

Rhabdomyosarcoma of the tongue base, its recurrence, and multiple lymph node metastases with imaging evidence

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ABSTRACT

Rhabdomyosarcoma (RMS) is an aggressive and fast-growing malignant tumor. RMS predominantly arises in the head and neck of infancy and children. Metastasis is usually via the blood vessel. We report a case of a recurred RMS of the tongue base with the metastasis to multiple lymph nodes in a 37-year-old female. On the follow-up examination using advanced imaging modalities after surgical treatment of RMS, the lymph nodes should be carefully evaluated like in other malignancies, such as a carcinoma, showing frequent lymph node metastasis. (*Korean J Oral Maxillofac Radiol* 2008; 38 : 225-8)

KEY WORDS : Rhabdomyosarcoma; Lymph node; Metastasis; Recurrence

Rhabdomyosarcoma (RMS) is an aggressive and fast-growing malignant tumor dominantly arising in the region of head and neck in children.¹⁻³ Less frequently, adolescents and adults may be affected.⁴

Adult RMS constitutes a very small fraction of soft tissue sarcomas, estimated between 2% and 5%.⁵ RMS potentially may arise in any anatomic location.⁴ However, the most common site is the head and neck, followed by the genitourinary tract/retro-peritoneum and extremities.⁴ A third of rhabdomyosarcomas occur in the head and neck.⁶ Despite this predilection for the head and neck, RMS arising within the oral cavity is rare.⁷⁻¹⁰ The most common sites involved are the tongue and palate.⁷⁻¹²

During the last 30 years, improvements in multimodal therapy have resulted in a cure rate of approximately 70% for patients with localized disease.¹³⁻¹⁵ Approximately 30% of patients experienced a recurrence, and 5-year survival rate after first recurrence remained poor at less than 20%.¹⁶

Metastasis is usually via the bloodstream and less commonly via the lymphatics.¹⁷ In the series of Quaglia et al.¹⁸ which included patients with distant metastatic disease, the rate of nodal disease was 28%.

Here we present a case of oral rhabdomyosarcoma of the

tongue base, its recurrence, and multiple lymph node metastases in a 37-year-old female.

Case Report

A 37-year-old woman was referred to Seoul National Dental Hospital in March 2007, complaining of muffled voice and dysphagia caused by a large mass of the tongue base.

On MRI, a large volume of mass in the tongue base was observed obstructing the airway (Fig. 1). The mass had an ill-defined margin and showed a heterogeneously high signal intensity on T2-weighted image. The lesion was infiltrating to the lateral oropharyngeal wall. The airway was almost obstructed by the large mass. Enhanced CT scans also showed a large heterogeneously well-enhancing mass in the tongue base (Fig. 2). Distant metastasis or lymph node metastasis was not observed on PET-CT.

The impression of this lesion was a malignant lesion with mixed high vascularity and cellularity in the tongue base without the evidence of necrosis or hemorrhage.

A biopsy under general anesthesia was implemented and it showed round to oval cells with hyperchromatic nuclei and scant cytoplasm aggregated, but they showed loss of cellular cohesion, forming "alveolar" spaces (Fig. 3). These tumor nests were separated by fibrovascular septa. Mass excision was done and only one adjacent lymph node was removed during the operation. The lymph node was negative.

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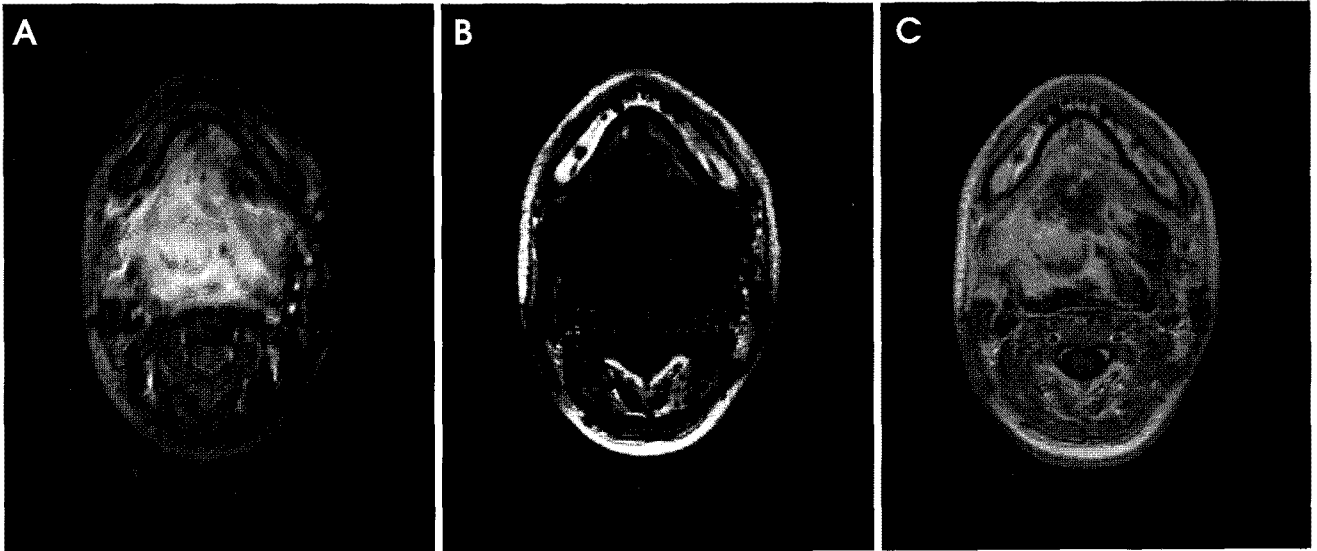


Fig. 1. Preoperative MRI. (A) T2-weighted image shows a large volume of mass of the tongue base obstructing the airway. The mass has a heterogeneously high signal intensity and an ill-defined margin. (B) T1-weighted image shows a homogeneously low signal intensity lesion. (C) Enhanced T1-weighted image shows a heterogeneously well-enhancing lesion.



Fig. 2. Preoperative enhanced CT image. A large heterogeneously well-enhancing mass can be seen in the tongue base. The lesion is infiltrating into the lateral oropharyngeal wall. The airway is almost obstructed by the large mass.

Two months after the first operation, several nodal masses of the right neck were noted on the follow-up examination and contrast enhanced CT scans revealed the recurrence of rhabdomyosarcoma with multiple lymph node metastases (Fig. 4). A heterogeneously enhancing soft tissue lesion, indicating a recurred mass, was observed at the pharyngeal area adjacent to the lower part of the posterior margin of the operation site,

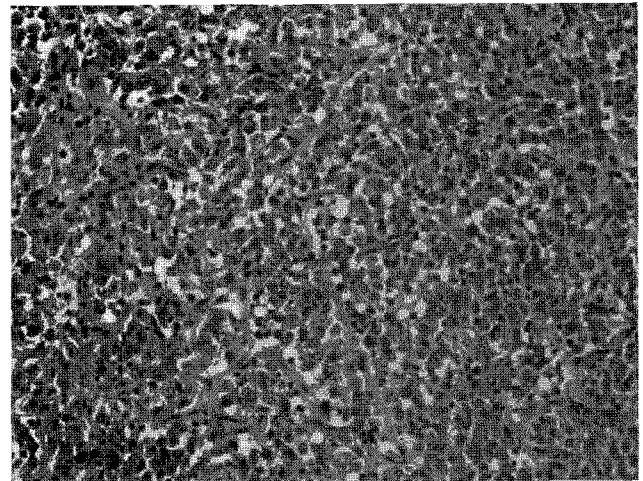


Fig. 3. Hematoxylin-eosin stain (original magnification $\times 200$). Round to oval cells with hyperchromatic nuclei and scant cytoplasm are aggregated, but they show loss of cellular cohesion, forming "alveolar" spaces. These tumor nests are separated by fibrovascular septa.

and it extended inferiorly along the esophagus. A recurrent lesion was also noted infiltrating the medial pterygoid muscle adjacent to the right pharyngeal wall. In particular, the lingual cortex of the right mandibular ramus was destroyed and the invasion into the marrow space was observed. Multiple lymph node metastases were seen on the levels I, II, and III of the right neck, and on the lateral retropharyngeal lymph node. Thereafter, although anti-cancer therapy was carried out, bleed-

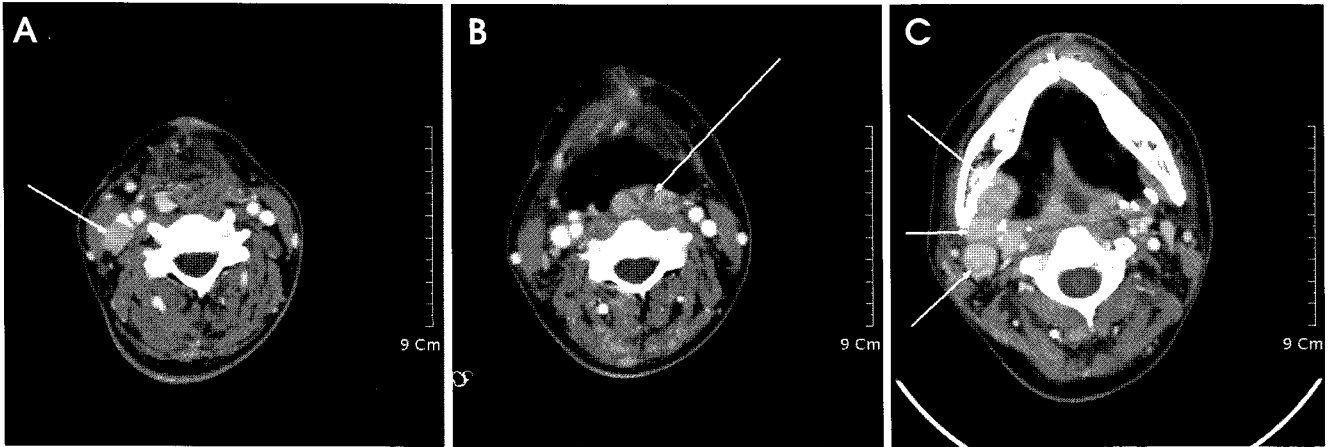


Fig. 4. Enhanced CT images, two months after operation. (A) Lymph node enlargement is seen on the level III of the right neck. (B) An enhancing soft tissue lesion, indicating a recurrent mass, is noted at the pharyngeal wall adjacent to lower part of the posterior margin of operation site. It is extended inferiorly along the esophagus. (C) Multiple enlarged lymph nodes are seen on the level I and II of the right neck.

ing had repeatedly occurred and the mass was continuously growing, so the chemotherapy was stopped and only supportive care and pain control were sustained. Finally, the patient was hopelessly discharged in January 2008.

Discussion

Rhabdomyosarcomas are thought to arise as a consequence of regulatory disruption of skeletal muscle progenitor cell growth and differentiation.¹⁹ It is now appreciated that the delicate balance between proliferation and differentiation, mutually exclusive yet intimately associated processes, is normally controlled in large part through the action of a multitude of growth factors.¹⁹

This tumor has been subdivided into three morphological types: pleomorphic, alveolar, and embryonal, the latter including a sub-type known as botryoid RMS.^{5,24} Among these RMSs, adult alveolar RMS is a highly malignant neoplasm with a significant incidence of metastatic recurrence and poorly documented due to its rarity.²⁰ Two thirds of head and neck rhabdomyosarcomas are of the embryonal type, and slightly fewer than a third consist of the alveolar type which has a poorer prognosis.^{6,21} Therapeutic success for adult RMS has been limited and long-term survival rates remain poor.^{5,22} Even in the International Rhabdomyosarcoma Study Group (IRSG) studies, an adverse effect of increasing age on outcome has been documented.²⁰ In IRS-IV, age was prognostically significant and, in particular, adolescent boys 10 years of age or older fared significantly worse than boys younger than 10 years.²³

Other investigators have also reported that increasing age is an adverse prognostic factor.²⁴ Our patient was 37-year-old and it might be related to the poor prognosis of the present case.

Apart from its poorer response to treatments, RMS in adults also appears to differ from children RMS in several other features.²⁰ Childhood RMS occurs with predilection in the head and neck and the genitourinary regions.²² In contrast, adult RMS arises predominantly in the extremities.²⁵ Although our patient was an adult, RMS occurred in tongue base. Another significant difference between childhood and adult RMS is the proportion of histopathologic subtypes.²⁰ Although the recognition and subclassification of RMS remains difficult with significant disagreement even among expert pathologists.²⁰ The relative paucity of embryonal tumors and the relative increase in alveolar and pleomorphic types are characteristic of all adult RMS series.^{5,22,24} Our case was an alveolar type.

By the time rhabdomyosarcoma is noticed, the patients in most cases already have large mass of tumors for the reason of fast-growing tendency of RMS. Therefore, early detection and diagnosis are important.¹ When a recurrence was detected in our patient two months after the first operation, multiple lymph node metastases were apparently seen on the level I and II of the right neck and lateral retropharyngeal lymph node. Hawkins et al.⁵ described twenty percent of the patients in the study group presented with recurrent disease, and 62% presented with regional or distant metastases. However, RMS metastasis in the region of head and neck is rare and to our knowledge, metastasis to lymph nodes is not common.

The most commonly reported MR imaging appearance of rhabdomyosarcoma in the head and neck is that of a homogeneous mass, isointense or minimally hyperintense relative to muscle on T1-weighted images and hyperintense relative to both muscle and fat on T2-weighted images, with postcontrast images showing enhancement of the tumor.²⁷ On MRI of our patient, a large volume of mass in the tongue base showed a mixed heterogeneous and homogeneous high signal intensity on T2-weighted image. Unfortunately, other relatively common tumors of the head and neck, such as nasopharyngeal carcinoma and lymphoma, may have a similar signal intensity characteristics to that of rhabdomyosarcomas.²⁸⁻³⁰

Generally, lymph node metastasis of RMS in infant and children is not common. However, nodal metastasis of rhabdomyosarcoma in an adult is more frequently found. Therefore, the possibility of lymph node metastasis should be considered and advanced imaging modality covering the neck nodes should be taken and thoroughly evaluated.

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