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Asymptomatic patient with "lumpy and bumpy" airways. A case of pulmonary MALToma

Abstract

Primary pulmonary lymphoma is a rare disease. The most frequent primary pulmonary lymphoma (PPL) is an extranodal marginal zone B-cell lymphoma of MALT. Approximately half of the patients are asymptomatic at diagnosis. In this article, we report a case of a 62-year-old male with benign prostatic hyperplasia (BPH) that was referred to us for a preoperative assessment. He had no respiratory complaints but, on evaluation, was detected to have a pulmonary MALToma. Our case highlights the fact that pulmonary MALTomas can present as lumpy and bumpy airways and also aims to showcase the importance of tissue diagnosis.

Key words: lymphoma, MALToma, cobble stone trachea

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Introduction

While gastric maltoma is a relatively well known entity, pulmonary MALToma is rare. It may not cause any symptoms and may be detected inadvertently when the patient is being evaluated for some other disease. Radiological findings are also non-specific [1]. We report a case of a 62-year-old male referred to us for preoperative assessment for benign prostatic hyperplasia (BPH), who, upon evaluation, was detected to have pulmonary MALToma. Radiologically, he had nodular lesions with peribronchovascular predominance. Fiberoptic bronchoscopy revealed multiple diffuse nodules along the wall of trachea, the right and left main bronchus, and in the deeper airways. Endobronchial and transbronchial biopsy confirmed the diagnosis. Our case is interesting because pulmonary MALTomas very rarely manifest as lumpy and bumpy airways or contain multiple, tiny nodules seen on fiberoptic bronchoscopy.

Case report

A 62-year-old male was referred to a chest clinic for preoperative assessment in preparation for surgery for BPH. He denied any history of fever, cough, shortness of breath, haemoptysis, or chest pain. The patient had been diagnosed with diabetes mellitus for 10 years which was well controlled on oral hypoglycemic drugs. He never smoked or consumed alcohol. On examination, digital clubbing was present. There was no pallor, icterus, cyanosis, edema, or lymphadenopathy. His vitals were as follows: BP was 120/70 mm Hg; pulse rate — 86/min; respiratory rate — 18/min; temp. — 98.6°F; and SpO_2 on room air — 95%. On auscultation of the chest, end inspiratory crepitations were present in bilateral infra-scapular areas. His hemoglobin was 15 gm/dL, total leucocyte count was 7800/cmm, and his liver and renal function tests were within normal limits. A chest radiograph revealed bilateral multiple small nodular opacities in the mid and

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lower lung zones. High resolution computed tomography (HRCT) of the thorax showed multiple nodular lesions of varying size predominantly in the bilateral lower lobes with peribronchovascular predominance. Narrowing of the right main bronchus with nodular bronchial wall thickening was also seen (Figure 1, 2).

Flexible fiberoptic bronchoscopy showed multiple diffuse nodules along the wall of the trachea, right and left main bronchus, and in the deeper airways (Figure 3). Bronchial and trans-bronchial lung biopsies were taken and sent for histopathological examination. As sarcoidosis was in the differential diagnosis, serum calcium, 24-hour urinary calcium, and angiotensin converting enzyme levels were sent for evaluation. Serum calcium was 9.5 mg/dL, 24-hour urinary calcium was 150 mg, and the ACE level was 50 U/L. An endobronchial biopsy revealed tissue lined by bronchial mucosa (pseudostratified columnar epithelium). Submucosal tissue showed infiltration with small to medium sized atypical lymphoid cells, centrocyte-like cells, lymphoepithelial lesions and Dutcher bodies. Transbronchial lung biopsy showed similar atypical lymphoid cells infiltrating into the lung parenchyma. IHC was positive for CD20, CD3, CD43, CD138, and Bcl2. Lymphoepithelial lesions were CK-positive. CD5 and CD10 were negative. No light chain restriction was present (Figures 4–6). Based on histopathological examination and immunophenotypical features, a diagnosis of extra nodal marginal zone B cell lymphoma of mucosa associated lymphoid tissue (Pulmonary MALToma) was made and the patient was started on the R-CHOP regimen. He had received 3 cycles of chemotherapy up until the time of the writing of this article. After 3 cycles of R-CHOP, chest x-ray and HRCT of the chest showed a significant resolution of nodular lesions.

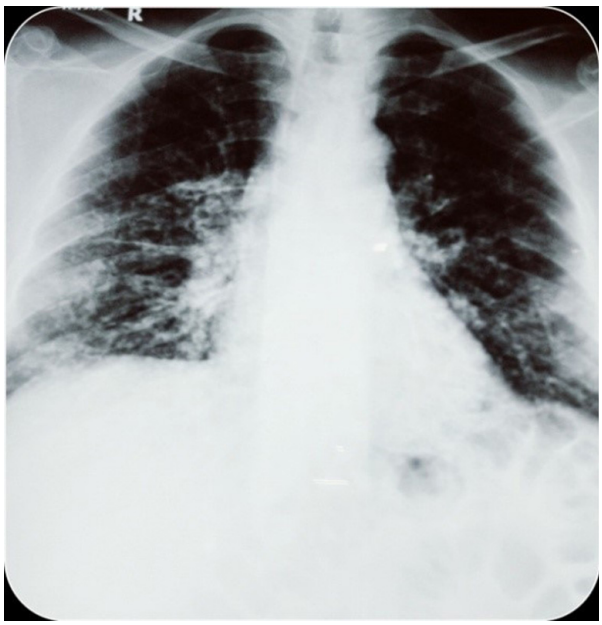

Figure 1. Radiograph of chest posteroanterior view

Discussion

Primary pulmonary lymphoma is a rare entity. It is defined as a lymphoma involving one or both lungs without any extrapulmonary or bone marrow involvement at the time of diagnosis or in the subsequent 3 months after diagnosis [2]. The most frequent primary pulmonary lymphoma (PPL) is extranodal marginal zone B-cell lymphoma of MALT [3]. MALT lymphomas account for less than 0.5% of all primary lung cancers. MALT lymphomas are low grade B-cell neoplasms [4]. They most commonly involve the gastric mucosa.

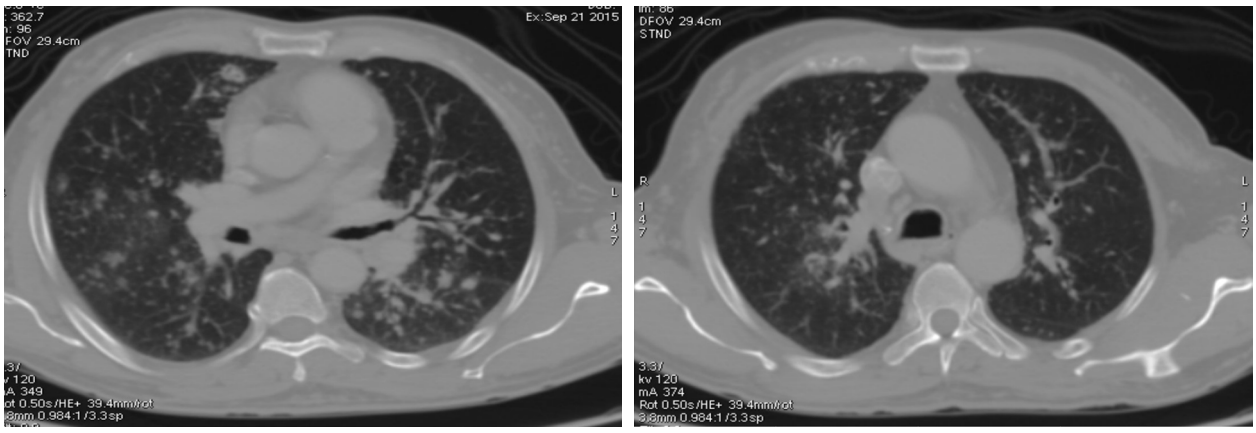

Figure 2. High resolution computed tomography of chest

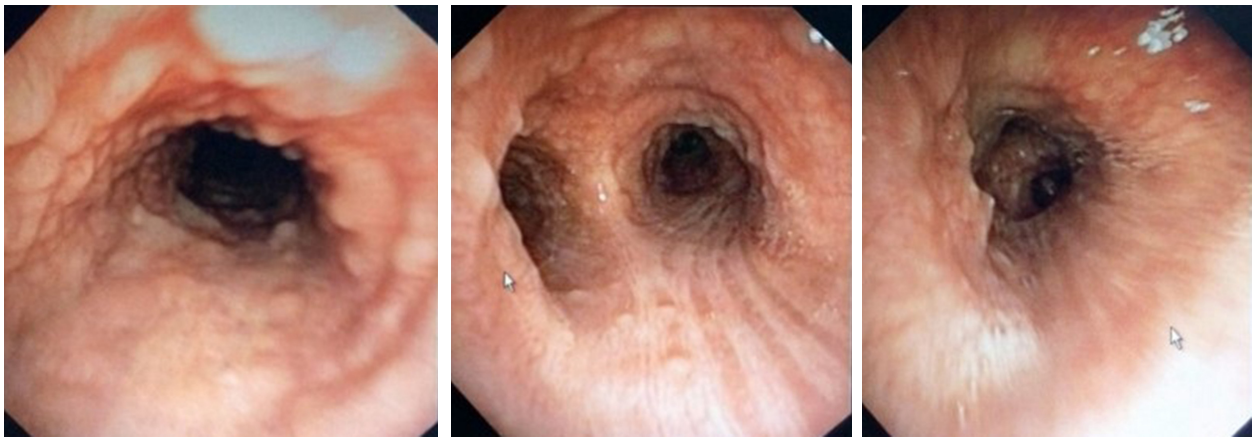

Figure 3. Bronchoscopy image of the trachea, carina, and right intermediate bronchus

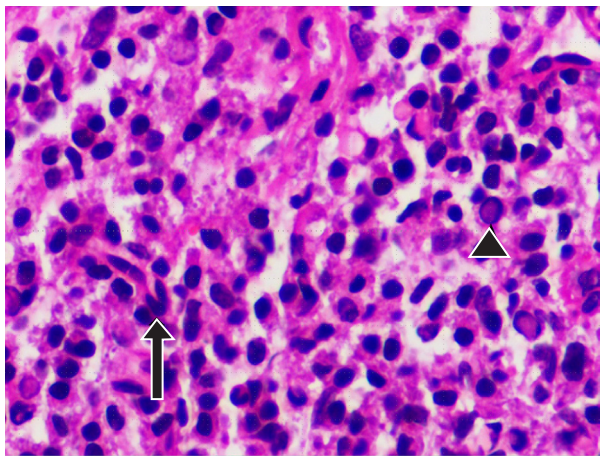

Figure 4. Endobronchial biopsy showing submucosal bronchial tissue with small to medium sized lymphoid cells, centrocyte-like cells (black arrow), lymphoepithelial lesions, and Dutcher bodies (arrowhead)

However, non-gastric site involvement may be seen in the large and small bowels, breasts, head and neck, lungs, dura matter, ocular adnexa, skin, parotid glands, prostate, and ovaries. They are believed to arise because of chronic antigenic stimulation associated with smoking, local chronic inflammatory disease, or autoimmune diseases. An association of gastric MALToma with Helicobacter pylori, ocular adnexal MALToma with Chlamydia psittaci, cutaneous MALToma with Borrelia burgdorferi, and hepatic MALToma with Hepatitis C virus has been proven. However, no such association of any infective agent with pulmonary MALToma is known [5].

It is most commonly seen in elderly patients with a slight male preponderance. It tends to remain localized to the lungs for long periods of time, follows an indolent course, and is associated with a good prognosis. Most of the patients are asymptomatic at diagnosis. Symptoms, when present, can be non-specific [1]. Radiological findings are also non-specific. The most common radiologic manifestation consists of a solitary nodule or a focal area of consolidation [6]. In our case, flexible fiberoptic bronchoscopy showed multiple diffuse nodules along the wall of the trachea, right and left main bronchus, and in the deeper airways. A nodular or lumpy appearance of the trachea and bronchi give the appearance of cobblestones. A cobblestone appearance of the trachea has been classically described in tracheobronchopathia osteoplastica (TPO) [7]. However, it can also be seen in sarcoidosis [8], relapsing polychondritis (RP) [9], amyloidosis [10], granulomatosis with polyangitis [11], and lymphoma [12]. In TPO and RP, nodules do not involve the posterior wall of the trachea [13]. Nodular lesions involving the trachea and bronchi are exceedingly rare in pulmonary MALToma [14–17].

Diagnostic yield of bronchial & transbronchial biopsy is higher when visible endobronchial lesions are targeted. A diagnosis of MALT type NHL is based on histological examination of surgical samples or bronchial/transbronchial biopsy material. Immunohistochemical analysis shows a B-cell phenotype (CD19, CD20), persistence of dendritic cells (CD21, CD35), and the presence of small reactive T lymphocytes (CD3) in alveolar wall infiltrate and around peribronchial nodules [18, 19].

Treatment depends upon the extent of the disease. Optimal management remains unclear. It has been suggested that since it has an indolent course, observation is preferable. However, this approach should be used with caution and only in highly selective patients, especially those in whom diagnostic biopsy was excisional. Surgical

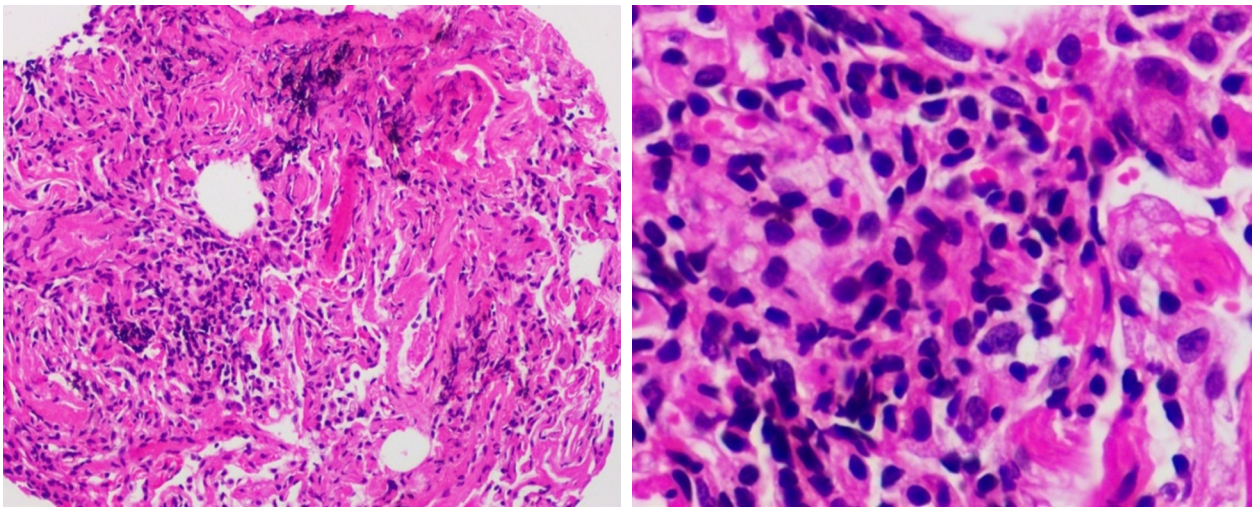

Figure 5. Transbronchial lung biopsy showing similar atypical lymphoid cells infiltrating into the lung parenchyma

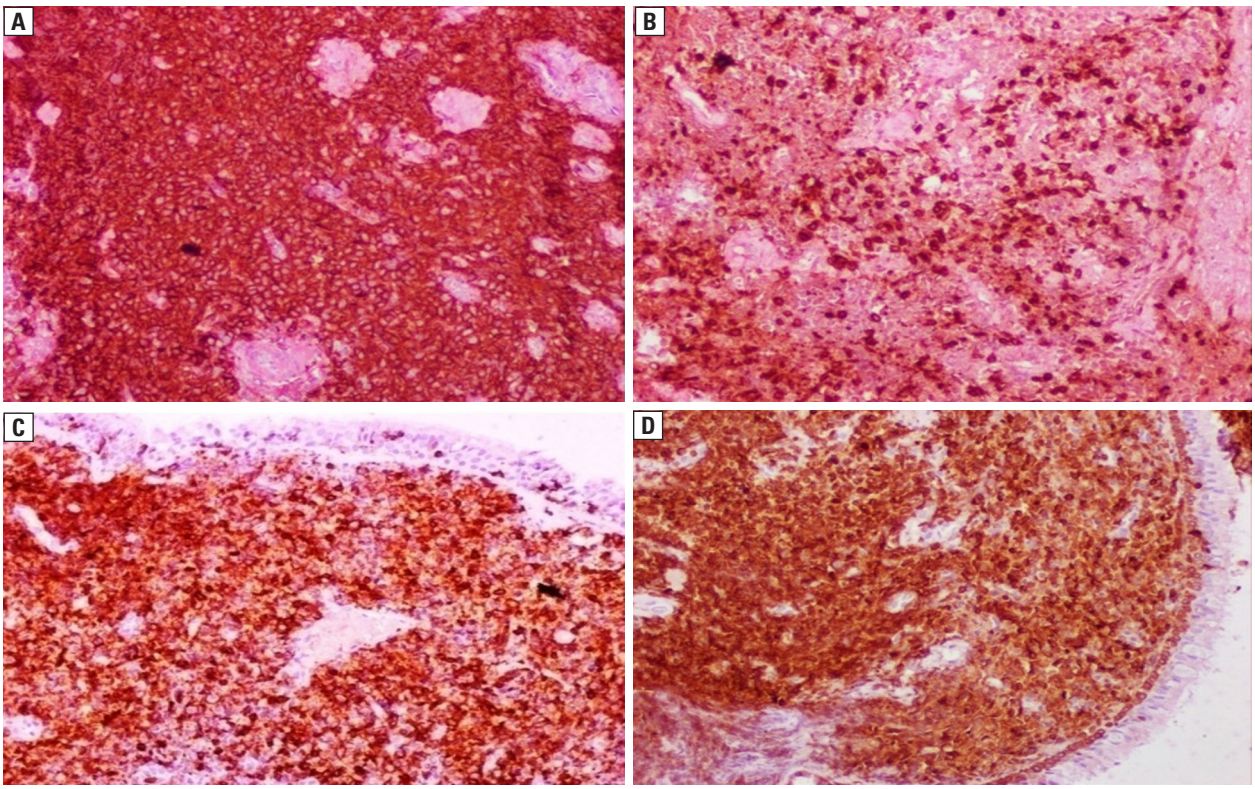

Figure 6. A. CD20 positive B cells; **B.** CD3 reactive T cells; **C.** CD43 positive B cells; **D.** Bcl2 positive B cells

resection is the treatment of choice for localized resectable pulmonary MALTomas. For unresectable disease, involved site radiation therapy (ISRT) or chemo-immunotherapy can be used [20].

Conclusion

The clinical and radiological presentation of our case closely mimicked sarcoidosis. However, biopsy confirmed the diagnosis of pulmonary MALToma. It is a rare disease. Non-specific symptoms and non-specific radiological findings make it difficult to diagnose. It can present with multiple nodular protrusions involving the trachea and bronchi. Histopathology confirms the diagnosis. This case highlights the importance of tissue diagnosis, especially when the radiology is suggestive of some other diseases.

Conflict of interest

None declared.

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