Clinical Case Report

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Extramedullary plasmacytoma of the trachea

A case report and review of the literature

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Abstract

Rationale: Extramedullary plasmacytomas (EMPs) are rare solitary soft tissue tumors characterized by monoclonal proliferation of plasma cells. Most lesions occur in the head and neck, but primary tracheal lesions are very rare.

Patient Concerns: In this report, we describe a case of tracheal EMP discovered in a 48-year-old man who presented with a history of progressive dyspnea.

Diagnoses: Computed tomography (CT) revealed a well-defined nodular mass in the posterior wall of trachea without signs of invasion of the tracheal walls. Then, a reddish mass occluding approximately 90% of the trachea was evidenced by bronchoscopic examination.

Interventions: The patient was treated with surgery followed by adjuvant radiotherapy to achieve better local control.

Outcomes: After the surgery, there was immediate symptomatic relief. There was no recurrence or metastasis during a 6-month follow-up.

Lessons: This study presents a rare case of tracheal EMP occluding approximately 90% of the lumen that was successfully managed by surgery followed by radiotherapy.

Abbreviations: EMP = extramedullary plasmacytoma, CT = computed tomography.

Keywords: case report, extramedullary plasmacytomas, trachea

1. Introduction

Plasma cell tumors consist of multiple myeloma (MM), extramedullary plasmacytoma (EMP), and solitary bone plasmacytoma (SBP). EMP is an extremely rare and discrete solitary mass of neoplastic monoclonal plasma cells, which accounts for approximately 3% of all plasma cell tumors.[1] It has been previously reported that almost 80% of EMPs are localized in the head and neck region, occasionally localized to the gastrointestinal tract, lungs, breasts, testes, and skin.[2] EMP of the trachea is an extremely rare entity. In this case report, we present a case of a male patient with EMP of the trachea who suffered from severe dyspnea but was treated successfully by surgery and radiotherapy.

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Both XZ and LS contributed equally to this work.

Informed consent of the patient has been obtained.

The authors report no conflicts of interest.

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2. Case history

A 48-year-old male was referred to pneumology department of the affiliated hospital of Hebei university due to severe dyspnea that had worsened continuously over a 2-month period. CT revealed a well-defined nodular mass in the posterior wall of trachea causing obstruction, without signs of invasion of the tracheal walls (Fig. 1). Flexible bronchoscopic examination was done, which evidenced a mass arising from posterior wall of trachea 7 cm below glottis occluding approximately 90% of the lumen (Fig. 2A). Surgical treatment was arranged and the tumor was excised almost completely by bronchoscopic electrocautery snare combined with CO_2 cryotherapy (Fig. 2B). After these procedures, there was immediate symptomatic relief. The histopathological study identified a diffuse infiltrate of neoplastic monoclonal well differentiated plasma cells (Fig. 3). The immunohistochemical profile demonstrated plasma cells express CD138, a marker characteristically positive for plasma cells (Fig. 4). Laboratory examinations, such as blood counts, protein electrophoresis, and serum chemistry, were within the normal range. Urine was negative for Bence Jones Protein. Bone marrow aspirate and bone marrow biopsy showed normal morphology. Further X-ray of the spine, pelvis femurs, humerus were performed after surgical excision, and no lytic bone lesion was observed, which eliminates MM. In the context of the clinical status and test results, a diagnosis of solitary EMP was made. The patient then had adjuvant radiotherapy at a dose of 50 Gy in 25 fractions on the area of tumor bed and surrounding area. The patient remains well, with no evidence of recurrence 6 months after treatment.

3. Discussion

Plasma cell neoplasms, characterized by neoplastic proliferation of single clone of plasma cells producing monoclonal

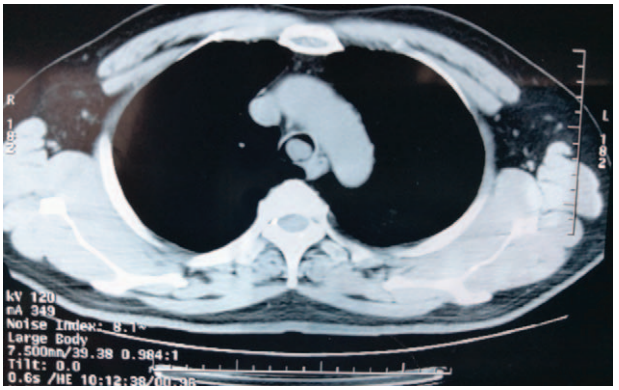

Figure 1. CT revealing a mass occupying almost the entire trachea space. CT = computed tomography.

immunoglobulins, can present as solitary (plasmacytoma) or multiple (MM) lesions. MM can present as multiple bony lesions, plasma cell infiltration of the bone marrow, and abnormal proliferation of monoclonal immunoglobulin in the blood, and damage multiple organ systems and lead to organ failure.[3] Solitary lesions, occurring in the bone, are called plasmacytoma of bone, which most frequently occurs in the axial skeleton, such as a vertebra.[4] When solitary lesions occur in the soft tissues, it is called EMP. Both of SBP and EMP may disseminate to MM but are distinguished by the presence of a single radiologic lesion and normal bone marrow.[1,4]

EMP is most commonly found in the head and neck region such as nasal cavity, paranasal sinuses, and oronasopharynx.[5] Rarely EMP can be located in trachea. Tracheal tumors are such uncommon and constitute around 1% to 2% of all respiratory tract tumors.[6] Squamous cell carcinoma and adenoid cystic carcinoma are the malignant tumors most common affecting the trachea.[7] EMP of the trachea, primarily occurring in 50-to-60-year-old male patients, represents around 3% of malignant tracheal tumors.[7,8] The pathogenesis of tracheal EMP is still unclear, but viral pathogenesis and chronic irritation may contribute to those lesions.[9] Airway obstruction is the most

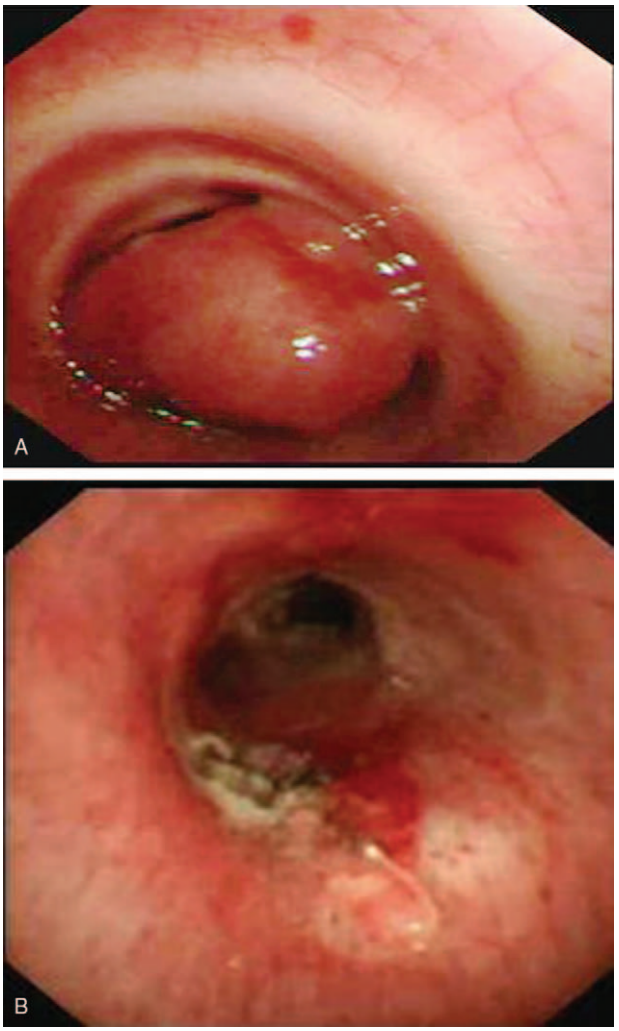

Figure 2. (A) Bronchoscopy shows an obstructive and fleshy tracheal mass markedly narrow the lumen. (B) Bronchoscopy shows that the tumor was excised almost completely.

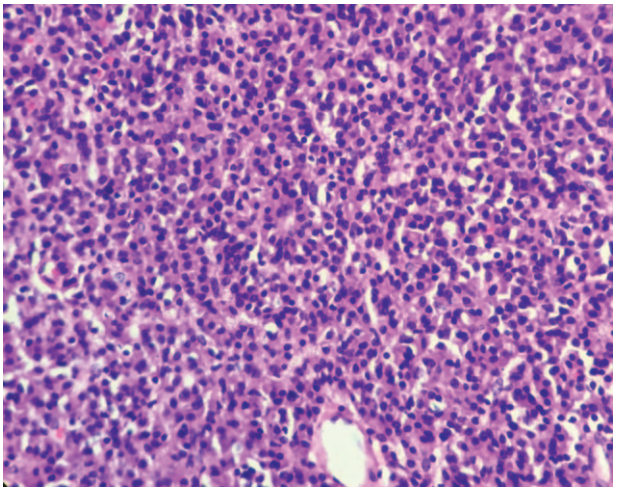

Figure 3. Photomicrograph of surgical specimen. Diffuse infiltrate of neoplastic monoclonal well-differentiated plasma cells is present. (H and E, 200).

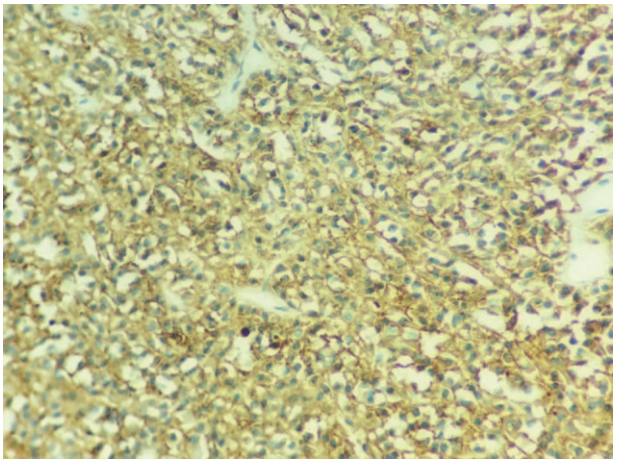

Figure 4. Neoplastic cells showed diffuse membrane positivity for CD138 at immunohistochemical staining. (IHC, ×200).

common symptoms of tracheal tumors. Clinically, in the early stages of development, patients may present with nonspecific symptoms such as chronic cough, dysphonia, hoarseness, hemoptysis, stridor, wheezing, and dyspnea, but dyspnea becoming evident when the narrowing of the airway is over 75%.[10]

CT scan is an essential diagnostic approach, which allows the lumen, airway wall, and mediastinal structures to be evaluated. Multiplanar reconstructions can be applied to evaluate the type, degree, and longitudinal extent of the airway narrowing as well as the location of the tumor and its distance from the cricoid cartilage to the carina.[6,11] Bronchoscopy can show an endoluminal view of the tumor.[6,11] In this case, the stenosis-to-lumen ratio was estimated at around 90% by both techniques. However, as there is lack of distinguishing clinical and radiological features of EMP, the final diagnosis is made through histological and immunohistochemical features, which demonstrate that the tumor is composed of neoplastic monoclonal plasma cells intensely positive for CD138 and showed kappa light chain restriction.[12] In addition, the diagnosis of EMP of the soft tissue has been based on the following criteria: pathological tissue evidence of monoclonal plasma cells involving a single extramedullary site; no bone marrow involvement; negative skeletal survey results; no anemia, hypercalcemia, or renal impairment caused by plasma cell dyscrasia; and low serum or urinary levels of monoclonal immunoglobulin.[13]

The current treatment options of tracheal plasmacytomas remain controversial including surgery and/or radiation therapy. Surgery is an effective method for treating trachea EMP, but that radical excision is often difficult because of the size of the tumor and the proximity of vital organs. EMP respond well to radiation therapy, with a local control rate of 90% to 100%. A radiation dose in the range 40 to 50 Gy delivered to the primary site of the EMP is usually recommended.[13] Moreover, Sasaki et al[14] found that radiotherapy combined with surgery produced an improved survival rates in the treatment of EMPs of the head and neck. Furthermore, the role of adjuvant chemotherapy in the treatment of EMP has not yet been established. Katodritou et al[15] has reported a case of extramedullary gastric plasmacytoma successfully managed with the combination of bortezomib and dexamethasone. In addition, Fukuhara et al[16] have reported a case of advanced gastric plasmacytoma using adjuvant chemotherapy involving bortezomib and autologous peripheral blood stem-cell transplantation after the resection. Lymph node metastasis and a larger primary tumor were reported as 2 independent poor prognostic factors for overall survival and disease-free survival.[17] The 5-year survival rates are between 30% and 82% in EMP. [18] In this case, the patient was treated successfully by bronchoscopic electrocautery snare combined with CO_2 cryotherapy followed by radiotherapy. There has been no recurrence or metastasis after a 6-month follow-up.

4. Conclusion

The novelty of this case was that the narrowing of the airway is severe, the tumor almost obstructing 90% of the lumen. Symptom resolution was observed after the resection. As the presence of positive and/or close margins, this patient received adjuvant radiotherapy. Even if the prognosis of EMP is relatively favourable compared with that of solitary plasmacytoma of bone or MM, a local recurrence or a progression to MM has been described in up to 20% of cases.[19] A long-term follow-up of radiological and electrophoresis assessment is recommended.

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