

# Isolated Extramedullary Plasmacytoma of the True Vocal Cord: A Case Report and Review of The Literature

Füsun TOKATLI\*, Nermin TUNÇBİLEK\*\*, Şemsi ALTANER\*\*\*, Zafer KOÇAK\*, Cem UZAL\*,  
Bengü DENİZLİ\*, Abdullah TAŞ\*\*\*\*

\* Department of Radiation Oncology, Faculty of Medicine, Trakya University,

\*\* Department of Radiology, Faculty of Medicine, Trakya University,

\*\*\* Department of Pathology, Faculty of Medicine, Trakya University,

\*\*\*\* Department of Otorhinolaryngology, Faculty of Medicine, Trakya University, EDİRNE

## ABSTRACT

Extramedullary plasmacytoma (EMP) is an uncommon neoplastic disorder arising from B-cell series lymphocytes and comprises 3% of plasma cell neoplasms and 0.1% of laryngeal carcinomas. Isolated EMP of the true vocal cord is an extremely rare lesion. Only 9 cases have been reported in the English medical literature. We report a case of isolated EMP of the right true vocal cord in a 55-year-old male with a 10-year history of intermittent hoarseness. Although this appears to be an isolated case, EMP of the true vocal cord must be included in the differential diagnosis in patients with history of longstanding hoarseness.

**Key Words:** Extramedullary plasmacytoma, True vocal cord

## ÖZET

### Gerçek Vokal Kordun İzole Ekstramedüller Plazmasitoması: Olgu Sunumu ve Literatür Derlemesi

Ekstramedüller plazmasitoma (EMP), B lenfositlerden orijin alan, plazma hücreli neoplazmaların %3'nü ve larenjeal karsinomların %0.1'ni oluşturan nadir bir hastalıktır. Gerçek vokal kordun izole EMP'sı oldukça nadir görülür. İngilizce tıp literatüründe bildirilmiş sadece dokuz olgu vardır. Bu sunumda, 10 yıldır aralıklı ses kısıklığı şikayeti olan 55 yaşındaki erkek hastada saptanan gerçek vokal kordda izole EMP bildirilmektedir. Uzun süreli geçmeyen ses kısıklığı hikayesi olan hastalarda ayırıcı tanıda gerçek vokal kordun EMP'sı da düşünülmelidir.

**Anahtar Kelimeler:** Ekstramedüller plazmasitoma, Gerçek vokal kord

## INTRODUCTION

Extramedullary plasmacytoma (EMP) are rare malignant neoplasms composed of plasma cells, 90 percent of which occur in the head and neck area, but generally representing only 1 percent of tumors of head and neck location (1,2). EMP makes up 0.1% of laryngeal carcinomas recorded in reports (2). The most commonly involved site is the supraglottis, followed by the true vocal cords, false vocal cords and ventricles. Isolated EMP of the true vocal cord is a very rare lesion, and only 9 cases were reported in the English medical literature, but none of them had regional lymph node metastasis (3-10).

We report a patient of EMP of the true vocal cord with regional lymph node metastasis who had complaint of intermittent hoarseness for the past 10 years; this is the first case of such cases (true vocal cord) reported in the literature.

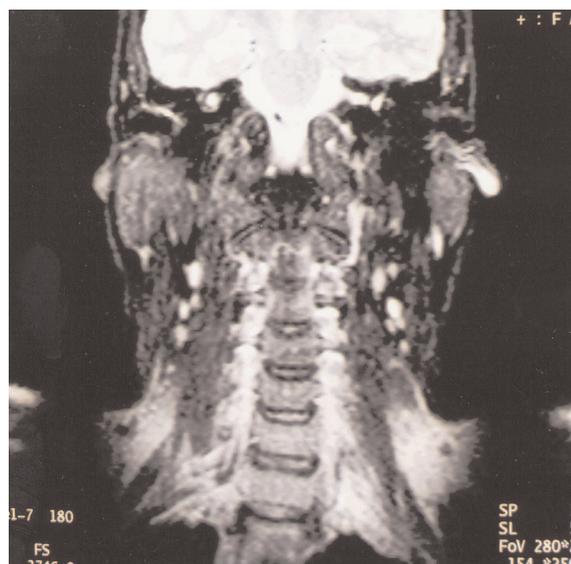
## CASE REPORT

A 55-year-old man presented with a 10-year history of intermittent hoarseness that had become severe and persistent in the previous three months.

He was not admitted to the hospital before. He had no dysphagia, stridor, or hemoptysis and had no history of previous trauma, surgery, or smoking. On examination, no enlarged lymph nodes were detected on the neck. Microlaryngoscopic examination revealed a thickening of the right true vocal cord, irregularity on the mucosa, and hemi-mobile cord fixation. To better evaluate the lesion and its extension, magnetic resonance imaging (MRI) was performed and showed a strong contrast enhanced annular tumoral mass originating from right vocal cord and bilateral metastatic multiple cervical lymph nodes in 1.5 cm diameter were detected (Figure 1A and 1B). The nodes, which are clinically non-palpable, showed the same signal intensity as the primary tumor. Histological examination of multiple biopsies from the right vocal cord revealed variation from mature to immature plasma cells (Figure 2A). Formalin-fixed, paraffin-embedded sections were stained with various NeoMarkers (Fremont, CA, USA) antibodies. The following immunoperoxidase stains were used with standardized avidin-biotin method. The tumor was expressed cytoplasmic kappa light chain and IgG (Figure 2B). The results of both serum and urine



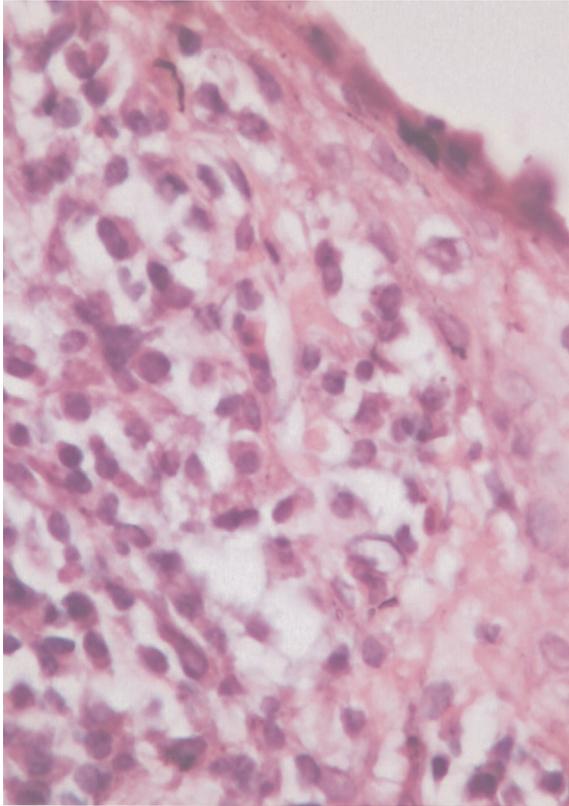
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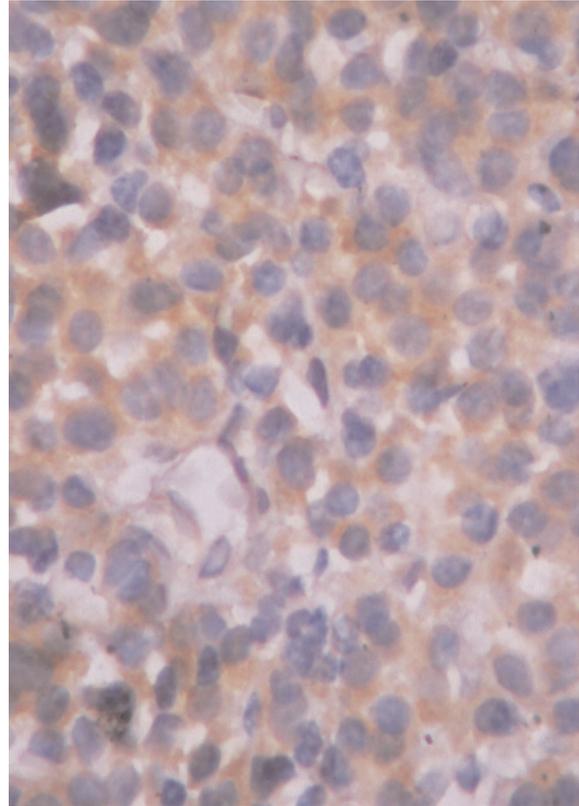
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**Figure 1-A.** Contrast-enhanced MRI shows a strong contrast fixation in the right true vocal cord.

**1-B.** Coronal T2-weighted fat-saturation image shows bilateral multiple cervical lymph nodes.



**A**



**B**

**Figure 2-A.** The tumor is composed of plasma cells that are surrounding squamous epithelium of the larynx. The mature plasma cells were containing a round eccentric nucleus without nucleoli, with abundant cytoplasm (H&E, x 400). **2-B.** Tumor reacted with anti-IgG antibody (Immunoperoxidase, x 400).

immuno-electrophoresis were within normal ranges as were those of beta-2-microglobulin and bone marrow biopsy. No osteolytic lesions were detected on skeletal survey. The full blood count and levels of serum calcium, creatinine and uric acid were within the normal range. As a result of these pathological findings, the tumor was diagnosed as extramedullary plasmacytoma. He was treated with external radiotherapy at a dose of 50 Gy in 25 fractions with Co-60 gamma rays to involved sites (the larynx and the upper and middle cervical lymph nodes) using two opposing lateral ports. Two years after the treatment, there were no tumor recurrence or systemic dissemination, while patient's voice improved considerably. MRI showed edema and thickness secondary to radiation in the right true vocal cord, and no pathologic lymph nodes were

detected. On endoscopic laryngeal examination, an edema in the right true vocal cord was the only finding.

## DISCUSSION

Plasmacytomas are a localized proliferation of plasma cells in the bone marrow and less frequently in extraosseous organs or tissues (11). Extraosseous plasmacytoma can present as a solitary lesion, referred to as extramedullary plasmacytoma or as a part of the process of disease dissemination.

Approximately 80 percent of solitary EMP occurs in the head and neck but they represent less than one percent of all head and neck tumors (1,2). Most

**Table 1.** Characteristics and management of patients with isolated EMP of the true vocal cord (modified from Rakover et al.)

No.	Author	Age/Sex	Symptoms	Site	Treatment	Follow-up
1	Ringertz (3)	59/Male	Hoarseness (4 months)	Left vocal cord	Radiotherapy	4 years, NED
2	Webb (4)	55/Female	Hoarseness (3 months)	Right vocal cord	Surgery	11 years, NED
3	Gorenstein et al. (5)	42/Male	Hoarseness (6 months)	Both true folds	Surgery	5 years, NED
4	Gadomski et al. (6)	54/Female	Hoarseness (1.5 months)	Glottis	Surgery + chemotherapy	Died 15 years later (reported tumor-free)
5	Kost (7)	43/Male	Hoarseness (3 months)	Left true vocal cord	Radiotherapy (70 Gy)	Laryngectomy 9 months later for radionecrosis, 7 years, NED
6	Nowak-Sadzi- kowska and Weiss (8)	50/Male	Hoarseness (3-6 months)	Glottis	Radiotherapy (60 Gy)	10 years, NED
7	Nowak-Sadzi- kowska and Weiss (8)	48/Male	Hoarseness (3-6 months)	Glottis	Radiotherapy (60 Gy)	10 years, NED
8	Rakover et al. (9)	38/Male	Hoarseness (12 months)	Right true vocal cord	Surgery + radiotherapy (50 Gy)	3 years, NED
9	Alexiou et al. (10)	69/Male	No data	Glottis	Surgery + radiotherapy (50 Gy)	5.2 years, NED
10	Present case	55/Male	Hoarseness (120 months)	Right true vocal cord	Radiotherapy (50 Gy)	2 years, NED

EMP: Extramedullary plasmacytoma, NED: No evidence of disease

cases occur in the nasal cavity, paranasal sinuses, nasopharynx and oropharynx. Plasmacytoma involving the larynx is very rare. The most common sites of the laryngeal EMP are, in order of decreasing frequency, the epiglottis, true vocal cords, false vocal cords, ventricles, arytenoids, and subglottic space (2). Our case presented as a vocal cord mass with radiologically detected lymph nodes. Up to date, we realized that only nine cases of isolated EMP of the true vocal cord were reported in the English medical literature (Table 1) (3-10). No clinically or radiologically involvement of regional lymph nodes were reported in these studies.

The symptoms of laryngeal plasmacytomas are like other tumor symptoms of the same site (2). The most common symptom is slowly progressive hoarseness (7,12). Dysphagia, stridor, hemoptysis, and / or choking sensations are symptoms associated with locally aggressive tumors (7,13). It is unusual for a patient to initially present with a mass in the head and neck region (14). Authors reported that approximately 20% of patients with EMP of the larynx suffer of metastasis to lymph nodes in the neck (7,15). Welsh et al. reported that 1 out of 8 cases of epiglottic plasmacytoma presented with a neck mass without other symptoms (16). Glottis has a poor lymphatic drainage; therefore metastasis to the cervical nodes is less than 5%. In the current case, although the tumor located in the true vocal cord, the patient had bilateral multiple metastasis to regional lymph nodes on cervical MRI, which were not clinically detectable. Since the metastasis was not proven by biopsy, it was not easy to distinguish them from reactive lymph nodes, which might disappear after the treatment of the primary tumor. However, MRI further improved our ability to predict lymphatic metastasis. It can easily detect nodes of 4-5mm and if they show the same signal intensity as the primary tumor on T1- and T2-weighted images they are probably invaded (17), as in our case. Furthermore, radioresponsiveness of the nodes suggested that they might be metastatic.

Generally the diagnosis is difficult due to unspecific symptoms and can only be confirmed histopathologically. The tumor is composed of varying well-to-poorly differentiated sheets of plasma cells. Immunoperoxidase stains demonstrate monoclonal

light-either kappa or lambda- or heavy chain immunoglobulin (1). As the treatment is different, it is important to distinguish plasmacytoma from a benign reactive plasmacytosis, pseudolymphoma and non-Hodgkin's lymphoma (1,12). In this present case, the uniformity of plasma cells and the immunohistochemical findings strongly supported for a plasmacytoma.

MRI has been reported as useful radiological study for the diagnosis of laryngeal plasmacytomas (18,19). The mass usually appears homogenous, with well-defined margins, located submucosally, and reveals slight to moderate enhancement after delivery of contrast material. These features are non-specific and radiology is used mainly to demonstrate disease extent. Other lesions need to be considered such as abscess, epithelial tumors, and chloroma. In the current case, the tumor was well defined, homogenous, located submucosally, and there was a strong contrast enhancement in the tumor.

Surgical excision, radiotherapy or a combination of these, constitutes effective therapy for EMP of the true vocal cord, and they produce excellent local and regional control rates (1,5,7,11). Small-localized tumors are best treated by surgery at the time of laryngoscopy. The larger lesions, however, necessitating major surgery, are best treated by radiotherapy (5,12). EMP's are radiosensitive tumors and their response to radiation is excellent (20). The doses required for the optimal management of the EMP of the true vocal cord is not well established, but most authors recommend doses of 45-50 Gy or more (7,11).

Although extremely rare, EMP of the true vocal cord should be considered in patients with history of longstanding hoarseness.

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## Correspondence:

Dr. Füsün TOKATLI  
Trakya Üniversitesi Tıp Fakültesi  
Radyasyon Onkolojisi Bölümü  
22030, EDİRNE

Phone: (0.284) 235 76 42 / ext:1065 - 4407

Fax: (0.284) 236 10 74

e- mail: fusun\_t@yahoo.com