Thoracic manifestation of Wegener’s granulomatosis: CT findings in 30 patients

Abstract  Our objective was to describe the CT findings of thoracic Wegener’s granulomatosis. At presentation, both conventional and thin-section CT scans were available in 30 patients with Wegener’s granulomatosis. Serial CT scans (range of intervals: 1–25 months, mean 4.5 months) were available in 20 patients. The initial and follow-up CT scans were analyzed retrospectively by two observers in terms of pattern and extent of parenchymal and airway lesions. Positive CT findings were seen in 29 of 30 (97%) patients at initial presentation. The most common pattern was nodules or masses seen in 27 of 30 (90%) patients. They were multiple in 23 of 27 (85%) patients, bilateral in 18 (67%), subpleural in 24 (89%), and peribronchovascular in 11 (41%) in distribution. Bronchial wall thickening in the segmental or subsegmental bronchi was seen in 22 (73%) patients. Large airways were also abnormal in 9 (30%) patients. Patchy areas of consolidation and ground-glass opacity were seen in 7 (23%) patients, respectively. In 17 of 20 (85%) patients in whom follow-up CT scans were available, the parenchymal or airway lesion showed complete or partial improvement with treatment. The CT findings of Wegener’s granulomatosis, although multiple and variable, consist mainly of bilateral subpleural or peribronchovascular nodules or masses and bronchial wall thickening in the segmental or subsegmental bronchi. Parenchymal and airway lesions improve with treatment in most patients.

Keywords  Lung · Granuloma · CT · Granuloma vasculitis · Wegener’s granulomatosis

Introduction

Wegener’s granulomatosis (WG) is characterized clinicopathologically by the triad of granulomatous necrotizing and ulcerative inflammatory process of the respiratory tract and the internal organs, generalized necrotizing granulomatous vasculitis, and a usually focal, necrotizing glomerulonephritis [1]. The lung is the most commonly involved organ and almost all patients have either pulmonary or upper airway involvement [2].
Wegener’s granulomatosis in the thorax presents with wide variety of radiologic findings [3, 4, 5, 6, 7, 8, 9, 10, 11]. More than one pattern of radiologic findings may be seen on CT scans at presentation or during the course of the disease. Because chest radiographs fail to show the exact pattern and the extent of thoracic involvement, CT is particularly useful in the assessment of thoracic involvement of the disease [12].

Recent improvement in the treatment of the disease with combined cytotoxic and corticosteroid therapy has led to good prognosis and long-term survival; however, serial CT findings of WG have been published in a limited number of papers [13]. The purpose of this study was to present the initial and follow-up CT findings of thoracic WG in a relatively large number of patients.

Materials and methods

Between March 1993 and April 2001, 30 patients with WG were seen in seven tertiary hospitals. The patients were identified retrospectively by computerized search of all patients’ records. They were 16 men and 14 women (age range 15–80 years, mean age 54.3 years). The diagnosis of WG was made in accordance with the 1990 American College of Rheumatology [14] and the Chapel Hill criteria [15]. Histologic verification was done with biopsies in 29 patients. The positive biopsy specimens were obtained in the lungs including airways (n=15 [13 patients]), paranasal sinuses or nasal cavity (n=15), kidneys (n=8), and skins including eyelids (n=2). C-antineutrophil cytoplasmic antibodies (c-ANCA) were evaluated in the peripheral blood at the time of the initial diagnosis in 28 patients. They were elevated in 24 patients. In the remaining 4 patients, they were not elevated.

Twenty-nine patients had symptoms and signs at admission including cough (n=15), nasal obstruction (n=11), fever (n=6), fatigue (n=6), dyspnea (n=4), hemoptysis (n=4), proptosis (n=3), headache (n=2), scleral ulcer (n=2), sputum (n=2), generalized edema (n=2), and diarrhea (n=1). The remaining 1 patient, in whom pulmonary abnormalities were detected incidentally on routine chest radiograph, was asymptomatic. The involved organs were the lungs or airways (n=29), the paranasal sinuses (n=26), kidneys (n=14), eyes (n=5), skin (n=1), and oral cavity (n=1). Both cyclophosphamide and corticosteroids were given in 23 patients and corticosteroids only were given in 7 patients.

Computed tomography scans were obtained with different machines, but mainly with three: GE 9800 or HiSpeed Advantage scanner (General Electric Medical Systems, Milwaukee, Wis.), a X-Vigor Scanner (Toshiba, Tokyo, Japan), and Somatom Plus 400 scanner (Siemens, Inselin, Germany). At initial evaluation, conventional CT scans were obtained from the lung apices to the middle portion of both kidneys with 7- to 10-mm collimation. The scans were obtained with (n=12) and without (n=18) intravenous injection of contrast medium (a total of 100 ml of 30 g iodine). Thin-section (1- to 2-mm thickness) CT scans were also obtained through the thorax at 20-mm intervals before (n=30, with full inspiration) and after (n=7, with full expiration) conventional CT scans. Expiratory thin-section CT scans were obtained only in 7 patients suspected clinically of airway involvement. Follow-up CT scans were available in 20 patients. When follow-up CT scans were performed more than twice in 1 patient, the last CT scans were analyzed. Intervals between the initial and follow-up CT scans ranged from 1 to 25 months (mean 4.5 months). The imaging data were reconstructed with bone algorithm. The scans were imaged by using lung (window width, 1500–2000 HU; window level, ~700–800 HU) and mediastinal (window width, 300–400 HU; window level, 0–20 HU) window settings.

Two independent chest radiologists of 2- and 7-year experiences, respectively, analyzed the CT scans retrospectively. Final decisions on the findings were reached by a consensus with a third reviewer who took a role of mediator when the two reviewers had different opinions. On initial CT scans, the patterns of parenchymal abnormalities were subdivided into being nodule (3 cm or less in diameter) or mass (>3 cm in diameter), consolidation (increased opacity with obscuration of underlying vessels), and ground-glass opacity (without obscuration of underlying vessels). When nodules <10 mm were identified, the presence of accompanying branching linear structures was also evaluated (centrilobular nodules and branching linear structures). The distribution of each pattern of parenchymal abnormality in each patient was classified as being in upper, middle, and lower lung zone; as being central, subpleural, and random; as being diffuse, patchy, and random; and as being peribronchovascular and random. Lesions were considered to be located in the upper lung zone when they were seen superior to the aortic arch, in the lower lung zone when seen inferior to the inferior pulmonary vein, and in the middle lung zone when seen between the two. Lesions were regarded central when they were located within 3 cm from the hilum, subpleural when located within 3 cm from the visceral pleura, and random otherwise. Lesions were regarded as diffuse when they were widespread and continuous, patchy when they were multiple in small pieces separated from each other, and random otherwise. Lesions were regarded as peribronchovascular when located along the bronchovascular structures in the pulmonary hilum and random otherwise. As for nodules or masses, the short-axis diameter of the smallest and largest lesions was recorded in lung window setting. When there were cavity changes in nodules, the number of patients who had cavitary nodule(s) was counted. The number of cavitary nodules among the total number of nodules was also counted. The presence of halo sign (nodules surrounded by areas of ground-glass opacity) was evaluated. The attenuation of nodules and masses in mediastinal window setting, when they were more than 20 mm in diameter, was compared with that of chest wall muscles and was subdivided into being low, iso, and high. The abnormalities of the large airways from the trachea down to the lobar bronchi were subclassified in its patterns into wall thickening, intraluminal nodule, and extraluminal mass. The walls of trachea as well as main and lobar bronchi were regarded as thickened when its wall thickness of >3.0 mm in thickness on lung window images [16]. The extent of large airway abnormalities was subclassified into diffuse (>3 cm in involved length) or focal (<3 cm). Bronchial wall was regarded as thickened in the segmental and subsegmental bronchi when their walls were >1.5 mm in thickness [16, 17]. Bronchi imaged perpendicilar or horizontal to the scan plane were chosen. Bronchi imaged oblique to the scan plane were not measured. When the bronchial wall thickness was difficult to measure on films, the images were magnified into a screen by using an overhead projector and bronchial wall thickness was assessed. Presence of bronchiectasis, hilar and mediastinal lymph node enlargement, as well as pleural and pericardial effusion was also evaluated. Bronchiectasis was regarded as present when the airways distal to lobar bronchi had diameters greater than those of accompanying arteries or did not show tapering with distal branching. Air trapping was regarded as present when expiratory scans showed mosaic hypopattenuation in lung parenchyma.

On the follow-up CT scans, overall extent and the extent of each pattern of the parenchymal abnormality and airway abnormalities were evaluated and classified into being totally disappeared, partially disappeared, stable, increased, or newly appeared.