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Laryngeal Extramedullary Plasmacytoma Progressing to Multiple Myeloma: A Case Report

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Abstract | **Case Report**

Background: Extramedullary plasmacytoma is a rare plasma cell neoplasm, representing less than 4% of all plasma cell disorders, with a predilection for the head and neck region. While Extramedullary plasmacytoma typically has a favorable prognosis with localized treatment, approximately one-third of cases progress to multiple myeloma within two years. ***Case Presentation:*** We report the case of a 50-year-old male presenting with acute laryngeal dyspnea due to a locally advanced laryngeal mass. Emergency tracheotomy was performed to secure the airway. Clinical evaluation and imaging revealed a pan-laryngeal tumor with cartilage destruction and extra-laryngeal extension. histopathological and immunohistochemical analysis confirmed the diagnosis of extramedullary plasmacytoma with KAPPA light chain restriction. Further investigations identified diffuse osteolytic lesions and 15% plasma cell infiltration in the bone marrow, leading to the diagnosis of multiple myeloma. The patient received chemotherapy (bortezomib, thalidomide, and dexamethasone) followed by autologous stem cell transplantation. He showed a favorable clinical and biological response, allowing for successful tracheostomy closure and continued maintenance therapy. ***Conclusion:*** This case highlights the diagnostic challenges of laryngeal plasmacytoma and the importance of considering EMP in patients with submucosal laryngeal masses. Given the potential for progression to MM, long-term monitoring and a comprehensive treatment approach remain essential for optimizing prognosis.

Keywords: Extramedullary plasmacytoma, multiple myeloma, laryngeal dyspnea, tracheotomy, autologous stem cell transplantation.

BACKGROUND

Solitary plasmacytoma is a clonal disorder of plasma cell origin, most frequently limited to one site of the axial skeleton and less frequently to a soft tissue area. The solitary plasmacytoma of bone, is considered to present a marked propensity to progress into multiple myeloma (MM), when compared to the solitary extramedullary plasmacytoma [1].

Extramedullary plasmocytoma (EMP) is a tumor of clonal plasma cells, that represents less than 4% of all plasma cell neoplasms, and 80% of them rises in the head and neck region, and most common sites are the nasal cavity, the nasopharynx, the paranasal sinus, and the larynx. [2] and approximately one third of them disseminate to multiple myeloma (MM) within 2 years from presentation [1]. In certain locations such as the head and neck, there is a 50-80% survival rate after 10 years [3]. However, the most important factor that influences the prognosis of the disease is the development to myeloma [4].

The current study report a case of extramedullary plasmocytoma evolving to multiple myeloma, circumstances of diagnosis and the treatment.

CASE PRESENTATION

A 50-year-old male patient, with no significant medical history, was admitted to the emergency department of CHU Hassan II in Fès for acute-onset laryngeal dyspnea, presenting as inspiratory bradypnea, stridor, and signs of respiratory distress, including nasal

flaring, suprasternal and intercostal retractions, and altered consciousness with a Glasgow score of 13/15. His oxygen saturation was 70% on room air and 80% under 15L/min oxygen therapy.

Emergency Management

The patient was immediately transferred to the operating room, where an urgent tracheotomy was performed under local anesthesia. A size 8 cuffed cannula was initially placed to secure the upper airway, followed by the placement of a double-sleeve PVC cannula (size 8.5).

Clinical History and Examination

Once stabilized, a thorough medical history was obtained, revealing no history of smoking (active or passive), alcohol consumption, or occupational exposure. However, the patient reported a two-year history of progressive dysphonia, exertional dyspnea that later occurred at rest, and progressive dysphagia, initially to solids and later to liquids, all evolving in a context of general health deterioration.

On general examination, the patient was conscious, hemodynamically and respiratory stable (WHO performance status: 0). His oxygen saturation was 97% on room air, the tracheostomy site was clean, and no palpable lymphadenopathy was detected. The rest of the physical examination was unremarkable.

An endoscopic examination was performed, revealing a large exophytic tumoral lesion causing significant obstruction of the laryngeal lumen. The lesion's surface appears irregular with a shiny aspect. The mass extends to adjacent laryngeal structures, leading to marked airway narrowing. Anatomical landmarks, particularly the vocal cords, are not visible; only the epiglottis and the right piriform sinus can be identified. Notably, the tumor remains immobile during phonation, which is suggestive of left vocal cord paralysis (Figure 1).

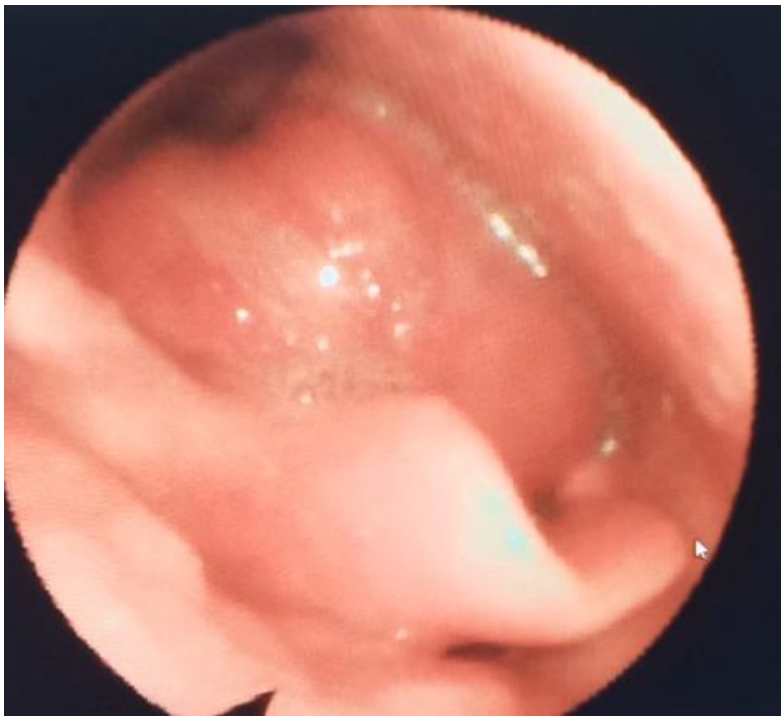

Figure 1: Nasofibroscopy showing an obstructive glotto-supraglottic tumoral process, significantly narrowing the laryngeal airway. The absence of vocal cord mobility during phonation suggests an associated paralysis

Paraclinical Investigations

A cervico-thoracic CT scan showed a pan-laryngeal tumoral process centered on the right vocal cord, which was locally advanced, crossing the midline inward and occupying almost the entire glottic space. It extended to the contralateral vocal cord, infiltrated both the anterior and posterior commissures, and invaded the ipsilateral aryepiglottic fold. Additionally, there was cricoid cartilage erosion posteriorly and thyroid cartilage destruction laterally, with extra-laryngeal extension to the prelaryngeal muscles. No distant metastases were detected on the cervico-thoracic CT scan (Figure 2).

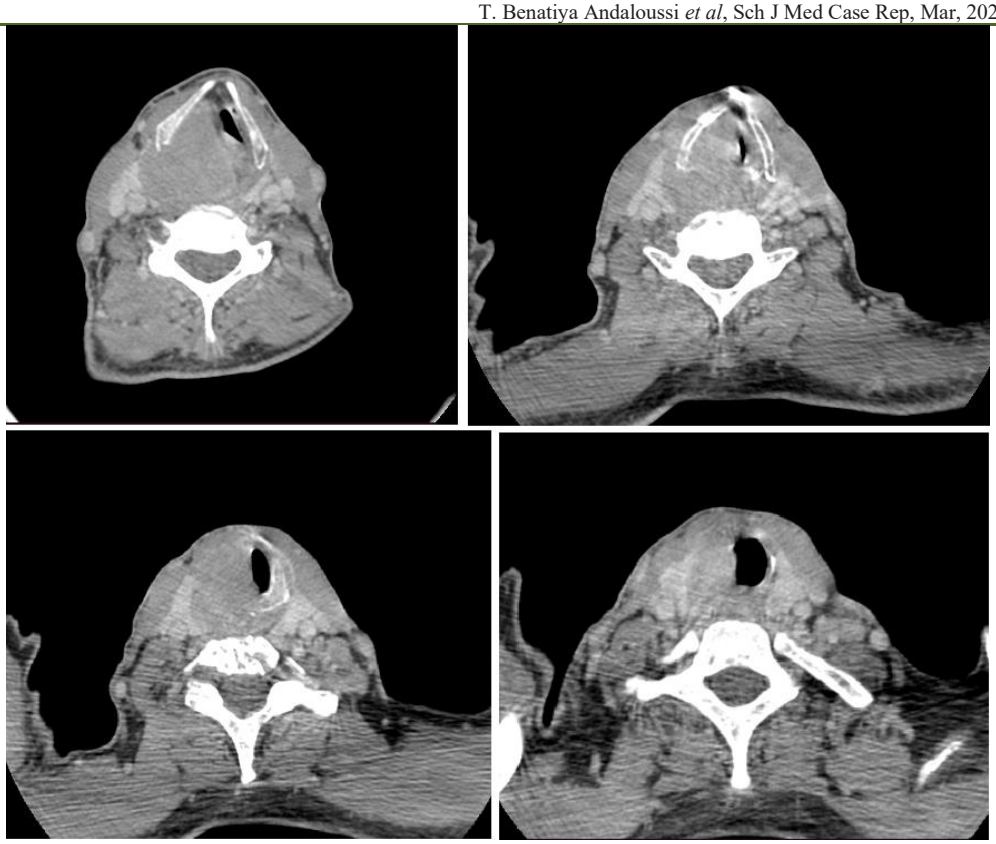

Figure 2: Axial view of the patient's CT scan showing a tumor centered on the right vocal cord, locally advanced, crossing the midline, occupying most of the glottic slit, contacting the contralateral vocal cord, infiltrating the anterior and posterior commissures, and invading the homolateral aryepiglottic fold. There is destruction of the posterior cricoid and lateral thyroid cartilage, with extension to the extralaryngeal pre-tracheal space

A direct laryngoscopy revealed a bulky, exophytic tumor obstructing the entire glottis, preventing visualization of anatomical landmarks. The piriform sinuses, tongue base, and vallecula were clear.

Three biopsies were performed:
The first two were inconclusive.

The third biopsy identified an undifferentiated tumor, and additional immunohistochemical analysis, correlated with FISH study findings, confirmed an extramedullary plasmacytoma with KAPPA light chain restriction (Figures 3, 4, 5).

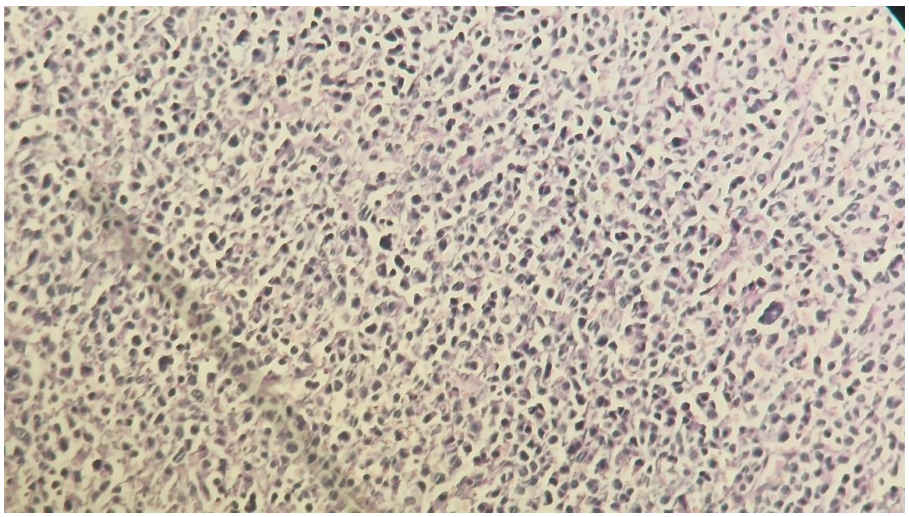

Figure 3: Tumoral proliferation arranged in sheets, composed of plasmacytic cells

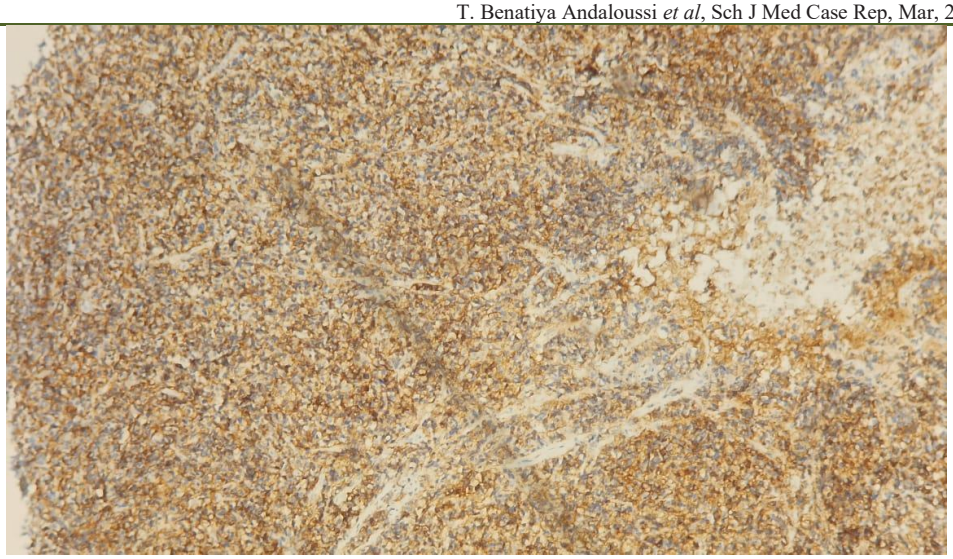

Figure 4: Immunohistochemical study showing CD138 expression by tumor cells.

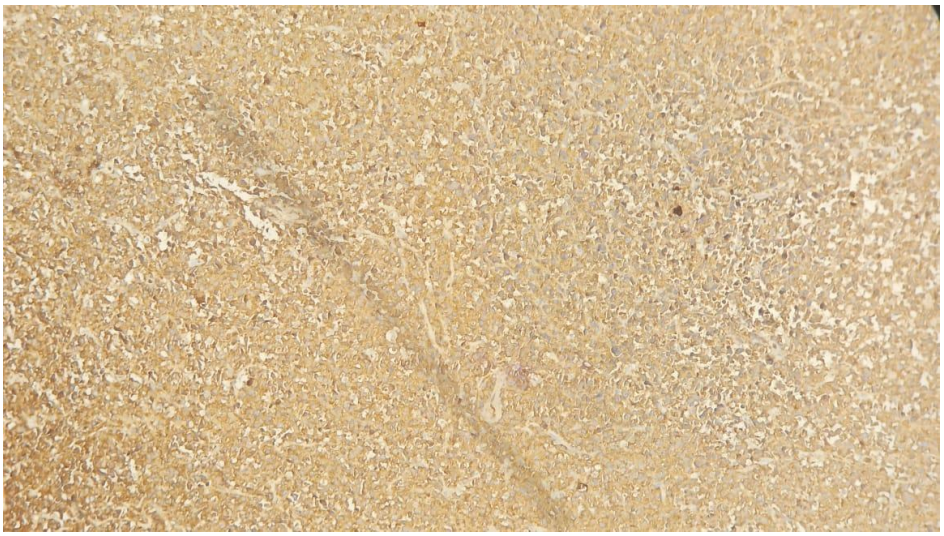

Figure 5: Immunohistochemical study showing kappa light chain expression by

Onco-Hematological Evaluation

The patient was referred to the onco-hematology department, where an extension workup revealed:

- A tissue lesion centered on the glotto-subglottic region, enhancing after contrast injection, measuring 48×64×86 mm.
- Lysis of the thyroid, cricoid, and right arytenoid cartilages.
- Infiltration of the right thyroid lobe.
- Complete obstruction of the laryngeal lumen.
- Bilateral jugulocarotid and submandibular lymphadenopathy, the largest measuring 15×9 mm.
- Multiple diffuse lytic lesions in the axial and peripheral skeleton.

Based on the histopathological findings and extensive osteolytic lesions, the diagnosis of multiple myeloma was confirmed. A bone marrow aspirate revealed 15% plasma cells, further supporting the diagnosis.

Treatment and Outcome

The patient underwent six cycles of chemotherapy with bortezomib, thalidomide, and dexamethasone over six months, with excellent clinical and biological response. He then received an autologous stem cell transplant.

Follow-Up and Decannulation

Three months later, the patient was admitted for a decannulation trial. An endoscopic examination was conducted, demonstrating complete regression of the tumoral process. The laryngeal landmarks appeared normal, with no signs of residual tumor involvement. However, there was persistent right vocal cord palsy, with the right cord remaining in a paramedial position. (Figure 6).

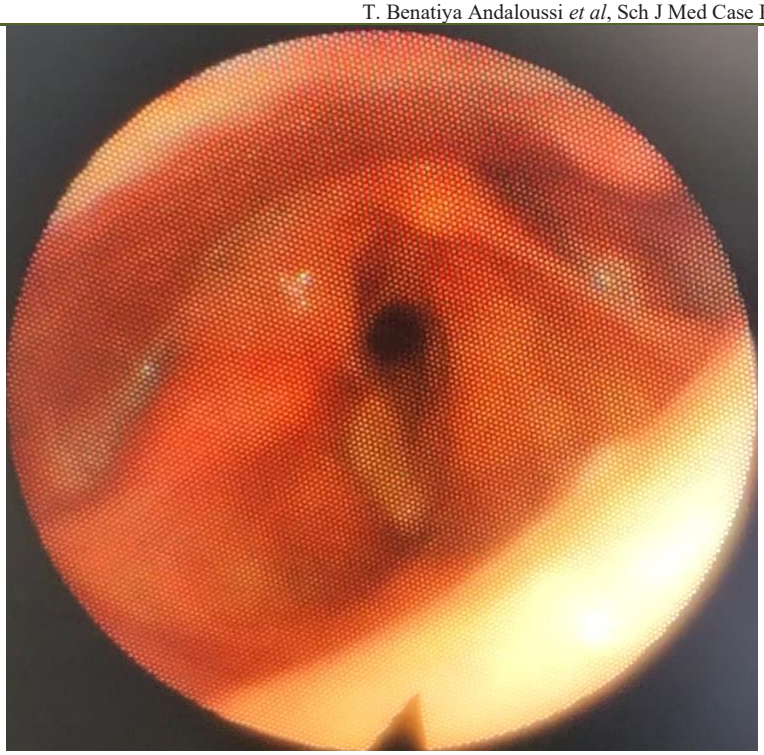

Figure 6: Nasofibros copy shows complete regression of the tumoral process, with persistent right vocal cord palsy in the paramedial position

The tracheostomy was successfully closed, and the patient tolerated room air well. He is currently on maintenance chemotherapy with bortezomib, showing good clinical and biological progression.

DISCUSSION

Solitary plasmacytoma is a rare malignant tumor characterized by the monoclonal proliferation of plasma cells [1]. It is most commonly classified as an extramedullary plasmacytoma (EMP) and primarily affects individuals between the ages of 40 and 70. EMP accounts for approximately 3% of all plasma cell neoplasms and less than 1% of head and neck tumors, with a significantly higher prevalence in men [5-7].

Although EMP typically achieves local control with treatment, there is a risk of progression to multiple myeloma (MM), particularly in cases with poorly differentiated tumor cells. Studies indicate that nearly one-third of EMP cases evolve into MM within two years [2].

Multiple myeloma (MM) is the most prevalent plasma cell dyscrasia, with a variable prognosis. It has an incidence of approximately four cases per 100,000 people and represents 1% of all malignancies [1,8]. The most critical prognostic factor in plasma cell neoplasms is the risk of progression.

MM, sometimes referred to in the literature as "metastatic" MM when presenting as a de novo laryngeal mass, is exceedingly rare, with only a few reported cases [9,10]. Distinguishing between an extramedullary manifestation of MM and a primary EMP is crucial, as it directly influences treatment strategies and prognosis. The diagnosis of MM requires at least 10% clonal bone marrow plasmacytosis along with evidence of end-organ damage attributable to myeloma, such as hypercalcemia, renal insufficiency, anemia, or bone lesions [11]. In our patient, the presence of osteolytic bone lesions and 15% plasma cells in the bone marrow smear favored the diagnosis of an extramedullary focus of MM rather than a primary EMP.

The management of plasma cell neoplasms typically involves radiotherapy, chemotherapy, or surgery. Given their high radiosensitivity, the standard treatment for EMP is radiotherapy [9], which achieves local control rates of 80–100% when administered at doses of at least 40 Gy. Surgery may be considered in cases of large tumors or extensive bone destruction; however, in laryngeal tumors, surgical intervention is often limited due to the need to preserve laryngeal function. Thus, radiotherapy remains the preferred treatment approach in such cases.

Conversely, multiple myeloma is considered a systemic disease requiring chemotherapy and, in selected cases, bone marrow transplantation [9,11]. Our patient was treated with chemotherapy and underwent a bone marrow transplant, achieving a favorable response and disease stabilization. He continues to do well approximately eight months after completing therapy.

Long-term follow-up is essential for patients with EMP due to the potential for MM progression.

Diagnostic evaluation should include deep tissue biopsy, immunophenotyping, and imaging techniques such as CT or MRI to assess local extension and detect occult lesions. Among plasma cell neoplasms, EMP generally has the most favorable prognosis, with a five-year survival rate ranging from 31% to 82%. However, continuous monitoring is crucial, as early detection of progression allows for timely therapeutic intervention, improving overall outcomes.

This case underscores the importance of considering EMP in patients presenting with laryngeal submucosal masses and highlights the necessity of deep biopsies to avoid false-negative diagnoses. Systematic evaluation and long-term follow-up remain critical for early detection of MM transformation and optimizing treatment strategies.

CONCLUSION

This case of extramedullary plasmacytoma evolving into multiple myeloma highlights the importance of early recognition and thorough diagnostic evaluation of laryngeal masses. While solitary plasmacytomas can be successfully managed with local therapy, their potential for progression to multiple myeloma necessitates long-term follow-up. In our patient, the initial presentation as a laryngeal mass eventually led to the diagnosis of systemic disease, underscoring the need for a multidisciplinary approach in plasma cell neoplasms. Early intervention and tailored treatment, including chemotherapy and autologous stem cell transplantation, can significantly improve patient outcomes. Vigilant monitoring remains crucial for detecting disease progression and optimizing long-term management strategies.

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