Aetiology of chronic suppurative lung disease

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Abstract

Forty one (1%) of 4000 children referred for respiratory disease had chronic suppurative lung disease not due to cystic fibrosis. Further investigations showed congenital malformations in six (15%), primary ciliary dyskinesia syndrome in seven (17%), 11 had immunological abnormalities (27%), and two bronchiectasis due to aspiration (5%). Therefore the underlying cause for the disease was found in 63%. Identification of predisposing causes may facilitate prevention of further bronchial damage.

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The respiratory tract is the commonest site of infection in children, usually by a virus, which rapidly clears. Some are infected with bacteria, and require antibiotic treatment, but a minority with aggressive organisms, abnormal host defence mechanisms, or inappropriate treatment develop permanent bronchial damage. These patients do not lose their symptoms, and develop a chronic productive cough. Demonstration of bronchiectasis provides a description of their bronchial morphology but not a diagnosis. We have reviewed our experience of bronchiectasis in a specialist paediatric respiratory service.

Aetiology of bronchiectasis

Diagnosis	*No of cases (n=41)*	*Bronchiectasis proved by*	*Surgery (n=18)*
Total congenital malformations:	6*		
Broncharterial malinosculation	2	Angiogram	2
Cystic adenomatoid malformation	1	Histology	1
Harmartomatous malformation	1*	Histology	1
Bronchomalacia	1	Bronchogram	–
Absent pulmonary valve syndrome	1	Bronchogram	1
Total primary ciliary dyskinesia	7**		
	*	2× Bronchogram	1
		1× Histology	1
	*	4× V/Q scan	–
Total primary immune defects:	8		
Neutrophil locomotion/chemotaxis defect	1	V/Q scan	–
Neutrophil chemotaxis defect, slightly reduced serum dependent killing	1	Bronchogram	1
Cell dependent neutrophil phagocytosis defect	1	CT scan	–
Serum dependent neutrophil killing defect, opsonisation defect	1	V/Q scan	–
Panhypogammaglobulinaemia	1	Bronchogram	1
Hyper-IgE syndrome, T helper/inducer reduction	1	Bronchogram	1
Ataxia teleangiectasia, low lymphocytes of all types, slightly reduced cell dependent neutrophil phagocytosis	1	V/Q scan	–
Moderately impaired T lymphocyte function	1	Bronchogram	1
Total secondary immune deficiencies:	3		
AIDS, IgG_2 deficiency	1	Bronchogram	–
Measles – adenovirus infection (Warner-Marshall syndrome)	1	Bronchogram	–
Measles – disseminated mycobacterium intracellulare infection	1	Histology	1
Total aspiration:	2		
Palm oil	1	Bronchogram	–
Pistachio nut	1	Bronchogram	–
Total without predisposing factors:	15		
Cystic fibrosis-like illness	1	V/Q scan	–
Pneumonia	12	7× Bronchogram	4
		2× CT scan	1
		3× VQ scan	–
Pertussis?	1	Bronchogram	1
Unknown	1	Bronchogram	–

*Additional immune defect.

Patients, methods, and results

PATIENTS

In an eight year period, over 4000 children with respiratory problems were referred to the paediatric respiratory department at the Royal Brompton National Heart and Lung Hospitals, London. In 41 children (1%) with chronic productive cough not due to cystic fibrosis, bronchiectasis was identified by bronchography, computed tomography, histology, or persistent radiological changes and matched ventilation and perfusion defects on isotope scans. Investigations to evaluate the severity of the lung damage and its causes are outlined in the table. Eighteen out of the 41 (44%) finally underwent lobectomy for localised disease.

CONGENITAL MALFORMATION

In six (15%) congenital malformations were detected, including arterial malinosculation,[1] cystic adenomatoid malformation, hamartomatous malformation, bronchomalacia – Williams-Campbell syndrome,[2] and absent pulmonary valve syndrome.[3]

PRIMARY CILIARY DYSKINESIA SYNDROME

Six had primary ciliary dyskinesia syndrome (PCDS), and one girl had completely absent cilia. Five of the seven children had situs solitus, and only two had dextrocardia.[4]

IMMUNE DEFICIENCY

Altogether 11 (27%) had immunological abnormalities: primary immune deficiency in eight, and three had secondary defects. Two patients with PCDS also had a serum dependent neutrophil killing defect, and the patient with hamartomatous malformation had a cell dependent neutrophil phagocytic defect.

FOREIGN BODY ASPIRATION

Two had bronchiectasis following aspiration episodes: one after a very delayed diagnosis of pistachio nut inhalation, and the second developed gross proximal bronchiectasis after having aspirated palm oil administered as a herbal remedy while unconscious, having had a convulsion. Altogether, a clear underlying cause for bronchiectasis was identified in 63% of cases.

WITHOUT UNDERLYING DISEASES

In 15 children (37%), no predisposing factors for chest infections were detectable. One girl

had lung disease similar to cystic fibrosis, with normal pancreatic function, and repeatedly negative sweat tests. We did not do genotype analysis which might have reintroduced the diagnosis of cystic fibrosis. Twelve had a history of severe pneumonia and one child had had a pertussis-like illness. Only one child with localised bronchiectasis had no detectable underlying abnormality, and no history of preceding aspiration or pneumonia.

Discussion

Chronic suppurative lung disease not due to cystic fibrosis is rare. Even in a specialist paediatric respiratory service, only 1% of referrals had obvious bronchiectasis. This is, nevertheless, an appreciable number of patients, many of whom had long delays in diagnosis. It is no longer necessary to do bronchograms which produce temporary deterioration in respiratory function, but non-invasive techniques including isotope ventilation and perfusion scans followed, if abnormal, by high resolution thin cut computed tomography of the lung should be employed for any child with unexplained chronic respiratory symptoms.

Major congenital lung malformation is uncommon and usually presents as an acute neonatal problem. Less severe malformations may occur more frequently; indeed, bronchial branch pattern defects occur in 0·3% of the population.[5] Some of our unexplained cases may have had minor congenital malformation leading to abnormal mucociliary clearance but this could only be established by bronchography or lobectomy. Two of our cases had defects identified at histology after surgery.

PCDS is frequently missed, particularly in those patients with levocardia and situs solitus.[4] Over the same period that we detected our seven cases with established bronchiectasis, we diagnosed an additional 12 with PCDS without irreversible lung damage. None of these have so far developed evidence of bronchiectasis over a 2–10 year period of follow up, using physiotherapy and frequent high dose, long course antibiotics.

We were surprised at the range of immunological abnormalities detected in our group of patients. However, we have identified the same range of abnormalities in other children presenting with recurrent respiratory tract infections but without evidence of bronchiectasis.[6] The serum dependent defects of neutrophil function and panhypogammaglobulinaemia are treatable and, if detected early, might have prevented irreversible lung damage.

Three patients had secondary immuno deficiencies. HIV infection is rare in childhood while measles is extremely common, and the most important cause of viral induced immunodeficiency worldwide. Delayed diagnosis of aspirated organic materials remains a worryingly frequent problem, and bronchiectasis is one of the consequences. Bronchoscopy should always be performed if there is any suggestion of foreign body aspiration. It is notable that pertussis does not appear as a significant cause of bronchiectasis other than possibly in one case. There is little evidence it has ever caused this problem in the antibiotic era. All the published literature on the association comes from a period before the availability of effective antibiotic treatment.

Bronchiectasis is only a descriptive label of the morphological defect in the airway. No stone should be left unturned in the pursuit of the underlying cause as specific treatment may fundamentally alter the natural history of the condition and prevent further lung damage.

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