Tuberculosis and Other Granulomatous Diseases of the Airway

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KEYWORDS

• Granuloma • Tuberculosis • Histoplasmosis • Wegener granulomatosis

KEY POINTS

- Granulomatous diseases of the airway are challenging lesions to diagnose and effectively manage because of their rarity and occurrence in different forms, each with unique clinical and radiological characteristics.
- Granulomatous diseases of the airway are not as common as granulomatous diseases involving the lung or other organs.
- Tuberculosis is still a major concern in underdeveloped countries, whereas histoplasmosis is relatively common in eastern and central United States.
- Most granulomatous diseases of the airway can be effectively managed conservatively with drug treatment and repeated airway dilatation.
- Surgical resection may be beneficial in cases presenting with localized airway obstruction or severe hemoptysis.

INTRODUCTION

Granulomas consist of compact aggregates of histiocytes (macrophages) known as "epithelioid histiocytes" because they have elongated cell borders and sole-shaped nuclei as opposed are to what is seen in "ordinary histiocytes," which have well-defined cell borders and round or oval nuclei.[1] The granulomatous inflammatory reaction itself relates to cell-mediated immunity and delayed hypersensitivity and represents the last line of pulmonary defenses. Pathologically, granulomas are characterized by the accumulation of blood-derived macrophages, epithelioid histiocytes, plasma cells, and multinucleated giant cells, representing fused macrophages. T lymphocytes can also be found around the periphery of granulomas. Granulomas can be either infectious or noninfectious in origin and necrotizing or nonnecrotizing.

Granulomatous diseases of the airway are often difficult to diagnose because they can occur in different forms, each with unique clinical and radiological features. Although airway involvement–related symptoms may be the initial clinical manifestation of disease, such involvement is, in most cases, indicative of a more generalized illness. The diagnosis of airway granulomatous disease can generally be established through radiographic and bronchoscopic examinations, whereas management can vary from observation alone to specific drug treatment, repeated airway

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dilatation, or surgical resection in cases presenting with localized and/or symptomatic airway obstruction or life-threatening complications, such as massive hemoptysis.

The 3 most common granulomatous airway disease processes that are likely to have an impact for thoracic surgeons are tuberculosis (MTB), histoplasmosis with secondary broncholithiasis, and Wegener granulomatosis (WG).

AIRWAY TUBERCULOSIS

MTB (*Mycobacterium tuberculosis*), called phthisis or the Great Plague, was the disease that initiated the birth of thoracic surgery near the end of the nineteenth century. It was one of the few remaining unsolved epidemics, its incidence having been accelerated by the "Industrial Revolution" throughout Europe with population shifts toward the cities, overcrowding, and people living in generally poor hygienic conditions. At the time, it was considered to be the most common cause of death worldwide.

For a long time, MTB was considered to be a disease occurring almost exclusively in underdeveloped countries. This concept is, however, no longer true because a significant number of cases are now diagnosed in industrial countries, a phenomenon that relates to an increased incidence of the AIDS syndrome, which predisposes to MTB, an ever-increasing geriatric population, and, finally, the occurrence of multiple-drug-resistant tuberculosis. In industrial countries, for instance, most MTB cases occur in the elderly population, whereas in developing countries, 80% of cases occur in people younger than 60 years of age.

Airway MTB can be secondary to direct implant of tuberculous bacilli from pulmonary lesions containing numerous bacilli into the airway, or it can occur, perhaps more commonly, in relation to local spreading along peribronchial lymphatic channels.[2,3] Isolated airway MTB is a much rarer entity that can, at times, be misinterpreted as being a primary airway neoplasm.[4]

The 4 commonest patterns of tuberculous airway disease that are pertinent to thoracic surgeons are those of (1) extrinsic airway obstruction by tuberculous lymph nodes, (2) endobronchial and endotracheal MTB with secondary stenosis, (3) tracheoesophageal (TEFs) or bronchoesophageal fistulas (BEFs), and (4) the so-called middle lobe syndrome (Table 1).

Table 1
Number of cases of airway tuberculosis seen by one of the authors (R.J.) at the Santosham Chest Hospital in Chennai, India over the past 20 y (Rajan Santosham, unpublished data)

Predominant Pattern of Disease	No. of Cases
Extrinsic obstruction by tuberculous lymph nodes	124
Endotracheal/endobronchial MTB with secondary stenosis	84
TEF/BEF	26
Middle lobe syndrome	84
Total	318

Extrinsic Airway Obstruction by Tuberculous Lymph Nodes

Although enlarged mediastinal nodes are common in patients with pulmonary MTB, it is only in a minority of cases (2%–5%) that they will cause airway compression severe enough to be symptomatic and/or require specific therapeutic interventions. Presenting symptoms are usually cough and stridor, which may be mistakenly attributed to asthma or even bronchogenic carcinoma (Fig. 1). In extreme cases, the involved nodes can even unload caseum into the tracheal or bronchial lumen, resulting in sudden respiratory deterioration or even death. Because of their yet poorly developed cartilaginous support, children are more susceptible to mediastinal lymph node compression than are the adults. Because thoracic imaging may not demonstrate clear-cut pulmonary involvement in this subset of patients, the preferred diagnostic method is that of bronchoscopy, which will confirm airway tuberculous granulomatous involvement through either endoluminal biopsies or positive bronchial washings.

Extrinsic airway obstruction is usually responsive to medical therapy, particularly that of corticosteroids. In the rare instance where the obstruction becomes clinically significant, surgical decompression may be necessary. Such operations are usually carried out through a right posterolateral thoracotomy and consist of nodal incision with either aspiration or curettage of nodal contents. Because some of these lymph nodes may contain a liquefied core, bronchoscopic debridement and drainage are sometimes possible.

Endobronchial and Endotracheal Tuberculosis with Secondary Stenosis

Endobronchial or endotracheal MTB (active or sequelae) is seen in approximately 10% to 20% of patients with pulmonary MTB,[5] and it can lead to cicatricial stenosis most commonly observed in the distal trachea or main stem bronchi.