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Hemangioma of the uvula: the most frequent vascular tumor with a rare localization - Case report with literature review

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ABSTRACT
Hemangioma is the most common tumor of the vascular origin, benign, frequently observed in children, and with preference for head and neck, but rarely having a uvular origin. In this organ, it presents potential risk of local trauma, hemorrhage and it may produce great discomfort for the patient. In this article we present the cases of two patients (34 year-old and 44-year old) with uvular hemangiomas; patient 1 was admitted for dysphagia, salivation and choking and patient 2 was admitted for the swelling of the uvula, dysphagia, salivation and difficulty in speaking. They both presented a history of this symptoms, with acute episodes of dysphagia. They did not report bleeding, pain or fever. Hemangiomas were histopathologically confirmed, thus raising the awareness that hemangiomas are included in the differential diagnosis when a patient presents an elongated, swollen uvula or uvular masses.

Keywords: venous hemangioma, cavernous hemangioma, uvula, benign vascular tumor

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Introduction

Hemangiomas are benign neoplasms that may be circumscribed. They usually have well-formed vessels, a wide variety of patterns, and clinical manifestations. They may be difficult to distinguish from hamartomatous conditions or malformations [1]. Hemangioma is one of the most common soft tissue tumors (7% of all benign tumors) and is the most common tumor during infancy and childhood. Most hemangiomas are superficial lesions that have a predilection for the head and neck region, but they may also occur internally, notably in organs such as the liver. The common capillary and cavernous hemangiomas of adults are more frequently encountered in women and may fluctuate in size with pregnancy and menarche; this suggests that the endothelial cells of these tumors may be responsive to circulating hormones. Although some vascular tumors regress altogether (e.g., juvenile hemangioma), most persist if untreated but have limited growth potential. Hemangiomas virtually never undergo malignant transformation; likewise, the concept of a benign metastasizing hemangioma is no longer accepted [2]. The genetic background of inheritable hemangiomas and vascular malformations is becoming known from

hereditary syndromes, but the somatic genetics of hemangiomas is still poorly understood [3].

Case report:

Case 1:

A 34-year-old man was admitted for dysphagia, salivation and choking. He described a 2–year history of dysphagia episodes and very loud snoring.

He did not report bleeding, pain or fever. No significant past medical history. Criteria for inclusion in the risk group for sexually transmitted diseases were absent. The oropharyngeal examination revealed a large volume uvula with irregular surface, presenting an angiomatous lesion, dark violet, that affected the whole extension, with elastic consistency and non-painful upon palpation. Upon inspecting it with posterior rhinoscopy and careful palpation of the soft palate and the tumor, it was suggested that the diagnosis would be a hemangioma limited to the uvula. The physical examination including cervical and facial areas did not show any associated abnormalities. Conventional surgery was performed.

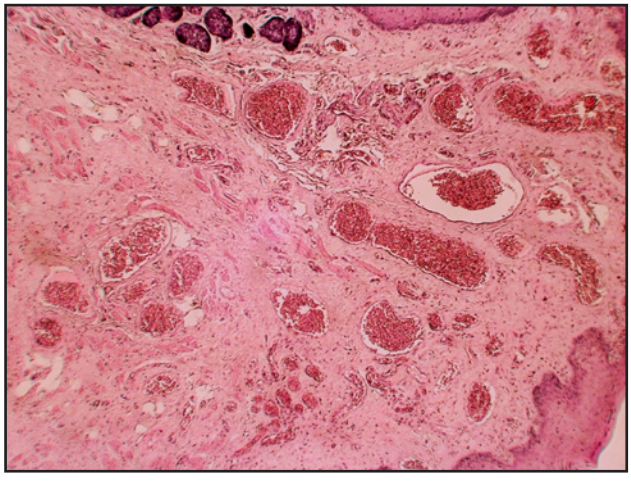

Figure 1. Connective stroma with numerous dilated blood vessels, the underlining endothelium is without atypia, the vessels are full of red blood cells. [H&E stain 4x]

Gross findings: post-surgery, we received a 1/0.8/0.6cm dark violet polypoid lesion with a polycyclic surface, with elastic consistency. When sectioned, the lesion presented a solid white aspect, with medium consistency.

Venous hemangioma was histopathologically confirmed.

On the fully included material we identified a connective stroma with numerous dilated blood vessels, the underlining endothelium was without atypia, the vessels were full of red blood cells (Figure 1). We remarked seromucous glands which were histologically normal (Figure 2).

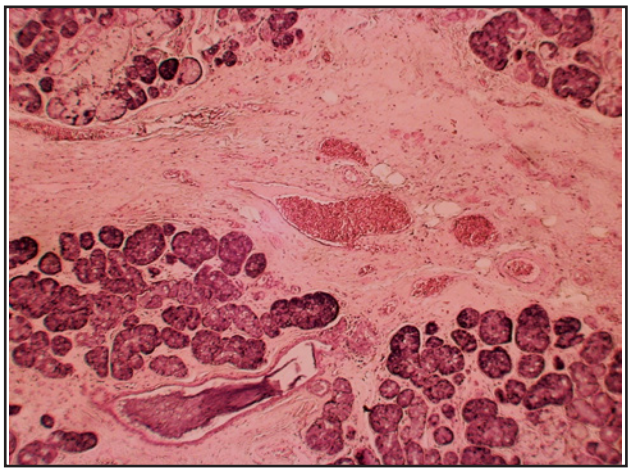

Figure 2. Dilated blood vessels full of red blood cells. Typical seromucous glands. [H&E stain 4x]

The stromal fragment was externally surrounded by a histologically normal stratified squamous epithelium. In the connective stroma we observed a mild lymphocytic infiltrate associating neutrophils; this infiltrate was solidly disposed. Malignancy was absent. Immunohistochemical study showed that CD34 stain highlighted the endothelial cells, supporting this diagnosis (Figure 3).

Case 2:

A 44-year-old man was admitted accusing the swelling of the uvula, dysphagia, salivation and difficulty in speaking. He described a 3–year history of episodes of dysphagia, choking and salivation.

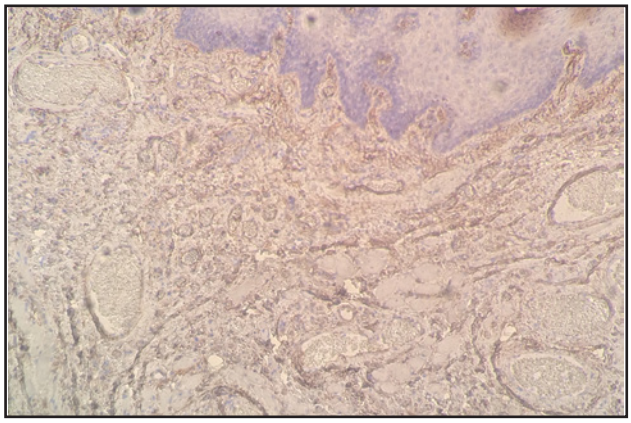

Figure 3. Positive CD34 stain. [4x]

He did not report bleeding, pain or fever. No significant past medical history. Criteria for inclusion in the risk group for sexually transmitted diseases were absent. The oropharyngeal examination revealed large volume uvula with regular surface, presenting an angiomatous lesion, yellow with violet spots, with elastic consistency and non-painful upon palpation. Upon inspecting it with posterior rhinoscopy and careful palpation of the soft palate and the tumor, it was suggested that the diagnosis would be cavernous hemangioma limited to the uvula. The physical examination including cervical and facial areas did not show any associated abnormalities. Conventional surgery was performed.

Gross findings: post-surgery, we received two nodular fragments (diameters: 0.5cm, 0.7cm), with regular, glossy surfaces. Both fragments where yellow with brown areas. When sectioned they presented a compact, translucent aspect, with little red and brown spots.

Cavernoushemangiomawashistopathologically confirmed. Microscopic examination revealed poorly circumscribed large cavernous spaces lined by flattened endothelium, separated by scant connective tissue, morphologically consistent with cavernous hemangioma (Figure 4, 5). Malignancy was absent.

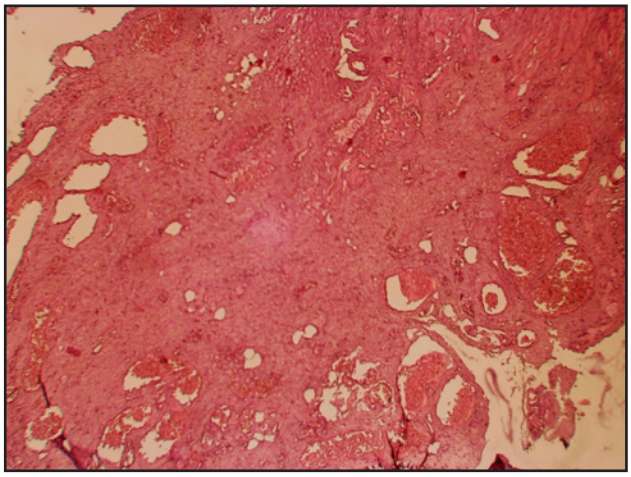

Figure 4. Poorly circumscribed large cavernous spaces lined by flattened endothelium, separated by scant connective tissue. [H&E stain 4x]

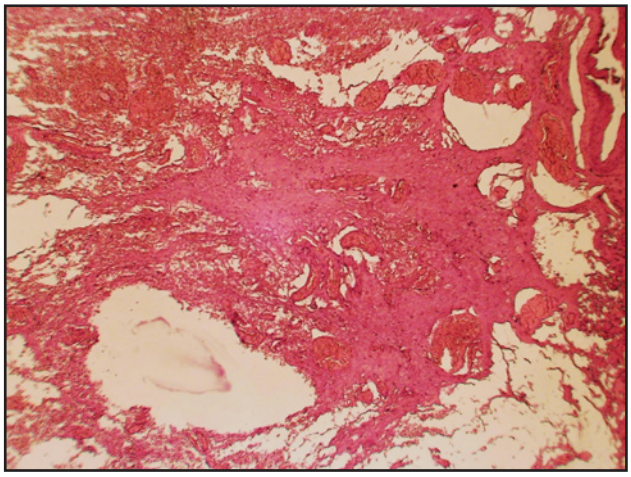

Figure 5. Poorly circumscribed large cavernous spaces lined by flattened endothelium, separated by scant connective tissue. [H&E stain 4x]

Discussion:

The most common tumor of vascular origin is the hemangioma. Hemangioma is a benign tumor, frequently observed in children, and with preference for head and neck [4]. In

the head, its preferred location is the face, oral mucosa, lips and tongue and it rarely affects the uvula [5]. Hemangiomas of the oral cavity are relatively rare with a prevalence rate of less than 1% (mostly involving lips, tongue, buccal mucosa, and palate) [6], and a prevalence of 8 in one thousand males and 4 in one thousand females [7]. Hemangiomas are rarely localized in the uvula. In this organ, they present potential risk of local trauma and hemorrhage [4]. This location can make it vulnerable to bleeding by local trauma [8]. Most of the hemangiomas can be diagnosed with clinical examination. Frequently, in about 95% of the cases, clinical history and physical examination are enough to differentiate a hemangioma from vascular malformations [4].

Venous hemangioma is composed of veins of variable size. Pure venous hemangiomas are rare and mainly present in adults. They are long-standing slowly growing tumors. The lesion is frequently ill defined and consists of dilated congested vascular spaces with areas of hemorrhage [9]. Cavernous hemangioma is a subtype of hemangioma. The involvement of the uvula by a cavernous hemangioma is extremely rare. To date, only 5 cases have been reported in the English literature [6]. Cavernous hemangiomas usually occur in young children but are far less frequent than capillary hemangiomas. They tend to be larger than other hemangiomas and frequently involve deep structures, particularly the liver. They do not regress and tend to be less well-circumscribed; they may cause local tissue destruction because of compressive effects. They require surgical removal and do not respond to medical therapy. On cut section, a sponge-like appearance is common. Morphologically they consist of large, dilated, blood-filled spaces, often with fibrotic walls. It may be vaguely lobular or consist of elaborately interanastomosing vascular spaces, resembling a sponge. The lining endothelium is usually flattened [10]. Pathological angiomatous growth of the uvula, starting in adult life, within a relatively short period, could result from previous vascular venous malformation, with quick expansion caused by local trauma, infection or hormone abnormalities, or hemangioma with clinical repercussion in a young adult [11]. Hemangiomas can present complications such as ulceration, hemorrhage, infection and thrombocytopenia [11]. Treatment of hemangioma is quite variable and may be customize for each patient. The therapeutic options are watch and see management, surgery, definite or preoperative embolization, laser, cryotherapy, radiotherapy or sclerotherapy, in addition to drug treatment with corticoids or fibrinolytic agents 1, 6, 7 [11]. In both our cases conventional surgery was performed. No complications were reported.

Conclusion:

Although rare, uvular hemangioma has to be included in the differential diagnosis when a patient presents an elongated or swollen uvula or a uvular mass because it may pose a threat due to its great risk of bleeding. Even though this tumor is benign, uvular localization may produce great discomfort to the patient because of possible obstructive oropharyngeal symptoms as dysphagia, choking, snoring, sleep apnea and difficulty in speaking. By reporting this cases we would like to raise the awareness that hemangiomas must not be forgotten in the process of diagnosing a uvular swelling or mass.

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