

Churg-Strauss Syndrome: The Spectrum of Pulmonary CT Findings in 17 Patients

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OBJECTIVE. The aim of this study was to evaluate the pulmonary CT findings in patients with Churg-Strauss syndrome to determine the frequency and nature of parenchymal abnormalities.

MATERIALS AND METHODS. CT examinations performed at the time of diagnosis in 17 patients with Churg-Strauss syndrome were retrospectively evaluated by two observers who reached a decision by consensus about the presence and nature of parenchymal abnormalities. High-resolution CT (1- to 3-mm collimation) was performed in 14 patients and conventional CT (6- to 10-mm collimation) was performed in three cases.

RESULTS. Predominant CT findings consisted of parenchymal opacification (consolidation or ground-glass attenuation) ($n = 10$), pulmonary nodules ($n = 2$), bronchial wall thickening or dilatation ($n = 2$), interlobular septal thickening ($n = 1$), and normal anatomy ($n = 2$). Parenchymal opacification was predominantly peripheral ($n = 6$) or random in distribution ($n = 4$).

CONCLUSION. The most common CT finding in patients with Churg-Strauss syndrome consists of areas of parenchymal opacification that may be random or peripheral in distribution. These findings are nonspecific.

Churg-Strauss syndrome is characterized by hypereosinophilia and a systemic necrotizing vasculitis seen almost exclusively in patients with asthma. It was first described in 1951 by Churg and Strauss [1] on the basis of the histologic criteria of tissue infiltration by eosinophils, necrotizing vasculitis, and extravascular granulomas. In fact, these three findings occur together only in a minority of patients [2]. Lanham et al. [2] suggested a clinical definition for the syndrome based on an extensive review of cases in the literature. According to this clinical definition, the diagnosis requires the presence of asthma, a peak peripheral blood eosinophil count of greater than $1.5 \times 10^9/l$, and a systemic vasculitis involving two or more extrapulmonary organs. In 1990 the American College of Rheumatology developed a set of criteria for diagnosis of Churg-Strauss syndrome [3]. The six criteria are asthma, eosinophilia of greater than 10% of the WBC differential count, neuropathy, migratory or transient pulmonary opacities, paranasal sinus abnormality, and extravascular eosinophils revealed on biopsy. A patient with vasculitis is considered to have Churg-

Strauss syndrome if at least four of these six criteria are present [3]. The prevalence of pulmonary opacities on chest radiography in previously published series has ranged from 26% to 77% [2, 4-7]. The radiologic appearance of the pulmonary abnormalities is diverse. To our knowledge, three isolated case reports of the CT appearances of the lungs in patients with Churg-Strauss syndrome have been published [8-10], but no systematic study of the CT findings of Churg-Strauss syndrome has been reported.

The aim of this study was to review the pulmonary CT findings in patients with Churg-Strauss syndrome to determine the frequency and nature of the parenchymal abnormalities.

Materials and Methods

The historical review included 17 patients with Churg-Strauss syndrome who presented to one of three institutions over the preceding 10 years and who had CT scanning of the chest performed at the time of initial diagnosis. The CT scanning was performed as part of the diagnostic workup in patients with clinically suspected parenchymal abnormalities. All 17 patients fulfilled at least four of the six American College of Rheumatology crite-

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ria for the diagnosis of Churg-Strauss syndrome. The patient population included seven women and 10 men who were 21–71 years old (47 ± 16 , mean \pm SD). CT was performed using an electron-beam CT scanner (Imatron, San Francisco, CA) ($n = 9$), a Somatom Plus scanner (Siemens, Erlangen, Germany) ($n = 5$), and a 9800 scanner (General Electric Medical Systems, Milwaukee, WI) ($n = 3$). End-inspiratory scans were obtained at 10-mm intervals with the patient supine. High-resolution CT (1- to 3-mm collimation) was performed in 14 patients and conventional CT (6- to 10-mm collimation) was performed in three patients. The images were photographed at a window level of -550 to -750 H and a window width of 1200–1800 H. The CT findings, including the pattern and distribution of parenchymal abnormalities and presence of any associated mediastinal or pleural abnormalities, were assessed by two observers who reached a decision by consensus about the presence, pattern, and distribution of parenchymal abnormalities. Both observers were aware of the diagnosis. The findings were recorded on a flow sheet that included presence, pattern, and distribution of parenchymal abnormalities (ground-glass attenuation, consolidation, nodules, interlobular septal thickening, and irregular lines), bronchial abnormalities (bronchial dilatation [defined as the presence of bronchi with an internal diameter greater than that of the adjacent pulmonary artery] and bronchial wall thickening), pleural abnormalities, and presence or absence of lymphadenopathy. Because some of the patients had conventional scans rather than thin-section CT scans, areas of ground-glass attenuation and consolidation were amalgamated as parenchymal opacification [11].

Results

Ten patients (59%) had parenchymal opacification, which had a predominantly peripheral distribution in six patients (Fig. 1) and was patchy with no predominant geographic distribution in the other four patients (Fig. 2). The predominant zonal distribution was lower zone in four patients, all zones in two patients, mid and lower zone in two patients, and upper zone or mid and upper zone in one patient each. In two patients (12%) the predominant abnormality consisted of pulmonary nodules. One patient had multiple cavitating nodules ranging from 1 to 3 cm in diameter (Fig. 3). The other patient had multiple nodules ranging from 5 mm to 3.5 cm, some of which contained air bronchograms. Small centrilobular nodules were also seen in two of the patients whose main CT finding was parenchymal opacification (Fig. 4). Bronchial dilatation or bronchial wall thickening, or both, was seen in six patients; in two patients this was the main CT finding and in the other four patients this finding occurred in addition to the presence of paren-

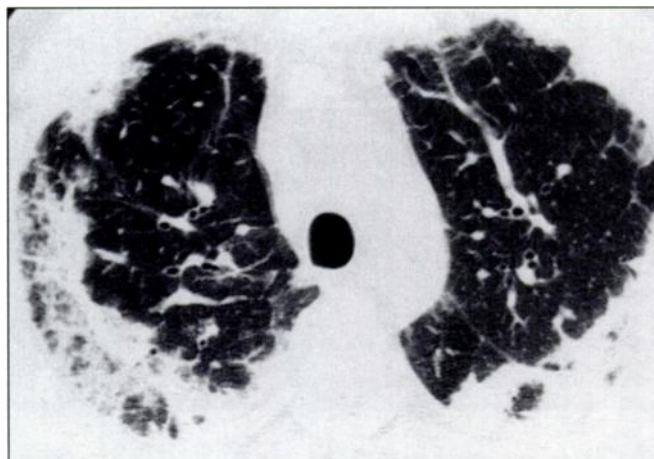


Fig. 1.—52-year-old man with Churg-Strauss syndrome. High-resolution CT scan (1.5-mm collimation) at level of aortic arch shows bilateral consolidation in peripheral distribution.

chymal opacification. The bronchial wall thickening was mild and diffuse in five patients. The bronchial dilatation affected all lobes in two patients and was confined to the lower lobes in one patient. The bronchial dilatation involved mainly the peripheral bronchi. Interlobular septal thickening was the predominant abnormality in one patient (Fig. 5). Patchy parenchymal opacification was also present. Small pleural effusions were seen in two patients. High-resolution CT revealed normal findings in two patients.

None of the 17 patients had any clinical or laboratory evidence of superimposed complications at the time of diagnosis. Specifically, none of the patients had cultures with findings positive for bacteria or fungi on sputum, bronchoscopy, or lung biopsy specimens.

Discussion

The lung is the organ most often involved in Churg-Strauss syndrome. Pulmonary consolidation is one of the six criteria used by the American College of Rheumatology to define Churg-Strauss syndrome [3]. In several reported series of patients with Churg-Strauss syndrome, pulmonary opacities were present 26–77% of cases [2, 4–7]. In a review of 138 cases by Lanham et al. [2], 74% of the patients had pulmonary opacities. In all of these previous reviews, pulmonary disease was revealed by chest radiography. The appearances reported were variable, including patchy or diffuse parenchymal changes and nodules. The parenchymal opacities are usually patchy in distribution, show no predilection for any lung zone, and are often

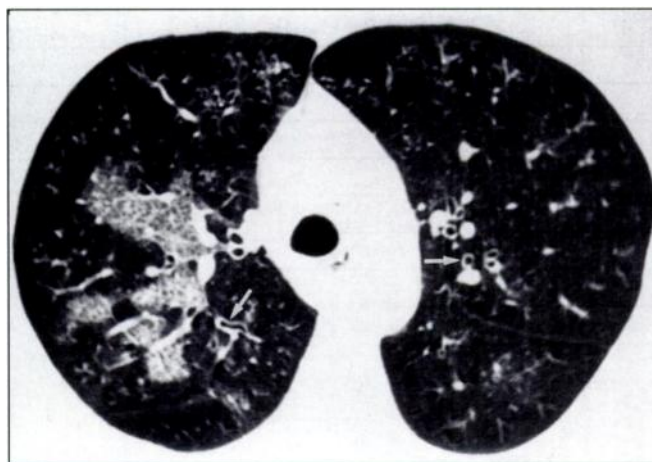


Fig. 2.—54-year-old woman with Churg-Strauss syndrome. High-resolution CT scan (1-mm collimation) at level of aortic arch shows patchy areas of ground-glass attenuation. Note bronchial wall thickening (arrows).

Pulmonary CT Findings in 17 Patients with Churg-Strauss Syndrome

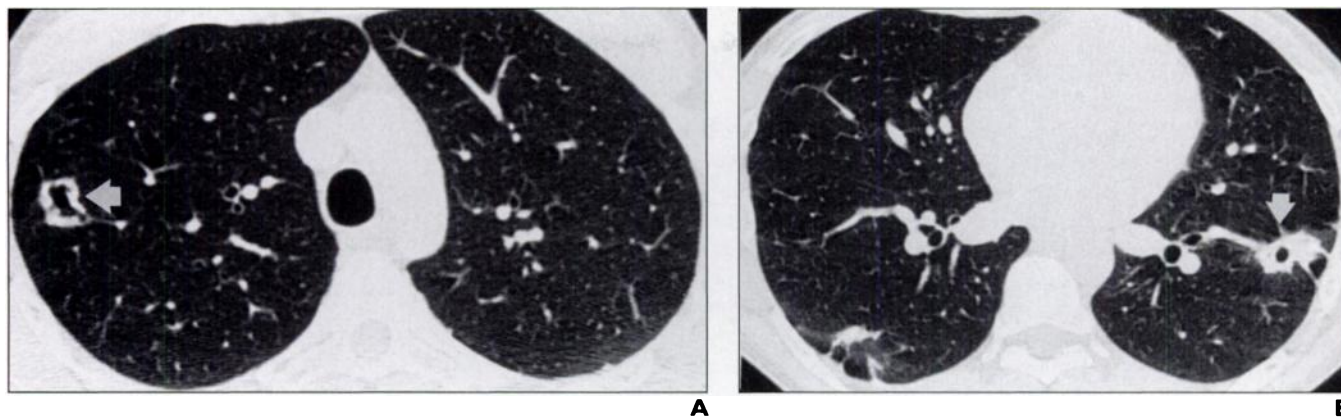


Fig. 3.—26-year-old man with Churg-Strauss syndrome. **A** and **B**, High-resolution CT scans (1.5-mm collimation) at level of aortic arch (**A**) and through lung bases (**B**) show bilateral cavitating nodules (arrows).

transient [2]. In some cases a peripheral symmetric distribution that is similar to the pattern seen in chronic eosinophilic pneumonia has been described [2]. Our findings indicate that on CT the most common abnormality consists of parenchymal opacification, which was present in almost two thirds of patients. In 60% of these patients, a predominantly

peripheral distribution was found. Parenchymal opacification in a predominantly peripheral distribution may be seen in various pulmonary conditions, including chronic eosinophilic pneumonia, bronchiolitis obliterans-organizing pneumonia, desquamative interstitial pneumonia, and pseudoalveolar sarcoid [11, 12]. In patients with asthma, the main differential diagnosis is chronic eosinophilic pneumonia, a condition that is seen with increased frequency in asthmatic patients and that may have an identical pattern and distribution [13, 14]. Patchy parenchymal opacification, as an isolated finding or in association with other parenchymal abnormalities, is seen in many of the infiltrative lung diseases [12].

To our knowledge, few reports of the pulmonary CT findings in patients with Churg-Strauss syndrome exist. Buschman et al. [8] described thin-section CT findings in one patient in whom peripheral pulmonary arteries were enlarged in comparison with adjacent bronchi. Some pulmonary arteries had an irregular stellate configuration, and interlobular septal thickening and patchy indistinct opacities were evident. Amato et al. [9] described interstitial disease with small parenchymal nodules on thin-section CT in one patient. Fluffy, discrete perivascular opacities were seen on CT of a child with Churg-Strauss syndrome [10].

These vascular and perivascular changes have been suggested to be directly related to

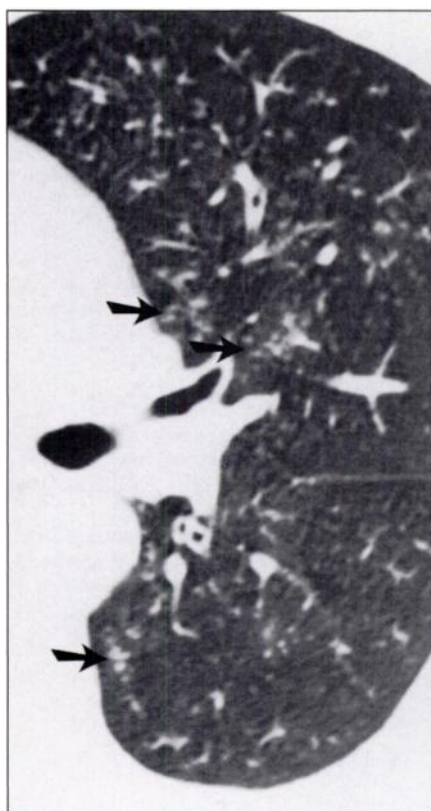


Fig. 4.—54-year-old woman with Churg-Strauss syndrome. High-resolution CT scan (1-mm collimation) of left lung reveals several small centrilobular nodules (arrows). Note bronchial wall thickening.

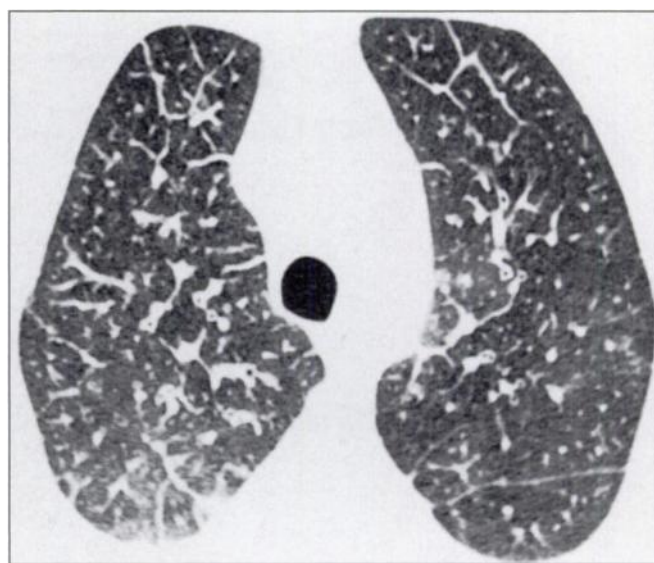


Fig. 5.—49-year-old man with Churg-Strauss syndrome. High-resolution CT scan (1-mm collimation) at level of aortic arch shows interlobular septal thickening and bronchial wall thickening. Findings were due to interstitial pulmonary edema associated with cardiac involvement in Churg-Strauss syndrome.

the pulmonary vasculitis. In the case reported by Buschman et al. [8], the CT findings correlated with the pathologic changes seen on an open lung biopsy from the same region of the lung. Marked expansion of the arterial adventitia by an eosinophil-rich inflammatory opacity was evident on histology, and obliteration of the arterial lumen and necrosis of a component of the arterial wall were found in the same areas.

In our study, 10 of the patients had lung biopsies, which in nine cases confirmed the presence of pulmonary vasculitis consistent with Churg-Strauss syndrome. However, no evidence of enlargement or stellate appearance of the peripheral pulmonary arteries was found on CT in any of our patients. Most scanning in this study was performed with narrow collimation, and thus subtle changes, if present in the study patients, would have been identified. Interlobular septal thickening was seen in only one patient, small centrilobular nodules were seen in two patients, and nodules measuring 0.5–3.5 cm in diameter were seen in two patients. Cavitation of nodules seen in one of the patients has been previously reported in a single case [2].

Bronchial dilatation or bronchial wall thickening, or both, was identified in 35% of patients. These findings are likely to be related to the patients' asthma, which is always present in Churg-Strauss syndrome [2]. Bronchial thickening and bronchial dilatation are commonly seen on thin-section CT scans in asthmatic patients [15, 16]. A recent study [17] found bronchial wall thickening in 44% and bronchial dilatation in 31% of asthmatic subjects—a rate significantly higher than that found in healthy control subjects.

The diffuse interlobular septal thickening seen in one patient was related to interstitial pulmonary edema attributed to cardiac involvement in Churg-Strauss syndrome [18]. In the original report by Churg and Strauss [1], cardiac failure was the cause of death in three (27%) of 11 patients. At autopsy, interstitial eosinophilic myocardial inflammation, coronary vasculitis, and pericardial involvement were common.

In this study 15 (88%) of 17 patients had pulmonary abnormalities revealed on CT, a finding that is slightly higher than the reported incidence on chest radiography [2]. This finding may be due to the selection bias in referring patients who have a higher rate of pulmonary abnormalities for chest CT. Two patients had no abnormal CT findings. One patient had bilateral pulmonary opacities documented previously by chest radiography that had resolved without treatment before CT.

This study shows protean pulmonary CT findings in patients with Churg-Strauss syndrome. The finding of enlarged stellate peripheral pulmonary arteries identified in the first reported case of high-resolution CT changes in Churg-Strauss syndrome [8] could not be confirmed in any of the patients in our series. This finding appears to be unusual and nonspecific for pulmonary vasculitis, contrary to findings of a previous report [8]. The most common finding, seen in almost two thirds of patients, was parenchymal opacification, with a predominantly peripheral distribution in 60% of the cases. Thus, parenchymal opacification in a predominantly peripheral distribution on CT in a patient with asthma should raise the possibility of Churg-Strauss syndrome. The diagnosis can only be suggested with reasonable confidence in asthmatic patients who have other findings, particularly peripheral neuropathy or sinusitis.

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