Giant Cell Angiofibroma (GCA), also referred to as Giant Cell-Rich Solitary Fibrous Tumor (SFT) with the new pathological classification, is a kind of soft tissue tumor that belongs to the solitary fibrous tumor group and is usually located at the orbital region.\(^1,2\) It is a different entity from nasopharyngeal angiofibroma, which also can be diagnosed in extra nasopharyngeal localizations.\(^3\) Although Giant Cell-Rich SFT is described as a primary tumor of the orbital region, there are many case reports of extraorbitally localized Giant Cell-Rich SFTs. Although rare, it can be encountered in the head and neck region, such as the oral cavity.\(^4\) To our knowledge, Giant Cell-Rich SFT was never identified in the sublingual region. We report the case of a 49 years old female who presented with painless, slowly growing mass in sublingual region, underwent excisional surgery and was diagnosed with giant cell-rich solitary fibrous tumor previously referred to as giant cell angiofibroma. In our report, we aimed to report the unusual localization of this rare tumor, examine the new nomenclature and classification of giant cell-rich solitary fibrous tumor or giant cell angiofibroma and review the literature regarding head and neck localization of this tumor.

**Case Report**

We present the case of a 49-year-old female who presented with painless, slowly growing mass in the sublingual region. A soft, mobile, painless mass with dimensions of 4x3x2 centimeters was palpated in the left sublingual region. The mass was in close proximity to the sublingual gland and initially was thought to be a sublingual gland mass. Our report, we aimed to report the unusual localization of this rare tumor, examine the new nomenclature and classification of Giant Cell-Rich SFTs or GCAs and review the literature regarding head and neck localizations of this tumor.

**Keywords:** Mouth floor; solitary fibrous tumors; sublingual gland neoplasms.
fied mass close to the midline in the submental area. MR imaging revealed a mass in the left sublingual region with close proximity to the sublingual gland with dimensions of 30x25x21 millimeters. The mass had a smooth surface, was isointense in T1A series, hyperintense in T2A series and had strong contrast enhancement (Fig. 1).

A Fine Needle Aspiration (FNA) Biopsy was performed and histologically multinucleated cells with hyperchromatic nuclei amongst vascular structures within fibrotic stroma were identified in a focal area between smooth muscle fibers. Transoral excision of the mass was planned. During the operation, the tongue was displaced with 0 silk sutures. Mandible was displaced inferiorly. Purple, smooth contoured, solid mass with dimensions of 4x3x2 centimeters was excised. The sublingual gland was identified during the surgery. No unexpected bleeding was encountered, and small mucosal bleedings were easily controlled with bipolar and monopolar cauterization (Fig. 2). No postoperative complications were encountered, and the patient was discharged the day after surgery.

Microscopic examination revealed ectatic vascular proliferations within a fibrotic stroma in x10 magnification and multinucleated giant cells in x40 magnification (Fig. 3). CD 34 was positive endothelially. Smooth Muscle Antigen (SMA), Estrogen and Progesterone receptors were negative. Ki 67 proliferation index was below 1%. After the definitive pathology report, patient was diagnosed with sublingual GCA or Giant Cell-Rich SFT. During the follow-up visits for six months, no recurrence was noted.

Discussion

GCA was first described by Dei Tos et al. in 1995. In their study, Dei Tos et al.[1] differentiated this tumor on the basis of histologic appearances from solitary fibrous tumor and suggested the designation “Giant Cell Angiofibroma of the orbit”. Although GCA was originally described as a tumor of the orbital cavity and periorbital soft tissues, reports of extraorbital localizations of this rare tumor started to accumulate in diverse regions, including oral cavity. [4] To our knowledge, GCA was never identified in the sublingual region.

Although previously GCA was recognized as a distinct entity, after similarities with Solitary Fibrous Tumor was detected pathologically,[5] it is now classified under solitary fibrous tumor and specified as a synonym with it.[2] In the recent update on the clinical, molecular and diagnostic features of the extrapleural solitary fibrous tumor, GCA is referred to as a giant cell-rich variant of SFT.[2] Previous case reports referred to this entity as Giant Cell Angiofibroma. More recent ones, such as one in which external auditory canal localization was reported, used the term Giant Cell Rich Solitary Fibrous Tumor.[6] In the light of the new classification, it was deduced that these terms were used interchangeably in recent reports, so these terms were used to refer to the same entity throughout our case report.

In a recent review of 36 cases of head and neck SFTs, only six cases showed angiofibroma-like histology and were therefore classified as giant cell angiofibroma like a solitary fibrous tumor. Of those six cases, only one case was localized extraorbitally, namely in the nasal cavity.[7] There are some pathological features that are diagnostic of this tumor, such as the absence of infiltrating growth pattern, fibroblastic spindle cells mingled with collagenous stroma, and prominent vascularity. The presence of multinucleated giant cells localized around pseudovascular spaces is another histopathological hallmark of this tumor. [4] In our case, vascular ectasia proliferation within a fibrotic stroma was identified, raising the suspicion of GCA. CD34 positivity is expected in most GCAs, as was detected in our case. Other immunohistochemical markers are bcl-2 and CD94. GCA is expected to be non-reactive with des-
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There is a report of a patient who was diagnosed with a vascular malformation of the parotid space after MRI, CT Scan and FNA were conducted. The patient was treated with a regimen of percutaneous sclerotherapy using ethanol and lauromacrogol for one year with no clinical response. After surgical excision, the final pathological report concluded that the mass was not a vascular malformation but a GCA. As is evident in this case, without surgical excision, it is difficult to diagnose this tumor. Clinical features vary and show similarities to other vascular tumors. In most the cases, imaging modalities cannot differentiate between various more common vascular tumors, let alone GCA. FNA usually indicates vascularity but is insufficient to make a definite diagnosis.

Although there are some reported cases with the recurrent disease of extraorbital GCAs in the literature; in our review, no report of recurrence was detected for this tumor in the head and neck area after total excision. Because of the rarity of this tumor and scarcity of the data, long-term follow-up was planned for our case.

**Conclusion**

Although a rare entity, possible diagnosis of GCA or Giant Cell-Rich SFT should be kept in mind in vascular tumors presenting with atypical features. Also, it should be kept in mind that in the current literature, although the classification has changed recently, Giant Cell Angiofibroma and Giant Cell-Rich Solitary Fibrous terms are used interchangeably and to define the same entity. When this diagnosis is suspected, it is advisable to refer to current pathological classification and newly identified markers. Although with current knowledge, surgical excision seems to be curative, more data are needed to solidify information regarding the clinical behavior of this tumor in the head and neck region. The authors have no conflict of interest to declare. No third party funding was received. Patient consent was obtained to publish clinical photographs. All authors have viewed and agreed to the submission.

**Disclosures**

**Informed consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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**Conflict of Interest:** None declared.

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