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Intraoral multifocal adult rhabdomyoma: a case report

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Abstract. *Intraoral multifocal adult rhabdomyoma: a case report.* Adult rhabdomyomas are rare, benign striated-muscle neoplasms that occur in the head and neck region. They are usually solitary, but can be multifocal. We report on clinical, radiographic and morphologic features of a rhabdomyoma in the floor of the mouth and the base of the tongue in a 65-year-old male. The patient presented with a painless mass in the right submandibular region. Clinical examination revealed diffuse enlargement of both sublingual and submandibular glands. Nasolaryngoscopy showed a hypervascular lesion on the right side of the base of the tongue. Radiological investigation showed the multilobulated aspect of the lesion, and an incisional biopsy of the submandibular tumour led to the diagnosis of adult-type rhabdomyoma. Surgery comprised of extirpation of both the sublingual and submandibular glands, as well as the lesion in the base of the tongue. Histological examination of the different lesions confirmed the diagnosis of adult-type rhabdomyoma. No recurrence has occurred in a follow-up period of 6 months. We compare this case with previous reports of adult rhabdomyomas in the literature.

Introduction

Extracardiac rhabdomyoma is an extremely rare striated-muscle neoplasm.[1-4] It accounts for approximately 2% of all myogenous neoplasms.[2,5,6] These neoplasms typically occur in the head and neck region, usually as a solitary mass, but occasionally multifocal.[3,5,7,8] We report on the clinical, radiographic, and morphologic features of a rhabdomyoma present in the floor of the mouth and the base of the tongue in a 65-year-old male.

Case report

An otherwise healthy man with no history of smoking or drinking was referred to our ENT department for a painless mass in the right submandibular region, which he first noticed approximately 2 years earlier. The progressive swelling had caused some minor dysphagia and globus sensation in the last few months. Incisional biopsy of the tumour in level II of the right neck had been performed prior to referral and had revealed an adult-type rhabdomyoma.

Upon palpation of the neck, the swelling was detected in the right submandibular region. No obvious cervical lymph nodes were palpable. Oral examination revealed diffuse enlargement of the sublingual mucosa. Nasolaryngoscopy showed a hypervascular lesion with normal overlaying mucosa on the right side of the base of the tongue.

A CT scan with contrast showed three sharp delineated lesions with homogeneous enhancement: one in the right sublingual region, one in the right submandibular region, and one in the right base of the tongue (Figure 1). On MRI, the three lesions had a homogeneous density with a slightly increased intensity relative to the surrounding muscle on both T1-weighted and T2-weighted images (Figures 2a and 2b). In gadolinium-enhanced sequences, the lesions had irregular outlines and increased intensity relative to the surrounding muscle (Figure 3). Enlarged cervical lymph nodes were not seen in these images. Surgery was performed in our department. The right submandibular lesion, the right sublingual lesion, and the lesion on the base of the tongue were all extirpated (Figure 4). The three hypervascular lesions were not continuous. The left submandibular gland exhibited possible clinical involvement and was extirpated at the time of surgery. The diagnosis of adult rhabdomyoma was

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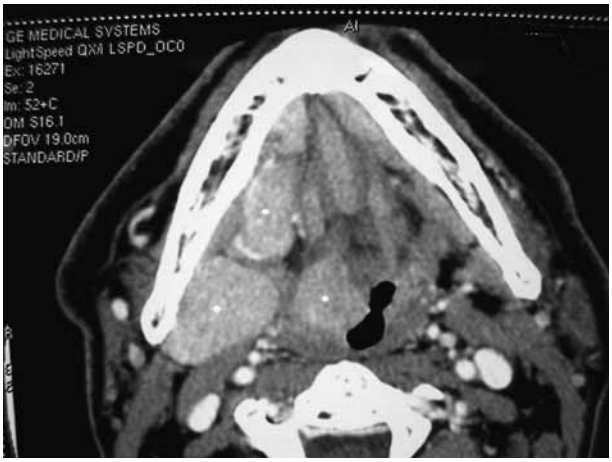

Figure 1
CT scan with administration of contrast shows three lesions that appear enhanced compared to the surrounding muscle, one in the right sublingual region, one in the right submandibular region and one in the right base of the tongue.

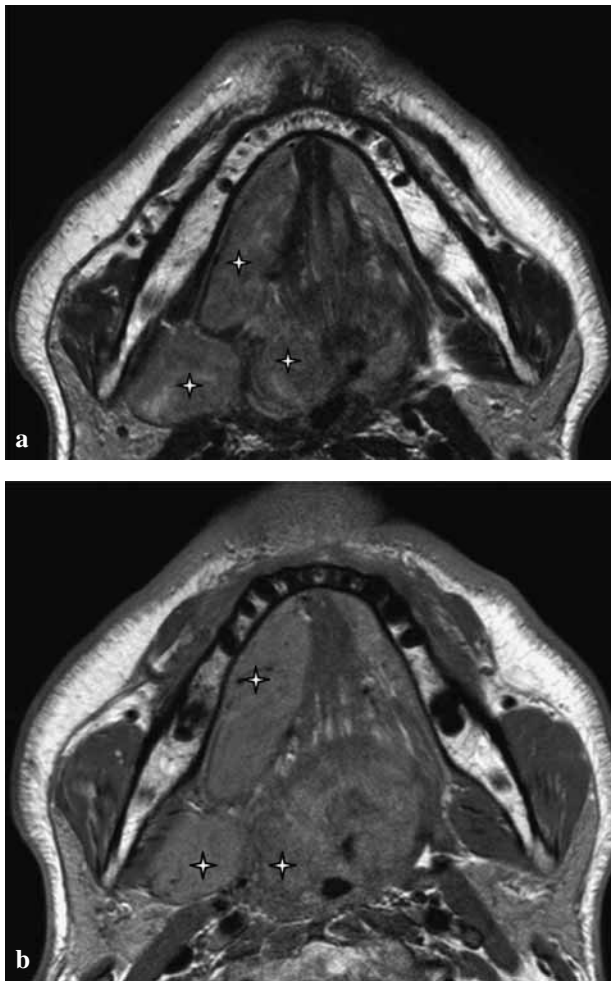

Figure 2
(a) Axial T2 - and (b) T1 - weighted MRI- images at the level of the base of the tongue. The three lesions are well delineated, and exhibit a homogeneous density with slightly heightened intensity relative to the surrounding muscle.

confirmed upon histological examination of sections from all three lesions. The tumour cells varied greatly in size and contained abundant, granular, eosinophilic cytoplasm. Multiple peripheral vacuoles were present (Figures 5a and 5b). No rhabdomyoma was ultimately found either within or adjacent to the left submandibular gland. The postoperative course was uneventful and follow-up at six months showed no clinical or radiographic recurrence.

Discussion

A rhabdomyoma is a benign tumour of skeletal or cardiac muscle origin.[1,2,4,7] The cardiac rhabdomyoma is regarded as a hamartoma and is seen most frequently in patients with tuberous sclerosis.[1,2,6] Cardiac rhabdomyomas often occur in children arising as multiple lesions in the septum and free walls of the ventricles, in association with other hamartomas in the brain, skin, subcutaneous tissue, kidney and other organs.[6]

Histopathologically, extracardiac rhabdomyoma is divided into fetal and adult types, according to the degree of cellular differentiation and maturity.[1,2,4] The adult type is the most common form of extracardiac rhabdomyoma. Adult rhabdomyomas,[7] first described by Pendl in 1897, are frequently encapsulated or well-circumscribed, firm, lobulated and homogenously tan-gray.[1,4,5,7] They occur in the head and neck region, predominantly in males[1] over the age of 40, and get their name due to the histological resemblance to mature skeletal muscle cells.[4,6] The fetal type[3,8] is considerably more rare. It occurs most commonly in the postauricular region, usually in males under the age of 4, and in the vulvovaginal region of adult women.[1,4] Histologically, fetal rhabdomyoma resembles developing skeletal muscle as seen in the fetus and is divided into myxoid and cellular variants.[3]

Although seventy-seven percent of extracardiac rhabdomyomas occur in the head and neck

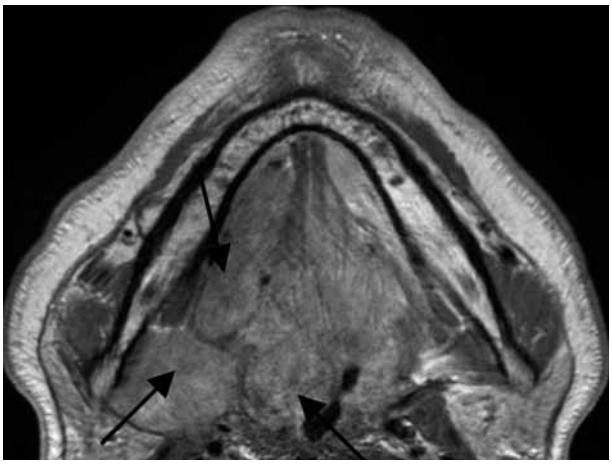

Figure 3
An axial T2-weighted image at the level of the base of the tongue with administration of contrast medium shows the same three lesions with homogeneous enhancement with heightened intensity relative to the surrounding muscle.

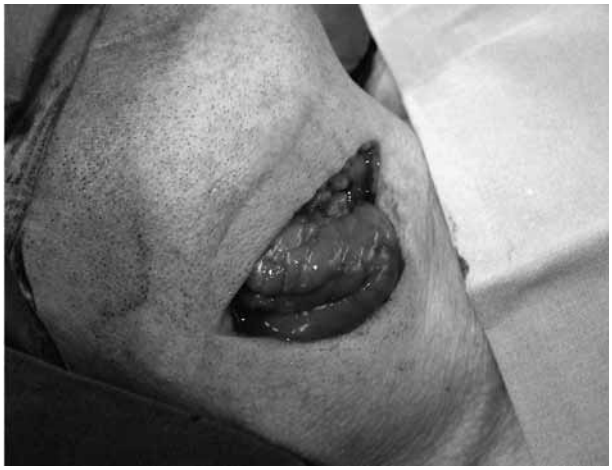

Figure 4
A preoperative image of the right submandibular lesion. The tumour was encapsulated and lobulated.

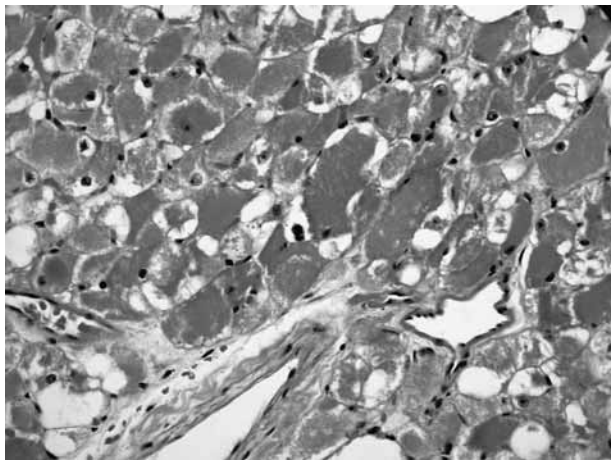

Figure 5a
The tumour cells vary greatly in size and contain abundant, granular, eosinophilic central cytoplasm. Multiple peripheral vacuoles are present.

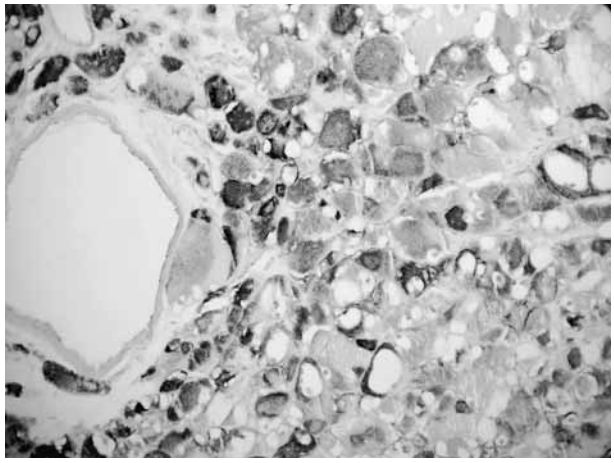

Figure 5b
Immunohistochemical studies show that tumour cells are desmin positive.

region, otolaryngologists rarely encounter them. Their signs and symptoms are usually non-specific and depend on both location and size.[7] Rhabdomyomas in the head and neck can cause a sensation of mass, airway obstruction, dysphagia, and hoarseness. The most common site of presentation is the tongue and sublingual region (33%), followed by the larynx (11%), pharynx (13%), anterior neck and sternocleidomastoid muscle (10%), postauricular area (9%), soft palate (8%) and orbit (5%). Lesions are rarely found in the cheek or lower lip.[6]

Extracardiac rhabdomyomas are usually unifocal. However, multifocality has been noted in 14-16% of cases.[3,5,7,8] Multifocality is nine times more likely to occur in men.[5] Due to the somewhat restricted anatomical distribution, it has been proposed that these legions originate in embryonic muscle groups, such as the 3rd and 4th branchial arches.[5,6]

A preoperative diagnosis of extracardiac rhabdomyoma is difficult to make based upon clinical history and examination alone; it requires specimens taken from an open biopsy, complemented by imaging techniques such as CT and MRI to determine tumour size

and extent.[1,9] In unenhanced CT, rhabdomyomas exhibit the same density as the surrounding muscle and have indistinct borders. Administration of contrast brought enhancement of various degrees. MRI showed slightly heightened intensity relative to the surrounding muscle on both T1- and T2-weighted images. Tumours also exhibited mild homogeneous enhancement and rim enhancement.[1]

The differential diagnosis of adult rhabdomyoma includes paraganglioma, granular-cell tumour, oncocytoma, acinic-cell carcinoma, rhabdomyosarcoma, hibernoma and normal skeletal muscle.[2-4,7-9]

The tumour cells vary greatly in size and contain abundant, granular, eosinophilic cytoplasm. Multiple peripheral vacuoles are present.[5] The tumour cells contain multiple, oval, vesicular nuclei either peripherally located or clustered centrally within the cytoplasm, leading to a "spider web" appearance.[1,2] Single, prominent, basophilic nucleoli are often present. Mitotic figures are not present. Intracytoplasmic inclusions and intranuclear inclusions have been described. Occasionally, cells have prominent cross-striations, which are best visualized with a PTAH stain. The cytoplasm contains abundant PAS-positive material. The background may be bloody; however, necrosis and cellular or proteinaceous debris are not observed. Immunohistochemical studies have shown myoglobulin positivity in tumour cells.[1,2,4,6]

Once the diagnosis of extracardiac rhabdomyoma has been made, a complete surgical resection of the tumour is advised.[1,2,4,5] Due to the benign nature of this tumour, complete resection should be curative.[5] The incidence of recurrence is low. When recurrence does occur, it is most likely due to incomplete resection.[1,2] Malignant transformation or locally aggressive behaviour has not been reported in these lesions.[6]

Conclusion

The only treatment for adult extracardiac rhabdomyoma is complete surgical excision. Although malignant degeneration has not been reported, a long-term follow-up is essential, as local recurrences do occur in instances of incomplete resection.

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