Churg–Strauss Syndrome
High Resolution CT and Pathologic Findings

Cleonice Isabela S. Silva, MD, Nestor L. Müller, MD, PhD,† Kiminori Fujimoto, MD, PhD,‡ Takeshi Johkoh, MD, PhD,§ Sérgio A. Ajzen, MD,* and Andrew Churg, MD¶*

Objectives: The purpose of this study was to evaluate high-resolution CT findings in 7 patients with Churg–Strauss syndrome and to compare the CT with the histopathologic findings.

Materials and Methods: High-resolution CT scans of 7 asthmatic patients (4 women, 3 men, age range, 34–62 years, mean 49 years) with Churg–Strauss syndrome were reviewed by 2 observers. Histologic specimens of lung obtained at surgical (n = 3) or transbronchial (n = 3) biopsy or autopsy (n = 1) were reviewed by an expert lung pathologist. The diagnosis of Churg–Strauss was based on clinical, laboratory, and histologic findings.

Results: Parenchymal and airway abnormalities included ground-glass opacities (n = 5), areas of air-space consolidation (n = 4), centrilobular nodules (n = 5), nodules 1–3 cm in diameter (n = 3), interlobular septal thickening (n = 4), bronchial wall thickening (n = 4), and areas of atelectasis (n = 1). Surgical biopsy (n = 3) and autopsy (n = 1) specimens demonstrated airspace disease in 3 patients, interlobular septal thickening in 3 patients, and airway abnormalities in 2 patients. Histologically, the airspace disease included eosinophilic pneumonia (n = 2) and small foci of organizing pneumonia (n = 1). The septal thickening was due to edema combined with numerous (n = 2) or few (n = 1) eosinophils. The airway abnormalities (n = 2) included muscle hypertrophy and large airway wall necrosis (n = 1) and eosinophilic infiltration of the airway walls (n = 1). Transbronchial biopsy (n = 3) demonstrated increased eosinophils.

Conclusion: The main high-resolution CT findings of Churg–Strauss syndrome consist of airspace consolidation or ground-glass opacities, septal lines, and bronchial wall thickening. These reflect the presence of eosinophilic infiltration of the airspaces, interstitium, and airways, and interstitial edema.

Key Words: lung diseases, lung computed tomography, computed tomography

(*J Thorac Imaging* 2005;20:74–80)

From the *Department of Diagnostic Imaging, University Federal of São Paulo, Brazil; †Department of Radiology, Vancouver General Hospital, Vancouver, Canada; ‡Kurume University School of Medicine, Kurume, Fukuoka, Japan; §Department of Radiology, Osaka University Medical School Suita, Osaka, Japan; and ¶Department of Pathology, University of British Columbia, Vancouver, Canada.

Reprints: Dr. Nestor L. Müller, Department of Radiology, Vancouver General Hospital, 899 W. 12th Avenue, Vancouver, BC V5Z 1M9 (e-mail: nmuller@vanhosp.bc.ca).

Churg–Strauss syndrome is a rare vasculitis of unknown etiology defined by the 1994 International Consensus Conference as an "eosinophil-rich and granulomatous inflammation involving the respiratory tract, and necrotizing vasculitis affecting small to medium sized vessels, and associated with asthma and eosinophilia."[1] The diagnosis can be made based on the presence of 4 or more of the following findings: 1) asthma, 2) peripheral eosinophilia greater than 10%, 3) peripheral neuropathy, 4) transient or migratory pulmonary opacities, 5) paranasal sinus abnormality, and 6) extravascular eosinophils on biopsy.[2]

The most common high-resolution CT manifestations consist of areas of ground-glass attenuation or consolidation in either a patchy or a predominantly peripheral distribution.[3–5] Less common abnormalities include small centrilobular nodules and, occasionally, larger nodules measuring 0.5 to 3.5 cm in diameter.[3] Recently, it has been shown that interlobular septal thickening is relatively common in Churg–Strauss syndrome, being seen in 9 to 16 patients reported by Johkoh et al.[5] There has, however, been limited information about the correlation between the CT and the histologic findings.

The aim of our study was to review the high-resolution CT findings in 7 patients with Churg–Strauss syndrome and to compare the CT with histopathologic findings.

MATERIAL AND METHODS

The patient population included 7 patients (4 women, 3 men, age range, 34–62 years, mean 49 years) who had a histologically proven diagnosis of Churg–Strauss syndrome and had undergone high resolution CT and lung biopsy or autopsy at 1 of our 3 institutions between 1995 and 2003.

All patients were asthmatics, and had peripheral eosinophilia and transient pulmonary opacities on chest radiography prior to CT. Histopathologic specimens of the lungs were acquired at surgical lung biopsy (n = 3), transbronchial biopsy (n = 3), or autopsy (n = 1) within 0 to 7 days (median 2 days) after the CT scan. High-resolution CT was performed on a variety of scanners using 1 to 2 mm collimation at 10 mm intervals (n = 3) or volumetrically on a multidetector CT scanner (n = 4). The CT scans were performed at end inspiration, with the patient supine, and reconstructed using a high-resolution edge-enhancing algorithm. The images were photographed at lung (level –600 to –700 HU, width 1000–1500 HU) and mediastinal windows (level 20–50 HU, width 300–500 HU).

The high-resolution CT scans were reviewed by 2 observers, who reached a decision about the findings by

consensus. The parenchymal findings were defined according to the criteria recommended by the Fleischner Nomenclature Committee.[6] The observers analyzed the presence, pattern, and distribution of areas of consolidation, ground-glass attenuation, nodules, interlobular septal lines, thickening of bronchovascular bundles, bronchial wall thickening, pleural thickening or effusion, and lymph node enlargement. Nodules were classified into small (less than 1 cm diameter) and large (equal or greater than 1 cm in diameter), and their distribution assessed in relation to vessels, bronchi, and secondary pulmonary lobule. Lymphadenopathy was considered present when the short axis diameter of the node was greater than 10 mm.

The pathologic specimens were reviewed by an expert lung pathologist.

RESULTS

HRCT Findings

All patients had abnormal parenchymal findings on high-resolution CT (Table 1). Parenchymal abnormalities included: ground-glass opacities (n = 5), areas of air-space consolidation (n = 4), centrilobular nodules (n = 5), nodules 1–3 cm in diameter (n = 3), interlobular septal thickening (n = 4), bronchial wall thickening (n = 4), and areas of atelectasis (n = 1). Other findings included pleural effusion (n = 4), mediastinal lymph node enlargement (n = 2), and pericardial effusion (n = 2).

Non-segmental bilateral areas of consolidation and ground-glass attenuation were the predominant abnormality in 4 patients (Figs. 1A, 2A). These abnormalities were bilateral and asymmetric and involved mainly the upper and middle lung zones in 3 patients, and the lower lung zones in 1 patient. A predominant distribution in the outer third of the lungs was seen in only 2 of these 4 patients.

In the remaining 3 patients, the predominant abnormalities consisted, respectively, of focal ground-glass opacities in the right upper lobe, bronchial wall thickening associated with areas of atelectasis, and bilateral septal lines.

Bilateral thickening of the interlobular septa was seen in 4 patients and was always associated with unilateral or bilateral pleural effusions. The patient in whom septal lines were the predominant finding had normal cardiac function on echocardiography (Fig. 3A). In 2 of the other patients, echocardiography confirmed the presence of left heart failure. The remaining patient had no other clinical findings of left heart failure and did not undergo echocardiography.

A few poorly defined centrilobular nodules (n = 5) in a patchy asymmetric bilateral distribution and 1 to 3 nodules 1–3 cm in diameter (n = 3) were a minor finding seen in patients with predominant airspace opacities (n = 4) or septal lines (n = 1).

Pleural effusions (n = 4) were bilateral in 2 patients and unilateral in 2. Two of the patients with bilateral effusions also had a small pericardial effusion. Two patients had 1 or 2 enlarged mediastinal lymph nodes.

Histologic Findings

Surgical biopsy (n = 3) and autopsy (n = 1) specimens demonstrated airspace disease in 3 patients, pulmonary vascular abnormalities in 3 patients, interlobular septal thickening in 3 patients, and airway abnormalities in 2 patients (Table 1). The airspace disease included eosinophilic pneumonia with granulomas in 1 patient, a few collections of necrotic eosinophils and macrophages representing the effects of steroid treatment on eosinophilic pneumonia in 1 patient, and small foci of organizing pneumonia in 1 patient (Fig. 1B). The vascular

TABLE 1. Churg–Strauss: HRCT and Pathologic Findings

No	Sex/Age	Airspace Consolidation	Ground-glass	Septal Lines	Nodules >1.0 cm	Centrilobular Nodules	Bronchial Wall Thickening	Pathology Material	Histologic Findings
1	F/54	Mainly peripheral	Mainly peripheral	+		+		Autopsy	Eosinophilic pneumonia with granulomas, vasculitis, edematous interlobular septa with few esoinophils, severe myocarditis, and necrotizing cardiac vasculitis.
2	F/38	Mainly peripheral	Mainly peripheral	+	+	+	+	Surgical lung biopsy	Occasional collections of necrotic eosinophils in airspaces and edema and necrotic eosinophils in interlobular septa.
3	M/52	Patchy	Patchy		+	+	+	Surgical lung biopsy	Small foci of organizing pneumonia and eosinophils.
4	M/41		Patchy	+		+		Surgical lung biopsy	Edema and numerous eosinophils in the interlobular septa and pleura.
5	F/34	Patchy			+			TBx skin Bx	TBx = Eosinophils. Skin Bx = Eosinophilic paniculitis.
6	F/59		Patchy	+		+	+	TBx	Eosinophils.
7	M/62						+	TBx skin Bx	TBx = Eosinophils. Skin Bx = Leukocytoclastic vasculitis with eosinophils.

F, female; M, male; Age, years; TBx, transbronchial biopsy; Skin Bx, skin biopsy.

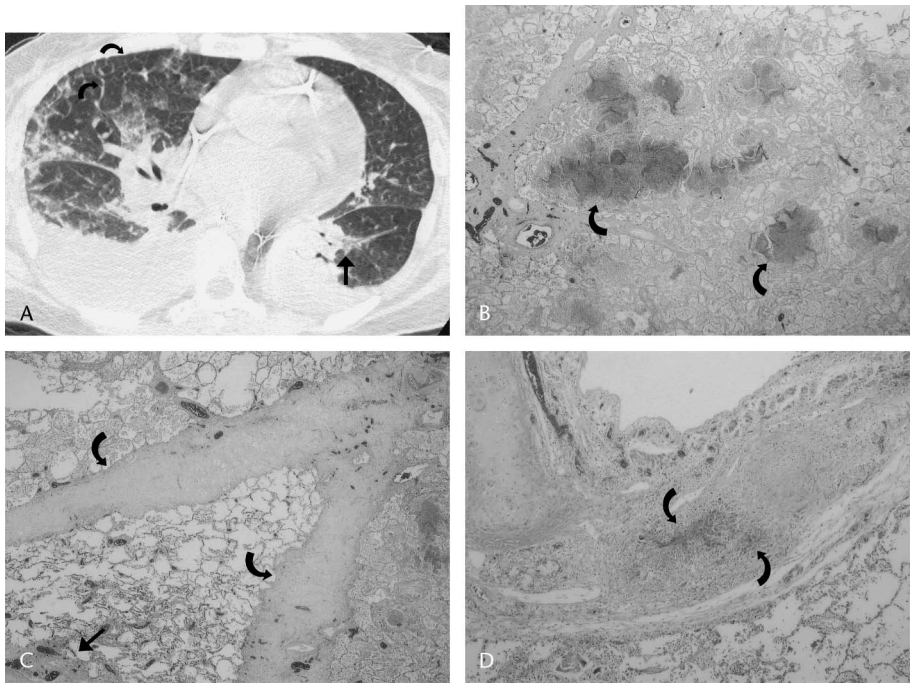

FIGURE 1. A 54-year-old woman with Churg–Strauss syndrome. A, High-resolution CT shows areas of ground-glass attenuation mainly in the peripheral regions of the right middle and lower lobes. Airspace consolidation and a small area of ground-glass attenuation (straight arrow) are seen in the left lower lobe. Also noted are thickening of the interlobular septa (curved arrows) mainly in the right middle lobe and bilateral pleural effusions. B, Photomicrograph of histopathologic specimen obtained at autopsy shows eosinophilic pneumonia pattern with large number of partially necrotic eosinophils (arrows) (H and E, × 50). C, Photomicrograph shows edematous and fibrotic interlobular septa (curved arrows) (H and E, × 50). A focus of very early eosinophilic pneumonia is present between the interlobular septa (straight arrow). Such a small focus could result in poorly defined centrilobular ground-glass opacities. A focus of well developed eosinophilic pneumonia with eosinophil necrosis is present to the right of the thickened septum. D, Photomicrograph shows necrotizing focus (early Churg–Strauss granuloma) in the wall of a large bronchus (H and E, × 75).

abnormalities included small vessel vasculitis (n = 1), infiltration by eosinophils, (n = 1) and old adventitial fibrosis (n = 1) (Fig. 2B). The septal thickening was due to edema (n = 3) combined with numerous eosinophils and minimal fibrosis (n = 2) or mild fibrosis and few eosinophils (n = 1) (Figs. 1C, 3B, and 3C). The airway abnormalities (n = 2) included muscle hypertrophy and large airway wall necrosis (n = 1) and eosinophilic and lymphocytic infiltration of the airway walls (n = 1) (Fig. 1D). Other findings included eosinophilic pleuritis in 1 patient (Fig. 3B) and severe myocarditis and necrotizing cardiac vasculitis in 1 patient.

Transbronchial biopsy specimens (n = 3) demonstrated increased eosinophils (n = 3).

High-Resolution CT—Pathologic Correlation

Histologic specimens from surgical biopsy (n = 3) or autopsy (n = 1) were available in 3 of the patients with predominantly consolidation and ground-glass opacities and the patient with predominantly septal thickening on high-resolution CT. The airspace disease in these 3 patients included eosinophilic pneumonia with granulomas in 1 patient, a few collections of necrotic eosinophils and macrophages in 1 patient, and small foci of organizing pneumonia and occasional eosinophils in 1 patient.

Eosinophilic pneumonia with granuloma formation was present in a patient with bilateral symmetric ground-glass opacities and consolidation (Fig. 1, Table 1 patient 1). This patient also had bronchial wall thickening, poorly defined centrilobular nodules, interlobular septal thickening, and bilateral pleural effusions on high-resolution CT. Autopsy in this patient also demonstrated small vessel vasculitis, large airway wall necrosis, markedly edematous, slightly fibrotic interlobular septa with a few eosinophils, and severe myocarditis and necrotizing cardiac vasculitis, the latter being the cause of death.

The patient with a few collections of necrotic eosinophils and macrophages in the airspaces on surgical biopsy also had eosinophilic infiltration of the airway walls and markedly edematous interlobular septa, some of which contained masses of necrotic eosinophils and necrotic macrophages (Fig. 2, Table 1 patient 2). This patient had been started on corticosteroid therapy 4 days prior to biopsy, and the necrosis presumably represents a treatment effect. The high-resolution CT performed prior to treatment demonstrated bilateral areas of consolidation and ground-glass attenuation, bronchial wall thickening, two 1–3 cm diameter nodules, a few small bilateral centrilobular nodules, extensive interlobular septal thickening, and right pleural effusion.

The patient with small foci of organizing pneumonia and some eosinophils on surgical biopsy had bilateral areas of consolidation and ground-glass opacification and several 1–3 cm diameter poorly defined nodules and a few centrilobular nodules on high-resolution CT (Table 1 patient 3). This patient was started on corticosteroids 1 week prior to the CT scan and surgical biopsy.

The patient with predominantly septal thickening on high-resolution CT was shown on surgical lung biopsy to have markedly edematous septa that contained numerous eosinophils and mild fibrosis (Fig. 3, Table 1 patient 4). The patient also had marked eosinophilic infiltration of the pleura. Other high-resolution CT findings in this patient included a few poorly defined small centrilobular nodules, bilateral pleural

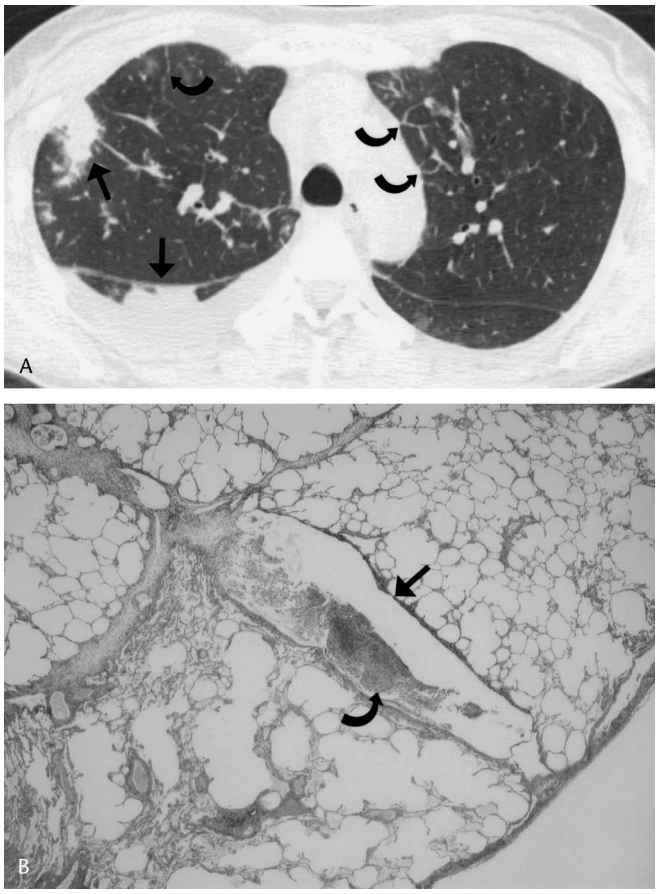

FIGURE 2. A 38-year-old woman with Churg–Strauss syndrome. A, High-resolution CT shows focal areas of consolidation in the right upper and lower lobes (straight arrows). Also noted are small areas of ground-glass attenuation and thickening of the interlobular septa (curved arrows). B, Photomicrograph of edematous interlobular septum contains masses of necrotic eosinophils; these represent a treatment effect of steroids (H and E, × 50).

effusions, and small pericardial effusion. This patient had no evidence of left heart failure at the time of the CT or on 1-year follow-up and had normal cardiac function on echocardiography.

The remaining 3 patients underwent transbronchial biopsy, which demonstrated increased eosinophils.

DISCUSSION

Churg–Strauss syndrome (allergic angiitis and granulomatosis) was first described in 1951 by Churg and Strauss in a study of 13 patients, 11 of whom died of the illness.[7] All 13 patients had severe asthma, marked peripheral eosinophilia, inflammatory, necrotizing granulomatous vasculitis,

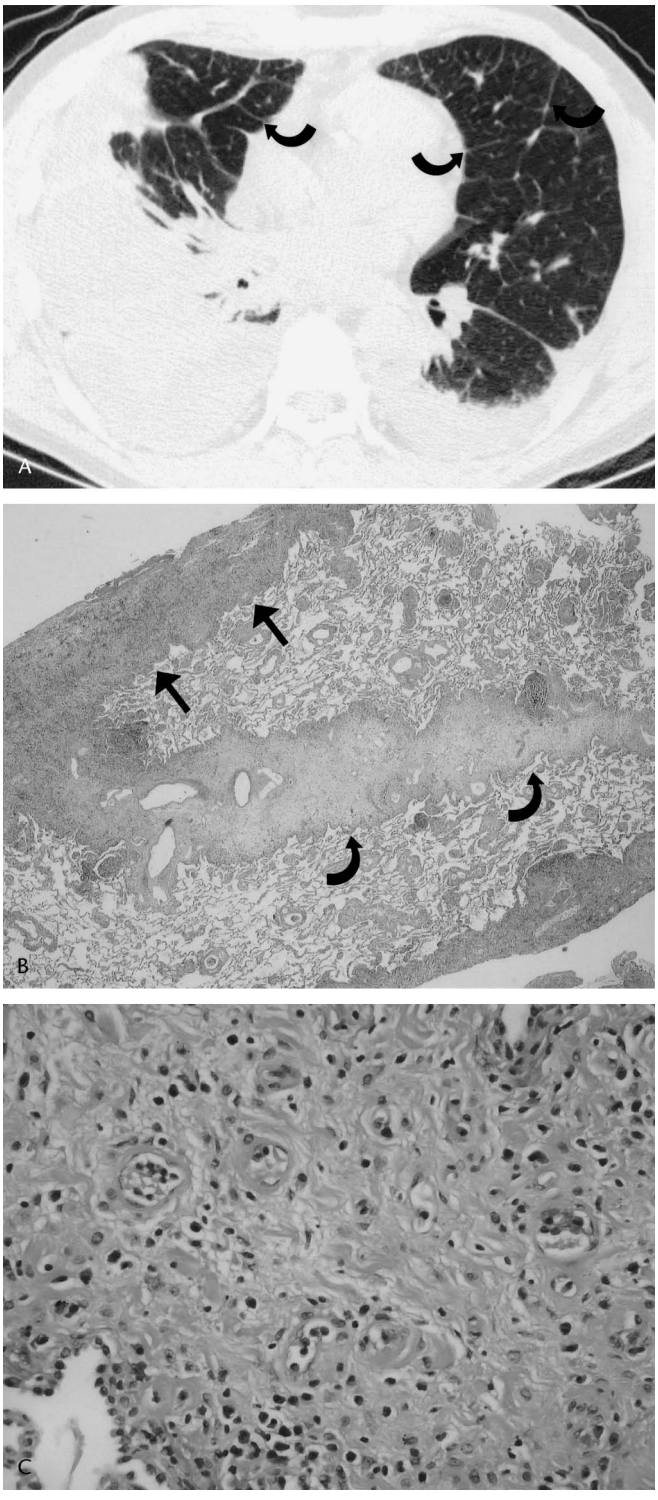

FIGURE 3. A 41-year-old man with Churg–Strauss syndrome. A, High-resolution CT shows thickening of the interlobular septa (arrows) and large right and small left pleural effusions. Note normal heart size and normal size of pulmonary vessels. B, Photomicrograph of histopathologic specimen obtained at surgical biopsy shows edematous and fibrotic interlobular septum (curved arrows). There is also a marked reaction with considerable eosinophilic infiltration of the pleura (straight arrows) (H and E, × 50). C, Higher power view of an interlobular septum shows lymphocytes and eosinophils in a fibrotic matrix (H and E, × 400).

and extravascular granulomatous lesions. The granulomatous lesions consisted of granulomatous proliferation of epithelioid and giant cells and contained necrotic eosinophilic exudates.[7]

The characteristic histologic features of fully developed vasculitis in Churg–Strauss syndrome consist of an eosinophilic vasculitis that may involve any organ, most commonly heart, kidney, gastrointestinal tract, central nervous system, skin, and lung.[1,7,8] The vasculitis typically involves small arteries, arterioles, small veins, venules, and capillaries. Outside the lung the vasculitis typically shows fibrinoid necrosis and an eosinophilic infiltrate of the vessel wall. In the lung the vasculitis may be necrotizing, but commonly is non-necrotizing and characterized by eosinophilic infiltration of the vessel walls.[8] The pulmonary parenchymal abnormalities in Churg–Strauss syndrome resemble those of chronic eosinophilic pneumonia and consist mainly of a prominent eosinophilic infiltrate and foci of eosinophilic necrosis. Distinguishing features include presence of granulomatous reaction around the necrotic foci and eosinophilic vasculitis in Churg–Strauss syndrome.[8] It should be noted that the classic description of the pathologic findings is based almost exclusively on autopsy material in patients with florid disease.[7,8] As seen in our study, transbronchial biopsy may only demonstrate increased eosinophils, and even surgical biopsy only shows some of the histologic findings characteristic of the disease. In patients with characteristic clinical manifestations, these findings are sufficient to make a confident diagnosis.

Early diagnosis of Churg–Strauss syndrome is important because these patients respond well to treatment with corticosteroids.[9,10] A retrospective clinical study with long-term follow-up of 96 patients diagnosed between 1963 and 1995 showed that treatment with corticosteroids alone or in combination with cyclophosphamide resulted in clinical remission in 91.5% of the patients.[9] However, 22 (26%) patients relapsed. Twenty-three patients died during follow-up; 11 of these deaths were directly due to vasculitis. Multivariate analysis demonstrated that the factors significantly associated with a poor outcome were myocardial and severe gastrointestinal tract involvement.

The current clinical emphasis is on diagnosing Churg–Strauss syndrome in the early prodromal or pre-vasculitic phase, because these patients respond best to treatment.[10] This phase is characterized by tissue eosinophilia manifested as eosinophilic pneumonia, eosinophilic gastroenteritis, or eosinophilic lymphadenopathy, but without vasculitis.[10] The histologic findings in the lungs in the pre-vasculitic stage are usually identical to those of chronic eosinophilic pneumonia.

In the current study, necrotizing small vessel vasculitis was only seen in the patient who underwent autopsy. This patient also had airway wall necrosis, eosinophilic pneumonia with granuloma formation, severe myocarditis, and necrotizing cardiac vasculitis, which was the cause of death. The remaining patients were diagnosed at initial presentation and had no evidence of vasculitis or extravascular granulomatous lesions on lung biopsy. The diagnosis of Churg–Strauss in all patients was based on the presence of 4 or more of the following findings: 1) asthma, 2) peripheral eosinophilia greater than 10%, 3) peripheral neuropathy, 4) transient or migratory pulmonary opacities, 5) paranasal sinus abnormality, and 6) extravascular eosinophils on biopsy.[2] One of the patients, who was started on treatment with high dose corticosteroids 4 days prior to surgical biopsy, demonstrated few collections of necrotic eosinophils and macrophages in the airspaces on surgical biopsy and masses of necrotic eosinophils and necrotic macrophages in the interlobular septa. The second patient, who received costicosteroid treatment 7 days prior to surgical biopsy, demonstrated only small foci of organizing pneumonia and rare eosinophils on surgical biopsy. The findings in both these patients are consistent with rapid response to treatment characteristic of the early phase of Churg–Strauss syndrome.

The most common radiographic manifestations of Churg–Strauss syndrome consist of bilateral nonsegmental areas of consolidation without predilection for any lung zone (Table 2).[11,12] The areas of consolidation can be transient, resembling Löffler syndrome, or predominantly peripheral, resembling chronic eosinophilic pneumonia.[11,12] Less common manifestations include a diffuse interstitial reticular or reticulonodular pattern and bilateral small and large nodular opacities.[11–13] Unilateral or bilateral pleural effusions are seen in approximately 30% of patients, and hilar or mediastinal lymphadenopathy in a small number of cases.[11,12,14]

Worthy et al reviewed the high-resolution CT findings at the time of diagnosis in 17 patients.[3] The most common abnormality, seen in 10 (59%) patients, consisted of bilateral areas of consolidation or ground-glass attenuation in either a patchy random or predominantly peripheral distribution (Table 2). Two patients had multiple 0.5 to 3.5 cm diameter nodules, 2 had small centrilobular nodules, 2 had bronchial wall thickening, and 1 had interlobular septal thickening. The

TABLE 2. Churg–Strauss Syndrome: Radiologic Manifestations

Chest radiograph
A. Common
Bilateral nonsegmental areas of consolidation*
Transient or predominantly peripheral*
Reticular or reticulonodular pattern
Unilateral or bilateral pleural effusions
B. Uncommon
Bilateral small and large nodular opacities
High-resolution CT
A. Common
Airspace consolidation or ground glass attenuation*
Patchy predominantly peripheral distribution*
Interlobular septal thickening
Centrilobular nodules
Bronchial wall thickening
Unilateral or bilateral pleural effusions
B. Uncommon
Large nodules
Increased vessel caliber
Mediastinal lymphadenopathy
Pericardial effusion

*Most common findings.

septal thickening in this patient was presumed to be due to interstitial edema secondary to cardiac involvement.[3]

Choi et al reviewed the high-resolution CT findings in 9 patients with Churg–Strauss syndrome.[4] The most common findings included bilateral ground-glass opacities (n = 9), centrilobular nodules mostly within the ground-glass opacities (n = 8); bronchial wall thickening (n = 7), and increased vessel caliber (n = 5) (Table 2). Air space consolidation was seen in 5 patients and was predominantly peripheral and surrounded by areas of ground-glass attenuation. Other findings included large nodules (n = 4), interlobular septal thickening (n = 2), hilar or mediastinal lymph node enlargement (n = 4), pleural effusion (n = 2), and pericardial effusion (n = 2).

Johkoh et al reviewed the CT findings in 16 patients with Churg–Strauss syndrome.[5] The most common findings included areas of ground-glass attenuation (n = 14), consolidation (n = 7), nodules (n = 9), interlobular septal thickening (n = 9), and bronchial wall thickening (n = 8) (Table 2). In the majority of patients, the areas of ground-glass attenuation and consolidation had no central or peripheral predominance and no upper or lower lung zone predominance. A peripheral distribution was only seen in 2 patients (13%). Four patients had enlarged mediastinal lymph nodes, and 2 had pleural effusion.

There is limited information on the correlation between high-resolution CT and histologic findings. Buschman et al described the high-resolution CT findings in 1 patient in whom peripheral pulmonary arteries were enlarged in comparison to adjacent bronchi.[15] Some pulmonary arteries had an irregular stellate configuration. Open lung biopsy demonstrated marked expansion of the arterial adventitia by an eosinophil-rich inflammatory infiltrate. The patient also had interlobular septal thickening on CT and was shown to have extensive eosinophilic infiltration of the interlobular septa at biopsy. Choi et al correlated the high-resolution CT with the lung biopsy findings in 2 patients.[4] One of the patients had patchy right lower lobe consolidation, diffuse reticulonodular opacities, bronchial wall thickening, cardiomegaly, bilateral hilar lymph node enlargement, and small bilateral pleural effusions. Open thoracotomy and biopsy of the right lower lobe revealed findings of eosinophilic pneumonia and asthmatic bronchitis with eosinophil infiltration and vasculitis in pleura and pericardium. A second patient had multiple bilateral variable-sized nodules and mediastinal lymph node enlargement. Thoracoscopic biopsy of a nodular lesion of the lung revealed a granulomatous lesion and necrotizing vasculitis with eosinophil infiltration. Mediastinal lymph node biopsy demonstrated reactive hyperplasia with eosinophil infiltration.

The high-resolution CT findings in the current study are similar to those of previous studies.[3–5] The main abnormalities consisted of ground-glass opacities, air-space consolidation, small centrilobular nodules, nodules 1–3 cm in diameter, interlobular septal thickening, and bronchial wall thickening. Correlation with autopsy or surgical biopsy specimens was available in 4 patients. These demonstrated that the histologic features that accounted for the ground-glass opacities and areas of consolidation seen in 3 of these 4 patients included eosinophilic pneumonia with granulomas in 1 patient, a few collections of necrotic eosinophils and macrophages in 1 patient, and small foci of organizing pneumonia and rare eosinophils in 1 patient. The main abnormalities on high-resolution CT in the other patient, who underwent surgical lung biopsy, were the presence of extensive bilateral septal thickening, bilateral pleural effusions, and small pericardial effusion. Histologic assessment showed markedly edematous septa that contained numerous eosinophils and mild fibrosis. This patient had no evidence of left heart failure at the time of the CT or on 1-year follow-up and had normal cardiac function on echocardiography. Choi et al also reported a patient with extensive septal thickening on high-resolution CT and normal cardiac function, but their patient did not have lung biopsy.[4] In our study, 3 of the 4 patients who had surgical biopsy or autopsy specimens available for review had interlobular septal thickening histologically due to edema, eosinophilic infiltration, and mild fibrosis.

Other findings seen in at least 1 of the surgical biopsy or autopsy specimens in our study included small vessel vasculitis, small vessel infiltration by eosinophils, or old adventitial fibrosis; airway muscle hypertrophy, large airway wall necrosis, or eosinophilic and lymphocytic infiltration of the airway walls; and severe myocarditis and necrotizing cardiac vasculitis. Transbronchial biopsy specimens performed in 3 patients demonstrated increased eosinophils. As previously postulated by Choi et al, the small vessel vasculitis with associated increased caliber of small arteries and arterioles may account for the centrilobular nodules seen on high-resolution CT. However, in contradistinction of the previous reports by Buschman et al[15] and Choi et al,[4] we did not see involvement of large vessels histologically or enlargement of large vessels on high-resolution CT scans performed prior to lung biopsy. Eosinophilic infiltration of the airway wall may account for the bronchial wall thickening seen in some patients with Churg–Strauss syndrome. However, bronchial wall thickening is a common finding in asthma,[16,17] and therefore not helpful in the diagnosis of Churg–Strauss syndrome.

Our study has several limitations. It is retrospective, includes a small number of patients, and does not provide a definite correlation between the high-resolution CT and pathologic findings. However, Churg–Strauss syndrome is rare. The incidence has been estimated to be approximately 1.8 to 3.3 cases per million person-years.[18]

Based on the results of our study, those of Choi et al,[4] and the case report by Buschman et al,[15] we conclude that the ground-glass opacities and consolidation seen on high-resolution CT reflect the presence of eosinophilic pneumonia. In patients who receive corticosteroid therapy, the eosinophilic infiltration resolves rapidly, and biopsy of patients on treatment may demonstrate only sparse eosinophils or organizing pneumonia. The interlobular septal thickening may be secondary to cardiac involvement and interstitial edema or, less commonly, be seen in patients with normal cardiac function and reflect the presence of marked septal eosinophilic infiltration and edema. Enlargement of vessels seen on high-resolution CT in some patients reflects the presence of vasculitis; vasculitis and enlargement of small vessels presumably also accounts for the centrilobular nodules seen in some patients. Pleural effusions may be secondary to cardiac involvement and left heart failure or be due to eosinophilic pleuritis. Mediastinal lymph node

enlargement, seen in a small number of cases, has been shown to be due to reactive hyperplasia and eosinophilic infiltration.[4,14]

REFERENCES

1. Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. *Arthritis Rheum*. 1994;37:187–192.
2. Masi AT, Hunder GG, Lie JT, et al. The American College of Rheumatology 1990 criteria for the classification of Churg–Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum*. 1990; 33:1094–1100.
3. Worthy SA, Müller NL, Hansell DM, et al. Churg –Strauss syndrome: The spectrum of pulmonary CT findings in 17 patients. *AJR Am J Roentgenol*. 1998;170:297–300.
4. Choi YH, Im JG, Han BK, et al. Thoracic manifestation of Churg–Strauss syndrome: radiologic and clinical findings. *Chest*. 2000;117:117–124.
5. Johkoh T, Müller NL, Akira M, et al. Eosinophilic lung diseases: diagnostic accuracy of thin-section CT in 111 patients. *Radiology*. 2000; 216:773–780.
6. Austin JH, Müller NL, Friedman PJ, et al. Glossary of terms for CT of the lungs: recommendations of the Nomenclature Committee of the Fleischner Society. *Radiology*. 1996;200:327–331.
7. Churg J, Strauss L. Allergic granulomatosis, allergic angiitis and periarteritis nodosa. *Am J Pathol*. 1951;27:277–301.
8. Churg A. Pulmonary angiitis and granulomatosis revisited. *Hum Pathol*. 1983;14:868–883.
9. Guillevin L, Cohen P, Gayraud M, et al. Churg –Strauss syndrome. Clinical study and long-term follow-up of 96 patients. *Medicine (Baltimore)*. 1999;78:26–37.
10. Churg A. Recent advances in the diagnosis of Churg–Strauss syndrome. *Mod Pathol*. 2001;14:1284–1293.
11. Lanham JG, Elkon KB, Pusey CD, et al. Systemic vasculitis with asthma and eosinophilla: a clinical approach to the Churg–Strauss syndrome. *Medicine (Baltimore)*. 1984;63:65–81.
12. Chumbley LC, Harrison EG Jr, DeRemee RA. Allergic granulomatosis and angiitis (Churg–Strauss syndrome). Report and analysis of 30 cases. *Mayo Clin Proc*. 1977;52:477–484.
13. Degesys GE, Mintzer RA, Vrla RF. Allergic granulomatosis: Churg–Strauss syndrome. *AJR Am J Roentgenol*. 1980;135:1281–1282.
14. Lesens O, Hansmann Y, Nerson J, et al. Severe Churg–Strauss syndrome with mediastinal lymphadenopathy treated with interferon therapy. *Eur J Intern Med*. 2002;13:458.
15. Buschman DL, Waldron JA Jr, King TE Jr. Churg–Strauss pulmonary vasculitis. High-resolution computed tomography scanning and pathologic findings. *Am Rev Respir Dis*. 1990;142:458–461.
16. Lynch DA, Newell JD, Tschomper BA, et al. Uncomplicated asthma in adults: comparison of CT appearance of the lungs in asthmatic and healthy subjects. *Radiology*. 1993;188:829–833.
17. Park CS, Müller NL, Worthy SA, et al. Airway obstruction in asthmatics and healthy individuals: inspiratory and expiratory thin-section CT findings. *Radiology*. 1997;203:361–367.
18. Watts RA, Scott DG. Classification and epidemiology of the vasculitides. *Baillieres Clin Rheumatol*. 1997;11:191–217.