

Bronchiectasis for Thoracic Surgeon: Classification, Etiologies, Diagnosis and Management

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Abstract

Bronchiectasis is an abnormal and irreversible dilatation of the bronchi. It is classified into two groups: localized and diffuse bronchiectasis. Local causes such as endobronchial process, foreign body or compressive lymph node are responsible of localized bronchiectasis. Cystic fibrosis, ciliary dyskinesia and immunity deficiency are the main etiologies for diffuse bronchiectasis. The medical treatment is indicated in all cases. Surgery is associated to well outcomes in localized forms and improve the daily life of patient in the diffuse forms when medical treatment is insufficient.

Keywords: *Bronchiectasis; Surgery; Anatomical Resection; Complication*

Introduction

Bronchiectasis is a chronic lung disease defined by an abnormal and irreversible dilatation and distortion of the bronchi [1]. Originally described by René Laënnec in 1819 [2], bronchiectasis has numerous causes. Chronic inflammation leads to chronic cough, sputum production and recurrent pulmonary exacerbation. Pathological mechanisms of bronchiectasis are well described in the vicious cycle of Cole and the diagnosis is evoked clinically, and radiologically confirmed by the CT scan. In countries of low socioeconomic level, bronchiectasis is secondary to parenchymal lesions of tuberculosis and respiratory infections during the childhood [3]. The management depends on the causes of bronchiectasis and its type: localized or diffuse bronchiectasis. When surgery is performed, it allows the disappearance or diminution of sputum production and avoidance the evolution towards destroyed lung and lung fibrosis.

Classification and etiologies (Table 1)

Bronchiectasis can be classified into two forms: localized and diffuse bronchiectasis. This classification was established in order to precise cases that must benefit of surgical treatment from cases where medical management is the best therapeutic option. No definition was made to characterize localized and diffuse forms of bronchiectasis. Bilateral lesions do not mean systematically diffuse bronchiectasis and in this case patient can benefit of surgery if lesions are localized to one lobe in each side. In the setting of bronchiectasis concerning all lobes of one side, patient probably can benefit of surgery (Pneumonectomy) owing to the risk of postoperative complications, and in this case medical management presents the best option.

Type of Bronchiectasis	Etiologies
Localized bronchiectasis	<ul style="list-style-type: none"> • Endobronchial obstruction: <ul style="list-style-type: none"> • Foreign body • Benign tumor (carcinoid tumor, hamartochondroma...) • Extrinsic compression: <ul style="list-style-type: none"> • Calcified lymph nodes (Brock syndrome)
Diffuse bronchiectasis	<ul style="list-style-type: none"> • Previous treated pulmonary tuberculosis • Recurrent respiratory infections during childhood (measles, pertussis...) • Cystic fibrosis • Primitive ciliary dyskinesia • Primitive immune deficiency • Gastroesophageal reflux disease • Systemic disorders (Rheumatoid arthritis...)

Table 1: Etiologies of bronchiectasis.

The localized bronchiectasis is most often secondary to a local cause by extrinsic compression of a lobar or segmental bronchus, as the case of Brock syndrome incriminated by the compression of the middle lobar bronchus, usually by adjacent calcified lymph nodes of tuberculosis origin (broncholithiasis). An endobronchial tumor is also responsible for focal bronchiectasis, especially a benign tumor with chronic development, such as a hamartochondroma, a lipoma or a carcinoid tumor [4]. Foreign body introduced accidentally in the bronchial tree is responsible of localized lesions of bronchiectasis. This cause is especially frequent in the right side at the level of lower bronchus.

Diffuse bronchiectasis can be the result of numerous causes. Repetitive respiratory infections during childhood have long been the leading cause of bronchiectasis. Measles, pertussis, bacterial and viral pneumonitis (adenovirus, influenza virus, and syncytial respiratory virus) were the main incriminated pathogens [5]. Currently these infections are less common especially in developed countries thanks to the extension of vaccination campaigns.

Genetic diseases are predominantly cystic fibrosis and ciliary dyskinesia. Cystic fibrosis in adults is responsible for 3-8% of bronchiectasis in prospective studies [6,7]. It is a genetic disease due to an abnormality of the gene coding for the CFTR protein (cystic fibrosis transmembrane receptor). Some criteria lead to the diagnosis of cystic fibrosis: predominance of bronchiectasis in the upper lobes, extrapulmonary manifestations such as recurrent nasal polyposis and pancreatic endocrine or exocrine insufficiency [8]. The diagnosis is confirmed by sweat test (positive if the chlorine concentration is greater than 60 mEq/L) and the search for mutation of the CFTR gene (located on chromosome 7) [8].

Primary ciliary dyskinesia is a heterogeneous group of diseases associating recurrent infections of the upper or lower airways with bronchiectasis, male infertility and abnormalities of the ciliary structure. The association of these affections with a situs inversus defines

the kartagener syndrome found in half of the cases of ciliary dyskinesia [9]. In this syndrome, lesions are predominated in the middle lobe, lingual, and lower lobes.

The existence of a congenital or acquired immunodeficiency favors the development of bronchiectasis [10,11]. The deficits in humoral immunity are the most common by deficiency affecting a class of immunoglobulins (IgA, IgG, IgM), or a subclass of immunoglobulins. In Good's syndrome, which associates thymoma with a deficit of humoral immunity, dilations of the bronchi are noted in the course of repeated pulmonary infections [12].

The existence of bronchiectasis in this setting of systemic disorder was demonstrated is recent knowledge thanks to the easy realization of high resolution CT. Rheumatoid arthritis (RA) is the most frequent systemic disorder predominated in women found in 30 to 50% of thoracic CT performed in these patients [13]. The association of bronchiectasis in other systemic disorders was reported such as disseminated lupus erythematosus and Sjögren's syndrome [14,15].

In inflammatory diseases of the digestive tract, bronchiectasis is more common in ulcerative colitis than in Crohn's disease [16]. They represent a quarter of the respiratory manifestations of these diseases and occur in 85% of cases while the digestive disease is known.

Clinical manifestations

The interrogation of patient is in the goal to search elements allowing to find one of the aforementioned etiologies. The main symptom in bronchiectasis is an old and daily sputum, variable quantitatively and qualitatively. Its importance ranges from a simple chronic cough to an abundant bronchorrhoea that is associated to a foul odor and impact on the patient's daily life. Hemoptysis is also frequent and can range from simple blood-streaked sputum due to the erosion of the inflammatory bronchial mucosa to massive hemoptysis linked to a rupture of the bronchial artery. Presence of dyspnea is variable and depends on the extent of the lesions of bronchiectasis. Most patients have a good general state and the deterioration is observed when bronchiectasis reaches the stage of severe chronic respiratory insufficiency or is complicated by a severe local or general infection. Physical examination is normal almost in all patients and aims to search especially extrapulmonary signs that orientate to different probably causes of bronchiectasis. Digital clubbing is observed more willingly in the extensive and old forms of bronchiectasis and old, with also the signs of right heart failure due to advanced respiratory failure.

Paraclinical examinations

If the clinical presentation makes it possible to evoke the diagnosis of bronchiectasis, paraclinical examinations especially thoracic CT allows confirmation of this diagnosis. Chest X-ray required in the first line can demonstrate direct signs of bronchiectasis like tubular opacities or in rail, or indirect signs essentially an atelectasis. The interest of thoracic CT is to confirm the diagnosis, determinate the type: cylindrical, varicose or cystic bronchiectasis, precise extension of lesions, appreciate the healthiness of the rest of the parenchyma in order to conduct the surgical procedure. This thoracic CT must be performed in fine sections (high resolution computed tomography: HRCT) with injection of the contrast product.

Fiberoptic bronchoscopy constitutes a method to explore the bronchial airways in the interest to search a local cause by an endobronchial process especially in a smoking patient, foreign body or extrinsic compression secondary to broncholithiasis or benign tumor. Origin of hemoptysis is also precised especially in bilateral lesions [17]. Bronchoalveolar lavage is performed to search bacterial or mycobacterial germs especially mycobacterium tuberculosis. When there is a large quantity of production of purulent sputum, this procedure can help in therapeutic option allowing the aspiration of this sputum in order to avoid bronchial congestion, atelectasis and pneumatoxis.

A cytobacteriological test of the sputum is requested in order to search for colonization by germs making evidence of a bronchial infection requiring treatment by antibiotic products. Also, looking for mycobacterium tuberculosis in sputum is done especially in regions when tuberculosis constitutes an endemic disease.

Surgical treatment

Goals and indications of surgical treatment

In bronchiectasis, surgery can be performed after failure of the medical treatment, or immediately in case of localized bronchiectasis. Indications of surgery are mainly massive daily bronchorrhea recurrent suppurative infections, hemoptysis, or occurrence of complications such as lung abscess, pneumothorax, or pyothorax. The goals of this surgery are to avoid a massive hemoptysis that can be life-threatening for patient, and to improve daily life of patient by removing the abundant sputum, and avoid occurrence of complications (lung abscess, kidney amyloidosis, empyema...).

Preoperative assessment

Evaluation of the respiratory function and co-morbidities of patients constitutes a mandatory phase before any surgical procedure. Measure of the FEV1 by spirometry (forced expiratory volume in 1 second) appreciate presence of obstructive or restrictive ventilator disorders, and DLCO (diffusion capacity for carbon monoxide) determinate the diffusing capacity of the lungs [21]. The assessment for lung parenchyma established by patient's exercise capacity (VO_2 max: peak oxygen consumption) is required in case of values lowered than 80% of at least one of the parameters (FEV1 or DLCO) [22]. The knowledge of the number of segments which will be resected according to the thoracic CT allows having a value of predictive postoperative FEV1 [23]. Others tests can be required in terms of the underlying co-morbidities. Lung scintigraphy provides essential informations to evaluate the appropriateness of a surgical excision procedure by quantifying the ventilation and perfusion of each lobe. However, these recommendations are more practiced in the setting of lung cancer where the tumor is resected with a healthy lung parenchyma. In the setting of chronic inflammatory lung diseases such as bronchiectasis and destroyed lung, the patient is already used to his respiratory reserve, and the intervention can be carried out with lower values of FEV1.

In order to reduce of postoperative complications, anemia caused by recurrent hemoptysis may require a preoperative blood transfusion and the infection is managed by antibiotics products, and the smoking cessation should be advocated [24]. The patient must be incriminated in the preparation for the surgery by explaining the surgical procedure, the various complications after the intervention, and the interest of a better respiratory kinesitherapy. Lastly, before the intervention, patient must be intubated preferentially by a double lumen endotracheal tube in order to protect the contralateral lung against spillage of secretions, and in the same time to ensure single lung ventilation for let enough space in the pleural space to the surgeon especially in the setting of the video-assisted thoracic surgery.

Approaches

Open surgery by posterolateral thoracotomy is the main approach in bronchiectasis [3,18]. This disease is usually associated to important dense pleural adhesions, calcified lymph nodes, and loss of anatomical landmarks, especially in patients with chronic inflammation observed in cases of tuberculosis. For these reasons, open surgery ensure an evident release of adhesions, a direct and quick control of vascular elements in case of significant bleeding, manual palpation of the parenchymal lung to appreciate extension of lesions, and avoidance of the contamination of the pleural space by opening the lesions of bronchiectasis. Muscle sparing in thoracotomy is of high importance for probably muscle interposition in case of bronchopleural empyema and postoperative empyema which are of a not negligible rate in bronchiectasis. Likewise, in chronic lesions, dissection through extrapleural plan can be helpful, and in some cases a rib resection in order to place retractor when there is a retracted intercostals spaces.

Video-assisted thoracic surgery (VATS) was reported in some series of bronchiectasis and was described safe and feasible in selected cases [19,20]. In the study of Schneiter and associates, among 48 patients, 21 patients were operated by exclusive videothoracoscopy and 10 patients by VATS through a mini-thoracotomy [21]. This minimally invasive approach has demonstrated many advantages after surgery: less chest pain, fast recovery, and short hospital stay. The rate of conversion is increased in chronic lesions of bronchiectasis second-

ary to old tuberculosis. The choice between open surgery and VATS is determined according to preoperative assessment on the HRCT of bronchiectasis and presence of lymph nodes. In some hospital centers, VATS is systematically performed and when there is a dense pleural adhesion conversion to thoracotomy is realized.

Surgical procedures

Whatever the surgical procedure performed, the principle is to have a complete resection of lesions when is possible to stop the vicious cycle described by Cole, and thereby slow the progression of the disease, and in opposite preserving the maximum of healthy lung [22]. The main surgical procedure remains lobectomy allowing an anatomical lung resection for avoidance of recurrence of lesions [23]. Pneumonectomy is associated with a high risk of morbidity and mortality, especially in this inflammatory and infectious disease, and should be avoided except in cases of destroyed lung. Segmentectomy is performed in patients with localized bronchiectasis to a segment and in patients that cannot support lobectomy owing to a less respiratory reserve. In bilateral bronchiectasis, the side to operate firstly is determined by presence of hemoptysis and extension of lesions [24]. In the setting of hemoptysis, patient must be operated firstly on the side that is responsible of this hemoptysis, because it can be life-threatening in the next time if not treated. In the setting of extension of lesions, the side where the bronchiectasis is predominated must be operated firstly because the surgeon is not sure if patient can be operated for the contralateral side according to general condition of patient and disappearance of symptoms after the first intervention [23]. After surgery of two sides, two lobes or six segments must be conserved in total [25]. The time between first and second surgical intervention must be delayed between three to six months in order to allow a well recovery of patient, and to appreciate the level of disappearance of clinical manifestations.

If surgery is retained for localized bronchiectasis, the question remains in doubt to diffuse bronchiectasis because of incomplete resection of lesions [22]. Some authors, even if the resection is incomplete in these settings, and when medical treatment is insufficient, surgery is performed because of improvement on the patient's symptoms and quality of life [26].

Postoperative complications and their management

Generally, surgery for infectious and inflammatory lung diseases and especially bronchiectasis is associated to infections and bleeding in postoperative phase more than surgical procedures performed for lung cancer because of infected underlying lung, dense pleural adhesions, multiples lymph nodes adherent to elements of pulmonary hile [27]. These complications are recorded with high rate in the setting of chronic inflammation and lesions of tuberculosis already treated. The infection is manifested by pneumonia and atelectasis at the level of parenchymal lung. The management is exclusively medical, based on antibiotic products, and in case of atelectasis the aspiration of endobronchial secretions using a fiberoptic bronchoscopy. Bleeding is manifested by hemothorax when the chest tube drainage brings a remarkable volume associated to clinical manifestations of hemorrhagic shock and a re-exploration for a hemostatic procedure is obtained. Prolonged air leakage is also a frequent complication after surgery of bronchiectasis and is managed in most cases by mobilization of the chest tube made during the intervention or added another chest tube drainage frequently in the anterior site. Bronchopleural fistula is a rare but a serious complication that occurs especially in the acute phase after surgery (2 to 3 weeks) and most commonly after a pneumonectomy more than after lobectomy [24]. It is associated as a consequent to postoperative empyema. The management can be a re-thoracotomy for closure of the bronchial stump in the acute phase or open window thoracostomy for dressing changes especially in the late phase with subsequent muscle plasties after sterilization of the pleural space.

Conclusion

Etiologies of bronchiectasis are multiple. Differentiation between diffused and localized bronchiectasis is essential in the therapeutic conduct. Medical management is indicated in all forms of bronchiectasis. However, localized bronchiectasis can benefit of a surgery in order to improve the quality of life of patients who suffer from bronchorrhoea. The intervention is usually an anatomical lung resection.

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