

Case Report

A primary extramedullary plasmacytoma of the tongue base evaluated by diffusion-weighted imaging: a case report and literature review

Guo-Qing Wei¹, Lan-Ying He², Shui-Hong Zhou², Bai-Shu Zhong³, Li-Fang Shen², Yong-Cai Liu², Yang-Yang Bao², Jiang-Tao Zhong², He-Ming Han²

Departments of ¹Hematology, ²Otolaryngology, ³Radiology, The First Affiliated Hospital, College of Medicine, Zhejiang University, Zhejiang Province, China

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Abstract: Primary extramedullary plasmacytoma (EMP) of the tongue base is rare. Diffusion-weighted imaging (DWI) and the apparent diffusion coefficients (ADCs) obtained by magnetic resonance imaging (MRI) may be used to differentiate benign from malignant lesions. No ADCs for an EMP of the tongue base have yet been reported. We report a 67-year-old male who presented with a 1-month history of a foreign body sensation in the pharynx. Physical examination revealed a round mass on the right base of the tongue. MRI showed that the mass was 0.8 cm in diameter. The mass was isointense on T1-weighted imaging and yielded a mixed signal on T2-weighted imaging. The mass was hyperintense on DWI and had an ADC ($b=1000 \text{ s/mm}^2$) of $1.23 \pm 0.316 \times 10^{-3} \text{ mm}^2/\text{s}$. The mass was strongly enhanced on contrast-enhanced T1-weighted MR images. The mass was excised via suspension laryngoscopy performed under general anesthesia. The immunohistochemical profile of the tumor (positive for CD138, CD79a, IgG, and kappa light chain) and a systemic examination that excluded multiple myeloma both indicated a diagnosis of EMP. A total of 18 EMPs of the tongue base (including our case) have been reported to date in the English-language literature, of which 7 underwent MRI examination. Only one report mentioned the features of T1- and T2-weighted imaging: hypointensity was evident on T1-weighted images and slight hyperintensity on T2-weighted images. DWI-MRI may help to differentiate malignant from benign lesions in the tongue base. Radiotherapy plays an important role in treatment.

Keywords: Extramedullary plasmacytoma, diffusion-weighted imaging, apparent diffusion coefficient, magnetic resonance imaging, radiotherapy

Introduction

Plasmacytomas are rare tumors characterized by monoclonal proliferation of plasma cells in the absence of multiple myeloma (MM). These tumors are classified as either medullary (i.e., developing within the medullae of long bones) or extramedullary [1]. Primary extramedullary plasmacytoma (EMP) is an uncommon malignant tumor accounting for approximately 5-10% of all plasma cell malignancies. Approximately 80-90% of EMPs occur in the head-and-neck region, principally in the paranasal sinuses, nasal cavity, oral cavity, and pharynx [2]. In general, the prognosis of head-and-neck EMP is relatively good [3]. Previously, we reviewed the available literature on laryngeal EMP and found

that prognosis becomes poor once the EMP has progressed to MM [2]. Thus, it is important to diagnose EMP early to ensure good outcomes.

Primary EMP of the tongue base is rare [1, 4]. A primary solitary EMP of the tongue base is an ovoid pedunculated mass that is always accompanied by submucosal or local swelling. Such a presentation may be readily confused with those of other diseases, including benign reactive lymphoid tissue proliferation, benign vascular disease, or lymphoma [1, 4]. Magnetic resonance imaging (MRI) has been used to differentiate these diseases [5]. Thus, MRI is valuable when diagnostic work-up of a mass at the base of the tongue is required [4].

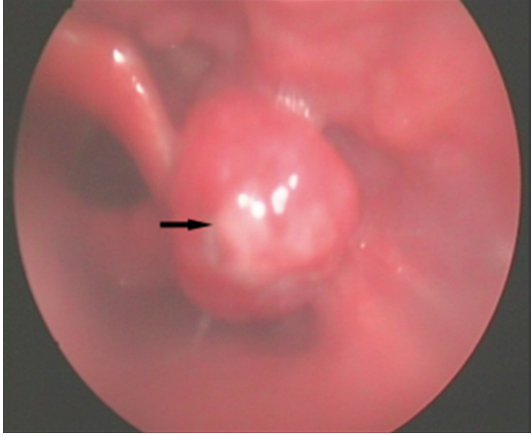


Figure 1. Laryngostroboscopy reveals a round mass at the right base of the tongue.

Diffusion-weighted imaging (DWI) and the apparent diffusion coefficients (ADCs) obtained by MRI can be used to differentiate benign from malignant lesions [4, 6]. Ramachandran et al. found that one EMP yielded a high DWI signal and a low ADC [6].

In the present study, we present an additional case of a primary solitary EMP of the tongue base and report the MRI and DWI findings and the ADCs. We also reviewed the English-language literature regarding primary solitary EMPs of the tongue base.

Case presentation

A 67-year-old male presented with a foreign body sensation in the pharynx for the past 1 month. He reported no dysphagia, stuffiness, shortage of breath, cough, sore throat, chills, or fever. His past medical history and his family medical history were generally unremarkable. However, he had undergone a right-bundle branch block and previously had a gastric ulcer, and his father had died of gastric carcinoma. He drank 750 mL beer/day for over 40 years but did not smoke. Physical examination revealed a round mass at the right base of the tongue (**Figure 1**). There was no cervical lymphadenopathy.

MRI of the head-and-neck region revealed a mass 0.8 cm in diameter at the right base of the tongue. The mass was isointense on the T1-weighted image and yielded a mixed signal on the T2-weighted image. The mass was hyperintense on DWI with an ADC ($b=1000$ s/

mm^2) of $1.23 \pm 0.316 \times 10^{-3} \text{ mm}^2/\text{s}$. On contrast-enhanced T1-weighted MR images, the mass was strongly enhanced (**Figure 2**). The structures of the larynx and adjacent tissues were normal. Neither the local nor regional lymph nodes were remarkable. The radiologist suspected a malignant lesion.

On May 20, 2016, the mass was excised via suspension laryngoscopy under general anesthesia. During the operation, a 1.0×0.8 cm mass was found at the right base of the tongue. A needle puncture yielded no obvious blood. A frozen section showed that the mass contained predominantly diffuse plasma cells. The post-operative histopathological results revealed diffuse infiltration of atypical plasma cells.

Immunohistochemically, the tumor was positive for CD138, CD79a, cyclin D1, melanoma-associated antigen (mutated) 1, Myc, kappa light chain, and Ki-67 (>70% of cells) (**Figure 3**) and was negative for CD30 (Ki-1), CD5, CD10, Bcl-2, Bcl-6, ALK, PAX-5, lambda light chain, CD3, and CD20.

We performed other laboratory tests to screen for systemic myeloma. These included a skeletal survey, serum and urine protein electrophoresis, and measurements of the erythrocyte sedimentation rate and the levels of β_2 -microglobulin, immunoglobulins, and urinary Bence-Jones proteins. All results were normal. Serum calcium and uric acid levels and renal function tests were normal. Bone marrow biopsy revealed a normocellular marrow of normal architecture; all hematopoietic elements were present at their normal ratios. Computed tomography of the chest, abdomen, and pelvis revealed no evidence of metastatic disease. The urine did not contain light chains. Thus, we diagnosed a primary solitary EMP of the tongue base.

The patient underwent radiotherapy (a total dose of 36 Gy in 18 daily fractions of 2 Gy each, 5 days a week for a total of 4 weeks). At the 4-month follow-up, he showed no sign of either systemic myeloma or local recurrence.

The institutional review board of the First Affiliated Hospital, College of Medicine, Zhejiang University (Hangzhou City, China) approved the present study (approval no.

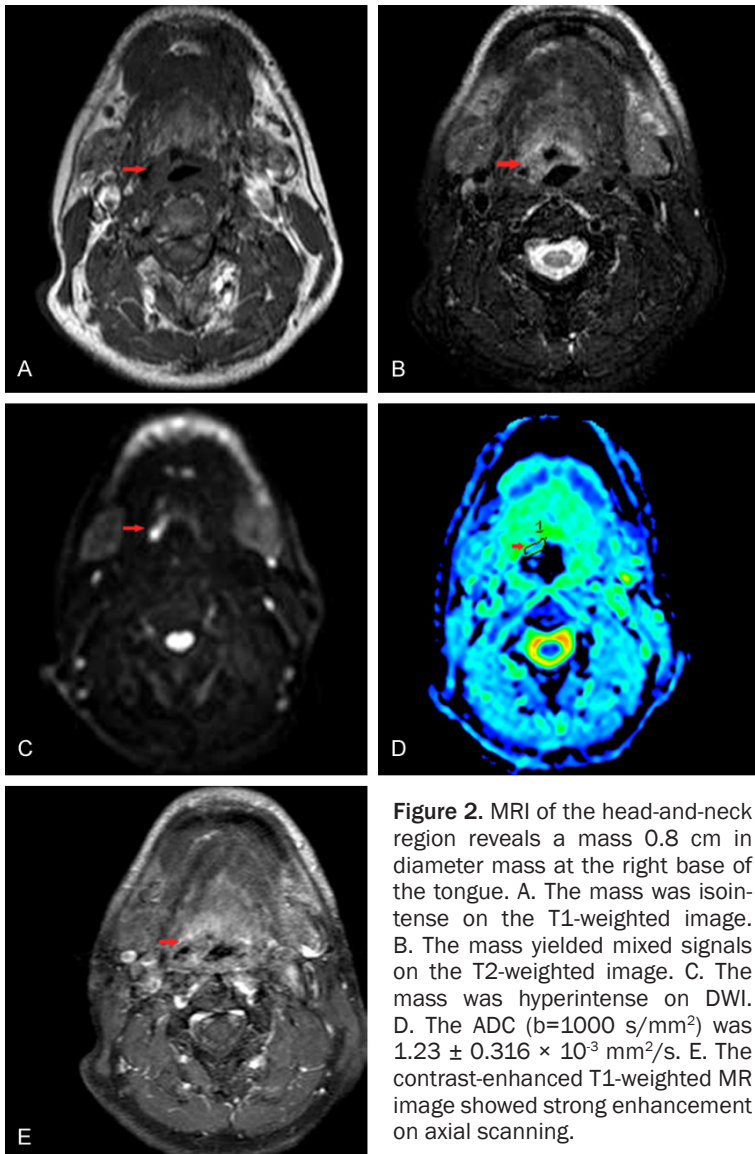


Figure 2. MRI of the head-and-neck region reveals a mass 0.8 cm in diameter mass at the right base of the tongue. A. The mass was isointense on the T1-weighted image. B. The mass yielded mixed signals on the T2-weighted image. C. The mass was hyperintense on DWI. D. The ADC ($b=1000 \text{ s/mm}^2$) was $1.23 \pm 0.316 \times 10^{-3} \text{ mm}^2/\text{s}$. E. The contrast-enhanced T1-weighted MR image showed strong enhancement on axial scanning.

the tongue base (Table 1) [1, 4, 7-20]. Eleven males and five females were affected (information on sex was available for only 16 cases). The male-to-female ratio was 2.2:1. The patients ranged in age from 30 to 87 years at the time of initial presentation, with a mean age of 63.9 years (age data were available for 15 patients). Of 11 cases for whom detailed symptoms were recorded, 4 (30.8%) presented with soreness of the tongue, 2 (15.4%) with swelling, 2 (15.4%) with enlarged cervical lymph nodes, 1 (7.7%) with burning mouth and dysphagia, 1 (7.7%) with recurrent epistaxis, and 1 (7.7%) with a foreign body sensation in the pharynx. Follow-up data were available for 13 cases. Ten patients were alive with no evidence of disease at the times of publication. Three patients had died from their disease.

Factors prognostic of EMP of the tongue base are unknown, considering the rarity of the condition. The prognosis of early-stage EMP is relative good, especially if the tumor is in the head-and-neck region [3]. Some studies found that light chain restriction may be

associated with prognosis. We found, in our recent review, that restriction of either lambda or kappa light chains afforded a relatively good prognosis. Of all cases reported, 50% had kappa light chain restrictions and 50% lambda light chain restrictions (detailed data were available for eight cases, none of whom progressed to MM). In our review (cited above), we found that the overall 5-year survival rate of patients with laryngeal EMP was 76.1% [2]. However, the 5-year overall survival rate decreased from 83.7% to 30.0% when these patients progressed to MM [2]. EMP progression to MM was the only (negative) prognostic factor identified for laryngeal EMP [2]. Jawad et al. found that the prognosis of older patients

2016344). Written informed consent was obtained from the patient.

Discussion

EMP is an uncommon tumor, accounting for 3% of all plasma cell neoplasms and <1% of all head-and-neck tumors 2. Primary EMP of the tongue base is rare [1, 4, 7-20]. We used MEDLINE to review the English-language literature, and we searched PubMed/Web of Science using the terms “plasmacytoma” or “extramedullary plasmacytoma” or “hemopoietic neoplasms” combined with “head and neck or tongue base or oral cavity”. We discovered only 18 cases (including our present case) of EMP of

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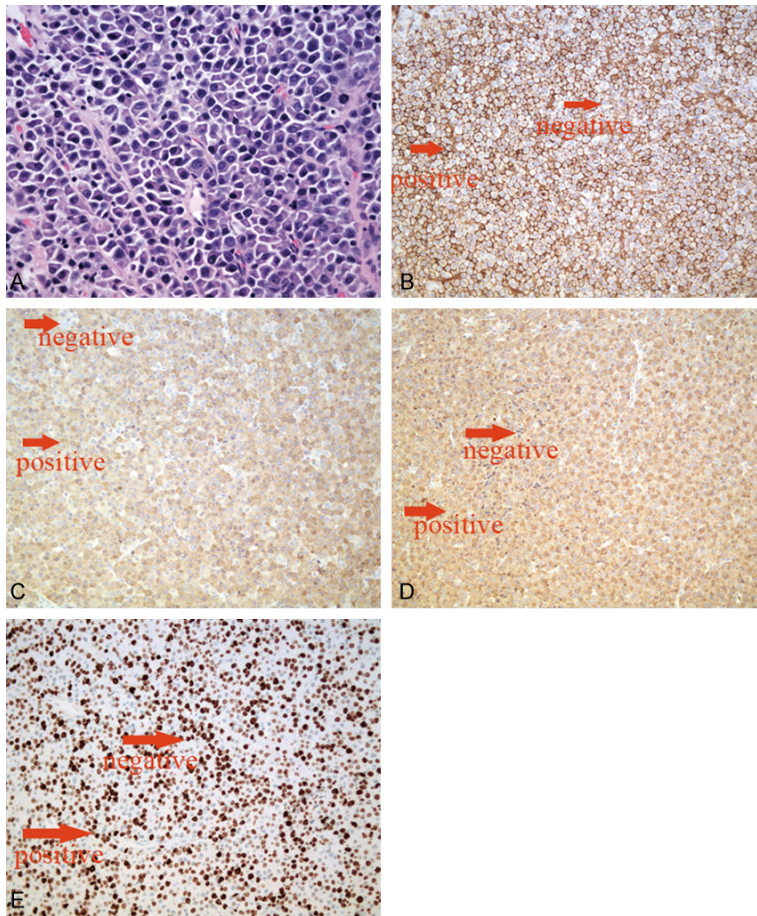


Figure 3. The postoperative histopathological and immunohistochemical results reveal diffuse infiltration of atypical plasma cells ($\times 400$). A. The result of hematoxylin-eosin staining. B. The immunohistochemical result showed that the tumor was positive for CD138 ($\times 200$). C. The immunohistochemical result showed that the tumor was positive for CD79a ($\times 200$). D. The immunohistochemical result showed that the tumor was positive for kappa light chain ($\times 200$). E. The immunohistochemical result showed that the tumor was positive for Ki-67 ($\times 200$).

with EMP was poor, attributable to progression to MM [21]. Thus, early EMP diagnosis and treatment are important to ensure good outcomes.

MRI is widely used to evaluate solitary plasmacytomas and to predict whether tumors have progressed to MM [21-24]. Vogl et al. described the characteristics of six primary EMPs in the head-and-neck region; the lesions were of moderate signal intensity on T2-weighted imaging, isointense or slightly hyperintense on T1-weighted imaging, and strongly enhanced on contrast-enhanced T1-weighted imaging [11]. The cited authors suggested that these MRI features could be used to differentiate EMPs from scars and inflammatory or edematous reac-

tions [11]. Of the published cases, seven underwent MRI examinations. Only two case reports mention the results of T1- and T2-weighted imaging (ours and that of Vogl et al.). Hama et al. found that the lesion was hypointense on T1-weighted imaging and slightly hyperintense, with a hypointense rim, on T2-weighted imaging [16]. The mass in our case was isointense on T1-weighted imaging and yielded a mixed signal on T2-weighted imaging. On contrast-enhanced T1-weighted MR images, the mass was strongly enhanced.

Recently, DWI has emerged as a relatively new functional imaging tool that measures the molecular (Brownian) motion of protons within living tissues [5]. The extent of molecular diffusion can be estimated and quantified in terms of the ADC [5]. DWI is widely used clinically to differentiate benign from malignant tumors, to diagnose lymph node metastasis, to detect recurrent lesions after radiotherapy/chemotherapy, and to predict the effect of treatment; DWI and ADC values are calculated. Only one of the reports

on EMPs provided an ADC [6]. A mass in the gastrocnemius was hypointense on DWI, and the ADC (range 0.88-1.1) was lower than that of the normal regional musculature (1.7). It was suggested that this feature might be useful for differentiating an EMP from a sarcoma [6]. Our present case is the first to provide an ADC for an EMP of the tongue base. The lesion had high signal intensity on DWI, and the ADC was $1.23 \pm 0.316 \times 10^{-3} \text{ mm}^2/\text{s}$. A malignant lesion was diagnosed. Thus, DWI-MRI may be useful for differentiating malignant from benign lesions.

An EMP of the tongue base may first present as an enlarged cervical lymph node (metastasis from an unknown primary tumor) [12]. Kole et al. (1998) used positron emission tomography/

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Table 1. Extramedullary plasmacytoma of tongue

Author	Age	Sex	Symptom	Physical examination	Treatment	Follow-up
Helmus (1964) [7]	NA	NA	NA	NA	NA	NA
WOODRUF (1979) [8]	87	M	NA	NA	Radiotherapy	Died at mon disseminated
Ye (1985) [9]	30	M	Enlarged cervical lymph node	Strawberry like, enlarged cervical lymph node	Chemotherapy was ineffective, disappeared after radiotherapy	51 mon, NED
Layton (1993) [10]	77	F	Soreness and ulceration of her mouth	Extensive oral ulceration	Biopsy+chemotherapy (melphalan+prednisolone)	1 yr NED
Vogl (1996) [11]	NA	M	NA	NA	Radiotherapy	NA
Kole (1998) [12]	71	F	Cervical lymph nodes	PET identified a small primary lesion on the base of the tongue	Surgery+radiotherapy	3 yrs NED
Liebross (1999) [13]	NA	NA	NA	NA	NA	NA
Webb (2002) [14]	65	M	Painful swallowing	The left of the midline of his tongue base and vallecula	Biopsy+radiotherapy (50 Gy)	Alive (time unavailable)
Wahner-Roedler (2003) [15]	49	M	Burring mouth and dysphagia	A cobblestone granular appearance in the tongue, soft palate and posterior pharynx	Azathioprine, one year later, cyclophosphamide and prednisone, 8 years later, oropharyngeal proliferation, received chlorambucil and prednisone, 12 yrs later, tongue base lymphoma, radiotherapy	20 yrs NED
Hama (2004) [16]	72	M	A tender swelling and ulceration in the tongue	Elastic hard mass with irregular surface of the tongue	Radiotherapy	Died during radiotherapy
Alwan (2010) [1]	65	M	Increasing odynophagia that was associated with a choking sensation during swallowing	The left side of the tongue base, intraoperatively, an ovoid pedunculated mass was seen arising from the tongue base on the left side	Surgery+radiotherapy	28 months NED
Garas (2010) [17]	63	M	Recurrent epistaxis	Identified coincidental asymmetrical nodular soft tissue swellings in the left and	Biopsy+radiotherapy (45 Gy)	1 yr, NED
Orme (2012) [18]	54	F	NA	NA	Radiotherapy	NA
Onal (2012) [4]	50	F	Recurrent pain in both sides of her tongue and an associated loss of taste sensation, tongue ulcerations, mild moderate bleeding swelling	A tender, superficial, erosive ulceration covered with white plaques on both sides of tongue	Biopsy+one cycle of chemotherapy (melphalan+prednisone), radiotherapy (50 Gy)	54 mon, NED
Bila (2013) [19]	78	F	Tongue swelling	An irregular tumor mass at the ventral left side of the tongue base	Biopsy+radiotherapy (40 Gy)	4 mon, NED
Skóra (2015) [20]	54	M	NA	NA	Radiotherapy (70 Gy)	0.7 yrs progression to MM, died of MM/1.8 yrs
	76	M	NA	NA	Radiotherapy (50 Gy)	NED, 4.4 yrs
Present case (2016)	67	M	Foreign body in the pharynx	Mass in the right base of tongue	Surgery+radiotherapy (36 Gy)	2 mon NED

NED: no evidence of disease; NA: not available.

computed tomography to identify the tongue base as the primary site of the mass and to confirm the mass as an EMP [12].

The principal treatment for EMP of the tongue base (14 case reports, including our case, provided detailed treatment data) is radiotherapy [1, 4, 8-12, 14-20]. Eleven of 16 cases received radiotherapy alone [8, 11, 14, 16-20]. Three patients died from their disease [8, 16], and the others lived for 4-54 months after treatment. Three patients initially received chemotherapy [4, 9, 15], which was unsatisfactory, and subsequently underwent radiotherapy (only one case report provided the dose: 50 Gy). These patients were alive without evidence of disease at 54 months [4], 51 months [9], and 20 years [15] after radiotherapy, respectively. The other three patients, including our case, underwent surgery and postoperative radiotherapy [1, 12] (the dose was provided only in our case report: 36 Gy). Although EMP of the tongue base is rare, radiotherapy plays an important role in treatment, as is also true of EMPs of other head-and-neck sites [25-28]. However, the optimal radiation dose remains controversial. Tournier-Rangear et al. recommended doses ≥ 45 Gy; the local control rate was 100% at doses ≥ 45 Gy versus 50% at lower doses ($P=0.034$) [25]. Chao et al. recommended a minimal radiation dose of at least 40 Gy [26]. Michalaki et al. found that a dose of 40-50 Gy was associated with a complete response [27]. Sasaki et al. suggested that variations in the radiation dose from 30 to 64 Gy did not affect the survival rate, and that surgery combined with radiotherapy enhanced overall survival [28].

Conclusions

DWI-MRI may be useful for differentiating malignant from benign lesions in the tongue base. Although EMP of the tongue base is rare, radiotherapy plays an important role in its treatment.

Disclosure of conflict of interest

None.

Address correspondence to: Bai-Shu Zhong, Department of Radiology, The First Affiliated Hospital, College of Medicine, Zhejiang University, Zhejiang Province 310003, China. Tel: 86-13588401891;

Fax: 86-571-87236894; E-mail: zhongbaishu-zh@163.com

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