Whipple’s disease presenting with lung involvement

C A Kelly, M Egan, J Rawlinson

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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Keywords: Whipple’s disease, pulmonary nodules, endobronchial lesions.

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. Polyrarthralgia 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.

Case report
A 31 year old man with no previous complaints presented with a four month history of a dry cough with progressive weight loss, malaise, and poor appetite. On examination he was cachetic weighing 52 kg with no specific abnormalities. Investigations revealed a normochromic anaemia, raised ESR, and an abnormal chest radiograph showing several discrete nodules. He was referred to the local urologist because of concern about possible pulmonary metastases from a teratoma or hypernephroma. No evidence of underlying malignancy was found. Cough became an increasingly troublesome feature and a repeat chest radiograph showed an alarming increase in the size of the pulmonary lesions (fig 1). At this stage there were no specific respiratory signs and pulmonary function was normal. There were no biochemical indices of malabsorption and he denied gastrointestinal symptoms. At bronchoscopic examination several raised yellow endobronchial lesions were seen mainly at the sub-carinae of the lobar divisions of the bronchial tree. A biopsy sample was taken of one of the lesions and sent for histological examination. This showed multiple foamy macrophages which were PAS positive and diastase resistant without granulomata or giant cells and with no evidence of malignancy (fig 2). In this clinical setting it was felt that a diagnosis of Whipple’s disease was justified, although there was insufficient material to confirm this with electron microscopy. He repeatedly refused to undergo upper gastrointestinal endoscopy.

Treatment with penicillin and tetracycline was commenced and over the next four months he made a gradual recovery with considerable improvement in the appearance of his chest radiograph. At this stage he agreed to have biopsy samples taken of the small bowel. Macroscopically the duodenum was normal and tissue from the second part of the duodenum was also normal microscopically with none of the features found in the endobronchial biopsy samples. He remains asymptomatic 15 months after presentation weighing 71 kg with a normal chest radiograph three months after stopping treatment.

Figure 1 Chest radiograph showing several large rounded lesions in both lung fields one month after presentation.
BOOK REVIEW


I very much enjoyed reading this book. It is, like the proverbial football match, an affair of two halves. All but one of the first 10 chapters is written by the editor, and in these he traces up to the 17th century the story of the struggle to understand the nature and purpose of breathing. After the customary coverage of Babylonian, Egyptian and Greek physiology, there is a chapter on Galen which helps to explain how his ideas, although wrong, became so authoritative. However, Dr Proctor's chief loves are clearly the English physiologists of the 17th century, particularly Mayow, and he devotes five chapters to them, providing a lot of detail on their lives and achievements, but unfortunately also with much repetition. He tackles the question of why they drifted into different fields of enquiry after coming so close to understanding the whole mystery, leaving it for Lavoisier, a century later, to provide the final essential missing piece in the jigsaw with the discovery of oxygen.

The second half of the book consists of eight chapters by various authors who bring the story up to date in their respective fields from pulmonary surfactant to regulation of breathing. These vary in quality; that by Permutt on the pulmonary circulation is outstanding. The book is lavishly illustrated and well produced, apart from the chapter on da Vinci in which the 14 illustrations, their legends, and their text references have been shuffled into total non-correspondence which is disappointing in a book that is so exorbitantly priced. – SF

NOTICE

1st Congress of Surgery of Bosnia and Herzegovina

This Congress, which was originally planned for 8–11 October 1995, will now take place in Sarajevo on 12–15 May 1996. For further information please contact the Congress Committee. Telephone: 387 71 44 55 22. Fax: 387 71 47 19 76.
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