

cytology is not reliable. It is important to assess preoperatively if there is suspect of angiolipoma to make a precise (and less invasive) surgical planning.

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## Conservative Surgical Treatment of Tongue Hemangiopericytoma

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**Abstract:** Hemangiopericytoma is a vascular tumor that is believed to arise from the Zimmermann's pericytes, smooth muscles cells localized around the blood vessels. This tumor presents as a slowly enlarging painless mass with a clear predilection for the musculoskeletal system. The aim of this work was to introduce a peculiar case of a tongue hemangiopericytoma managed by conservative surgical treatment.

**Key Words:** Hemangiopericytoma, mesenchymal tumor, neoplasm of the tongue

**H**emangiopericytoma (HPC) is a vascular tumor described initially by Stout and Murray<sup>1</sup> in 1942; it is believed to arise from Zimmermann's pericytes, smooth muscles cells localized around the blood vessels. Hemangiopericytomas of the cervicofacial region account from 9.4% to 28%; it can occur at any age, without any sex

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predilection. The most frequently involved sites reported in the literature are nasal and paranasal region, oral cavity, parotid gland, orbit, scalp, and the infratemporal fossa.<sup>2</sup>

This tumor presents as a slowly enlarging painless mass with a clear predilection for the musculoskeletal system. The diagnosis of certainty can be troublesome because of the close likeness to other spindle cell neoplasms such as malignant fibrous histiocytoma, solitary fibrous tumor (SFT), synovial sarcoma, and mesenchymal chondrosarcoma.<sup>3,4</sup>

The aim of this work was to introduce a peculiar case of a tongue HPC managed by a conservative surgical treatment.

## CLINICAL REPORT

On December 2010, a 77-year-old woman was referred to our department by her general practitioner for a painless mass in the apex of the tongue, localized in the left paramedian region, which had appeared 4 years before (Fig. 1).

There was no history of breathing or swallowing difficulty. Oral examination revealed a bulging mass with normal mucosa. The remainder head and neck examination did not reveal any lymphadenopathy or pathological findings.

A magnetic resonance imaging was performed to evaluate the dimension of this neoplasm. A vascular flow was detected in the apex of the tongue of 3-cm diameter (Fig. 2).

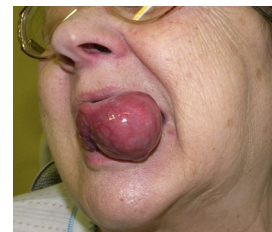
The lesion was approached under general anesthesia through a transoral paralateral glossotomy. To preserve the tongue's function, the tumor was resected encapsulated (Fig. 3).

Microscopically, the tumor was composed of hyaline stroma and spindle cells distributed in sheets and cords with voluminous, round, and ovoid nuclei. The architectural pattern was "staghorn" or "antler-like." The mitotic rate was low (<1 per 10 high-power fields). There was no evidence of necrosis or cytological atypia. The lesion was positive for CD34 and negative for actin and synaptophysin.

On the basis of this pathological finding, the diagnosis of HPC was made. The postoperative course was uneventful; the patient was discharged 7 days after the surgical procedure, and good mucosal healing was observed after 20 days. At 1-year follow-up, clinical and radiological examination did not reveal pathological relapse (Fig. 4).

## DISCUSSION

Although HPC occurs more frequently in the adults, a minority rate is reported for the pediatric population; there are 2 forms of this soft tissue tumor: the congenital/infantile and the adult. These 2 variants present different histological features and clinical behavior. The pediatric form (occurs at birth or first year of life) occurs more frequently in the subcutaneous tissue of the head and neck region or the extremities with a large size; however, it is associated to a more benign clinical behavior than the adult variant.<sup>5</sup>



**FIGURE 1.** Clinical view of the lesion: neoplasm of hard consistency.

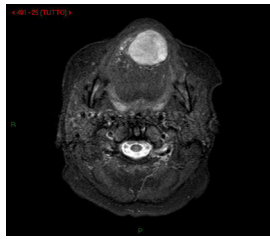


FIGURE 2. T2-weighted magnetic resonance slide of the lingual lesion.

The pathological diagnosis requires microscopic examination and immunohistochemical and ultrastructural techniques. Microscopically, it is characterized by staghorn or antler-like vessels; HLA-DR, factor VIII, and vimentin are usually positive, whereas S100 protein, CD34, smooth muscle actin, and Leu-7 are variable. Electron microscopic examination confirms the origin of these cells from the pericytes, which are peculiar cells in the walls of venules and capillaries, separated from the endothelial cells by the basement membrane.<sup>6-8</sup>

Distinction with SFT of the oral cavity was the most difficult differential diagnosis because SFTs can sometimes display extensive areas with a growth pattern that is virtually indistinguishable from HPC. However, morphologic patterns, such as herringbone formations, neurofibroma-like and schwannoma-like areas, and diffuse sclerosing areas, which may be present in SFT, are not generally associated with HPC.<sup>9</sup> Furthermore, in our case, the pathologist described multilobular fashion, a feature usually not found in SFTs. Also, cellularity can be used for differential diagnosis: in the SFT, it varies from area to area and is inversely related to the amount of collagen; the feature observed in our case showed a homogeneous pattern of cellular distribution.

Enzinger and Smith<sup>10</sup> defined possible histologic criteria to predict the clinical outcome: mitotic activity, cellularity, necrosis, and hemorrhage. The lesion is considered as high-grade tumor if it presents more than 3 mitotic figures per 10 high-power fields, increased cellularity, necrosis, and hemorrhage.

The clinical behavior of this tumor is very uncertain; some tumors are considered as benign lesions, whereas others are malignant variants with a metastatic course. The most common site of metastasis is the lung; however, it has been observed that the youngest patients have the lowest rate of malignancy. The histological feature of this tumor, which is considered a borderline neoplasm and therefore necessitating a close follow-up, does not predict the clinical course; instead, cytological atypia, frequent mitosis, and tumor necrosis are associated with malignant behavior.<sup>2,3</sup>

Given the relatively low incidence of this tumor and its unpredictable behavior, several doubts can arise regarding the best choice for a successful treatment. The most frequent therapy reported by the literature is surgery. Several authors have described adjuvant post-operative radiotherapy/chemotherapy for high-grade tumors or for incomplete resections. However, long-term follow-up is necessary to assess long-term outcome, detecting any potential relapses.<sup>6-8</sup>

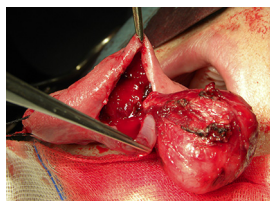


FIGURE 3. Enucleation of the capsulated neoplasm.



FIGURE 4. Healing of the tongue 1 year after surgery.

We treated this case with a conservative surgical procedure. We preserved the function of tongue. This peculiar case adds knowledge to the scientific literature regarding the complex field of head and neck HPC.

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## Gorham Disease in the Maxilla

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**Abstract:** Gorham disease is a rare condition that is characterized by the proliferation of thin-walled vascular channels associated with

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