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A Case Report of Tracheal Extramedullary Plasmacytoma

Kun Wang
Yunchao Huang
Anning Chen

Department of Cardiothoracic Surgery, Third Hospital of Kunming Medical College, Kunming 650118, Yunnan Province, China.

Correspondence to: Yunchao Huang
Tel: 86-871-821 6465
E-mail: huangych2001@yahoo.com.cn

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CJCO http://www.cjco.cn
E-mail: 2008cocr@gmail.com
Tel (Fax): 86-22-2352 2919

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Introduction

Extramedullary plasmacytoma (EMP) is an uncommon tumor that often develops outside the bone and arises from clonal proliferation of atypical plasma cells before EMP is diagnosed. Multiple myeloma (MM) must be excluded by performing laboratory tests such as serum protein electrophoresis, bone marrow biopsy and skeletal imaging examinations. A bone marrow biopsy should show no evidence of multiple myeloma, and less than 3% of plasma cells. Monoclonal bands of serum protein and Bence-Jones protein in the urine can sometimes be detected. EMP can involve any extraosseous organs, but it predominantly affects the head and neck areas. Any extra-osseous organ may also be involved[1,2]. Tracheal involvement is a rare finding. Only a few cases of primary tracheal extramedullary plasmcytoma have been reported[2-4], Here we present a rare case of truly localized tracheal extramedullary plasmacytoma without evidence of myeloma elsewhere.

Case Report

A 38-year-old male patient was admitted to our hospital in February 2006, with symptoms of cough and hemoptysis for over 1 month. The patient had exertional dyspnea during the last two weeks. He had received an X-ray examination at a local hospital before his hospitalization, resulting in a normal chest radiograph. Thoracic CT showed a mass in the left lateral wall of the trachea (Fig.1). Tracheoscopy confirmed a movable neoplasma with a pedicle from the right lateral tracheal wall, 6~7 cm below the glottis. A bronchoscopic punch biopsy was conducted with the pathologic evidence revealing extramedullary plasmacytoma (EMP). The cytologic findings of bone marrow aspiration were normal. There was no pathologic finding on SPECT examination of the cranium, pelvis, vertebral column, or the extremities. The results were normal for both serum and urine electrophoresis performed to search for the presence of monoclonal immunoglobulin or immunoglobulin fragments. Complete blood count, blood biochemistry, and urinalysis results were within the normal limits.

After all these investigations, the diagnosis of multiple myeloma was excluded, and solitary EMP was confirmed. The lesion was surgically excised after which the histopathologic findings reported the specimens to be a colloidal goiter and the tracheal mass as EMP without lymph node invasion. He was not treated with radiotherapy, and there has been no recurrence after a six-month follow-up.

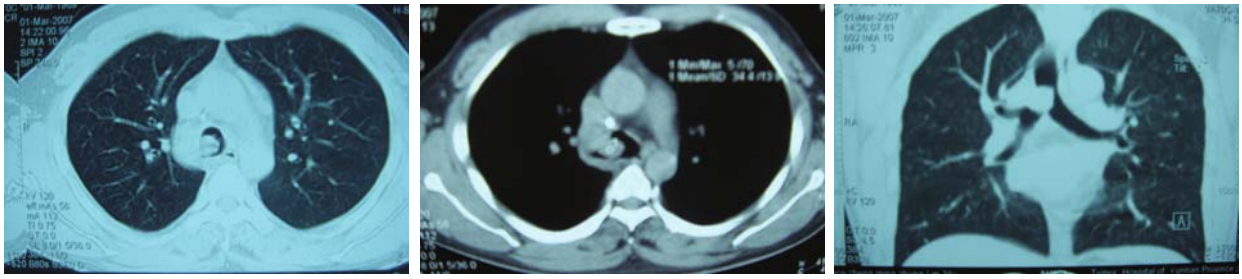

Fig.1. CT image of the patient on admission.

Discussion

Primary tracheal tumors are rare, and the trachea is an exceedingly rare site of extramedullary plasmacytoma. Plasmacytoma is an immunoproliferative, monoclonal disease of the B-cell line which is classified as a B-cell neoplasm by the World Health Organization. It originates as a clone of malignant, transformed plasma cells.

The plasma cell is a highly mature B-cell form that is capable of immunoglobin synthesis and secretion. This is the origin for monoclonal plasma cell foci located outside the bone marrow and, without a generalized plasma cell disorder such as myelomatosis or plasma cell leukemia, it is called EMP. Of the reported EMP cases, 80% were in the upper aerodigestive (UAD) tract. Male patients have EMP in the UAD region 3 times more commonly than female patients. Most cases of EMP in the UAD area occur between the fifth and sixth decades, and arise in the mucosa of the head, or neck region, especially in the upper respiratory tract, including the nasal cavity, the paranasal sinuses, and the oronasopharynx[1-4]. Rare cases of primary EMPs have been described in the larynx, hypopharynx, glands of the UAD tract (parotid gland, submandibular gland, thyroid, and mandibular region), trachea, esophagus, cervical lymph nodes, middle ear, and mastoid. Non-UAD regions for EMPs include gastrointestinal and urogenital tracts, breast, conjunctiva, musculature, retroperitoneum, skin, lung parenchyma, bronchus and pleura, and pituitary gland[5,6].

For tracheal tumors, especially with an exfoliated pedicle, intubation anesthesia is one of the most important risk factors. As in this case, when the tumor was located near the glottis, exfoliation of the tumor could cause asphyxia. Considering the tracheal resection, both through the mouth and operating area intubation are necessary. In this case, we performed a right thoracotomy, disassociated the trachea, incised the trachea, and intubated a duct to the left main bronchus. All these skills provided a safe air way to ensure the complete resection and anastomosis of the tumor.

For the patients who have lost the opportunity of an operation, the current treatment of choice is radiotherapy because these tumors are highly radiosensitive. A radiation dose of 4,000 cGy to 5,000 cGy over 4 to 5 weeks was associated with a less than 5% risk of local recurrence. No local recurrences were observed with radiation doses of at least 4,500 cGy[7,8]. For extramedullary plasmacytomas at sites other than head and neck, if possible complete surgical removal should be considered[7]. The diagnosis of patients with extramedullary plasmacytomas is usually made after surgical excision. It is recommended that patients with positive surgical margins should receive adjuvant radiotherapy, but adjuvant radiotherapy is not recommended for patients who have undergone complete surgical excision with a negative margin. There is no evidence that adjuvant chemotherapy improves the patient outcome[7].

As a result, extramedullary plasmacytoma has a relatively favorable prognosis, and a solitary extramedullaty plasmacytoma can be a curable disease by meticulous staging. At least 70% of these patients remain disease-free at 10 years. Among the cases treated for EMP in the UAD tract; 61% of the patients had no MM or recurrence; however, in 22% there was a recurrence, and in 16% there was progression to MM., In conclusion, the annual follow-up is very important for extramedullary plasmacytoma patients[4,9].

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