Case Report
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Churg-Strauss Syndrome with Cardiac Involvement: A Case Report with CT and MRI Findings[1]

심근병발을 동반한 Churg–Strauss 증후군: CT와 MRI 소견에 대한 증례 보고 1예[1]

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This is a case report of Churg-Strauss Syndrome (CSS) associated with cardiac involvement which is demonstrated in chest CT and cardiac MRI (CMR) without specific cardiac symptoms. A 32-year-old woman had a 3-year history of bronchial asthma, chronic sinusitis, and otitis media. The patient had various typical findings of CSS. The patient had no specific cardiac symptoms or signs such as chest pain, palpitations, syncope, or murmur, but she had diffuse low attenuation lesions in the inner wall of the left ventricle (LV) in contrast-enhanced CT. This corresponded to the area of subendocardial hyperenhancement in delayed contrast-enhanced CMR images. She was treated with steroids for 2 months. Follow-up delayed contrast-enhanced CMR of the LV showed a decrease in the size of the subendocardial enhancement area, and she had no symptoms. Therefore, the radiologist and clinician both should pay careful attention to observe possible cardiac involvement in case of CSS.

Index terms
Churg-Strauss Syndrome
CT
Magnetic Resonance
Cardiomyopathy
Lung Abnormalities

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INTRODUCTION

Churg-Strauss Syndrome (CSS) is a rare multi-systemic disease that involves the upper respiratory tract, skin, nervous system, gastrointestinal tract, kidneys, and heart (1-3). CSS is also known as allergic granulomatosis and angiitis (2). CSS was first reported by Churg and Strauss in 1951 and is characterized by necrotizing vasculitis, extravascular granuloma formation, and eosinophilic infiltration of various organs (2, 4, 5). Although the exact etiology of CSS is unknown, it is considered most likely related to an autoimmune disorder. The manifestations of cardiac involvement in patients with CSS include acute pericarditis (32%), acute heart failure (47%), and myocardial infarction and myocarditis, which account for one-half of the causes of death associated with CSS (1). We report a case of CSS associated with cardiac involvement without specific cardiac symptoms or signs, such as chest pain, palpitations, syncope, or murmur.

CASE REPORT

A 32-year-old woman had bronchial asthma for 3 years. She underwent a right mastoidectomy and tympanoplasty for chronic sinusitis and otitis media. She presented to the outpatient clinic for evaluation of progressive dyspnea on resting for an approximately 2 week period, which prevented her from sleeping. Other symptoms included numbness of the extremities, multiple skin rashes, and purpura on her right wrist (Fig. 1).

An electrocardiogram showed a sinus rhythm with a heart rate of 87 beats per minute (bpm) and moderate voltage criteria for left ventricular hypertrophy, which may be a normal variant. The patient had no cardiovascular symptoms or signs, such as chest pain, palpitations, murmur, or syncope. Initial laboratory testing showed an increased white blood cell (WBC) count and eosinophils on the differential count (WBC, 15100/μL; eosinophils, 31.7%). A four day follow-up laboratory finding

demonstrated more than a 10% increase in eosinophils. A biopsy obtained during a tympanoplasty for chronic otitis media at that time revealed infiltrating clusters of eosinophils in the middle ear.

The initial chest radiography showed multifocal patchy and non-segmental areas of consolidation mainly located in the periphery of both lungs (Fig. 2A). A follow-up chest radiography on the 4th hospital day demonstrated worsening of consolidation, despite treatment with antibiotics (Fig. 2B).

A chest CT scan (Aquilion 64; Toshiba Medical System, Tokyo, Japan) demonstrated a multifocal patchy consolidation and ground glass opacity in the peripheral area of the bilateral lungs without zonal predominance in coronal view (Fig. 2C). In the contrast-enhanced chest CT scan, diffuse low attenuation lesions were observed in the inner wall of the left ventricle (LV) from the base toward the apex. The same finding was noted in the short axis view of the contrast-enhanced CT scan (Fig. 3A). No pericardial effusion was noted. Multiple lymph node enlargements were observed in the mediastinum, which is considered to be reactive hyperplasia. Multiple ill-defined low attenuated lesions were also observed in the liver. Cardiac magnetic resonance (CMR) was performed using a 3-T MR system (Achieva, Philips Medical Systems, Netherlands). Delayed contrast-enhanced short-axis CMR images of the LV showed diffuse subendocardial enhancement of the LV from the base toward the apex (Fig. 3B). These findings were also found in delayed contrast-enhanced 4- and 2-chamber long-axis CMR images. The cardiac wall motion was relatively well-preserved, and no cardiac wall thickening was seen.

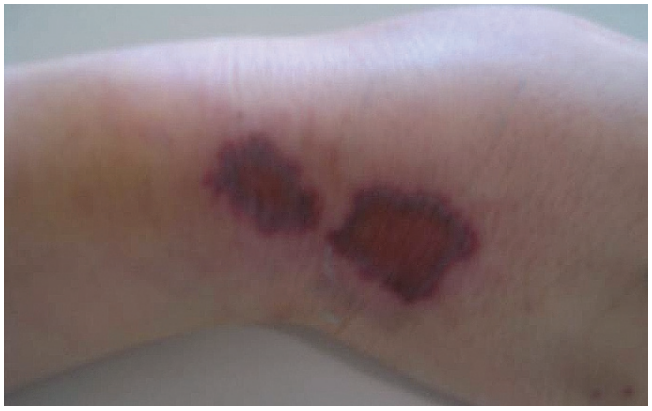
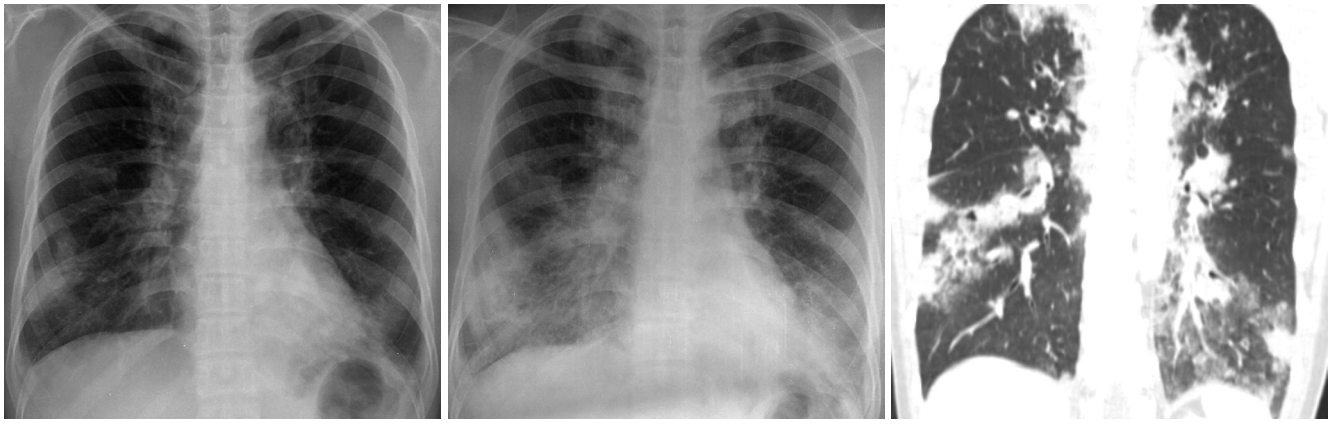


Fig. 1. Multiple skin rashes and purpura on the right wrist of the patient.

In comparing CT with MR images, the low-attenuated lesions of the LV inner wall in contrast-enhanced CT scans corresponded with the area of subendocardial enhancement in delayed enhanced CMR images (Fig. 3). As these lesions were not confined to the coronary artery territory and had a predominantly subendocardial location, we concluded that these lesions represented eosinophilic cardiomyopathy.

The most significant finding in a review of her medical history was a 3-year history of asthma, biopsy-proven eosinophilic infiltration in the middle ear, skin rash, purpura, neuropathy, peripheral eosinophilia, and abnormal radiological findings in the lung and LV in chest CT scans and CMR images. Therefore, we confirmed CSS with cardiac involvement.



A B C

Fig. 2. A 32-year-old woman with Churg-Strauss Syndrome.
A. Initial chest radiography shows bilateral multiple patchy and non-segmental consolidation, more prominent in the lower lung zone.
B. Chest radiography obtained 5 days later shows an increase in the size and extent of multiple consolidations despite antibiotic therapy.
C. The lung window of coronal CT scans obtained 5 days after initial chest radiography shows a multi-focal patchy distribution of consolidation and surrounding ground glass opacity in the peripheral area of both lungs without zonal predominance.

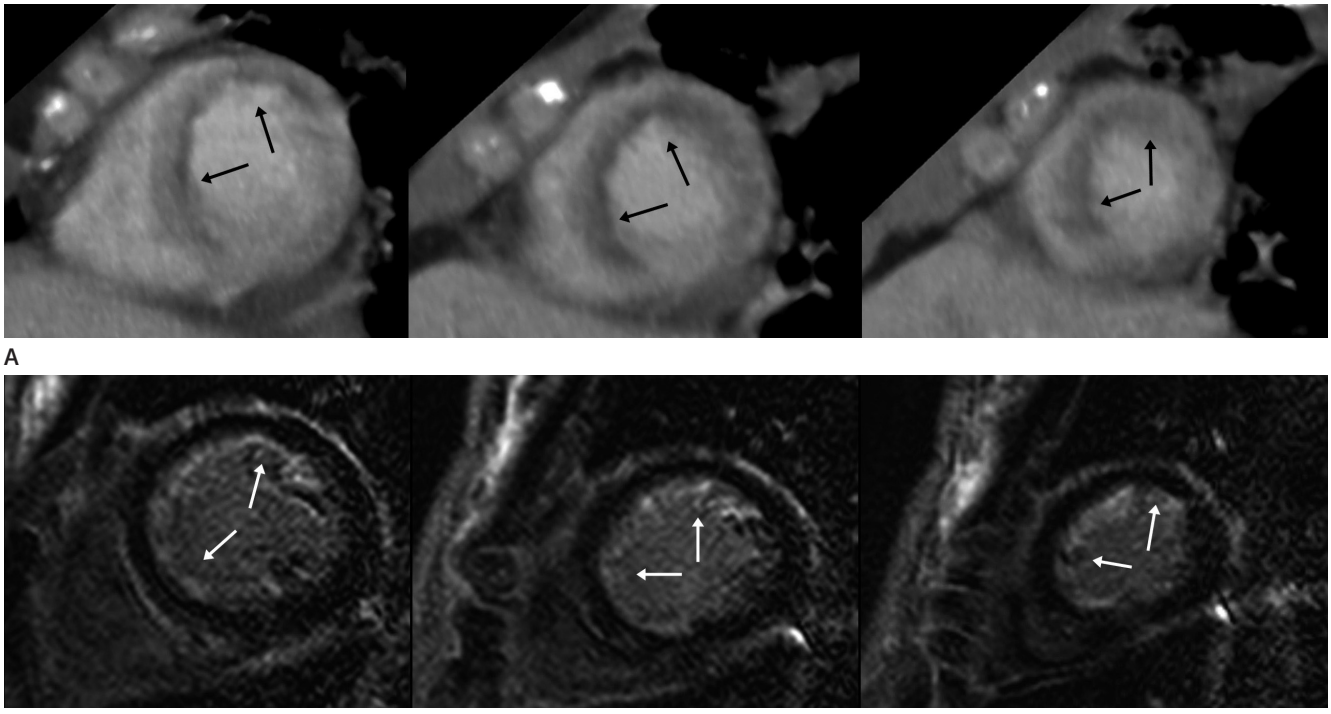


Fig. 3. Contrast-enhanced chest CT scans.
A. Short axis views show diffuse low attenuation lesions (black arrows) in the inner wall of the LV from the base toward the apex.
B. Delayed contrast-enhanced CMR images of the LV obtained from the base toward the apex. Diffuse subendocardial enhancement (white arrows) is observed in the LV that is not limited to a vascular distribution.
Note.—CMR = cardiac magnetic resonance, LV = left ventricle

She was under oral steroids therapy (1.0 mg/kg daily) for 2 months. At the time of follow-up, she was asymptomatic, and her functional status had improved. The peripheral eosinophil count decreased to 6.0% at that time, and the follow-up chest radiography was normal. Three year follow-up CMR demonstrated a decrease in the size of the subendocardial enhancement area (Fig. 4), and she had no symptoms.

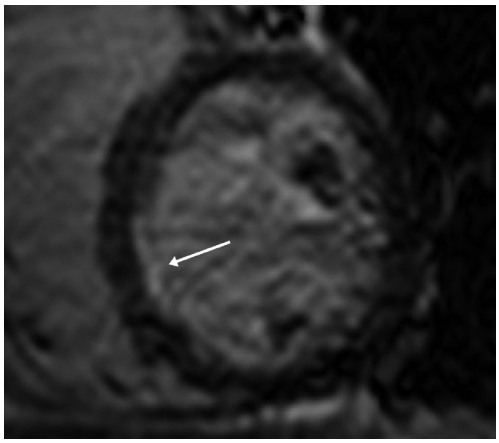


Fig. 4. Follow-up cardiac magnetic resonance imaging. Remaining subendocardial enhancement (arrow) is still observed in the interventricular septum of the LV in the delayed contrast-enhanced CMR of the LV, but the extent of enhancement is decreased.
Note.—CMR = cardiac magnetic resonance, LV = left ventricle

DISCUSSION

CSS was first described in 1951 as an independent disease that could be differentiated from polyarteritis nodosa (4). The presence of four or more of the following six criteria establishes a diagnosis of CSS with a sensitivity of 85% and a specificity of 99.7%: asthma, eosinophilia of more than 10% in a WBC differential count, mono- or poly-neuropathy, migratory or transient pulmonary opacity, paranasal sinus abnormality (such as sinusitis), and vasculitis or extravascular eosinophils at biopsy (2, 3, 5, 6). The essential etiology of CSS is thought to be vasculitis (7).

This patient had eosinophilia, a neuropathy, a pulmonary opacity, and a paranasal sinus abnormality. She also had multiple ill-defined low-attenuated lesions in the liver and a diffuse lower attenuation lesion in the inner wall of the LV in the contrast-enhanced CT scan and delayed subendocardial enhancement in the LV in the CMR image which we thought represent-

ed an eosinophilic infiltration. Thus, we confirmed CSS with cardiac involvement.

Clinically, there are three distinct phases associated with CSS. The first is the prodromal phase consisting of allergic rhinitis, nasal polyposis, or bronchial asthma (often preceded by allergic rhinitis). The second phase is marked peripheral blood eosinophilia and eosinophilic tissue infiltration (lung or gastrointestinal tract) or chronic eosinophilic pneumonia, which may recur over a period of years. The third phase is a life-threatening systemic or small-vessel vasculitic phase (3). A skin rash is one of the most common features of the vasculitic phase of CSS (3); this patient was thought to be in the third phase.

Cardiac involvement is the major cause of death in patients with CSS (8), which accounts for 48% of deaths attributed to CSS, including acute pericarditis, constrictive pericarditis, cardiac failure, and myocardial infarction with or without pericardial effusion (4). As the heart is a primary target organ in CSS causing a grave prognosis, delayed treatment can lead to intractable cardiac failure (2).

It is known that infiltrating eosinophils can damage the endocardium and vascular endothelium, which promotes the development of endomyocardial fibrosis. In CSS, this eosinophilic infiltration predominantly involves endocardium compared to the other non-ischemic cardiac myopathies, such as viral myocarditis which predominantly involves middle or epicardial layers of myocardial wall (7). Additionally, cavity dimensions and systolic function are often preserved, and subtle echocardiographic abnormalities can easily be overlooked in CSS (8). Cardiac magnetic resonance may provide additional information regarding myocardial damage even in the presence of preserved cardiac wall motion and cavity size (8). Thus, it has been advocated that endomyocardial biopsies be performed liberally and serially to assess disease activity in the myocardium (2).

In this case, the patient had neither cardiac symptoms or signs nor unusual findings in electrocardiography. However, the patient had radiological cardiac manifestations in CT and CMR, which was thought to be eosinophilic cardiomyopathy. As noted above, cardiac involvement in CSS requires long-term therapy and has a poorer prognosis than CSS without cardiac involvement (3, 5). Therefore, the radiologist and clinician should both pay careful attention to observe possible cardiac involvement and must make an accurate diagnosis.

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심근병발을 동반한 Churg-Strauss 증후군: CT와 MRI 소견에 대한 증례 보고 1예[1]

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특별한 심장 이상 소견은 없었으나 흉부 CT와 심장 MRI에서 심근을 침범한 Churg-Strauss 증후군(이하 CSS)을 보고하고자 한다. 32세 여자가 3년 동안의 기관지 천식, 만성 부비동염과 중이염 병력이 있었고, 전형적인 Churg-Strauss 증후군의 소견이 다수 보였다. 환자는 흉통, 심계항진, 실신, 잡음과 같은 특별한 심장 이상의 증상 및 징후는 없었으나, 흉부 조영증강 CT에서 좌심실의 내측 벽에 미만성 저음영 병변이 있었고, 이들 부위는 조영증강 심장 MRI의 지연기에서 조영증강되는 부위와 일치하였다. 환자는 두 달간 스테로이드를 복용하였고, 치료 후 시행한 심장 MRI에서 좌심실 벽의 조영증강되는 부위의 범위가 감소하였다. 이에 영상의학과 의사와 임상의는 CSS 환자에서 심근 침범 여부를 주의 깊게 관찰하여야 한다.

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