

## CASE REPORT

# A case report of extramedullary plasmocytoma—rare pathology in the larynx

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## Key Clinical Message

Extramedullary plasmocytoma is a rare finding. The diagnosis is made by histological and immunohistochemical examination. Hematological evaluation is mandatory to rule out multiple myeloma. Radiotherapy is treatment of choice with good results.

## KEYWORDS

imaging of the larynx, larynx, microlaryngeal surgery, radiation therapy, voice/ dysphonia

## 1 | INTRODUCTION

Direct laryngoscopy is mainstay in the workup of patients suffering from hoarseness. This examination usually give vital information in discriminating between functional complaints and anatomical abnormalities. In this case study, we present laryngoscopic findings and subsequent investigations of a rare cause of slowly progressive hoarseness. The differential diagnosis upon presentation included laryngocele, amyloidosis, papillomatosis, and squamous cell carcinoma. Only after multiple diagnostic interventions, we diagnosed the patient with

extramedullary plasmocytoma (EMP). This rare pathology represents less than 1% of plasma cell neoplasms in the head and neck region.<sup>1-3</sup>

Plasmocytoma is a solitary lesion, which can be found in bone marrow or soft tissue. When situated outside the bone marrow, it is termed EMP. The lesion consists of localized malignant plasma cells that secondarily can lead to amyloid deposition.<sup>1</sup> Eighty percent of all EMP are found in the head and neck region, most commonly in the nasal cavity, paranasal sinus, nasopharynx, and larynx. With this report, we aim to create more awareness for this rare pathology.

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## 2 | CASE REPORT

A healthy 45-year-old man was seen in our outpatient clinic because of slowly progressive hoarseness, which started 1.5 years earlier. There were no other symptoms concerning breathing or swallowing. He is a nonsmoker and does not use his voice professionally. Laryngoscopy showed a cystic lesion arising from the right ventricle (Morgagni's sinus) that obscured full view of the right vocal cord. The vocal cords showed normal mobility with some limitation due to mass action. A computed tomographic (CT) scan of the neck was performed and showed a slightly lobular lesion with a diameter of 1.3 cm in the right anterior glottic region (Figure 1; axial and sagittal view of a slightly lobed lesion with a diameter of 1.3 cm in the right anteriorly glottis region). During subsequent microlaryngeal surgery, the lesion was seen to originate from the right ventricle where it presented as a smooth purple lobular tumor (Figure 2 the lesion originating from the right ventricle, presenting as a smooth purple lobed tumor). The vocal cords and supraglottic structures were not affected. The lesion was removed, and additionally the surface of the wound was evaporated using a CO<sub>2</sub> laser, laser setting 6 Watt, depth 0.1, diameter 1 mm, repeated pulse, and off time 0,1 s. Histopathologic examination revealed a plasmocytoma with numerous closely spaced plasma cells (Figure 3A overview of the biopsies with surface lining by both squamous and respiratory epithelium, and dense subepithelial infiltrates (hematoxylin and eosin staining, 50x magnification) consisting of (B) plasma cells (hematoxylin and eosin staining, 200x magnification) that showed immunohistochemical expression restricted to Lambda light chains). Immunohistochemical staining showed positivity for CD138 and restricted expression of lambda light chain.

To exclude systemic disease, an extensive workup was performed at the department of hematology This consisted of urine and blood analysis, bone marrow biopsy, and positron emission tomography (PET) scan. No other evidence for lymphoreticular malignancies was found.

Accordingly, the patient was diagnosed with extramedullary plasmocytoma of the larynx. Treatment consisted of radiotherapy in 25 sessions of 2 Gy each. Within weeks after treatment, the symptom of hoarseness and all clinical signs of the tumor had disappeared.

## 3 | DISCUSSION

In this case study, histopathologic examination revealed plasmocytoma. Recognizing extramedullary plasmocytoma is challenging because of its rare presentation (<1% of the plasma cell neoplasms in the head and neck region).<sup>1-3</sup>

Our differential diagnosis included laryngocele, amyloidosis, papillomatosis, and squamous cell carcinoma (SCC). Based on direct laryngoscopic findings, the diagnosis of a laryngocele was considered first, since it is a herniation of the laryngeal saccule that extends upward within the false vocal fold. It is normally filled with air or mucus. Computed tomographic findings were not consistent with this entity. Second in the differential diagnosis was primary amyloidosis. It is an extracellular deposition of amyloid. It appears as a submucosal lesion with poorly defined borders. Computed tomographic scan typically demonstrates marked thickening of laryngeal soft tissues. In this case, amyloidosis was histologically excluded. Next to be considered was laryngeal papillomatosis. It appears as a polypoid lesion localized in the vocal cords or ventricular pleats. Histopathology shows central fibrovascular cores covered by squamous epithelium. Last in the differential diagnosis was a SCC, which is often recognized as an exophytic or ulcerated lesion showing possible invasion of the adjacent structures on CT scan. Both were not present in our case.

Extramedullary plasmocytoma is a solitary lesion in soft tissue, consisting of malignant plasma cells that can lead to an amyloid deposition.<sup>1</sup> Extramedullary plasmocytoma is most commonly found in the head and neck region (80%). The mean age is 60 years, and EMP is more prevalent in men.<sup>3,4</sup> Clinical symptoms relate to the location of

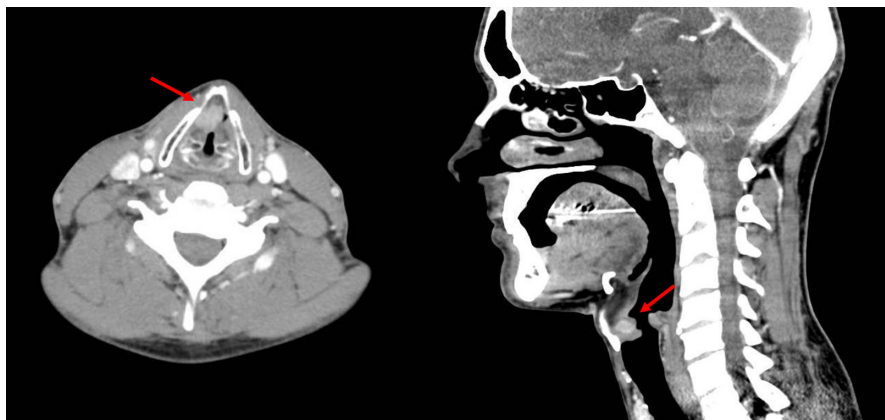
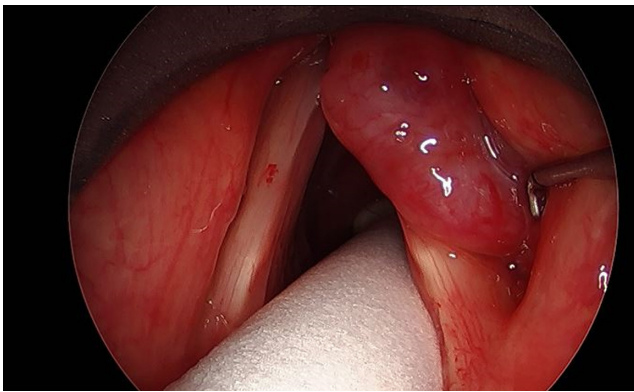


FIGURE 1 Axial and sagittal view of a slightly lobed lesion with a diameter of 1.3 cm in the right anteriorly glottis region.

the lesion and infiltration in adjacent structures. In the larynx, EMP mostly affects the epiglottic, ventricle, or vocal cords. Similar to our case, the most common symptom of EMP is slowly developing hoarseness.<sup>1–6</sup> Laryngoscopy can show a polypoid lesion or submucosal swelling of the soft tissue.<sup>2</sup> Other case reports also describe a mucosa-covered purple/reddish mass lesion.<sup>1–5</sup> A CT scan can be performed, which shows a homogenous laryngeal mass, possibly with calcifications, areas with low densities in the cartilage or infiltrative growth pattern, typical for EMP. MRI can show a laryngeal mass with hypo intense on T1-weighted images and hyperintense on T2-weighted images.<sup>5</sup> However, confirming the diagnosis is not possible by imaging methods and is based on histological and immunohistochemical evaluation. Immunophenotyping is positive for either kappa or lambda light chains and in 15% amyloid is detected.<sup>4</sup> Since EMP can occur as a presentation of a more generalized disease, further localizations should be ruled out. This is done by performing



**FIGURE 2** Lesion originating from the right ventricle, presenting as a smooth purple lobed tumor.

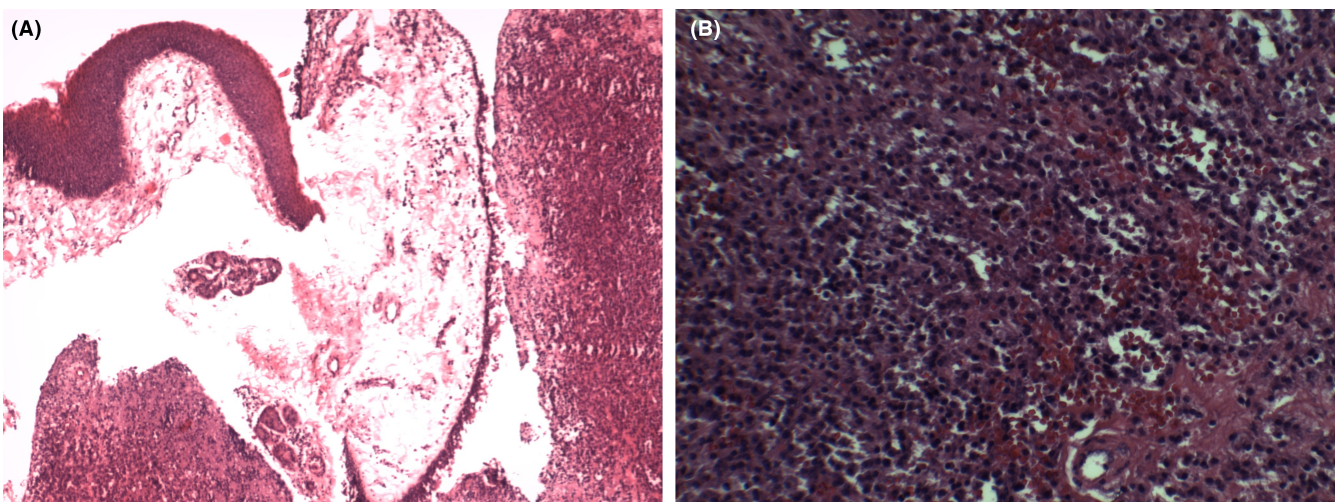
peripheral blood samples, PET scan and bone-marrow biopsy.<sup>1,2,4</sup> In our case study, no further localizations were found making radiotherapy the treatment of choice.

Extramedullary plasmocytoma is highly radiosensitive, and local radiotherapy (40–60Gy) is advised as primary treatment.<sup>1,2,4,6</sup> Unfortunately, optimal radiation dose is still undetermined.<sup>6</sup> In case of small lesions, surgical resection, with preservation of laryngeal function, can be performed with adjuvant radiotherapy.<sup>2,6</sup> However, recurrence is seen in up to 30%.<sup>1</sup>

Extramedullary plasmocytoma is known as locally aggressive yet nondisseminating.<sup>4</sup> Nonetheless, metastasis in the cervical lymph nodes are reported, but do not negatively affect prognosis.<sup>4</sup> Elective treatment of the neck is not routinely advised since there is no evidence for positive effect on prognosis.<sup>4,6</sup> Prognosis is mainly dependent on the possible future progression to multiple myeloma. In 10%–30% of patients with EMP, the disease will progress to multiple myeloma within 2 to 3 years. Therefore, long-term follow-up is indicated.<sup>2,4</sup> In our case study, the current follow-up length is currently 20 months, with no evidence of disease.

## 4 | CONCLUSION

Hoarseness due to extramedullary plasmacytoma is a rare finding. The diagnosis is typically made by histological and immunohistochemical examination. Additional hematological evaluation is mandatory to rule out multiple myeloma. Radiotherapy is treatment of choice with good results in the majority of patients. However, long-term follow-up is necessary because of the possible development of multiple myeloma.



**FIGURE 3** (A) Overview of the biopsies with surface lining by both squamous and respiratory epithelium, and dense subepithelial infiltrates (hematoxylin and eosin staining, 50x magnification) consisting of (B) plasma cells (hematoxylin and eosin staining, 200x magnification) that showed immunohistochemical expression restricted to Lambda light chains.

## CONSENT STATEMENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

## AUTHOR CONTRIBUTIONS

**Cornelia Gloria Florence van Lanschot:** Conceptualization; data curation; formal analysis; investigation; methodology; writing – original draft. **Lara H Böhmer:** Conceptualization; supervision; validation; writing – review and editing. **Rosita L Ten Berge:** Data curation; writing – original draft; writing – review and editing. **Jeroen G Vinke:** Conceptualization; methodology; project administration; supervision; validation; writing – review and editing.

## FUNDING INFORMATION

None.

## CONFLICT OF INTEREST STATEMENT

None to declare.

## DATA AVAILABILITY STATEMENT

Data available on request due to privacy/ethical restrictions.

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