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# Rhabdomyoma of the base of the tongue

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**Abstract**

The histopathological and imaging findings of a rhabdomyoma of the base of the tongue were studied. An immunohistochemical examination of the tumour cells showed positive immunostaining for myoglobin, desmin, and striated muscle actin, but negative immunostaining for smooth muscle actin. Electron microscopy showed many glycogen granules and mitochondria in the tumour cells. The T2-weighted and contrast-enhanced magnetic resonance images (MRI) clearly delineated morphological features of this tumour, but T1-weighted MRI and computed tomography (CT) images showed no important features. These findings are typical for an adult extracardiac rhabdomyoma located in the head and neck region, and they will be useful for diagnosis of this tumour.

**Key words: Rhabdomyoma; Tongue; Pathology; Radiology**

## Introduction

Rhabdomyoma is a rare benign tumour of striated muscle origin and is classified as either cardiac or extracardiac. Although extracardiac rhabdomyomas are most often found in the head and neck region,[1] otolaryngologists rarely encounter them. The macroscopic features of an extracardiac rhabdomyoma are a reddish-brown colour, multiple lobules, and a soft consistency,[1] and its histopathological features are cross-striation and jackstraw-like crystalline structures.[2] However, computed tomography (CT) and magnetic resonance imaging (MRI) features and immunohistochemical features have not yet been established. In this report, we focus on the histopathological and imaging findings important for the pre-operative diagnosis of an extracardiac rhabdomyoma.

## Case report

A 51-year-old man was admitted to our hospital on May 13, 2001 with a swelling in the right submandibular region and dysphagia without associated pain. He had a 14-year history of gout and resulting chronic renal dysfunction, but no particular family history. The tumour was firm and estimated to be the size of a man's thumb. No other particular findings were noticed in the oral cavity. A flexible fibrescopic examination revealed a mass protruding on the right side from the base of the tongue to the pharyngeal space (Figure 1). No cervical lymph nodes were palpable. CT and MRI demonstrated a multilobular mass extending from the right side and base of the tongue to the pharyngeal space and right submandibular region, with a maximum diameter of 60 mm (Figure 2 and 3). On CT, this mass showed a homogeneous density equal to that of the surrounding muscles (Figure 2(a)) and was slightly enhanced with iodine contrast media (Figure 2(b)) without bone destruction. On MRI, it showed a homogeneous density nearly equal to that of the surrounding muscles on the T1-weighted images (Figure 3(a)) and slightly greater than that of the surrounding muscles on the T2-weighted images (Figure 3(b)). On contrast-enhanced images, its irregular outline as well enhanced with gadolinium (Figure 3(c)). The multilobular shape of the tumour was delineated well, especially on the T2-weighted and contrast-enhanced MR images. Cervical lymph node swelling was not evident on these images. These findings suggested that the tumour was not malignant. The pathological findings by fine needle aspiration biopsy showed few mitotic cells. However,

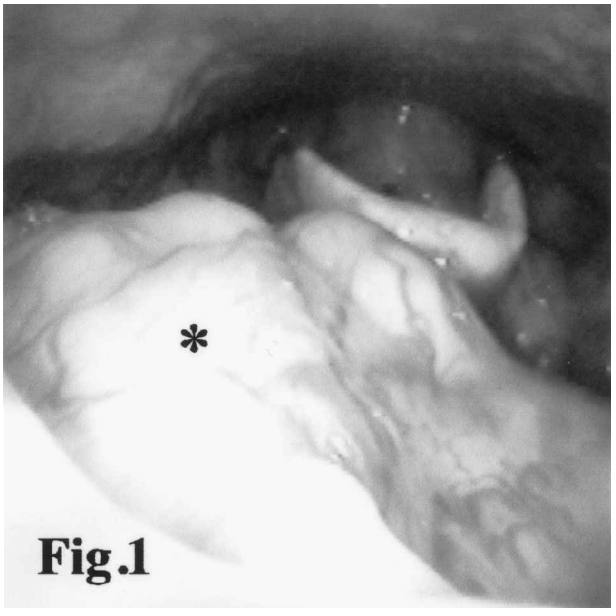


Fig. 1

Laryngeal fibrescopic examination before treatment. A mass protruding on the right side from the base of the tongue to the anterior oropharyngeal wall was noted (*).

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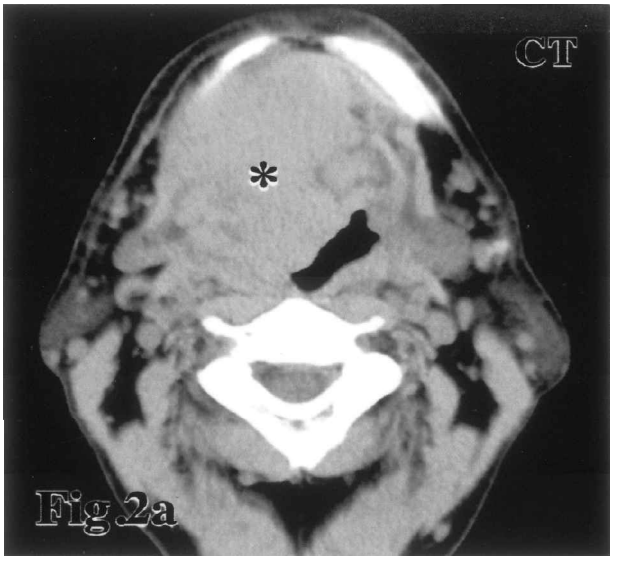
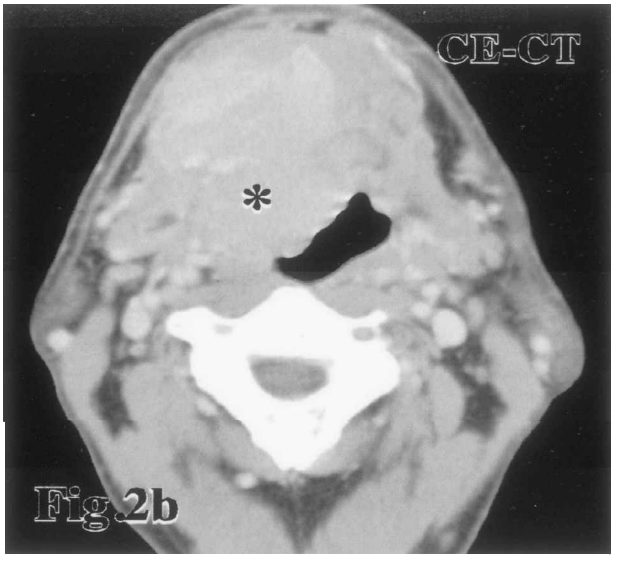


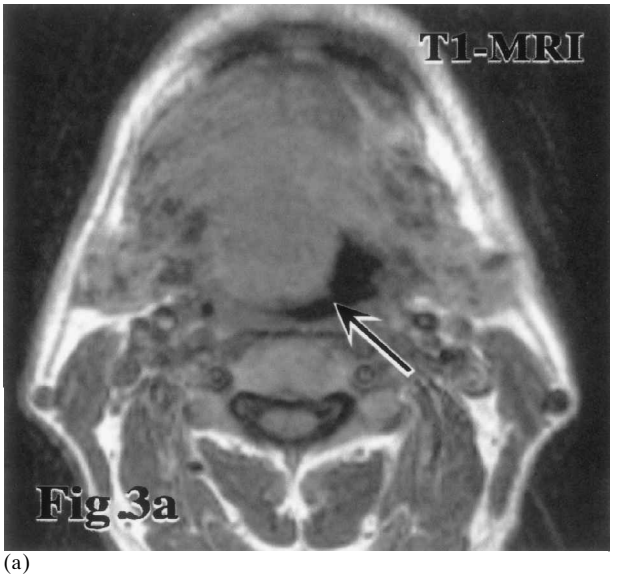

(a)



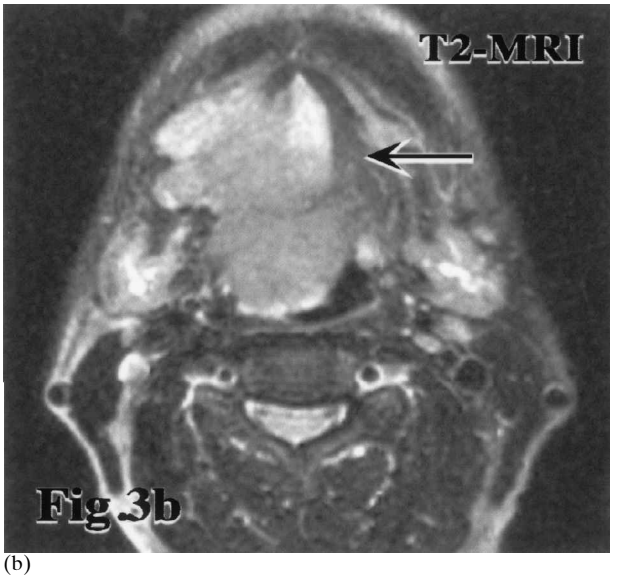

(b)

FIG. 2

Axial computed tomography (CT) images at the level of the base of the tongue. (a) Unenhanced CT shows a mass of uniform density relative to the surrounding muscles. It protrudes from the right side of the base of the tongue to the oropharyngeal space. The border of the mass is indistinct. (b) On contrast-enhanced CT, the mass is very slightly enhanced. Its multilobular shape and border are better delineated.




(a)

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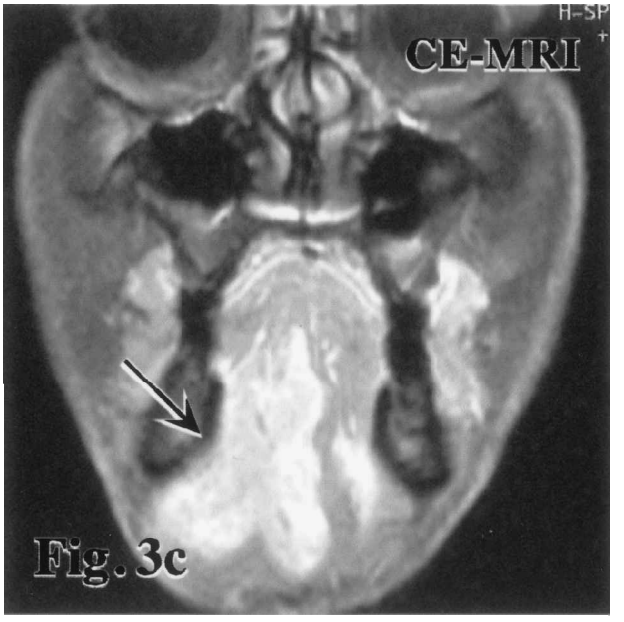


(c)

FIG. 3

Magnetic resonance images. (a) An axial T1-weighted image (TR/TE, 500/14) at the level of the base of the tongue shows a mass of homogeneous density nearly the same as that of the surrounding muscles. The border of the mass is indistinct. (b) An axial T2-weighted image (TR/TE, 4487/103) at the level of the base of the tongue shows a mass of greater intensity than that of the surrounding muscles. Its multilobular shape and border are well delineated (arrow). (c) A coronal enhanced T1-weighted image with fat saturation (TR/TE, 856/12) shows a well-enhanced mass extending from the right side of the tongue to the right submandibular region. Its multilobular shape and border are well delineated (arrow).

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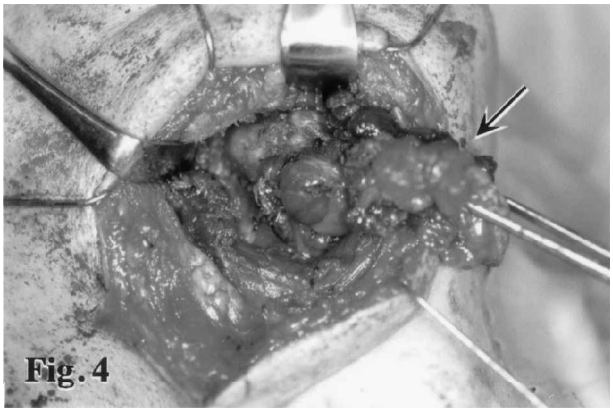


FIG. 4

The operative findings. The tumour (arrows) was reddish-brown, multilobular, soft, and fragile. The tumour penetrated the entire depth of the tongue without definite boundaries. Because the tumour broke apart and bled easily, its total dissection was not possible.

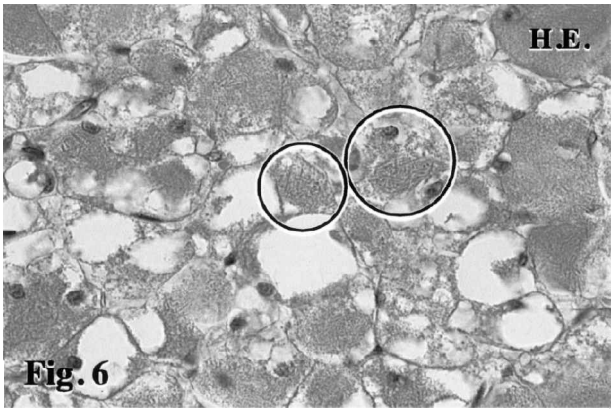


FIG. 6

Light microscopy section (H & E; ×80). The tumour is tightly composed of medium to large, round or polygonal cells. They are eosinophilic and some contain jackstraw-like crystalline materials (in circles).

dyspnoea was a concern because the tumour was huge and protruding into the pharyngeal space, so the tumour was removed.

On May 14, 2001, with the patient under general anaesthesia, a tracheotomy was performed to remove the risk of post-operative dyspnoea, then the tumour was dissected via the right submandibular region. The tumour was reddish-brown, multilobular, soft, and fragile (Figure 4). The tumour penetrated the entire depth of the tongue without definite boundaries. Because the tumour broke apart and bled easily, its total dissection was not possible. However, it was possible to reduce the tumour volume sufficiently. The extirpated tumour was soft, without a tegmen, and multilobular with a size of 70 × 35 × 25 mm (Figure 5).

Histopathological examination revealed that it was not encapsulated and was tightly composed of medium to large, round or polygonal cells. They were eosinophilic and surrounded by thin fibrous septa and capillaries. Some striated muscle tissue was also noticed in the tumour. The tumour cells had granulated cytoplasm and some contained thin eosinophilic (jackstraw-like crystalline) materials, which are thought to represent Z-band materials.[3] Neither atypical nuclei nor mitosis was observed (Figure 6). Staining with periodic acid-Schiff stain showed many glycogen granules in the cells. Electron microscopy also revealed many glycogen granules and mitochondria in the cells. Strands of myofilaments with Z-bands were also seen (Figure 7).

Immunohistochemical examination showed tumour cells with positive immunostaining for myoglobin, desmin and striated muscle actin, but negative immunostaining for smooth muscle actin (Figure 8) and S100. These findings indicated that this tumour was an adult extracardiac rhabdomyoma. After the surgery, the swelling in the right submandibular region and the dysphagia disappeared. Tumour regrowth was not apparent 12 months after the surgery (Figures 9 and 10).

## Discussion

Rhabdomyoma is a benign tumour of skeletal or cardiac muscle origin, accounting for only two per cent of all striated tumours.[3] Cardiac rhabdomyoma, the more common type, is associated with tuberous sclerosis.[3] The rarer extracardiac rhabdomyoma is primarily located in the head and neck region. Histopathologically, extracardiac rhabdomyoma is divided into fetal and adult types

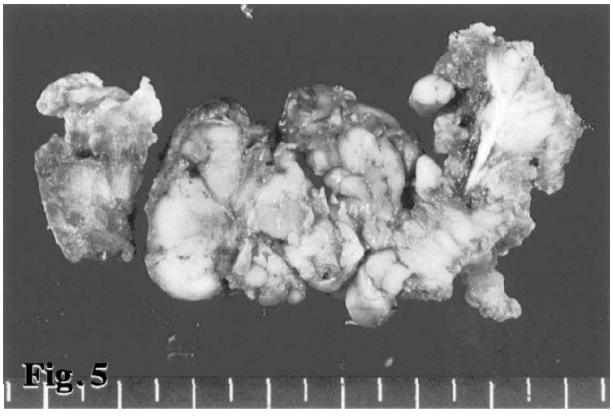


FIG. 5

The resected tumour. The tumour was soft, without a tegmen, and multilobular with a size of 70 × 35 × 25 mm.

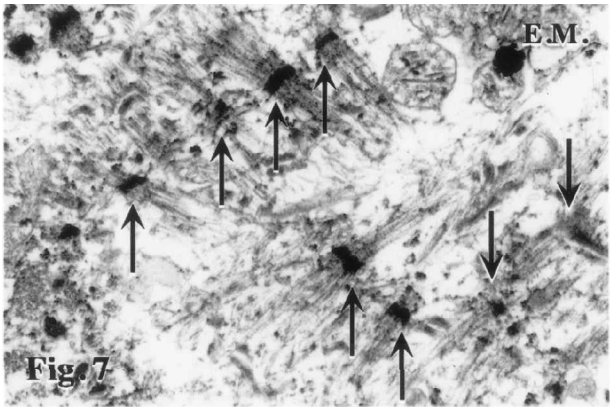


FIG. 7

Electron microscopic examination (×15 000). Strands of myofilaments with Z-bands (arrows) were noted.

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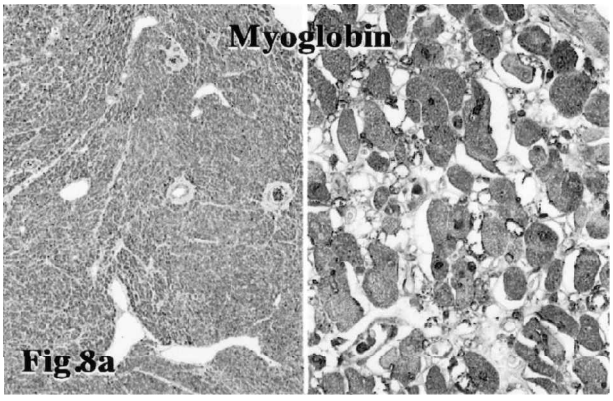
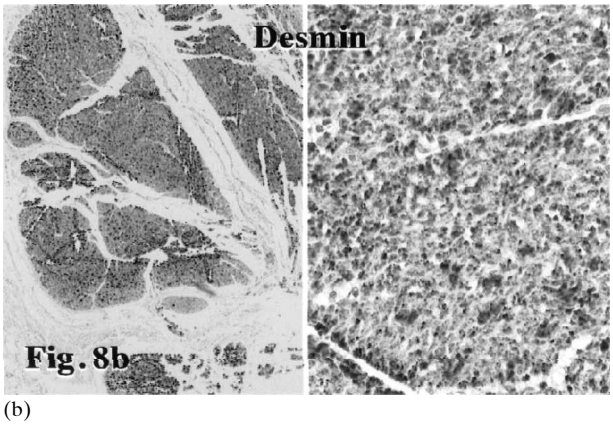



(a)

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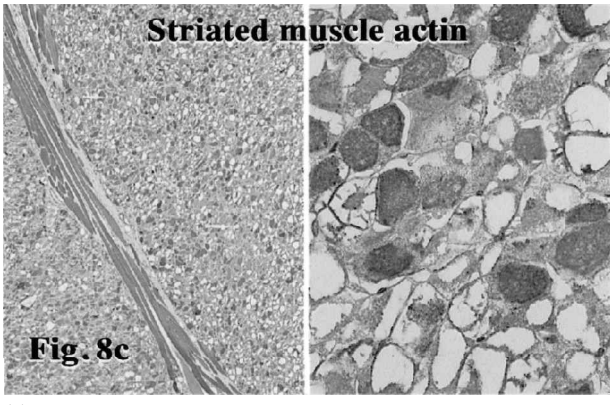


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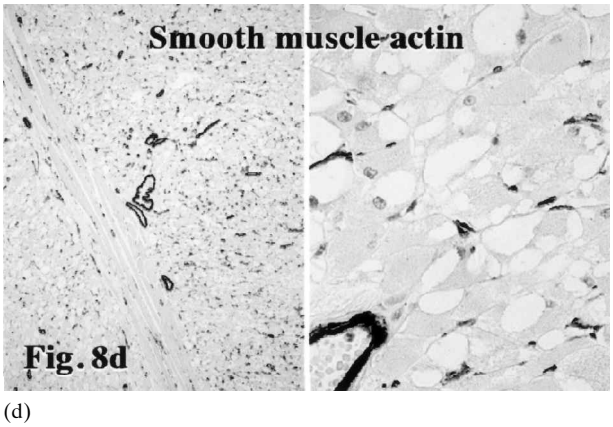


(d)

FIG. 8

Immunohistochemical examination. Left side shows the lower magnification (×4) and right side shows the higher magnification (×30). The tumour cells stained positive for myoglobin (a), desmin (b) and striated muscle actin (c), but not for smooth muscle actin (d).

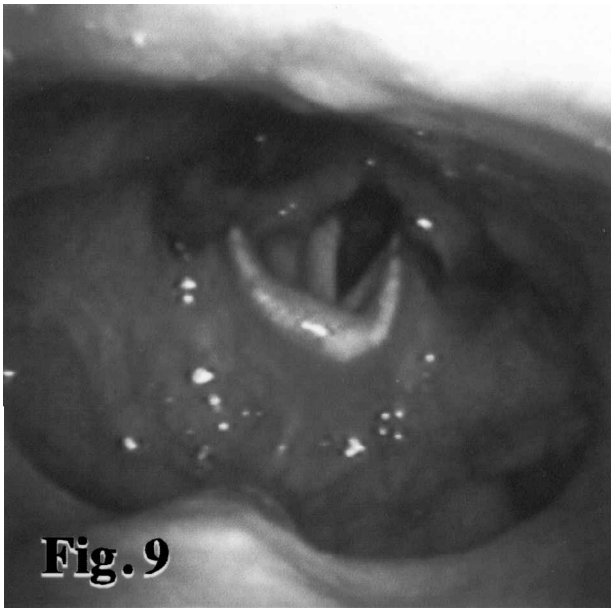


FIG. 9

Laryngeal fibrescopic examination after surgery. The mass previously protruding from the base of the tongue wall has been removed.

according to the degree of cellular differentiation and maturity.[1] The fetal type occurs most commonly in the post-auricular region of children and in the vulvovaginal region of adult women. Fetal extracardiac rhabdomyoma is composed of immature spindle cells that show positive immunostaining with primitive and mature muscle markers.[13] The adult type occurs most commonly in the head and neck region, particularly in the pharynx, the floor of mouth, and the base of the tongue, of adults, with a 3:1 male predominance.[4] This type of tumour may cause a sensation of mass, airway obstruction, dysphagia, and

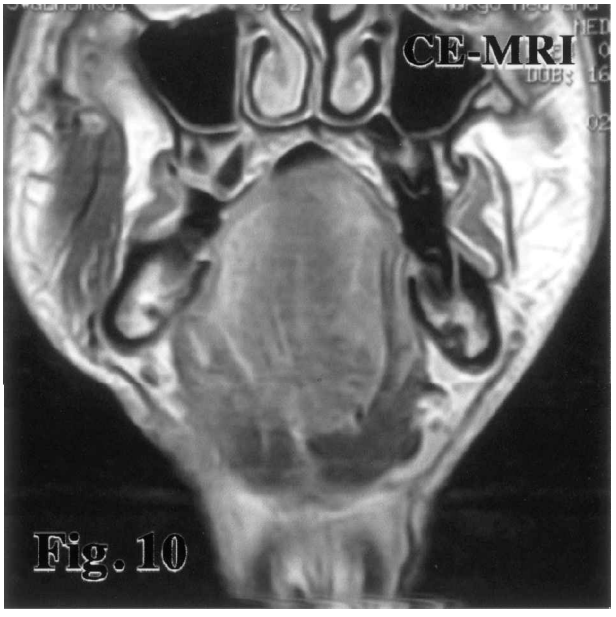


FIG. 10

Coronal enhanced MRI taken 12 months after the surgery. No recurrence of the tumour is seen.

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hoarseness[4–6] such as seen in this case, although its growth is so slow that it may grow very large before it becomes symptomatic.[4]

Histopathologically, rhabdomyoma is composed of large round cells with eosinophilic and glycogen-rich cytoplasm. Cross-striations and jackstraw-like crystalline structures are especially typically of this tumour and helpful in diagnosis.[2–4,7] However, cross-striations and jackstraw-like crystalline structures are obscure in light microscopy, thus electron microscopy and immunohistochemical findings are important for diagnosis. In electron microscopy, the presence of strands of myofilament with Z-bands is a feature of this tumour.[2] In immunohistochemistry positive staining for myoglobin, desmin, and striated muscle actin, which are markers for mature muscle cells, is characteristic of this tumour.[2–4] These two characteristics indicate a histopathological diagnosis of an adult extracardiac rhabdomyoma.

A pre-operative diagnosis is difficult to make: these examinations require specimens taken from an open biopsy rather than from fine needle aspiration. In general, CT and MRI are useful modalities for determining tumour size, extent, and nature. However, features revealed by radiological imaging have been reported in only a few cases of adult rhabdomyoma,[2,6–11] and the diagnostic usefulness of imaging in cases of these tumours has not been fully established. Rhabdomyoma has been reported to show the same density as surrounding muscle and to have indistinct borders when studied by unenhanced CT[10] and administration of contrast media brought enhancement of various degrees.[2,9,11] On the other hand, MRI showed a mass with slightly heightened intensity relative to the surrounding muscle on T1- and T2-weighted images.[6–9] With administration of contrast media, the tumour showed mild homogeneous enhancement[6,8,9] and rim enhancement.[9] On MRI, the tumour showed nearly the same density as surrounding muscle on T1-weighted images and heightened intensity on T2-weighted images. Enhancement was homogeneous and strong, but no rim enhancement was seen. The multilobular shape and the border of the mass were delineated well on T2-weighted images and contrast-enhanced MR images. While T1-weighted MR images and CT images revealed no important features of this tumour, T2-weighted and contrast-enhanced MR images are especially useful for determining the size, extent, and nature of the tumour, including its multilobular feature. These findings will facilitate a pre-operative diagnosis of this tumour type.

The only treatment for adult extracardiac rhabdomyoma is surgical excision. It was not possible however, to dissect this tumour totally. Although malignant degeneration has not been reported, the local recurrence rate is 42 per cent, mostly from incomplete resection,[4] indicating that long-term follow-up is important.

- **Rhabdomyomas are rare and are classified as either cardiac or extra-cardiac with the majority of the latter cases occurring in the head and neck region**
- **This paper presents the histopathological and radiological findings of a tongue base rhabdomyoma**
- **It is anticipated that these findings will help in the pre-operative diagnosis of such lesions in the future**

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