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CASE REPORT

Non-secretory multiple myeloma expressed as multiple extramedullary plasmacytoma with an endobronchial lesion mimicking metastatic cancer: A case report

Seul Bi Lee, Chi Young Park, Hee Jeong Lee, Ran Hong, Woo Shin Kim, Sang-Gon Park

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Seul Bi Lee, Chi Young Park, Hee Jeong Lee, Sang-Gon Park, Department of Hemato-oncology, Chosun University Hospital, Gwangju 501-717, South Korea

Ran Hong, Department of Pathology, Chosun University Hospital, Gwangju 501-717, South Korea

Woo Shin Kim, Department of Laboratory Medicine, Chosun University Hospital, Gwangju 501-717, South Korea

Corresponding author: Sang-Gon Park, MD, Assistant Professor, Department of hemato-oncology, Chosun Univ Hosp, 365 Pilmun-daero, Dong-gu, Gwangju 501-717, South Korea. sgpark@chosun.ac.kr

Abstract

BACKGROUND

Non-secretory multiple myeloma (MM) is a rare condition that accounts for only 3% of MM cases and is defined by normal serum and urine immunofixation and a normal serum free light chain ratio. Non-secretory MM with multiple extramedullary plasmacytomas derived from endobronchial lesions is extremely rare and can be misdiagnosed as metastasis of solid cancer.

CASE SUMMARY

A 36-year-old man presented with progressive facial swelling and nasal congestion with cough. Various imaging studies revealed an endobronchial mass in the left bronchus and a large left maxillary mass with multiple destructive bone metastatic lesions. He initially presented with lung cancer and multiple metastases. However, pathologic reports showed multiple extramedullary plasmacytomas in the left maxilla and the left bronchus. There was no change in the serum and urine monoclonal protein levels, and no abnormalities were observed in laboratory examinations, including hemoglobin, calcium, and creatinine levels. The bone marrow was hypercellular, with 13.49% plasma cells. The patient was diagnosed with non-secretory MM expressed as multiple extramedullary plasmacytomas with endobronchial lesions in a rare location. Radiation therapy for symptomatic lesions with high-dose dexamethasone was started, and the size of the left maxillary sinus lesion dramatically decreased. In the future, chemotherapy will be administered to control lesions in other areas.

CONCLUSION
We present a rare case of non-secretory MM with multiple extramedullary plasmacytoma with an endobronchial lesion.

Key Words: Maxillary mass lesion; Destructive bone metastatic lesion; Multiple extramedullary plasmacytoma; Endobronchial lesion; Non-secretory multiple myeloma; Case report

Core Tip: Endobronchial and maxillary mass lesions without abnormality on laboratory examination are easily misdiagnosed as metastases of primary lung cancer or other head and neck malignancies. Histopathological studies are required to avoid erroneous diagnoses.

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INTRODUCTION

Multiple myeloma (MM) is a mature B cell neoplasm that accounts for 10% of all hematologic malignancies and is defined by the presence of ≥ 10% of clonal plasma cells in the bone marrow or biopsy-proven extramedullary plasmacytoma and the presence of related tissue or organ damage[1]. Symptomatic MM is defined by the presence of a monoclonal protein in the serum or urine, plasma cells in the bone marrow (at least 10%), and presence of related organ disorders (hypercalcemia, renal insufficiency, anemia, and bone lesions)[1,2]. MM is primarily observed in older patients and considered difficult to treat[3]. Over the past decade, the median survival of patients with myeloma has increased with the development of therapeutic agents, including immunomodulatory drugs (thalidomide and lenalidomide) and proteasome inhibitors (bortezomib). High-dose therapy followed by autologous stem cell transplantation (ASCT) has also contributed to this improvement in the survival rate[4].

In most patients, plasma cell proliferation is restricted to the bone marrow. However, in some cases, extramedullary plasma cell proliferation is also observed in other tissues, such as the nasal cavity, lung, and pleura[5]. Endobronchial plasmacytoma has been reported in several cases. Although exceedingly rare, according to published reports, most cases were reported in solitary lesions or advanced MM[6]. Here, we report the first diagnosed case of multiple extramedullary plasmacytoma with endobronchial lesions in non-secretory MM.

CASE PRESENTATION

Chief complaints

A 36-year-old man presented to our hospital for evaluation of progressively worsening facial swelling and nasal congestion. He was initially diagnosed with a nasal polyp with sinusitis and underwent polyp removal with antibiotic therapy.

History of present illness

The patient visited our hospital for further evaluation after redeveloping facial swelling and nasal congestion, this time accompanied by a gradually worsening cough with sputum and blurred vision in the left eye.

History of past illness

The patient had no previous medical history.

Personal and family history

The patient is a non-alcoholic and non-smoker. He has no family history.

Physical examination

The patient showed painful facial swelling and blurred vision. The patient's respiratory rate was 22 breaths per minute, blood pressure was p (B) = 15.99/10.7 kPa, and oxygen saturation in room air was 90%.

Laboratory examinations

Based on laboratory findings, the hemoglobin level was 11.9 g/dL (normal range, 12-16 g/dL), and the creatinine level was 0.7 mg/dL (normal range, 0.5-1.3 mg/dL). Monoclonal proteins could not be detected by serum and urine protein immunofixation electrophoresis. The albumin level was 3.77 g/dL (normal range, 3.5-5.5 g/dL) and beta-2-microglobulin level was 4.2 mg/L (normal range, 0.0-2.4 mg/L). The lactate dehydrogenase level was 271 U/L (normal range, 125-220 U/L).

Imaging examinations

Computed tomography of the neck and thorax revealed a solid mass occupying the left maxillary sinus and an endobronchial lesion in the left main bronchus (Figure 1).

Positron emission tomography/computed tomography revealed a hypermetabolic mass in the left maxillary sinus extending to the left ethmoid sinus and nasal cavity and multiple hypermetabolic metastatic nodules in both the cervical and left supraclavicular areas. Multiple hypermetabolic osseous metastases had spread to the sternum, ribs, right scapula, right humerus, thoraxic and lumbar spines, pelvic bone, and left femur. Focal hypermetabolic nodular lesions in the left main bronchus were also observed (Figure 1). Primary lung cancer with multiple bone metastases was initially suspected, but double primary lung cancer with maxillary sinus cancer was excluded. We immediately performed a pathologic examination of the maxillary sinus mass and the endobronchial mass using bronchoscopy. Bronchoscopic findings showed a 1.5-cm protruding mass with pedicles arising from the anterior wall of the left proximal main bronchus (Figure 1), which was suspected to be primary lung cancer. Initially, the maxillary mass had pathologic findings of monomorphic plasmacytoid cytoplasm (positive for CD138, kappa light chain, and negative for CD3, CD20, and lambda light chain) (Figure 2), and it was subsequently diagnosed as a plasmacytoma, which was confirmed by the bronchoscopic biopsy result (Figure 2).

Further diagnostic workup

The patient was referred to the hematology department to undergo an evaluation for systemic MM. Biochemical tests revealed normal calcium and creatinine levels, and serum and urine immunofixation were negative for monoclonal proteins. However, the bone marrow biopsy from the iliac crest showed hypercellularity for his age with diffusely infiltrated plasma cells (13.49%) (Figure 3). Although the patient had no anemia and the serum creatinine levels were normal, a diagnosis of non-secretory MM was considered based on the bone marrow biopsy findings and the multiple lesions, including confirmation of the maxillary sinus and endobronchial lesions as plasmacytoma.

FINAL DIAGNOSIS

The patient was finally diagnosed with non-secretory MM, expressed as multiple extramedullary plasmacytomas with an endobronchial lesion. The International Staging System stage at the time of diagnosis was II.

TREATMENT

The left maxillary sinus mass extended to the nasopharynx and left ethmoid sinus, which had caused severe facial edema and blurred vision in the left eye. Moreover, endobronchial lesions also caused severe respiratory distress symptoms. Therefore, we started high-dose steroid therapy with dexamethasone 40 mg for 4 days. Palliative radiation therapy of the left maxillary sinus lesion was performed simultaneously. After steroid administration, facial edema dramatically decreased, and respiratory distress symptoms improved (Figure 4). We continued radiation therapy on the symptomatic lesions to a total dose of 15 Gy.

OUTCOME AND FOLLOW-UP

Finally, the facial mass and symptoms almost regressed, and the patient will subsequently undergo chemotherapy with bortezomib, thalidomide, and dexamethasone, followed by ASCT.

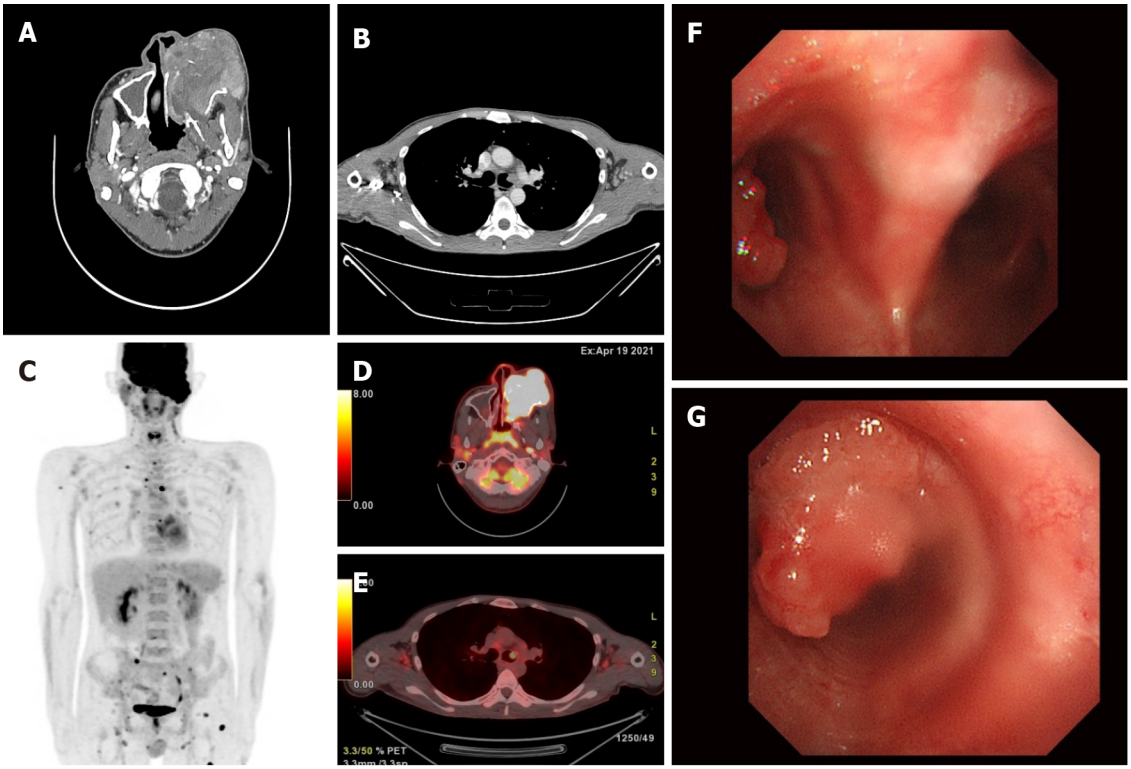

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Figure 1 Imaging at admission. A: Contrast-enhanced neck computed tomography shows a bulky mass in the left maxillary sinus extending to the orbit, nasal cavity, ethmoid sinus, infratemporal fossa, and pterygopalatine fossa; bone destruction extends to the nasal cavity; B: Contrast-enhanced chest computed tomography shows an enhanced nodule approximately 0.8 cm in size in the left main bronchus; C-E: 18F-fluorodeoxyglucose positron emission/computed tomography shows a large expansile hypermetabolic mass in the left maxillary sinus and hypermetabolic focal activity in the nasopharynx, multiple metastatic lymphadenopathies in both cervical and left supraclavicular areas, and multiple osseous metastases. There is a focal hypermetabolic nodular lesion in the left main bronchus; F and G: Bronchoscopy shows a 1.0-cm sized nodular lesion with pedicles arising from the anterior wall of the left main bronchus.

DISCUSSION

In the initial stage of diagnosis, our patient was strongly considered as having primary lung cancer with multiple bone metastases or double primary lung cancer with maxillary sinus cancer. Our patient's laboratory tests showed normal results. For this reason, we excluded the possibility of plasmacytoma or non-secretory MM. However, the biopsy confirmed an extramedullary plasmacytoma. In addition, bone marrow examination showed more than 10% plasma cell infiltration without alterations in serum or urine paraprotein and immunoglobin subtype. Thus, we diagnosed the patient with non-secretory MM based on bone marrow examination and biopsy results.

Extramedullary plasmacytoma is a variant of a plasma cell tumor involving organs outside the bone marrow without any sign of systemic involvement (primary solitary plasmacytoma) or secondary to MM[7]. The differential diagnosis of plasma cell dyscrasias is vital because these diseases may exhibit diverse clinical courses and prognoses. Extramedullary plasmacytoma is most often located in the upper respiratory tract and nasopharynx, and involvement of the lower respiratory tract is rarely observed[8]. Endobronchial plasmacytoma is a rare manifestation of extramedullary plasmacytoma[6], with very few cases reported in the literature. Most endobronchial plasmacytoma cases were solitary plasmacytomas with no systemic involvement of MM[9-12]. Our patient showed systemic involvement of a plasma cell malignancy.

Extramedullary plasmacytoma is associated with adverse prognoses in patients with newly diagnosed and relapsing MM[13]. Almost all patients show multiple extramedullary plasmacytomas as the terminal event of their MM[13,14], whereas this patient showed multiple extramedullary plasmacytomas at the initial diagnosis of MM.

Non-secretory MM is a rare variant that accounts for 1%-5% of all cases of MM. It is characterized by the absence of monoclonal gammopathy in the serum and urine[15]. In this case, monoclonal gammopathy was not observed, and there was no organ dysfunction. Due to the inability to detect monoclonal proteins, it is difficult to establish an accurate diagnosis, and misdiagnosis of this condition as a solitary plasmacytoma delays systemic treatment[16,17].

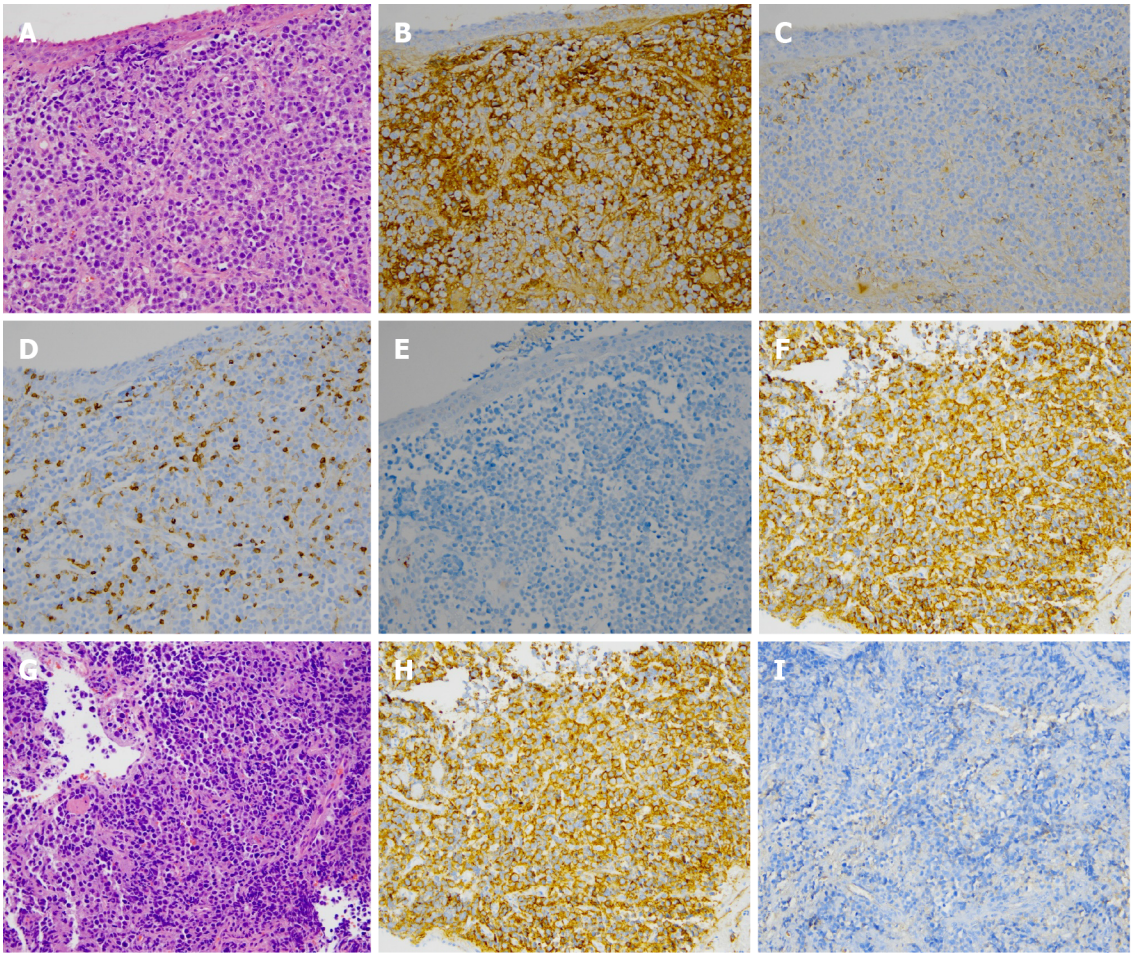

Figure 2 Microscopic examination of the specimen using hematoxylin and eosin staining and immunohistochemistry staining. A-F: In the bronchus, plasmacytoid large atypical cells are densely infiltered beneath the surface epithelium. These cells are immunoreactive for kappa-light chain (B) and CD138 (F) but not for lambda-light chain (C), CD3 (D), and CD20 (E); G-I: The lesion in the nasal cavity also shows densely packed plasmacytoid cells and is positive for kappa-light chain (H) and negative for lambda light chain (I).

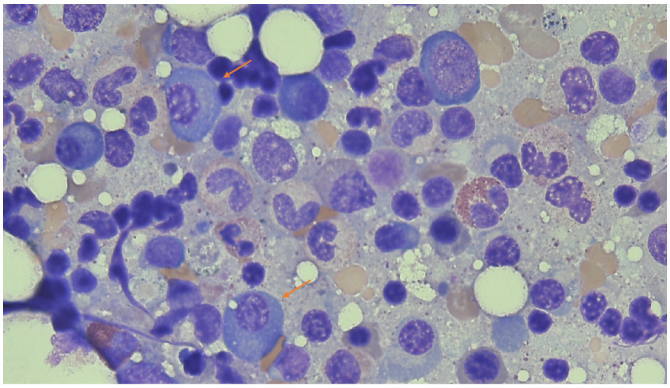

Figure 3 Bone marrow biopsy. Plasma cells are present in the biopsy. Arrow: Plasma cell.

CONCLUSION

Initially, this patient was diagnosed with primary lung cancer with multiple metastases because there was no reversal of the A/G ratio or increase in serum monoclonal protein levels. However, bone marrow and tissue biopsy results showed systemic involvement of MM. Thus, we present a case of non-secretory MM expressed as multiple extramedullary plasmacytoma with an endobronchial lesion.

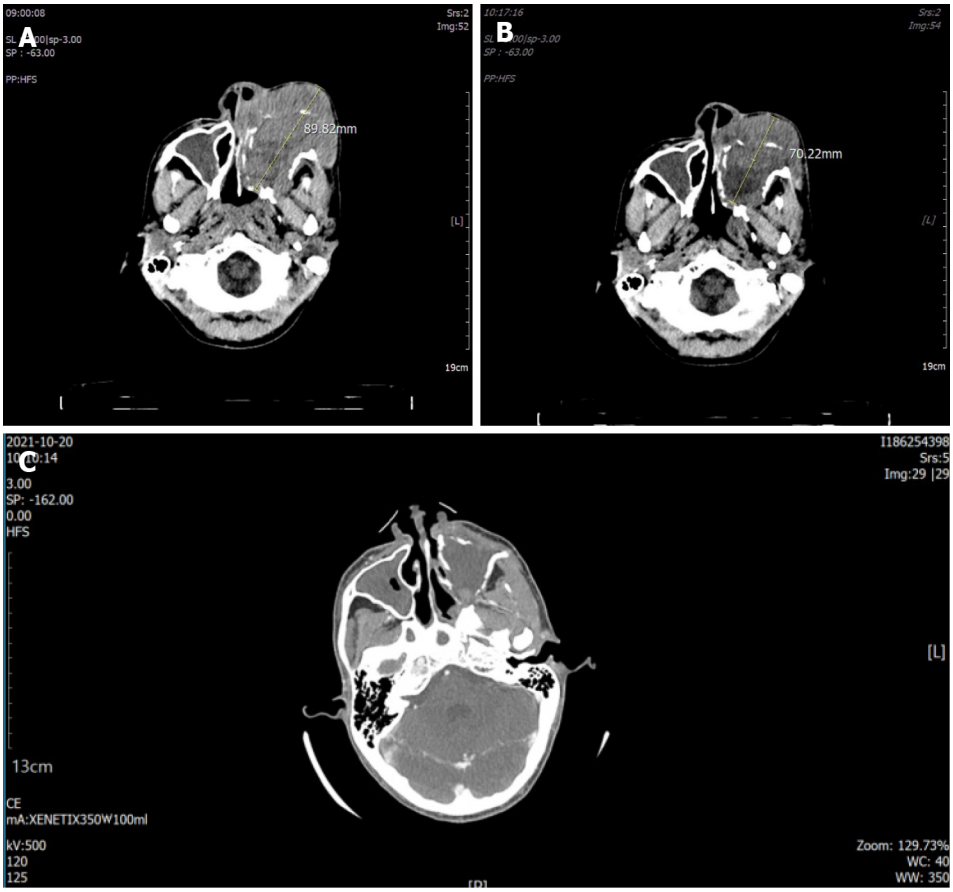

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Figure 4 Imaging after radiotherapy. A: Before radiotherapy; B: During radiotherapy; and C: After completion of radiotherapy, bulky mass of the left maxillary sinus decreased after radiotherapy.

Such cases are extremely rare and can be easily misdiagnosed as solid cancers of the upper respiratory tract until histologic confirmation. These clinical situations are extraordinarily heterogeneous, and care must be taken before making a diagnosis. These cases should be considered as having high-risk myeloma systemic involvement and treated appropriately.

FOOTNOTES

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ORCID number: Seul Bi Lee 0000-0001-8086-4631; Chi Young Park 0000-0001-5216-7257; Hee Jeong Lee 0000-0001-8295-

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