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Bronchiectasis in Central Australia: A young face to an old disease

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KEYWORDS

Bronchiectasis; Indigenous; HTLV-1; Recurrent pneumonia

Summary

Background: Bronchiectasis remains a significant cause of morbidity among specific populations world wide, including many indigenous groups. Data on prevalence in Australian adults are lacking. Indigenous children in Central Australia have the highest rates of bronchiectasis in the world. Outcomes for these individuals after they become adults are not currently available.

Methods: We performed a retrospective case review of the presentation and likely aetiology of adult patients presenting to the Alice Springs Hospital with a primary diagnosis of bronchiectasis.

Results: Sixty-one patients and 166 admissions were identified. Fifty-nine patients were indigenous (97%). Mean age was 42 ± 15 years. Forty-three patients (70%) had past histories notable for recurrent respiratory tract infections. No predisposing factors could be identified in 11 patients (18%). Human T-cell lymphotropic virus type 1 (HTLV-1) serology was positive in 72% of those studied. Eight (13%) patients died during the study period. Conclusion: Bronchiectasis remains a significant cause of morbidity and mortality in Central Australia, with notably different patient characteristics and disease aetiology to other published cohorts. Recurrent respiratory infection is the major cause of illness. Associated factors include indigenous ethnicity, HTLV-1 positivity and childhood in a remote region.

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Introduction

Bronchiectasis is an anatomical diagnosis describing irreversibly dilated bronchii, associated with bronchial wall distortion, inflammation, and destruction. It was a common

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disease in the pre-antibiotic era although its prevalence is felt to have fallen in developed countries¹ following the introduction of vaccination and antibiotics and the decline in tuberculosis.² Current data is lacking and specific populations, particularly indigenous people of Alaska, the South-west Pacific, and South-east Asia continue to suffer high rates of disease.^{3,4} Indigenous children living in Central Australia have amongst the highest reported rates of bronchiectasis in the world,⁵ though the consequences of bronchiectasis for these individuals after they enter adulthood have not been studied.

Our experience suggested the Central Australian adult population served by our hospital experience significant morbidity due to bronchiectasis and, moreover, that patient features differ significantly from cohorts representative of urban populations. We therefore examined data from a large cohort of patients presenting with bronchiectasis to Alice Springs Hospital (Northern Territory, Australia) over a 12-month period.

Methods

A retrospective review of all adult patients admitted to Alice Springs Hospital between July 1, 2004 and June 30, 2005 was performed. Alice Springs Hospital is a 164 bed facility and is the only hospital servicing the Central Australia Region (approximately 1,000,000 square kilometres). The area has a population of approximately 47,000 people, of whom 40% are indigenous. The closest referral centres are Darwin and Adelaide, both over 1500 kilometres from Alice Springs.

All adult patients with a discharge diagnosis of bronchiectasis were identified by analysis of hospital admission data for the study period. The diagnosis of bronchiectasis was considered to be confirmed if at any time during or prior to the admission a patient with typical clinical features of bronchiectasis demonstrated classic radiographic changes of bronchiectasis on either chest X-ray (CXR) (tram-tracking, ring shadows, cystic change) or high-resolution computerised tomography (HRCT) of chest (airway dilatation, bronchial wall thickening, cystic change). Patient histories were then reviewed for clinical, microbial, spirometric, and demographic data. All information recorded, including microbial and laboratory results, were obtained in the routine care of each patient with no subsequent investigations performed to aid our study.

Statistical analysis was performed using the Microsoft Excel 2004 for Mac (Microsoft Corporation, Redmond, WA, USA).

The project received approval from the Central Australian Human Research Ethics Committee. The requirement of informed consent for use of retrospective data was not required by the committee.

Results

During the study period, 69 patients were admitted to Alice Springs Hospital with a primary diagnosis of bronchiectasis. Eight patients had no radiographic evidence of bronchiectasis on either CXR or HRCT. The remaining 61 patients form

No. of patients	61
Male/female	32 (52%)/29 (48%)
Age at admission (yr \pm SD)	42 ± 15
Background	
Indigenous	59 (97%)
Non-indigenous	2 (3%)
Location (childhood)	
Remote	51 (84%)
Urban	3 (5%)
Unclear	7 (11%)
Location (current)	
Remote	43 (70%)
Urban	18 (30%)

the basis of this report. Demographic data are recorded in Table 1.

The initial diagnosis of bronchiectasis had been made by CXR in 29 patients (47.5%), HRCT in 29 patients (47.5%), and plain CT chest in three (5%). Eleven patients had not had CT imaging of the chest at the time of our report. No patients had undergone bronchoscopy or bronchography.

Twenty-seven patients (44%) were current smokers, 17 were non-smokers (28%), 10 were ex-smokers (16%), and smoking status was not recorded in seven (11%).

Eight patients died during the 12-month-study period. Seven deaths were due to respiratory illness and one due to complications of non-related trauma.

Aetiology

Forty-three patients (70%) had past histories notable for recurrent childhood respiratory infections. Three patients, one indigenous and two non-indigenous, had severe chronic obstructive pulmonary disease (COPD) with bronchiectasis noted on HRCT. One patient had respiratory syncitial virus pneumonia documented in infancy. A further patient had severe Nocardia pneumonia requiring intubation and tracheostomy. Bronchiectatic changes were correlated with the distribution of consolidation seen during their pneumonic illness. Two patients had conditions predisposing to recurrent aspiration (Down's syndrome, severe intellectual disability). No predisposing factors were identified in 11 patients. Factors identified in the development of bronchiectasis are summarised in Table 2.

Relatively, few patients had investigation for underlying disorders. Six patients were tested for $\alpha 1$ -antitrypsin deficiency; all were normal. Immune investigations included anti-nuclear antibodies in seven, with two positive, at low titres. In the absence of other rheumatic symptoms this was thought to be due to acute infection. Serum protein electrophoresis and immunoglobulin levels were normal in all six patients studied. Rheumatoid factor was negative in all seven patients studied. Aspergillus precipitans were negative in three patients, though serum immunoglobulin E was noted at $> 1000 \, \text{ng/mL}$ in one patient. A. fumigatus was

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