Rare disease

Spontaneous pneumothorax in a patient with granulomatosis with polyangiitis

Hasan Kahraman,¹ Mehmet Fatih Inci,² Mahmut Tokur,³ Gozde Yildirim Cetin⁴

¹Department of Chest Diseases, Sütçü İmam University, Kahramanmaraş, Turkey

²Department of Radiology, Sütçü İmam University, Kahramanmaraş, Turkey

³Department of Chest Surgery, Sütçü İmam University, Kahramanmaraş, Turkey

⁴Department of Rheumatology, Sütçü İmam University, Kahramanmaraş, Turkey

Correspondence to Dr Hasan Kahraman, drhasankahraman@hotmail.com

Summary

Granulomatosis with polyangiitis (GPA) (Wegener's) is a multiorgan system disease of unknown aetiology characterised by granulomatous inflammation, tissue necrosis and vasculitis. The characteristic lung parenchymal lesions of GPA are firm spherical nodules that may cavitate. Pneumothorax (PX) can develop as a quiet rare complication of cavitary nodules. Our case admitted to our clinic with the diagnosis of GPA showing cavitary pulmonary mass. While taking immunosuppressive treatment, spontaneous PX on left lung was developed. A closed chest tube was inserted to the left lung for expansion of PX. Even after 30 days, the left lung did not re-expand and wedge resection with thoracotomy was conducted and the closed chest tube was still in the left lung. On the seventh day, empyema emerged as a complication and, with appropriate treatment, the patient became well. In GPA patients taking immunosuppressive medication, PX is a serious complication and requires aggressive therapy with broad-spectrum antibiotics.

BACKGROUND

Granulomatosis with polyangiitis (GPA) (Wegener's), is a disease of unknown aetiology characterised by necrotising granulomatous angiitis of the upper and lower respiratory tracts, systemic small-to-medium-sized vasculitis and necrotising glomerulonephritis. Other organ systems that may be involved in GPA include; joints, eyes, skin, nervous system, heart and, less commonly, the

gastrointestinal tract, lower genitourinary tract, parotid glands, thyroid, liver or breast.¹ Depending on the case series, the most commonly reported chest radiograph findings are nodules that may cavitate, patchy or diffuse opacities that sometimes with an air bronchogram and lobar or segmental atelectasis. Other less common findings include pleural effusions, hilar adenopathy and spontaneous pneumothorax (PX) that results from rupturing of the cavitary nodules.^{2 3}

Figure 1 Chest radiogram showing a lobulated mass, with a diameter of 9 cm, presenting cavity and clear boundaries in the upper zone of the left lung.

Figure 2 Chest radiogram presenting cavitary mass and pneumothorax in the left lung.

Figure 3 CT showing cavitary mass and pneumothorax in the left lung.

CASE PRESENTATION

A 32-year-old man with complaints of cough, hearing loss and right-side facial paralysis applied to an otolaryngologist in August 2011 and partial response was detected with short-course low-dose corticotherapy. In November 2011, swelling and pain of both ankles emerged and he attended to orthopaedia and then consulted the rheumatology clinic. He was hospitalised at the rheumatology clinic and diagnosed as GPA with pulmonary involvement and treatment was started with cyclophosphamide 1 g monthly and prednisone 80 mg daily. Then, he was admitted to our University Hospital Chest and Rheumatology Clinics in January 2012 with complaints of cough, sputum and shortness of breath.

On physical examination, breath sound in the left upper lung zone was coarse and rales could be heard. Examinations of the heart and abdomen were normal. His chest radiogram showed a lobulated mass, with a diameter of 9 cm, presenting cavity and clear boundaries in the upper zone of the left lung (figure 1). The blood analysis showed haemoglobin 15.2 g/dl, platelets 147 K/ μ l,

C reactive protein 90 mg/dl and erythrocyte sedimentation rate 77 mm/h. In the urinalysis, only microhaematuria was present. In sputum analysis, acid-fast bacteria of both smears and cultures were negative. The result of C-antibodies to neutrophil cytoplasmic antigens (ANCA) test was 26 (N 0–20 U/ml). The treatment of the patient had been continued. In April 2012, he was admitted to our Emergency Department with sudden onset complaints of dyspnoea and dry cough.

INVESTIGATIONS

On physical examination, the breath sound was diminished on the left side. Blood pressure was 120/80 mm Hg, pulse rate was 110/min. Plain chest film was taken and cavitary mass and PX were detected in the left lung (figure 2). He was hospitalised, a pleural drainage was applied and CT was taken (figure 3). The pleural fluid presented exudate characteristics and the serial cultures for blood, sputum and pleural fluid were negative. Cyclophosphamide treatment was stopped, corticosteroid dose was diminished to 8 mg/day and antibiotic was added. After 30 days, without pulmonary re-expansion, the patient was evaluated by the department of thoracic surgery, which performed wedge resection with thoracotomy. On pathological investigation of the specimen, there was transmural inflammation of the vessel wall with destruction of the elastica and multinucleated giant cells in the necrotic area (figure 4A,B). After the operation, PX did not resolve because of generation of pleural leakage and, on the seventh day, pleural effusion became purulent and *Pseudomonas aeruginosa* was cultured from empyema material. According to the culture results, imipenem/cilastatin therapy was added, and the patient improved and the tube was removed after the 45th day after operation. He was discharged from the hospital after 3 months (figure 5), and the outpatient treatment has been continuing for GPA.

DISCUSSION

GPA is a complex immune-mediated disorder which involves many interrelated pathogenic pathways including genetic, cell-mediated, neutrophil-mediated, humoral and environmental. A part of this immune response is directed to previously secured epitopes of neutrophil granule proteins and leads to high titre of ANCA which is found in 80–90% of all patients.⁴

Figure 4 (A) There is a transmural inflammation of the vessel wall with destruction of the elastica. (B) Multinucleated giant cells are seen in necrotic areas.

Figure 5 Chest radiogram presenting elevated left diaphragm, displacement of heart and mediastinum to the left, and persevered partial pneumothorax on the left.

Hearing loss in GPA generally results from chronic otitis media leading to tympanic membrane abnormalities or fusion of ossicles. Less frequently, vasculitis may also cause sensorineural deafness. Hearing loss, in any form, is found in about 30–40% of patients. In our patient, the first symptom was variable bilateral hearing loss and with corticosteroid treatment, it first decreased partly and then deteriorated and finally, permanent hearing loss occurred.

The lungs are major target organs of GPA and pulmonary nodules, either single or multiple are most common manifestations with a size that ranges from 0.3 to 12 cm.² Central necrosis of nodules may present and then cavitate in 25–50% of patients.⁵ In our case, mass with central necrosis, in a diameter of 9 cm, was present in the upper zone of the left lung. PX is quite rarely seen in GPA patients and emerged from rupturing of cavitory nodules present near the pleura. Owing to the high frequency of complications of even small PX, caution is needed in cases taking immunosuppressive treatment.⁶ It is accepted that infections are common and cause potentially severe complication of all cyclophosphamide-included regimens. The reported frequency of cyclophosphamide-associated infection varies according to the stage and the severity of the underlying disease, the degree of immunosuppression and concomitant glucocorticoid regimens.⁷ For this reason, the treatment of PX should include the installation of pleural drainage for at least 2 weeks and appropriate antibiotic treatment. In patients using cyclophosphamide, the antibiotic treatment should cover opportunistic pathogens like

Pseudomonas, *Escherichia coli*, *Klebsiella* and *Staphylococcus aureus* which are frequently seen as infectious pathogens.⁷ The prognosis is poor especially for infectious complications and is greatly dependent on the basal situation of the patient. In our case, PX emerged during cyclophosphamide and glucocorticoid treatment. Even though our patient had been taking antibiotic therapy, on the seventh day, empyema appeared and in culture *P aeruginosa* was detected. Then, antibiotic therapy was changed and he became well.

In conclusion, PX, although seen quite rarely in GPA, is possible especially in cases with cavitory nodules which are particularly close to the pleura. In this kind of immunosuppressed situation, PX is a serious complication and requires aggressive therapy with broadspectrum antibiotics.

Learning points

- ▶ Pneumothorax (PX), although seen quite rarely in GPA, is possible especially in cases with cavitory nodules which are particularly close to the pleura.
- ▶ In GPA patients taking immunosuppressive medication, PX is a serious complication and requires aggressive therapy with broadspectrum antibiotics.
- ▶ In patients using cyclophosphamide, the antibiotic treatment should cover opportunistic pathogens like *Pseudomonas*, *Escherichia coli*, *Klebsiella* and *Staphylococcus aureus*, which are frequently seen as infectious pathogens.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Hoffman GS, Kerr GS, Leavitt RY, et al. Wegener granulomatosis: an analysis of 158 patients. *Ann Intern Med* 1992;**116**:488–98.
2. Cordier JF, Valeyre D, Guillevin L, et al. Pulmonary Wegener's granulomatosis. A clinical and imaging study of 77 cases. *Chest* 1990;**97**:906–12.
3. Daum TE, Specks U, Colby TV, et al. Tracheobronchial involvement in Wegener's granulomatosis. *Am J Respir Crit Care Med* 1995;**151**:522–6.
4. Tarabishy AB, Schulte M, Papaliadis GN, et al. Wegener's Granulomatosis: clinical manifestations, differential diagnosis, and management of ocular and systemic disease. *Surv Ophthalmol* 2010;**55**:429–44.
5. Yousem SA. Wegener's granulomatosis. In: Churg AM, Myers JL, Tazelaar HD, Wright JL (eds) *Thurlbeck's pathology of the lung* 3rd edn. New York: Thieme, 2005:371.
6. Garcia MB, Tirado VV, Suárez LA, et al. Spontaneous pneumothorax in Wegener's granulomatosis: case report and literature review. *Semin Arthritis Rheum* 2011;**41**:455–60.
7. Sezer I, Kocabas H, Melikoglu MA, et al. Spontaneous pneumothorax in Wegener's granulomatosis: a case report. *Mod Rheumatol* 2008;**18**:76–80.

Copyright 2012 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Kahraman H, Inci ME, Tokur M, Cetin GY. Spontaneous pneumothorax in a patient with granulomatosis with polyangiitis. *BMJ Case Reports* 2012; 10.1136/bcr-2012-007765, Published XXX

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow