Whipple’s disease presenting with lung involvement

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Abstract

A 31 year old man presented with non-specific symptoms and a chest radiograph showing several pulmonary nodules which rapidly enlarged. Bronchoscopic examination showed endobronchial lesions, histological examination of which led to the diagnosis of Whipple’s disease. The patient made a gradual recovery with prolonged antibiotic therapy and the pulmonary lesions reduced in size. This is the first reported case of Whipple’s disease presenting with large pulmonary nodules and endobronchial lesions and is notable for the absence of gastrointestinal features.

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Whipple’s disease was first described in 1907 and usually presents at an advanced stage with features of malabsorption. Confirmation of its suspected bacterial aetiology has recently been obtained and the responsible organism identified. Polyrthralgia is a well recognised feature of the disease, while pulmonary manifestations are less common and often develop late in the course of the disorder. Pulmonary features are usually those of basal lung opacities, with or without pleural disease, and a necropsy study has shown a high prevalence of clinically unsuspected pleural disease. A single case presenting with lung symptoms has been described. However, discrete pulmonary lesions of this size have not been previously reported and endobronchial lesions have never been noted.

Discussion

The diagnosis of Whipple’s disease is usually made on the basis of histological examination of tissue obtained from biopsy samples of the small bowel. Although material obtained from the lung has been of diagnostic value in an earlier case, abnormal bronchial histology has not been previously reported in this condition. Three cases of Whipple’s disease with sarcoid-like granulomata in the lung have been reported. Two did not show PAS positivity.

Figure 1 Chest radiograph showing several large rounded lesions in both lung fields one month after presentation.
and there is some doubt as to whether the granulomata were due to direct infection or resulted from a delayed hypersensitivity reaction. Differential diagnosis should include Wegener's granulomatosis, although this diagnosis was rejected in our patient in view of the negative anticytoplasmic antibody test and his benign clinical course.

We feel that the concept that dissemination of infection to organs outside the gastrointestinal tract occurs late in the evolution of the disease should be challenged, as diarrhoea and biochemical evidence of malabsorption may be absent while involvement elsewhere may be significant, as our case suggests. Our patient did not consent to having biopsy samples taken of the small bowel until several months of treatment had been completed, in spite of being persistently requested to do so. The absence of diagnostic changes at this stage is not surprising and is consistent with his excellent clinical and radiological response to antibiotics. We suggest that this case represents a unique presentation of Whipple's disease and that pulmonary manifestations of this disorder should be extended to include pulmonary nodules and endobronchial lesions which may be mistaken for features of malignancy. Pulmonary involvement may be the presenting feature of the condition and offers the opportunity to diagnose the disease at a stage where response to treatment is likely to be both more rapid and complete than if it is delayed until manifestations of gastrointestinal involvement appear.