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Case report

Tracheomegaly in association with rheumatoid arthritis

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Abstract. Herein we present a case of tracheomegaly seen in a patient with rheumatoid arthritis. To the authors' knowledge, and from a review of the literature, this combination has not been previously described.

Key words: Tracheomegaly – Rheumatoid arthritis – Thorax – CT

Introduction

Tracheobronchomegaly is a distinctive condition that consists of marked dilation of the trachea and central bronchi, in association with chronic respiratory tract infections. It probably results from a congenital defect of the elastic and muscle fibers within the tracheal and bronchial walls. It is occasionally seen together with connective tissue disease.

Case report

A 57-year-old Caucasian man who had rheumatoid arthritis for 20 years complained of progressive shortness of breath and cough for 2 years. He had smoked 35 pack/years tobacco, but he was a non-smoker for the previous 6 years. His arthritis involved hands (primarily proximal interphalangeal joints), elbows, knees, temporomandibular and ankle joints, and he had long-standing subcutaneous nodules over the elbows and hands. He was treated with nonsteroidal anti-inflammatory drugs for approximately 20 years and with prednisone for approximately 6 years. General physical examination revealed arthritic deformities of both hands and rheumatoid nodules over elbows. There was no cyanosis, pallor, edema, clubbing, or lymphadenopathy.

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Examination of the chest disclosed bilateral diffuse end-inspiratory crackles. Certain laboratory tests included the following: Hb, 13.4 g/dl; white blood count, 12.9×10^9/l; platelet count, 604×10^9/l; erythrocyte sedimentation rate, 75 mm in 30 min and 116 mm in first hour; rheumatoid factor, 198 (normal: < 32 IU/ml); anti streptolysin-O, < 200; C-reactive protein, 0.085; acid resistence bacteri, negative. Chest radiograph showed right upper lobe pleural thickening, bilateral hyperinflation, and reticulonodular opacities on the other areas, but it did not show tracheomegaly. Additionally, CT showed tracheomegaly as well as destructive and emphysematous areas. Also, high-resolution CT (HRCT) showed bronchiectasis, interlobular septal thickening, honeycombing, and mosaic perfusion areas. Mediastinal slices showed aortic valve and lumen calcifications. The intrathoracic trachea was dilated from thoracic inlet for 3 cm with a slightly deformation on cross-sectional configuration. The maximum coronal diameter was 39 mm and the maximum anteroposterior diameter 49 mm. Dilatation did not extend into the major bronchi. The maximum diameters of the left and right main bronchi were normal. The walls of the trachea showed no evidence of calcification, thickening, or pouching. Mediastinal lymphadenopathy was not detected (Fig. 1).

Discussion

Pleuropulmonary manifestations in rheumatoid arthritis have been well reported and include pleural abnormalities, diffuse interstitial fibrosis, necrobiotic nodules, pulmonary arteritis, obliterative bronchitis, and cardiac enlargement [1]. Other less common manifestations are pneumothorax due to erosion by parenchymal nodules, amyloidosis, pulmonary vasculitis, and drug-related problems [2]. Bronchiectasis has also been reported to be more common in rheumatoid arthritis [3]. Rheumatoid nodules in the trachea has been reported by Ip et al. [4].

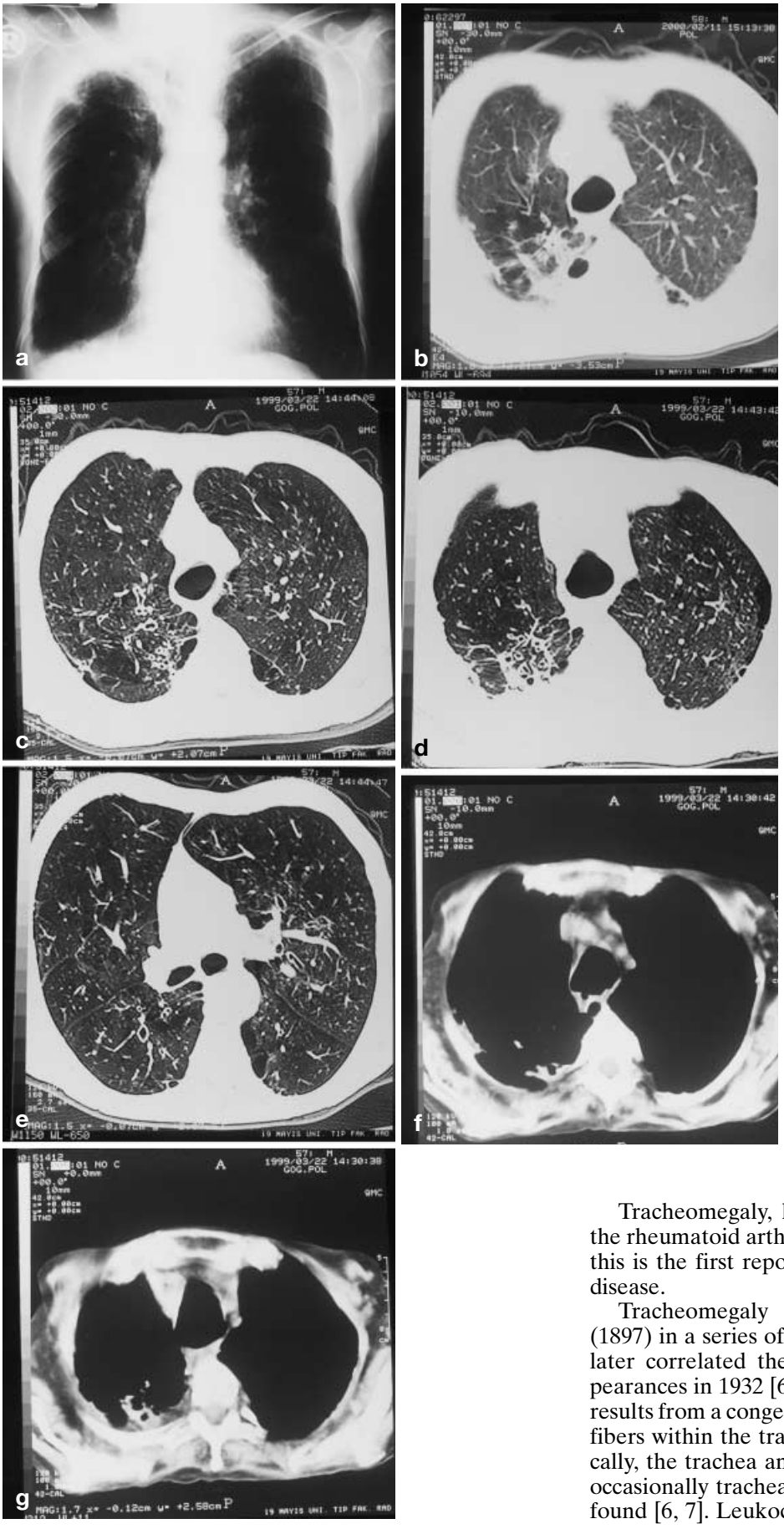

Fig. 1 a–g. A 57-year-old man with tracheomegaly. **a** Posteroanterior radiograph of chest shows pleural thickening in right upper lobe, bilateral hyperinflation, depressed diaphragm, and decreased pulmonary vascular markings. **b** Conventional CT scan (lung window) in upper lobe level shows tracheomegaly, destructive areas, and parascatriciel emphysema posterior segment in the right upper lobe. **c–e** High-resolution CT scans at three levels: **c** honeycombing in right posterior segment in the right upper lobe; **d** interlobular septal thickening in different areas; **e** scan through midzone shows mosaic attenuation and bilateral tubular bronchiectasis. **f,g** Conventional CT scans (mediastinal window) at two levels shows prominent dilatation of trachea, and pleural thickening and retractions in upper zones

Tracheomegaly, however, has not been reported in the rheumatoid arthritis to date. As far as we are aware, this is the first report of tracheomegaly in rheumatoid disease.

Tracheomegaly was first described by Czyhlarz (1897) in a series of autopsy studies [5]. Mounier-Kuhn later correlated the endoscopic and radiographic appearances in 1932 [6]. Tracheobronchomegaly probably results from a congenital defect of the elastic and muscle fibers within the tracheal and bronchial walls [7]. Typically, the trachea and central bronchi are involved, but occasionally tracheal or bronchial involvement alone is found [6, 7]. Leukocytoclastic vasculitis often is report-

ed within rheumatoid nodules and in adjacent tissues and is believed to be due to immune complex deposition and pathogenetically responsible for the development of the nodules [8].

Radiographically, the condition is characterized by a greatly increased tracheal caliber, measuring 35–50 mm or more in diameter [9]. The diameter of the trachea is normally 10–27 mm in adults (mean 19.5 mm in males, 17.5 mm in females) [10]. This compares with the values described for CT by Vock et al. and they give the mean maximum normal coronal diameter of the trachea as 21.8 mm for men and 19.4 for women [11].

The pathogenesis of tracheomegaly in rheumatoid arthritis remains speculative. We found only four case reports of tracheomegaly in association with a connective tissue disorder, two of these being associated with ankylosing spondylitis [12] and one with acquired cutis laxa in a child [13], and the other with Ehlers-Danlos syndrome in the original description by Mounier-Kuhn [6]. We are unaware of any case of tracheomegaly occurring in association with rheumatoid arthritis. Padley et al. [14] and Fenlon et al. [12] postulated that tracheobronchomegaly and ankylosing spondylitis may represent part of a spectrum of connective tissue disorders, and that their association may not be by chance.

Following our observations in the case described, we reviewed the literature for cases of tracheomegaly with rheumatoid arthritis and with connective tissue disorders. Our data support this suggestion. It is possible that tracheomegaly and rheumatoid arthritis are both parts of the spectrum of connective tissue disease, and as such their association may not be by chance.

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