Title	Dysfunctional allele of the mannose binding protein (MBP) gene in rheumatoid arthritis (RA) - a preliminary study
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BRONCHIOLITIS OBLITERANS WITH ORGANISING PNEUMONIA (BOOP).

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BOOP is a recently established disease entity characterized pathologically by the presence of granulation tissue plugs in the lumens of small airways extending into alveolar ducts and alveoli, and clinical response to steroid therapy. However very little is known of the clinical features in Chinese patients. Six cases (mean age 55±7.2; 5F, 6NS) had been diagnosed at QMH and TGH and described here. The time of onset was 3.8±2.4 weeks with cough, dyspnoea, fever and type I respiratory failure. Clinical examination revealed crackles on auscultation in 6/6. Mean FEV_1/FVC and KCO were 82.8±13.28 and 76.5±25.49% respectively. Chest radiography showed consolidation in lower zones (4/6), upper zones (1/6), and left mid zone (1/6). High resolution computerized tommography showed ground glass appearances and consolidation (6/6) and pleural effusion (1/6). Diagnostic evidence was provided by histology (open lung 4/6, transbronchial biopsies 1/5) and radiology (HRCT 1/6). One case had *Mycobacterium tuberculosis* yielded in standard sputum culture but others had no positive microbiology. Therapy with rapid response (symptomatically and radiologically) to prednisolone (± aziothioprine) was achieved in all cases. Past medical history of rheumatoid arthritis (2/6), scleroderma (1/6), polymyositis (2/6), dermatomyositis (1/6) and breast cancer was noted. Our experience suggests that BOOP in Chinese patients may be strongly associated with rheumatological diseases and the female sex and these results differ from the Western experience. More cases need to be analyzed for further disease characterization on Chinese patients.

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DYSFUNCTIONAL ALLELE OF THE MANNOSE BINDING PROTEIN (MBP) GENE IN RHEUMATOID ARTHRITIS (RA) - a preliminary study. Carman Ho , Chan D, Mok CC, Chan SY*, Lau YL*, Wong RWS, Lau CS. Departments of Medicine and Paediatrics*, University of Hong Kong, Queen Mary Hospital, Hong Kong.

The MBP is a lectin secreted by the liver as an acute phase protein and is capable of activating the classical complement pathway independent of antibody and C1q. We have recently shown MBP deficiency may be an additional risk factor for SLE, probably due to impaired immune complex clearance. As RA is associated with immune complex deposition within the patient's joint, we have determined whether a dysfunctional allele of the MBP gene is associated with susceptibility to RA.
74 consecutive patients with definite RA and 123 healthy subjects were studied. The dysfunctional allele is characterised by the point mutation GGC → GAC at nucleotide 230 of exon 1. This was determined by the loss of the BanI restriction site in the PCR product. Serum levels of MBP was determined by ELISA. The MBP A-230 allele was present in 28 (38%) RA patients and in 28 (23%) controls (chi-square=5.16, p=0.02, OR= 1.77 (1.07-2.57, 95% ci). The presence of a dysfunctional MBP allele is confirmed by low phenotypic expression of MBP in the serum (controls: 370 (<10-655) vs RA 254 (78-352) ng/l; median (5th-95th percentile).
Our preliminary results suggest that the presence of a dysfunctional allelic form of MBP may be a minor risk factor for RA. However, more patients need to be studied. On the other hand, as MBP has been shown to bind with glycosylated immunoglobulins to form immune complexes in RA, MBP deficiency may be protective and this may explain the apparent low incidence of vasculitic complication of RA in Southern Chinese.