Spontaneous Pneumomediastinum in a Case of Mixed Connective Tissue Disorder

Col V Vardhan*, Maj V Marwah⁺, Col SP Rai[#], Col AP Patil^{**}, Col R Murlidhar⁺⁺, M Pethe^{##}

MJAFI 2008; 64 : 367-368

Key Words : Mixed connective tissue disorder; Interstitial lung disease; Spontaneous pneumomediastinum

Introduction

Spontaneous pneumomediastinum refers to dissection of free air from the airways or lung parenchyma into the mediastinal structures without any antecedent thoracic trauma or surgical intervention. It is a rare condition associated with several pulmonary disorders such as bronchial asthma and emphysema and it can also occur in apparently healthy individuals [1]. Very rarely intestinal lung disorders may be associated with a spontaneous pneumomediastinum [2]. We describe a patient with mixed connective tissue disorder (MCTD) who developed pneumomediastinum during treatment.

Case Report

A 36 yrs old male, a known case of MCTD on treatment with oral steroids and methotrexate for six months, presented with a two month history of progressively worsening exertional breathlessness. He had no wheeze, chest pain, fever or other systemic complaints. There was no significant occupational exposure to any organic or mineral dust and he did not have any pets at home. He had bilateral basilar gravity dependent end inspiratory crackles on examination. A chest radiograph showed reticulonodular opacities in both mid and lower zones. High resolution computed tomogram of the chest showed reticulonodular opacities with honeycombing in lower lobes. Spirometry revealed a restrictive pattern of moderate severity. Arterial blood gas analysis on room air revealed mild hypoxemia (PaO₂ 64.3 mm Hg). Pulse oximetry showed oxygen saturation of 98% on room air and after six minute of walk he desaturated to 80%. Fibreoptic bronchoscopy was normal and transbronchial lung biopsy was inconclusive. Post bronchoscopy check chest radiograph did not reveal any evidence of pneumothorax or pneumomediastinum. Serum anti nuclear antibody, ribonucleoprotein (RNP), anti Smith antibody (anti Sm) and anti U₁ ribonucleoprotein (U₁ RNP) were positive. A diagnosis of MCTD with interstitial lung disease was made and tablet hydroxychloroquine 400 mg twice

a day was added to oral steroids.

After 19 days of fibreoptic bronchoscopy and transbronchial lung biopsy, he noticed a nasal intonation of voice and a diffuse swelling over face, chest and the right side of the neck. There was no history of any associated chest pain, bout of vigorous coughing or severe physical exertion. On examination he was hemodynamically stable. The swelling was soft, non tender and a crepitus was palpable. Chest examination revealed bilaterally symmetrical breath sounds with bilateral basilar end-inspiratory crackles. Hamman's crunch was positive, chest radiograph showed free mediastinal air along the left cardiac border (Fig.1) and neck radiograph revealed subcutaneous emphysema around the soft tissues of neck (Fig.2). There was no pneumothorax. Lateral radiograph of the chest revealed lucencies in retrosternal space (Fig.3). A computed tomogram of thorax showed bilateral reticulonodular interstitial opacities and evidence of pneumomediastinum and translucencies in pre vertebral space (Fig.4). He was diagnosed to have a spontaneous pneumomediastinum. Patient was managed with

Fig. 1 : Chest radiograph showing free mediastinal air along the left cardiac border and continuous diaphragm sign

*Reader (Department of Internal Medicine), AFMC, Pune, ⁺Graded Specialist (Respiratory Medicine), MH Namkum. ^{##}Senior Resident (Respiratory Medicine), [#]Senior Advisor (Medicine & Respiratory Medicine), ^{**}Classified Specialist (Medicine & Respiratory Medicine), ⁺⁺Senior Advisor (Clinical Immunology & Medicine), Military Hospital (CTC), Pune.

Fig.2 : Neck radiograph (AP view) showing subcutaneous emphysema around the soft tissues of neck

Fig.3 : Lateral radiograph of the chest showing lucencies in retro-sternal space

Fig. 4 : Computed tomogram of thorax showing bilateral reticulonodular interstitial opacities with pneumo-mediastinum and translucencies in pre vertebral space.

oxygen inhalation and antibiotics. His subcutaneous emphysema resolved completely in the next one week.

Discussion

MCTD is an overlap syndrome characterized by combinations of clinical features of systemic lupus erythematosus, systemic sclerosis, polymyositis and rheumatoid arthritis. These patients have very high titers of circulating auto antibodies to nuclear ribonucleoprotein (RNP) antigen. Approximately 85% patients have pulmonary involvement which is often asymptomatic while some develop interstitial lung disease. We could not find any case of spontaneous pneumomediastinum in MCTD in literature. Yamanishi et al [3,4], suggested that the weakening effect of steroids on lung interstitial tissues might cause pneumomediastinum. Alternatively, fibrosis itself leads to cyst formation and rupture of such cysts can result in pneumomediastinum. Alveolar rupture and subsequent air leak into the perivascular interstitium

eventually leads to pneumomediastinum under various conditions. Honeycombing and violent cough are considered to be predisposing factors to the rupture.

Conflicts of Interest

None identified

References

1. Pierson DJ. Pneumomediastinum. In : Murray JF, Nadel JA, editors. Textbook of Respiratory Medicine. 2nd ed. Philadelphia: WB Saunders company, 1994;2250-65.
2. O'Connor I, Thomas GO. Spontaneous pneumomediastinum in a patient with fibrosing alveolitis. Respiratory Medicine 1993; 87: 313-14.
3. Yamanishi Y, Maeda H, Konishi F, Hiyama K, Yamana S, Ishioka S, et al. Dermatomyositis associated with rapidly progressive fatal interstitial pneumonitis and pneumomediastinum. Scand J Rheumatol 1999; 28:58-61.
4. Kono H, Inokuma S, Nakayama H, Suzuki M. Pneumomediastinum in dermatomyositis: association with cutaneous vasculopathy. Ann Rheum Dis 2000; 59:372-6.

ONLINE MANUSCRIPT MANAGEMENT SYSTEM

MJAFI announces the availability of Trial version of Online Manuscript Management System by the end of October 2008. The system will enable online interaction between the Authors, Editors, Referees and the Readers. We request all the Readers, Authors and Referees to register at **mjafi.org** by the end of November 2008. Registration is a simple one page proforma with separate pages for Readers, Authors and Referees. A detailed help file is available on the website for assistance. The Online Manuscript Management System is based on Open Journal System developed and licensed under the terms of the GNU General Public License. We are confident of migrating completely to online article submission and review by 01 January 2009 and solicit your cooperation for the same.