

Extramedullary Plasmacytoma of the Tonsil: Clinical, Imaging and Histopathological Features of a Rare Tonsil Tumor

Tonsilde Ekstramedüller Plazmasitom: Nadir Tonsil Tümörünün Klinik, Görüntüleme ve Histopatolojik Özellikleri

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ABSTRACT

Extramedullary plasmacytoma of the tonsil is very rare. Although it has some characteristic histopathological features, clinical presentation and imaging findings constitute a diagnostic challenge, and it may be confused with more common tumors of the tonsil. We aimed to demonstrate clinical, histopathological, magnetic resonance and diffusion-weighted-imaging findings of a patient with an extramedullary plasmacytoma of the tonsil. These findings increase the awareness of this rare tumor, and provide valuable information for considering plasmacytoma in the differential diagnosis of tonsil tumors.

Keywords

Extramedullary, plasmacytoma, palatine tonsil, magnetic resonance imaging, diffusion magnetic resonance imaging

ÖZET

Ekstramedüller plazmasitom tonsilde oldukça nadir görülür. Karakteristik histopatolojik özelliklerine karşın, görüntüleme ve klinik bulguları tanısal zorluklar oluşturur ve tonsilin daha sık görülen tümörleri ile karıştırılabilir. Tonsilde ekstramedüller plazmasitomu olan olgumuzu klinik, histopatolojik, manyetik rezonans ve diffüzyon ağırlıklı görüntüleme bulgularıyla birlikte sunduk. Amacımız nadir görülen bu tümörün farkındalığını arttırmak ve tonsil tümörünün ayırıcı tanısında düşünülmesini sağlamaktır.

Anahtar Sözcükler

Ekstramedüller, plazmasitom, palatin tonsil, manyetik rezonans görüntüleme, diffüzyon manyetik rezonans görüntüleme

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INTRODUCTION

Plasmacytomas are abnormal proliferation of the plasma cells in the bone marrow. Plasmacytoma growing outside the bone marrow, extramedullary plasmacytoma, is extremely rare. It may occur without multiple myeloma.^{1,2} Extramedullary plasmacytoma represents only less than 1% of all malignancies in the head and neck region, commonly affected areas are nasopharynx and paranasal sinuses.³ Extramedullary plasmacytoma are rarely seen in palatine tonsils.⁴ Although it has some characteristic histopathological and immunohistochemical features, clinical presentation and imaging findings of tonsil plasmacytoma constitute a diagnostic challenge and may be confused with more common tumors of the tonsil.³⁻⁵ In this case report, we aimed to present clinical, histopathological and magnetic resonance imaging (MRI) and diffusion-weighted imaging (DWI) findings of the extramedullary tonsil plasmacytoma.

CASE REPORT

A 77-year-old man presented with the complaints of lump sensation in the throat and dysphagia. The right tonsil was enlarged with a superficial ulceration (Figure 1). The patients' past medical history, physical examination and laboratory tests were unremarkable. MR imaging of the neck (1.5 T MRI system, Philips Achieva, Best, the Netherlands) revealed a well-defined, homogeneous mass, measuring 25x19x47 mm in the right tonsil. The mass showed a signal isointense to muscles on T1 weighted (T1W) image. It was slightly hyperintense on T2-weighted (T2W) images. On contrast-enhanced T1W image, the mass demonstrated diffuse mild enhancement. Diffusion-weighted imaging was obtained in axial orientation using an echo-planar imaging sequence (Figure 2).

Excisional biopsy of the tonsil demonstrated a solid, white nodular tumoral mass. Histopathological examination showed the lesion was consisting of atypical and multinucleated plasmacytes under the squamous epithelium of the right tonsil. Immunohistochemical staining showed that tumor cells were positive for CD138 and lambda light chain protein. They were negative for CD20 and kappa light chain protein which was excluding B cell non-Hodgkin lymphoma. Histopathological and immunohistochemical studies were consistent with plasmacytoma (Figure 3).

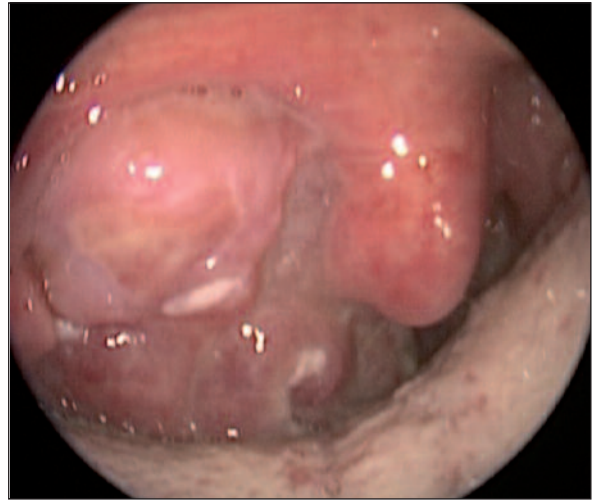


Figure 1. Endoscopy shows superficially ulcerated hypertrophic right tonsil.

DISCUSSION

We presented clinical, histopathological and imaging findings of a case with extramedullary tonsil plasmacytoma. Although it is rare, and both clinical and imaging findings resemble other more common tumors of the tonsils, some features of this rare tumor may suggest this diagnosis. Once it is included in the differential diagnosis, it may be identified by typical histopathological findings.⁴⁻⁷

Although imaging findings of the head and neck extramedullary plasmacytoma were reviewed and reported to be nonspecific, the imaging findings of the tonsil plasmacytoma have not been reported thoroughly yet.⁵ In our case, the lesion was well-defined, bulky, homogeneous, isointense to muscles on T1 weighted images and slightly hyperintense on T2-weighted images.

The diffusion-weighted imaging findings of head and neck plasmacytomas have not been reported previously. Diagnosis and differentiation of the tonsil tumors is difficult on conventional CT and MR imaging because both malignant and benign pathologies may show similar homogeneous appearance. Diffusion-weighted imaging may give valuable information about tumor diagnosis and histology.⁸ A previous study that compared diffusion-weighted imaging findings of normal tonsils and tonsillar squamous cell carcinomas found a higher apparent diffusion coefficient (ADC) values in the squamous cell carcinoma group. All tonsil tumors in their study demonstrated ADC values higher than $0.82 \times 10^{-3} \text{ mm}^2/\text{s}$.⁸ In our case, the mean ADC of the tonsil tumor

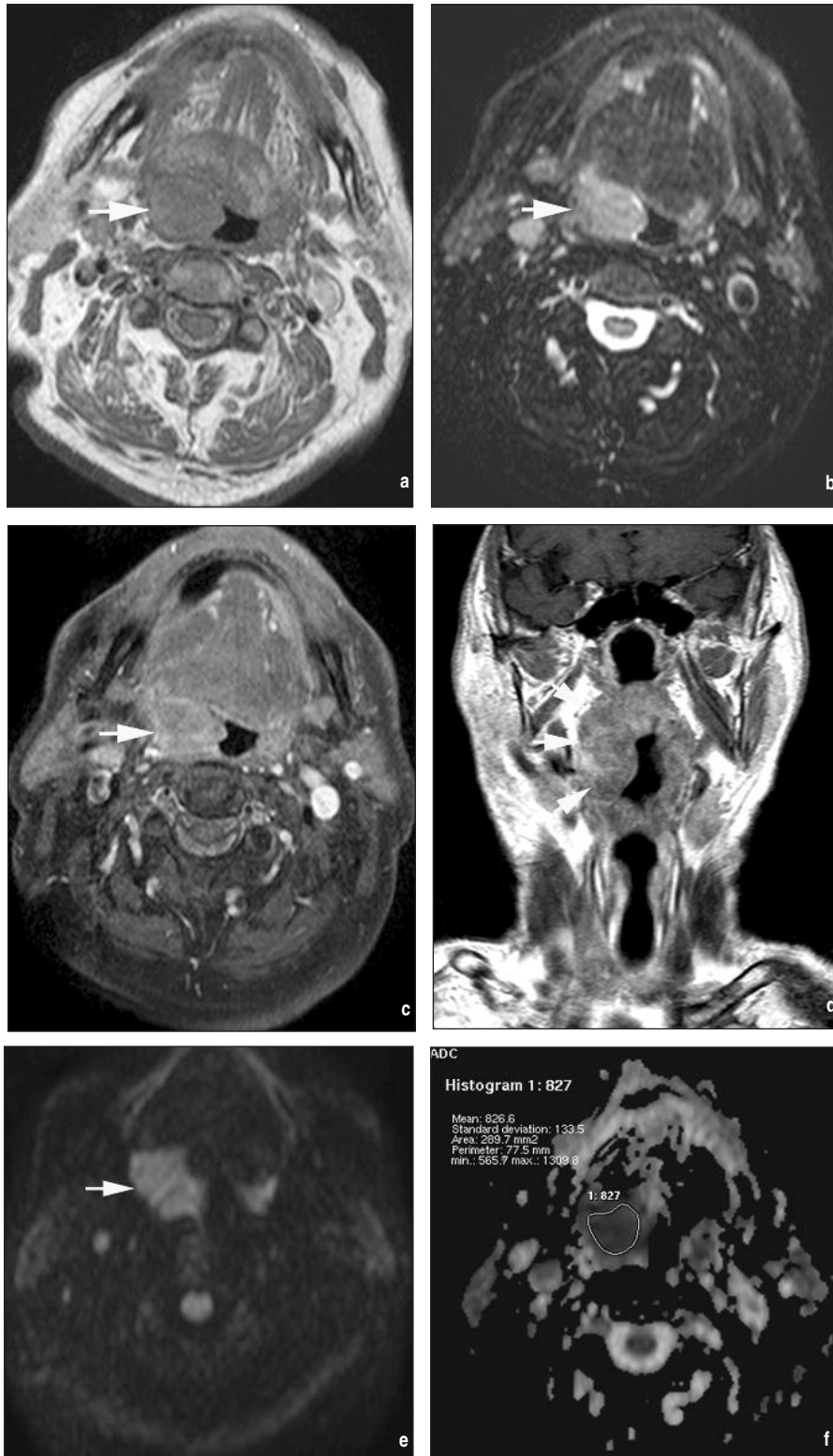


Figure 2a-f. Images of a 77-year-old man with extramedullary plasmacytoma of the right tonsil. **a.** Axial T1-weighted image demonstrates right tonsil mass isointense to muscle. **b.** It is slightly hyperintense on axial T2-weighted image with fat-suppression. **c, d.** The mass shows mild enhancement after contrast injection on axial gadolinium-enhanced T1-weighted fat-suppressed and coronal gadolinium-enhanced T1-weighted images. **e.** Axial diffusion-weighted image shows hyperintensity and **f.** ADC maps with the region of interest drawn around the primary tumor shows low ADC values.

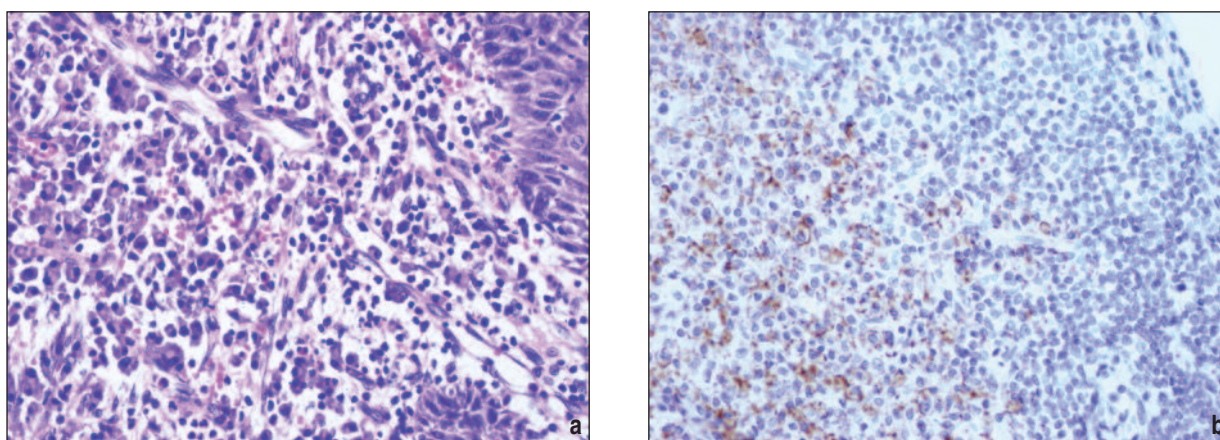


Figure 3a,b. a. Photomicrograph shows tumor cells consisting of typical and atypical plasmacytes beneath the tonsillary epithelium (hematoxylin and eosin stain). b. Immunohistochemical examination demonstrates that the tumor cells were positive for plasma cell marker, CD138.

was $0.827 \times 10^{-3} \text{ mm}^2/\text{s}$, and considering the above mentioned study, the calculated ADC value was lower than squamous cell carcinoma of the tonsil and higher than the normal tonsils. The lower calculated ADC value is probably due to densely packed cells in plasmacytoma when compared the cells that contain necrosis in squamous cell carcinoma. The ADC values of the tumor in our case were similar to the recently reported primary gastric plasmacytoma, $0.863 \times 10^{-3} \text{ mm}^2/\text{s}$.⁹ Although further studies are required, obtaining diffusion-weighted images may support diagnosis.

Plasmacytomas of the head and neck region are rare and before treatment it should be distinguished

from multiple myeloma.³ Treatment of extramedullary plasmacytoma could be with local radiation therapy and/or surgery. This tumor is highly radiosensitive and radiation therapy is accepted as a primary treatment by some authors. Long-term post-treatment follow-up recommended for detection of conversion to multiple myeloma.³⁻¹⁰

In conclusion, extramedullary plasmacytoma should be considered in the differential diagnosis of tonsil masses. Bulky, homogeneous, well-circumscribed tumors with slight T2 hyperintensity, mild contrast enhancement and low ADC values may help suggesting extramedullary plasmacytoma.

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