International Journal of Otorhinolaryngology and Head and Neck Surgery
Vahl JM et al. Int J Otorhinolaryngol Head Neck Surg. 2019 Nov;5(6):1705-1707
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20194955

Rhabdomyoma of the tongue base

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Received: 13 August 2019
Accepted: 01 October 2019

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ABSTRACT

Extracardiac rhabdomyoma is a very rare condition with potentially multifocal manifestations including the head and neck region. We present a case of an 81-year-old patient who presented as a medical emergency because of ambiguous, progressive throat pain and dysphagia. After diagnostic work up including magnetic resonance (MR) scan and biopsy a rhabdomyoma of the tongue base was diagnosed. The tumor was completely excised by transoral laser microsurgery and the patient's symptoms vanished. This case report is described in context of the current literature with a discussion of diagnostic steps and treatment options of rhabdomyoma in the head and neck region. Rhabdomyoma is a rare, but nevertheless relevant differential diagnosis of head and neck tumors.

Keywords: Mesenchymal tumor, Soft tissue tumor, Head and neck, Tuberous sclerosis, Tumoral laser surgery

INTRODUCTION

Rhabdomyomas are rare myogenous, slowly growing, circumscribed but not encapsulated benign tumors. They are divided into cardiac and extracardiac tumors, representing 2% of striated muscle tumors.[1,2] Etiologically the genes tuberous sclerosis 1 (TSC1), coding for hamartin, and TSC2, coding for tuberin, have been identified to be frequently associated with the genesis of rhabdomyomas from embryonal myoblasts, especially in infantile cardiac tumors.[3] Interestingly cardiac rhabdomyomas tend to partly or fully regress after birth presumably due to lack of maternal estrogen.[3] Extracardiac rhabdomyomas are further divided into three subtypes: fetal and adult type tumors (75%), which are mainly found in the head and neck and genital type tumors (14%), found almost exclusively in the vagina and vulva of middle aged women.[1,4] The average age to be diagnosed with adult extracardiac rhabdomyoma is between 40-60 years (male/female:3/1).[1,5] Fetal extracardiac rhabdomyomas usually become symptomatic by the age of 3 years or less.[5] While fetal rhabdomyomas histopathologically present immature muscle differentiation, the adult form presents mature skeletal muscle structures.[1] Furthermore multifocal extracardiac rhabdomyomas are documented too, but are extremely rare.[2,4,6] The affection of the tongue base has been rarely described.[6,7]

CASE REPORT

An 81-year-old man presented with progressive, left sided throat pain, radiating to the ipsilateral ear for few days to an ENT physician during emergency service. Endoscopic examination of the throat revealed a massive tumor of the left tongue base with intact mucosa. Since no relevant infectious parameters were present a cervical magnetic resonance imaging (MRI) was performed (Figure 1). The tumor of the left tongue base presented as a 4 × 3 × 3.5 cm sized, spherical formation was suspected to be a solid and single neoplasm of unknown dignity for example a lymphoma or a struma of the tongue base. A panendoscopy with biopsy was performed and the tissue was histopathologically examined, showing a solid tumor formation with tightly arranged cells. The cell nuclei were isomorphic and round. The majority of the cells presented an optically empty cytoplasm with vacuoles, while some cells showed an eosinophilic cytoplasm.

Mitotic activity was not observed. Histochemical periodic acid-Schiff (PAS) staining reaction revealed PAS-positive cytoplasm, which was diastase sensible, correlating to intracytoplasmatic glycogen. Immunhistochemically the muscle specific proteins desmin and actin were expressed, while staining for S100B-protein (i.e., marker of granular cell tumor) was negative (Figure 2), concluding the diagnosis of a rhabdomyoma. Subsequently a carbon dioxide laser (CO_2) laser-based, total excision of the tumor was performed. After healing of the operating field the initial symptoms vanished. A follow up period was planned.

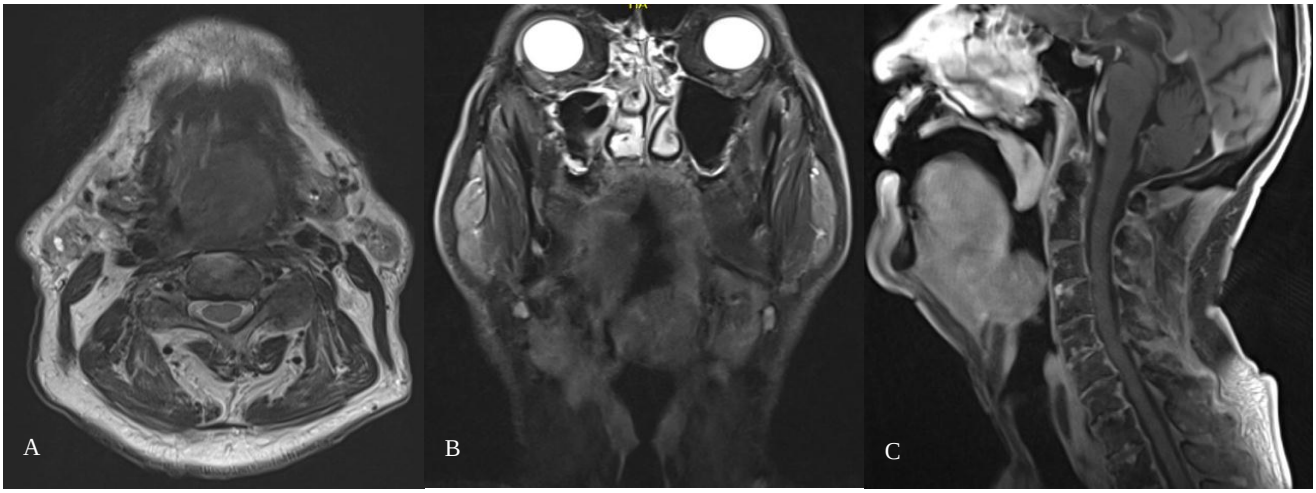

Figure 1: MRI of the rhabdomyoma of the tongue base, measuring 4 × 3 × 3.5 cm, shown in a (A) transversal T2 weighted, (B) frontal T1 weighted and (C) sagittal T1 weighted section.

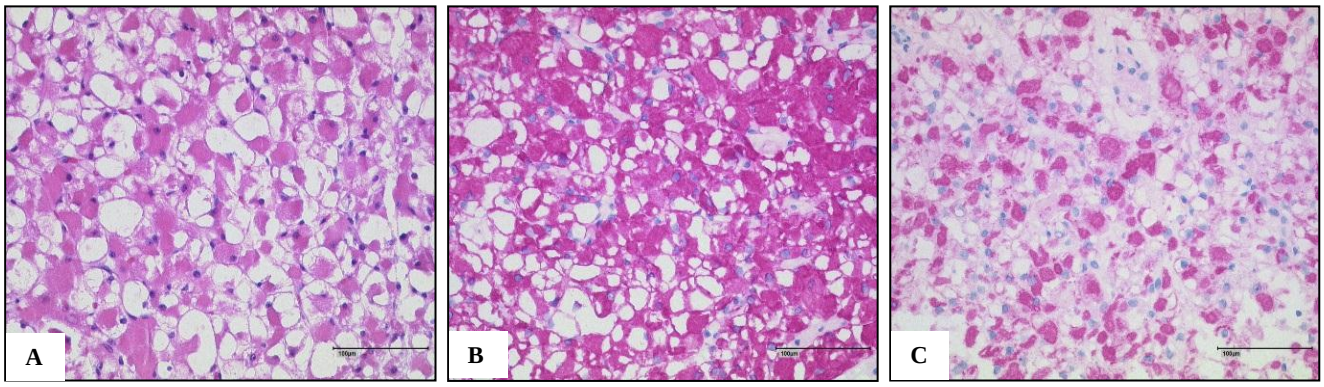

Figure 2: Histology of (A) adult rhabdomyoma showing closely packed polygonal cells with deeply eosinophilic vacuolated cytoplasm (hematoxylin and eosin). The tumor cells are positive for (B) desmin and (C) muscle specific actin (200X, scale bar 100 µm).

DISCUSSION

Rhabdomyomas in the head and neck are rare tumors and have been reported at variable locations like the lip, tongue, tongue base, soft palate, cheek, orbita, nasopharynx, larynx, the parapharyngeal and paratracheal space, hypopharynx or even multifocal.[2,4-11] Extracardiac rhabdomyoma cannot be diagnosed by clinical examination only. Histopathological examination is necessary to secure the diagnosis and to exclude differential diagnoses like granular cell tumor, hibernoma, oncocytoma, paraganglioma, histiocytosis and of course rhabdomyosarcoma.[1] Tissue for histopathological analysis can be collected by direct or needle biopsy/cytology.[4,10] Before deciding on the definitive treatment, the extent of the rhabdomyoma should be determined by proper imaging methods like sonography, MRI or CT.[6] For rhabdomyoma of the head and neck a surgical approach is most common.[2,8] Complete resection is required to prevent recurrence.[6] Remarkable tumor regression has been observed in cardiac rhabdomyomas with off label use of everolimus and sirolimus, inhibitors of the mTOR pathway, in context of tuberous sclerosis. This offers treatment options for non-operable rhabdomyomas.[12,13] Furthermore mTOR-Inhibitors like temsirolimus seem to slow down tumor growth and metastasis via antiangiogenic effects in mice bearing human rhabdomyosarcoma.[14] It is of great interest that the mTOR pathway is also activated in extracardiac rhabdomyomas.[15] In light of these findings mTOR-Inhibitors seem to be a promising option in the therapy of

extracardiac rhabdomyomas, which are critical to excise. Further trials could provide helpful insights.

CONCLUSION

Although extracardiac rhabdomyomas are extremely rare, the proper diagnosing and complete surgical treatment is crucial for the patients.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Vahl JM, Baumann L, Hoffmann TK. Rhabdomyoma of the tongue base. Int J Otorhinolaryngol Head Neck Surg 2019;5:1705-7.