Bronchocentric Granulomatosis

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• Disclosure:
• Nothing to disclose
Definition

• In 1973 A. Liebow coined the term “bronchocentric granulomatosis” (BCG) to describe one of several pulmonary angiitis and granulomatous syndromes.

• He defined the lesion in purely pathologic terms as necrotizing granulomatous inflammation centered in the airways.

• In 1975 Katzenstein et al noted that BG is a distinctive tissue reaction with mucosal destruction of undetermined etiology.

• BCG has since emerged as a relatively nonspecific pathologic response to various forms of airway injury.

Myers JL. CHEST 1989;96:3-4
Etiology

- BCG is not a disease but rather a descriptive pathologic morphologic diagnosis
- Approximately half of all cases are associated with asthma and allergic bronchopulmonary aspergillosis (ABPA)
- Other cases are idiopathic or associated to mycobacterial or fungal infections
- BCG is considered to represent a histopathologic manifestation of fungal hypersensitivity
- The incidence and prevalence of BG are unknown

King TE et al. Up to Date 2017
• BCG is characterized by peribronchial and peribronchiolar necrotizing granulomatous inflammation.

• Destruction of airway walls and adjacent parenchyma leads to granulomatous replacement of mucosa and submucosa by palisading, epithelioid, and multinucleated histiocytes

• Lumen of affected airway may contain necrotic debris and in case of ABPA mucoid impaction.

• Pulmonary eosinophilia is often present

Bronchocentric granulomatosis in a patient with ABPA. Photomicrograph shows palisading granulomas (arrows) around the bronchioles with disruption of the wall with areas of organization and dense intraluminal eosinophilic content.
Bronchocentric granulomatosis in a patient with ABPA. High power photomicrograph shows palisading granulomas (arrows) around the bronchioles with disruption of the wall with areas of organization and dense intraluminal eosinophilic content.
• BCG does NOT involve the pulmonary arteries
• Other different conditions such as granulomotosis with angiitis (Wegener’s), necrotizing sarcoid granulomata and pulmonary lymphomatoid granulomatosis often include granulomatosis with and angiocentric component
• It has been proposed that the diagnosis of BCG should be reserved for isolated idiopathic cases and for those secondary to ABPA

Allergic Bronchopulmonary Mycosis (ABPM)

- Most cases (>90%) of ABPM are due to Aspergillus infection (ABPA).
- But in addition to Aspergillus, hypersensitivity mediated allergic bronchopulmonary mycosis (ABPM) may develop from other fungal infections, in particular Candida albicans and Bipolaris species.
- Notably in such cases history of asthma is reported only in 32% of affected patients.

Imaging Findings

• Imaging manifestation of BCG varies

• Presentation include single and multiple pulmonary nodules, air-space consolidation, and pulmonary or hilar mass
  - Pulmonary nodule or mass (40%-60%)
  - Consolidation (27%-40%)
  - Mucoid impaction (20%)

• Metabolic activity on FDG-PET has been reported

Ward S et al. Clinical Radiology 2000;55;296
Hurwitz LM et al. CHEST2005;128:1018
BCG secondary to allergic bronchopulmonary mycosis (Bipolaris sp) in a 15 year old girl with cough and recurrent hemoptisis. CT shows an oval shaped hyperdense, non-enhancing oval shape infrahilar mass (white arrows) in the right lung with middle lobe atelectasis and bronchiectasis.
Chest MRI in the same patient demonstrates the oval shaped mass and bronchiectasis with low signal intensity on all sequences and no contrast enhancement (white arrows). Gross surgical specimen demonstrates ectatic bronchi filled with purulent and mucoid material.
Right middle lobe lobectomy

Bronchiectasis with eosinophils, saprophytic fungal hyphae, acute suppurative inflammation, and peribronchial chronic inflammation and fibrosis.

Post obstructive pneumonia including areas of chronic inflammation, foamy histiocytes, microabscesses, foreign-body-type/granulomatous reaction and organization
Whole mount view of one representative section of the specimen. Minimal identifiable lung parenchyma is present. There is significant bronchiectasis (white arrow) and marked peribronchial fibrosis as well as associated parenchymal hemorrhage.
Within the mucin, there is a significant quantity of degenerated eosinophilic infiltrate enmeshed in inspissated laminated mucus in the lumen. High lighted are also charcot leyden crystals and stain of the mucus plug demonstrates saprophytic septate branching fungal hyphae Bipolaris sp.
24 y/o male with history of asthma and eosinophilia. Chest radiograph show bilateral hilar irregular dense opacities and left upper lobe cavitation.
Allergic Bronchopulmonary Aspergillosis with Bronchocentric Granulomatosis

Chest CT in the same patient 2 years later shows consolidation, nodules, and mucoid impaction. Path: ABPA with necrotizing granulomas in the airways.
Bronchocentric granulomatosis in a 35-year-old man with ABPA and a longstanding history of asthma. (A) PA radiograph shows consolidation of the right upper lobe. 
(B,C) Axial CT images obtained with a soft-tissue window depict opacification of the right upper lobe and several hyperattenuating tubular mucoid impactions in dilated bronchi.

Bronchocentric Granulomatosis

BCG in a 38-year-old man with asthma and recurrent pneumonia. (A, B) Axial contrast-enhanced CT chest images (mediastinal windowing) demonstrate left upper lobe pneumonia and right perihilar branching endobronchial soft tissue mass.
(C) High-power photomicrograph (original magnification X 200; H-E stain) of a transbronchial biopsy specimen shows eosinophilic cellular debris and bronchocentric granuloma formation (arrows).
BCG and Chronic Necrotizing Bronchopulmonary Aspergillosis

• In addition to ABPA, BCG can rarely develop in patients with chronic necrotizing bronchopulmonary aspergillosis (CNBPA)

• CNPA typically occur in patients with mild degree of immunodeficiency

• CNBPA and BCG have different immunologic mechanisms.

• CNBPA occurs in the setting of immunosuppression, whereas BCG is more commonly a hypersensitivity reaction

Yousem SA. Human Pathol 1997;28:650
Hemoptysis in a 31 y/o male with AIDS, with a cavitary lesion in the left upper lobe (black arrows). CT shows a pseudoaneurysm in the left cavity wall (yellow arrow). LUL resection was performed. Pathology reveal BCG and necrotizing bronchopulmonary aspergillosis due to Aspergillus niger.
Bronchocentric Granulomatosis

Adult female patient with history of refractory asthma. Chest CT depicted numerous small pulmonary nodules and patchy irregular parenchymal opacities. Lung biopsy demonstrated necrotizing granulomas surrounding small caliber airways.
Bronchocentric granulomatosis in a 51 y/o male with cough-asthma. CT shows multiple small nodules in the right lung (white arrows). Lung biopsy demonstrates bronchioles with granulomatous inflammation and obliterative changes. The centers of the bronchioles contain necrotic debris (black arrows).

Courtesy Santiago Martinez M.D.
Summary

• BCG is a destructive granulomatous lesion of the bronchi and bronchioles
• BCG is not a disease but rather a descriptive pathologic morphologic diagnosis
• Most cases are associated with asthma or ABPA
• Imaging manifestation includes single or multiple pulmonary nodule, mass or consolidation
References

- Myers JL. Bronchocentric granulomaatosis. Disease or diagnosis? Chest 1989;96:3-4
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