Granulomatosis with polyangitis (Wegener’s)

A 51-year-old woman was admitted to our hospital with general fatigue, fever and cough. Upon examination, sinusitis, papules of the planta, polymorph leucocytosis, hypoalbuminaemia and an elevated level of PR3-antineutrophil cytoplasmic antibodies (ANCAs) were found. Her chest radiograph (figure 1A) and thoracic CT (figures 1B 2A) demonstrated a large area of consolidation with air bronchogram and cavitation in the right lung field. Specimens of biopsy from skin, lung and kidney showed necrotising granulomatous vasculitis. The patient was given a diagnosis of Wegener granulomatosis (WG) and treated with corticosteroid and cyclophosphamide. The consolidation resolved with residual scarring and was transformed into cystic lesions (figure 2B).

The lung is the most commonly involved organ in WG, and the most representative CT findings are multiple nodules, occasionally with cavitation. Areas of consolidation and ground-glass attenuation are also frequent findings, which represent granulomatous changes and pneumonia. In untreated disease, the lesions tend to increase in size and number, and sometimes progress with coalescence of adjacent opacities. The large consolidation observed in the present case may be explained by this mechanism, and cavities may represent necrosis or bronchiectasis.

Learning points

- WG may take a variety of forms, including a large area of consolidation.
- Physicians should consider WG in the differential diagnosis of consolidation.
- WG may regress with extensive cystic change.

Figure 1  Chest radiograph (A) and coronal view of thoracic CT (B) at initial presentation.

Figure 2  Thoracic CT at initial presentation (A) and post-treatment (B).
On treatment, the lesions disappear completely in the majority of cases, whereas occasionally they may regress with scarring constituted by linear opacities and bronchiectasis. The post-treatment extensive cystic change was also considered to be rare.

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Correction notice This article has been corrected since it was published Online First. The title has been amended to ‘Granulomatosis with polyangitis (Wegener’s)’.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Accepted 19 April 2010
Published Online First 29 July 2010


REFERENCES

Pulmonary puzzle

All that wheezes is not asthma: the value of curves

CLINICAL PRESENTATION
A 64-year-old woman, never smoker, with a history of fully treated tuberculosis at 20 years of age attended our department for lung function testing.

She had recently experienced several episodes of intermittent breathlessness and wheeze presumed to be due to asthma and was referred to a respiratory physician following an emergency department visit during one of these episodes. She did not have any recent weight loss, night sweats, purulent sputum or haemoptysis. Her dyspnoea and wheeze (inspiratory and expiratory) had been refractory to inhaled corticosteroids and both short and long acting β2 agonists.

Lung function using American Thoracic Society criteria (Sensormedics, Yorba Linda, California, USA) showed mild air flow obstruction with forced expiratory volume in one second (FEV1) of 1.67 l (82% predicted), forced vital capacity (FVC) of 2.92 l (120% predicted) and a FEV1/FVC ratio of 57%. Total lung capacity was normal (110% predicted). Her maximum flow-volume curves had a peculiar and reproducible appearance, particularly in the inspiratory phase (figure 1). There was no change following the use of a bronchodilator.

QUESTION
What is the diagnosis and which specific clinical sign might she have had?
See page 567 for the answer

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Competing interests None.

Patient consent Obtained.

Ethics approval Ethics approval was provided by the Sydney Local Health District Human Research Ethics Committee.

Contributors All authors contributed to the conception, design, analysis and interpretation of data, carried out critical appraisal of the manuscript, and approved the final version to be published.

Provenance and peer review Not commissioned; externally peer reviewed.

Published Online First 23 November 2011

Thorax 2012;67:564. doi:10.1136/thx.2011-201097

Figure 1 Maximal flow-volume loops before (‘Pre’) and after (‘Post’) bronchodilator. Reference lines (with SD bars) are linked to plethysmographic total lung capacity. There is a long expiratory tail with marked reduction in the end inspiratory flow.
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Thorax 2012 67: 563-564 originally published online July 29, 2010
doi: 10.1136/thx.2009.133496

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