

Angioleiomyoma of the oral cavity extended to submandibular space; an unusual tumor in an unusual deep-seated space: a case report

A. MINNI, A. DE CARLO, R. ROUKOS, G. ILLUMINATI¹, B. CERBELLI²

Department of Sensory Organs, ¹Department of Surgery Francesco Durante, ²Department of Pathology; Sapienza University of Rome, Rome, Italy

Abstract. – Angioleiomyoma (AL) is a benign neoplasia originating from smooth muscle and very uncommon in the oral cavity. The most frequent subtype in the oral cavity is the vascular one. AL usually occurs in the extremities: only around 12% are found in other areas such as head and neck. It presents as an asymptomatic, slow growing nodule lodging in the palate, tongue or lips. The diagnosis is essentially by histological exam and special specific stains are helpful to confirm the origin and to distinguish it from other tumors. We present a case of AL found in unusual site: attached to the submandibular region in a deep-seated space.

Key Words:

Angioleiomyoma, Rare tumor of oral cavity.

Introduction

Vascular leiomyoma or angioleiomyoma (AL) is a benign smooth muscle tumor that occurs most frequently in the uterine myometrium, gastrointestinal tract and skin. Because of the paucity of the smooth muscle, occurrence in the oral cavity is rare and it is approximately about 0,065%¹. Stout² suggested that this type of tumor origins from the tunica media of the blood vessels wall, whereas Glass³ considered a possible origin from ductus lingualis, exactly from the circumvallate papillae.

The oral AL can appear at any age, but the most prevalence is between 40-60 years old especially in men. It spreads out mainly in the lips, tongue and soft palate, but there have been cases of location in the cheek and retromolar trigon⁴⁻⁶.

The best option for treatment is surgical excision and there has been only one case of recurrence just 2 weeks after surgery⁷.

Nowadays there are only 48 cases of oral AL reported in the literature. We present a case of

AL found in an unusual site, developing from the floor of the oral cavity deep to the submandibular space.

Case Report

The patient is a 54 years old woman that recovered in our Hospital because of a cranial trauma with haemorrhage and fracture of the cranium teca. During the recovery she claimed a right swelling under the mandible in the last 2 years with no other symptoms. Thus, she underwent occasionally a MRI of the neck that demonstrates the presence of a submandibular mass of about 37×37×35 mm, which likely came from the floor of the oral cavity. The mass displaced the submandibular gland, the tongue and the oropharynx airway compressing the right tonsil, without any phenomena of infiltration neither of soft tissues nor of bone (Figures 1 and 2). Furthermore, our evaluation revealed a swelling of about 3 × 3 cm in diameter, firm at touch, mobile at superficial and deep layers and it didn't present any unusual coloration. There was not associated lymphadenopathy or abnormality of the skin. A ultrasound-guided FNA yielded markedly blood-stained aspirate: the cytology showed red cells only.

The lesion was completely removed in general anaesthesia appreciating the difficult detachment and isolation of the lingual nerve that entered the mass and the hypoglossal nerve, which was isolated.

Histological examination revealed a well-demarcated and encapsulated nodule of smooth muscle tissue punctuated with thick-walled vessels, capillary and venous type vessels.

The inner layers of smooth muscle of the vessel are arranged in an orderly circumferential fashion, and the outer layers spin and swirl away from the vessel, merging with the less well-ordered peripheral muscle fibers (Figure 3).

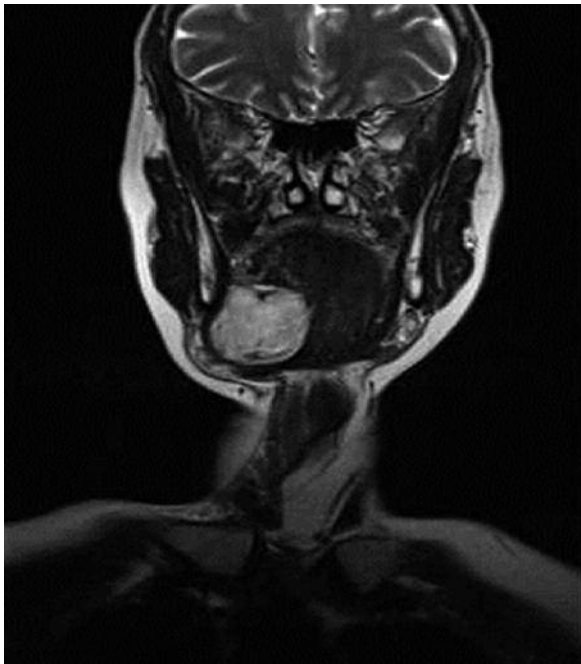


Figure 1. Image showing the mass dislocating the submandibular space.

Leiomyocytes were spindle with blunt-end, cigar-shaped nuclei and abundant pink cytoplasm; neither necrosis or mitotic figures were seen. The neoplasms stained positive for smooth muscle actin (AML) (Figure 4).

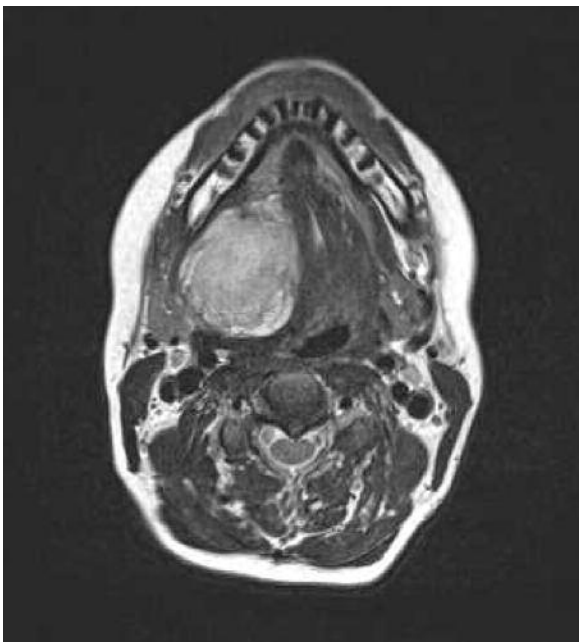


Figure 2. Image showing the mass displacing the tongue and oropharynx.



Figure 3. 10x Haematoxylin-eosin staining for endothelial layer of vessels.

With all these features final diagnosis was of AL. Six months after excision of the lesion no signs of any recurrence are observed.

Discussion

Vascular leiomyomas are rare in the oral cavity because of the paucity of the smooth muscle. It is claimed that the dose of topical estrogen and mechanical factors such as external injuries take part in the occurrence of angioleiomyoma, but it is not clear yet⁸. When they arising like an asymptomatic nodule, the most frequent sites are lips, tongue and palate. Uncommon is the development in the cheek and retromolar trigon as leiomyoma located in the bone. Radiographically the oral intraosseous leiomyoma is characterized

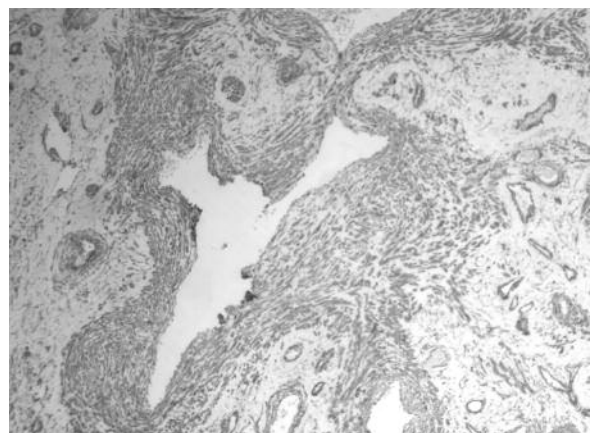


Figure 4. 5x smooth muscle actin (AML) staining for leiomyocytes surrounding vessels.

by an uni or multilocular radiolucency, generally with a sclerotic border without cortical displacement⁹. It can cause mobility of teeth and occasionally dysphagia and dysgeusia¹⁰. In the head and neck AL is the most usual smooth muscle tumor in the turbinates and larynx¹¹.

Their peak incidence is the fourth and fifth decades and they are unusual in children and old patients where myosarcoma is more likely. They occur more commonly in male subjects and are usually painless; the possible pain may be explained by an intratumoral vasoconstriction with follow local ischemia or a compression of somatic nerve in the tumor⁸.

The World Health Organization (WHO) distinguished three types of leiomyoma: leiomyoma (solid), angiomyoma (vascular leiomyoma), and epithelioid leiomyoma (leioblastoma). The angiomyoma has the higher frequency with a 74%, followed by solid leiomyoma with a 25% and there is only one case in literature of epithelioid leiomyoma¹². There have been cases of granular cell leiomyoma of the oral cavity as well¹³.

The vascular leiomyoma (AL) is characterized by a proliferation of mesenchymal cells, with eosinophilic cytoplasm and nuclei with a typical cigar-like aspect, and a single layer of endothelial cells, visible with the Haematoxylin-eosin coloration, which lined the vascular spaces. In the same lesion is possible to notice an heterogenic cell population formed by two or more different histological patterns⁹. Furthermore the immunohistochemistry is important to diagnose a leiomyoma, but even to differentiate other mesenchymal tumors characterized predominantly of fusiform cells such as myofibroma, myopericitoma, inflammatory myofibroblastic tumor and histiocytoma⁹. In our case the neoplastic cells showed a positive immunoreaction to actin. Smooth muscle actin corresponds to the alpha fraction of the actin chain; it is an immunomarker of smooth muscle but it could have an immunoreaction in skeletal muscle¹⁴. Other immunomarkers are S-100 protein, Vimentin, which is a structural protein of the cytoplasm filaments of mesenchymal cells and Desmin, that is a type III intermediate filament near the Z line in sarcomeres¹⁴. The endothelial cells present immunoreactivity against anti-CD34 antibody. CD-34 is a transmembrane protein that is broadly expressed by vascular endothelium¹⁴.

It is useful for making a differential diagnosis between smooth muscle tumor like leiomyoma and myofibroma the Masson Trichrome Stain. It

is observed that smooth muscle lesions are less abundant of fibrous tissue surrounding the smooth muscle cells and in the septa between the smooth muscle masses. Moreover, the smooth muscle masses appear red while the fibrous tissue is blue on low-power view¹⁵. This tumor is difficult to diagnose before surgery, especially when it occurs in the head and neck region as a painless mass. In our case neither FNA nor RMI examination was diagnostic but both pointed toward a vascular lesion. RMI was useful in demonstrating the deep location and the abnormal vascularity within the solid tumor.

Anyway, AL is usually found in a superficial location and a deep-seated lesion close to the submandibular gland similar to the present case was not previously reported. In most cases, the tumor is completely resected and preoperative embolization could occur to minimize the risk of possible hemorrhage. In our case we preferred a submandibular approach to remove the mass because of its extension. The submandibular gland was detached from the tumor as the lingual nerve that entered the lesion as the hypoglossal nerve which was stressed by it. In six months we don't have any recurrence and the patient recovers her taste and function of the tongue.

In conclusion, AL arising from the space deep to region of the submandibular gland is a rare tumor; with the awareness of this tumor, AL should be included in the differential diagnosis of a mass in the submandibular triangle.

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