Short paper

Localised pulmonary resection for bronchiectasis in hypogammaglobulinaemic patients

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Abstract

Background – Bronchiectasis and pulmonary infections are common in patients with hypogammaglobulinaemia. Despite intravenous gammaglobulin treatment and appropriate antibiotics, a subgroup of patients remains with persistent localised pulmonary infection in segments where bronchiectasis had developed before appropriate treatment. As such localised pulmonary suppuration (segmental or lobar) may serve as a focus for progression of bronchiectasis, surgical resection of the involved segments may be considered. The outcome of pulmonary resection in four such patients is reported.

Results – Surgery was well tolerated except for one postoperative empyema. Information on follow up is available from 3.5 to 5 years. All patients experienced considerable reduction of symptoms including cough, sputum production, antibiotic use, and hospital admissions.

Conclusions – Surgical resection of localised bronchiectatic segments should be considered in patients with hypogammaglobulinaemia with persistent localised suppuration and symptoms refractory to medical treatment.

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Bronchiectasis and pulmonary infections are common in patients with hypogammaglobulinaemia.^{1,2} High dose intravenous gammaglobulin administration results in physiological circulating levels of immunoglobulins and thus patients on this therapy should be fully immunocompetent.³⁻⁶ Indeed, such patients have fewer pulmonary infections and show considerable improvement.^{6,7} However, some patients, particularly those who have developed bronchiectasis before treatment of their immunodeficiency state, have persistent localised pulmonary infection and refractory symptoms.

Surgical resection used to be a common means of attempting to control symptoms and preventing spread of bronchiectasis by eradication of suppuration in truly localised areas.⁸⁻¹⁰

Today surgical resection is rarely attempted unless medical treatment has failed even in immunocompetent patients.^{11,12} Resection is not generally considered appropriate for patients with bronchiectasis resulting from hypogammaglobulinaemia or ciliary dysformation because of the high probability of recurrence.¹² However, patients with hypogammaglobulinaemia receiving intravenous gammaglobulin replacement therapy should be fully immunocompetent and perhaps more suitable for resection of localised disease. We describe four patients who, despite aggressive medical treatment (intravenous gammaglobulin replacement, physiotherapy, and broad spectrum antibiotics), had persistent pulmonary infection with localised bronchiectasis who underwent successful lobectomy.

Methods

PATIENT SELECTION

Four of 65 patients with hypogammaglobulinaemia followed at the Immunology Clinic at the Hospital for Sick Children were considered for surgery on the following criteria: (1) documented panhypogammaglobulinaemia despite monthly intravenous gammaglobulin replacement; (2) persistent or recurrent pulmonary infections unresponsive to antibiotic therapy; and (3) localised bronchiectasis involving one lobe only as demonstrated by high resolution computed tomographic (CT) scanning.

All patients were receiving monthly intravenous gammaglobulin (Sandoglobulin 0.6 g/kg/m²). At each clinic visit complete blood counts and quantitative immunoglobulin measurements were taken and the state of health reviewed. At follow up visits after resection patients and family were questioned on cough, sputum production, purulent exacerbations, hospital admissions, days off work or school, and general level of activity. Follow up pulmonary function tests, chest radiographs, and high resolution CT scans were available in all patients.

Results

The cases are summarised in the table. The first case is presented in more detail to illustrate the clinical course of a representative patient.

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Summary of patients

Patient no.	Current age (years)	Before surgery		After surgery		Length of follow up (months)
		Symptoms	Lab testing	Surgery: complications symptoms	Lab testing	
1	15	Cough + + + Sputum + + + Purulent exacerbations + + +	PFTs: FVC 100%, FEV ₁ 103% CT: RML bronchiectasis	RML lobectomy No complications Cough + Sputum + Purulent exacerbations +	PFTs: FVC 106%, FEV ₁ 97% CT: no bronchiectasis, mild bronchial thickening LLL	48
2	9	Cough + + + Sputum + + + Purulent exacerbations + + +	PFTs: FVC 99%, FEV ₁ 109% CT: LLL bronchiectasis, atelectasis, and consolidation	LLL lobectomy No complications Cough + Sputum + Purulent exacerbations +	PFTs: FVC 92%, FEV ₁ 101% CT: no bronchiectasis, bronchial wall thickening RLL	43
3	28	Cough + + + Sputum + + + Purulent exacerbations + + +	PFTs: FVC 57%, FEV ₁ 57% Po ₂ 10.9 kPa CT: LLL bronchiectasis, mild RLM bronchial dilation	LLL lobectomy Complication: empyema Cough + Sputum + Purulent exacerbations + Two successful pregnancies	PFTs: FVC 45%, FEV ₁ 43% Po ₂ 10.5 kPa CT: left sided pleural thickening, RML and LUL bronchial dilation	72
4	39	Cough + + + Sputum + + + Purulent exacerbations + + +	PFTs: FVC 96%, FEV ₁ 90% CT: LLL bronchiectasis	LLL lobectomy No complications Cough + Sputum + Purulent exacerbations +	PFTs: FVC 92%, FEV ₁ 88% CT: mild cylindrical bronchiectasis of lingula	42

CT = computed tomographic scan of chest; RML = right middle lobe; LLL = left lower lobe; PFTs = pulmonary function testing reported as percentage predicted normalised for age.

Cough: + mild; + + moderate; + + + frequent and severe.

Sputum: + ≤ 2 tbsp/day; + + > 2 tbsp/day; + + + > 1.2 cup/day.

Purulent exacerbations (required antibiotics): + < 1 year; + + 2–6 year; + + + > 6 year.

CASE 1

A one year old boy with recurrent infections was diagnosed as having acquired hypogammaglobulinaemia. Treatment was initiated with intramuscular gammaglobulin replacement but frequent infections of the eyes, nose and chest continued. At 9 years intravenous gammaglobulin therapy was commenced with a decrease in ocular and nasal infections, but cough productive of purulent sputum continued with right middle lobe consolidation and atelectasis. High resolution chest CT scanning indicated bronchiectasis limited to the right middle lobe and, at 11 years, right middle lobectomy was performed without complication. Postoperatively there was a significant diminution of sputum production and cough. Four years after surgery he continued to have minimal pulmonary symptoms, and results of pulmonary function tests were within normal limits. Chest CT scanning demonstrated mild peribronchial thickening in the right lower lobe which had not progressed in more than one year.

Discussion

In each of the cases resection of a localised bronchiectatic segment caused a considerable diminution of symptoms, requirement for antibiotic therapy, need for medical care, and improvement in the quality of life. The lobectomies were physiologically well tolerated with the exception of case 3 who developed an empyema which rapidly resolved with appropriate therapy. Our experience suggests that surgical resection of localised bronchiectasis should be considered in patients with hypogammaglobulinaemia who have continued to have symptoms refractory to conventional medical therapy including gammaglobulin replacement. All our patients have progressed to develop bronchial thickening in previously unaffected segments as seen by high

resolution CT scanning, and whilst the clinical significance of these abnormalities is uncertain, it is possible that bronchiectasis might develop in these areas. Based on the rate of progression of symptoms preoperatively, it is possible that bronchiectasis may have developed in additional areas even if surgical resection had not been undertaken. Only case 4 has developed obvious bronchiectasis which is cylindrical, limited to one segment of the lingula, and has not progressed further in the three years of follow up. The improvement seen in the four patients following their resections has continued for 3.5–5 years.

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