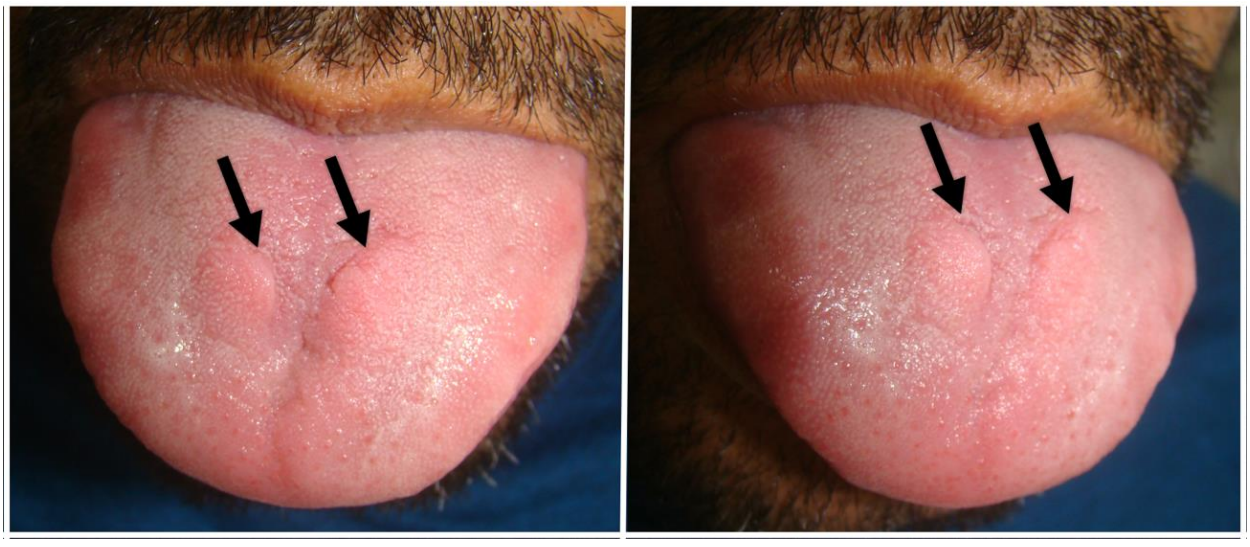

Figure 1. Clinical appearance of the hamartomatous lesion (arrows).

The diagnosed hypotheses were inflammatory fibrous hyperplasia and traumatic neuroma. After the incisional biopsy in one of the lesions, the fragments were submitted to histopathologic analysis. The microscopical

sections revealed, fibrous connective tissue, a large number of well-formed nerve bundles, grouped and closely intertwined with blood vessels of medium and small size (Figure 2). In the face of these characteristics, the diagnosis of NVH was established, and the patient preferred not to remove the remaining lesions.

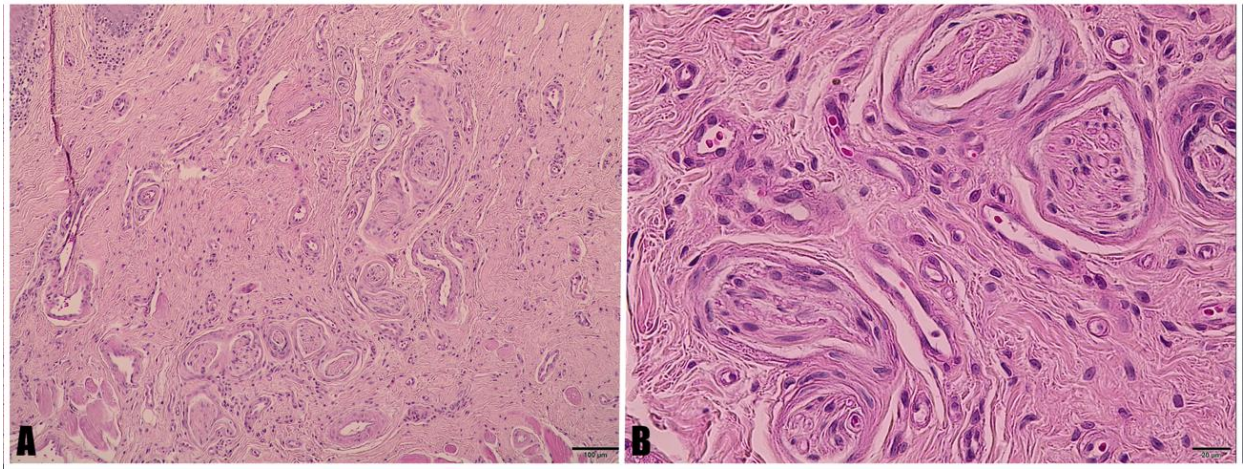

Figure 2. A. At low magnification, multiple nerve bundles and blood vessels are in subepithelial connective tissue. (H&E staining, magnification 10x). B. At high magnification, nerve bundles and blood vessels are closely intertwined (H&E staining, magnification 40x).

Table 1. Clinical data of neurovascular hamartomas in the literature.

Author	Age/gender	Initial diagnosis	Evolution time	Size	Location
[6]	29-year-old/male	Neurovascular hamartoma	N/A	11 x 7 cm	Left upper back and included the scapular area.
[1]	34-year-old/female	Fibrous hyperplasia;	Approximately 6 months to years.	0,4 cm	Corner of mouth
	20-year-old/female	Fibrous hyperplasia; Pyogenic granuloma.	Approximately 6 months to years.	1,8 cm	Tongue, middle, lateral

7-year-old/male	Fibrous hyperplasia;	Approximately 6 months to years.	0,6 cm	Buccal mucosa, area of corner of mouth
14-year-old/male	Fibrous hyperplasia;	Approximately 6 months to years.	0,8 cm	Gingiva, area 31–32
65-year-old/female	Papiloma	Approximately 6 months to years.	0,4 cm	Tongue, tip
59-year-old/male	Pyogenic granuloma	Approximately 6 months to years.	0,7 cm	Tongue
68-year-old/male	Lipoma, mucocele	Approximately 6 months to years.	1,0 cm	Lower lip
58-year-old/female	Fibrous hyperplasia;	Approximately 6 months to years.	1,7 cm	Tongue, lateral aspect
36-year-old/male	Fibrous hyperplasia;	Approximately 6 months to years.	0,5 cm	Tongue, dorsal aspect
59-year-old/female	Traumatic ulcer	Approximately 6 months to years.	2,5 cm	Tongue, lateral aspect
12-year-old/male	Fibrous hyperplasia	Approximately 6 months to years.	0,8 cm	Tongue, lateral aspect
23-year-old/male	Mucocele	Approximately 6 months to years.	0,7 cm	Lower lip
18-year-old/female	Rule out tumor	Approximately 6 months to years.	0,8 cm	Tongue, dorsal aspect
76-year-old/female	Rule out tumor	Approximately 6 months to years.	0,7 cm	Palate
67-year-old/male	N/A	Approximately 6 months to years.	0,5 cm	Tongue
46-year-old/female	Fibrous hyperplasia	Approximately 6 months to years.	0,5 cm	Buccal mucosa
51-year-old/female	Fibrous hyperplasia	Approximately 6 months to years.	0,8 cm	Tongue, dorsal aspect
63-year-old/male	Fibrous hyperplasia	Approximately 6 months to years.	1,0 cm	Tongue, tip
50-year-old/female	Fibrous	Approximately 6	0,25 cm	Tongue, tip

		hyperplasia	months to years.		
	44-year-old/female	Fibrous hyperplasia	Approximately 6 months to years.	0,4 cm	Lower lip
	46-year-old/female	Fibrous hyperplasia	Approximately 6 months to years.	0,5 cm	Tongue, anterior dorsal
	63-year-old/male	Fibrous hyperplasia	Approximately 6 months to years.	0,5 cm	Upper lip
	61-year-old/female	N/A	Approximately 6 months to years.	N/A	Lower lip
	N/A/male	Fibrous hyperplasia	Approximately 6 months to years.	1,0 cm	Lower lip
	6-year-old/female	Fibrous hyperplasia	Approximately 6 months to years.	0,5 cm	Buccal commissure
[2]	14-year-old/male	Neurovascular hamartoma	One and a half year	2×1.5 cm	Two finger breadths below the zygoma.
	4-Months/male	Hemangioma	4 months	N/A	Left kidney
[4]	1-Month/N/A	Hamartomatous nevoid lesion such as epidermal nevus or a vascular lesion.	4 and a half months	5,5 x 3,5 cm 2,5 x 2,0 cm	Left scapula
[7]	58-year-old/male	Undefined tumor of the small intestine	N/A	6,5 cm	Small bowel mucosa
[8]	91-year-old/male	Neuromuscular and vascular hamartoma of the small bowel	One year	2.5 x 2.0 x 2.0 cm	Small bowel
[3]	3-year-old/female	Traumatic lesions and benign neoplasms	N/A	0.1 to 2 cm	Dorsal left
	16-year-old/male	Traumatic lesions and benign neoplasms	N/A	0.1 to 2 cm	Dorsal right

[9]	73-year-old/male	Hamartomatous diaphragms of the small bowel	One year	N/A	Bowel wall
	76-year-old/male	Hamartomatous diaphragms of the small bowel	One year	N/A	Bowel wall
[10]	28-year-old/female	Squamous cell papiloma	6 months	N/A	Central surface of the tongue and the adjacent mucosa of the floor of mouth

Discussion and Conclusion

Traditionally, hamartomas appear as asymptomatic exophytic growths at birth or in early childhood, formed by disorganized but normal endogenous tissue [11], although they can be identified later, as in the case of the patient in this report. Elements described in hamartomatous lesions include smooth muscle, skeletal muscle, blood vessels, gland structures, fat tissue, nerves, lymphatic tissue, connective tissue, skin appendages, glial elements, and epithelium [2, 3].

According to Caruso et al (2018)[7], NVH is an extremely rare lesion with a macroscopic pattern of "neoplastic" appearance and clinical manifestations such as intense bleeding, which is caused by the abnormal mixing of the normal components of local native tissue. On the other hand, Junaid et al (2014)[1] indicate that histology consists of an abnormal mixture of cells and tissues located in specific locations, secondary to developmental errors. They do not develop as part of the inflammatory or tumor process and exhibit self-limiting proliferation.

Oral NVH presents in a wide age range, with a mean age of 44 years at diagnosis, with a predominance in females. Lesions predominantly occur on the tongue, followed by the oral mucosa and lower lip, however, lesions can be observed in other areas (Table 1). Eventually NVH can be interpreted as lesions associated with the HPV virus, like squamous papillomas (10). However, despite the possible polypoid architecture, histological analysis is required to establish the final diagnosis. In the pediatric population, these hamartomas may be described as part of genetic syndromes, especially oral-facial-digital syndrome [1, 3]. NVH can also be observed in the intestinal region, however, the hamartomatous nature of these lesions is questionable, since the

characteristics may be part of the histological spectrum of Crohn's disease, or even other enteral lesions [7].

Histologically, there are numerous nerve fibers associated with blood vessels of different calibers. In the immunohistochemical examination, the S-100 protein helps to identify the nervous component and CD34 helps to identify the vascular component. When talking about the differential histological diagnosis of tongue hamartoma, we can include reactive or traumatic lesions and benign neoplasms. Oral NVH remains traumatic hamartomas. The main difference is vascular proliferation. In neurovascular hamartoma, vascular proliferation and nerve components are closely linked, whereas in traumatic neuroma, these two components are separate, and the neural component is dominant [2, 3].

Thus, considering that this rare lesion has specific features, we need to consider it in the diagnosis of oral entities, especially on the tongue. There are reports that are described in the table, where neurovascular hamartoma on the tongue was initially diagnosed as fibrous hyperplasia, pyogenic granuloma, papilloma, traumatic ulcer, among others. Therefore, it is important to know the histopathological characteristics associated with the lesion.

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