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DIFFUSE PANBRONCHIOLITIS: THE HONG KONG EXPERIENCE

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Diffuse panbronchiolitis (DPB) is a recently recognised and rare chronic progressive suppurative airway disease predominantly encountered in Japan. Patients suffer from chronic sinobronchial sepsis progressing rapidly to respiratory failure and death unless treated with empirical macrolides. Little is known of the features of DPB in Chinese patients. Seven Chinese DPB patients (3F; mean age \pm SD, 48 \pm 18.6 yrs; all never smokers) were studied. Lung function assessment showed typical obstructive pattern (n=5) and air trapping (n=7). Typical bronchiolar infiltration by lymphocytes and plasma cells, and accumulation of foamy macrophages in the intra-luminal tissue were detected in open lung biopsy (n=2). Chest radiographs and HRCT revealed hyperinflation, diffuse nodules, bronchial thickening and dilatation, peripheral hypo-attenuation, and bronchiectasis (n=7). Radiological improvement, manifests as reduction in the nodular density and bronchial thickening, and persistence of other abnormalities such as air-trapping, were not accurately depicted by the classical Nakata or Akira classifications. The other "characteristic" features including: HLA-B54 (n=0), IgG subclass deficiency (n=0), raised CD4/CD8 T-lymphocyte ratio (n=0), cold haemagglutinaemia (n=0), raised IgA and IgG (n=0), and rheumatoid factor (n=0), were not present. Treatment with erythromycin led to excellent response in symptoms, lung function indices, and radiologically. We report the only non-Japanese Mongoloid series of well characterized DPB patients who have uncharacteristic investigation profiles. Our experience should help alert other clinicians in the investigation and management of DPB in non-Japanese patients.

ERYTHROMYCIN IS HIGHLY EFFICACIOUS IN CHRONIC BRONCHIECTASIS

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Bronchiectasis patients often suffer from regular sputum production, recurrent exacerbations, and progressive airway destruction. Erythromycin (EM) reduces mucus production in bronchorrhoea and improves lung function in diffuse panbronchiolitis, another suppurative airway disorder predominantly encountered in Japan. We have conducted this double-blind placebo-controlled study (500mg b.i.d.) to evaluate the effects of 8 week administration in steady state bronchiectasis. The patients (EM: 8F, mean age 50.2 \pm 15.1 yrs., n=11; and placebo: 10F, 59.3 \pm 15.6, n=10), who were mostly chronically infected with *Pseudomonas aeruginosa*, were assessed at 3 consecutive weekly visits before commencement of EM or placebo following which they were re-assessed at 4 and 8 weeks. The mean \pm SD for sputum volume, FEV₁, and FVC were 41.7 \pm 34.4, 0.9 \pm 0.6, 1.3 \pm 1.2; 30.2 \pm 25.1, 0.9 \pm 0.6, 1.2 \pm 0.7; and *30.3 \pm 27.4 ml/24h, *1.2 \pm 0.7 l, and *1.9 \pm 1.2 l respectively at 0, 4, and 8 weeks after EM therapy (*p<0.05 when compared with week 0). These improvements were not seen in the placebo patients. There were no significant changes in 24h sputum total bacteria, commensal bacteria, *Ps. aeruginosa*, leukocyte density, interleukin-1 α and -8, tumor necrosis factor α , and leukotriene B4. Low dose erythromycin therapy appears to reduce sputum volume and improve lung function without any effects on sputum inflammatory or microbial indices. Further studies are required to evaluate the long term effects of erythromycin therapy in bronchiectasis.