Concomitant presentation of myocarditis and non-bacterial thrombotic endocarditis in a patient with granulomatosis with polyangiitis

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Abstract

Granulomatosis with polyangiitis (GPA) is a necrotizing vasculitis. The etiology of the disease is not completely known. In this report, we describe a 47-year-old male who presented with ongoing dyspnea. He is a known case of GPA for 3 years based on skin biopsy and positive antineutrophil cytoplasmic antibody. The electrocardiogram showed changes suggestive for myocarditis. A transthoracic and trans esophageal echocardiogram revealed left ventricular systolic dysfunction, thickened left atrial and ventricular wall and vegetation on mitral valve. Blood cultures were negative. Myocarditis and nonbacterial thrombotic endocarditis (NBTE) were diagnosed.

Introduction

Granulomatosis with polyangiitis (GPA) is a necrotizing vasculitis of small vessels that affect respiratory tract and the kidneys. The etiology of the disease is not completely explained yet, but it seems that both environmental and genetic factors play roles in this disease. GPA involve many organs; among them kidneys, ear, nose, throat and lungs are more frequently involved; but eyes, cartilages, skin and other organs can also be affected. In a patient presenting with clinical findings suggestive for GPA, the confirm diagnosis is made by a tissue biopsy or sufficient positive titer of antineutrophil cytoplasmic antibody (c-ANCA) which is highly specific for GPA. Suggested treatment for GPA consists of IV glucocorticoids and oral cyclophosphamide. About 50-70% of patients experience relapsing episodes. According to different studies, echocardiographic abnormalities due to cardiac involvement in GPA is relatively common, including systolic dysfunction, pericardial effusion, valvular involvement and so far. Nonbacterial thrombotic endocarditis (NBTE) is a rare manifestation of GPA and to our knowledge its concomitant presentation with myocarditis is not reported to date. Herein, we report a case of GPA, who diagnosed with both myocarditis and NBTE.

Case Report

This patient was a 47-year-old male, who was diagnosed with GPA 3 years earlier with recurrent episodes of polyarthritis, petechiae and abnormal renal function test which lead to end stage renal disease (ESRD). The diagnosis of GPA was made based on skin biopsy and c-ANCA titer of 31.5 (greater than 9.0 is considered as positive). He was started on immunosuppressive therapy (prednisolone and azathioprine), but he has discontinued treatment after few months. No, He does dialysis 2 times a week. After months of being relatively healthy, he presented to our outpatient clinic with increased shortness of breath and orthopnea, which limited his daily activities. On physical examination, he was alert and afebrile, diminished breath sounds at both sides specially left side and moist rale up to half of both lungs. He was admitted to our hospital for more evaluation.

Laboratory data, including complete blood count (CBC), blood urea nitrogen (BUN), creatinine (Cr), serum electrolytes and erythrocyte sedimentation rate (ESR) are shown in Table 1.

| Table 1. Laboratory data, including complete blood count (CBC), blood urea nitrogen (BUN), creatinine (Cr), serum electrolytes and erythrocyte sedimentation rate (ESR) |
|---|---|---|---|---|
| Hemoglobin | 13.5 | g/dL | 13.5 | g/dL |
| White blood cells | 7,500 | /μL | 7,500 | /μL |
| Erythrocyte sedimentation rate (ESR) | 50 | mm/h |
| Creatinine | 3.5 | mg/dL |
| Blood urea nitrogen (BUN) | 45 | mg/dL |

Chest X-ray showed small bilateral pleural effusion also perm catheter for dialysis (Figure 1).

The ECG showed: sinus tachycardia, T wave inversion, LHV voltage criteria (Figure 2).

Transthoracic echocardiography (TTE) was done and it revealed LV systolic dysfunction and a small and irregular vegetation on anterior or mitral valve leaflet (Figure 3).

Transesophageal echocardiography was also performed (TTE) and it confirmed previous findings.

Several blood cultures were negative and no growth was observed after five days of incubation. No sign or symptom compatible with cancer was mentioned. No embolic event was occurred. We requested serological testing (including Anti cadiolipinAb, lupus anticoagulant test, ANA, Anti ds-DNA, p-ANCA, C3, C4 and CH50). All were within normal limits, except: c-ANCA: 20 (greater than 9.0 is considered as positive)

The patient underwent emission computed tomography (99m Tc-MIBISPECT) scanning (Figure 4) after 10 days of immunosuppressive therapy (60 mg/d prednisolone and 100 mg/d azathioprine). It showed persistent hypo activity in apical anteroseptal segment and reversible defects in the mid to apical portion of anterior wall, hypo activity of septum, LV dilation and dramatically increased global LV function to 50%.

The diagnosis was myocarditis and non bacterial thrombotic endocarditis (NBTE) due to relapse of the disease.

According to insignificant mitral regurgitation and no obvious embolic event patient was discharged with increased dose of immunosuppressive drugs, 80 mg ASA and conventional heart failure management. He was strongly recommended to a complete course of treatment, close symptoms and echocardiography follow up.

Discussion

In echocardiography, mobile masses on valve constructions usually are vegetations...
from subacute bacterial endocarditis, tumors, or (rarely) thrombi. In the absence of symptoms, benign tumors such as fibroelastomas often are considered. The finding of a large, intracardiac mass caused by NBTE, without known malignancy or systemic lupus erythematosus or anti phospholipid antibody syndrome, is rare. Vegetations produced by NBTE are found in ~1.2% of autopsies, but the reported prevalence varies widely (0.3% to 9.3%) and predominantly involves patients with coexisting malignancy, sepsis, burns, or disseminated intravascular coagulation. However, autoimmune disease are found in

Table 1. Complete blood count, blood urea nitrogen, creatinine and erythrocyte sedimentation rate.

<table>
<thead>
<tr>
<th>Tests</th>
<th>Result</th>
<th>Units</th>
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<tbody>
<tr>
<td>Red blood cell</td>
<td>3.66</td>
<td>mL/cumm</td>
</tr>
<tr>
<td>Hemoglobin</td>
<td>8.5</td>
<td>g/dL</td>
</tr>
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<td>Hematocrit</td>
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<td>%</td>
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<tr>
<td>Platelet</td>
<td>113</td>
<td>cumm</td>
</tr>
<tr>
<td>Mean corpuscular hemoglobin concentration</td>
<td>29</td>
<td>g/dL</td>
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<tr>
<td>Mean corpuscular hemoglobin</td>
<td>23.2</td>
<td>pg</td>
</tr>
<tr>
<td>Blood urea nitrogen</td>
<td>60</td>
<td>mg/dL</td>
</tr>
<tr>
<td>Creatinine</td>
<td>12.5</td>
<td>mg/dL</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate</td>
<td>40</td>
<td>mm</td>
</tr>
</tbody>
</table>

Figure 1. Chest X-ray shows bilateral pleural effusion and perm catheter for dialysis.

Figure 2. Electrocardiogram shows tachycardia, T wave inversion and LVH voltage criteria.

Figure 3. Transthoracicechocardiograhy shows left ventricle systolic dysfunction and a vegetation on anterior mitral valve leaflet.

Figure 4. Computed tomography scanning shows findings compatible with myocarditis.
60% of patients who undergo surgical resection for an intracardiac mass caused by NBTE. Among autoimmune disease, antiphospholipid syndrome (APS) and systemic lupus erythematosus (SLE) are more frequently associated with this condition. NBTE in patients with GPA is rarely reported in literature and in some cases has been presented by embolic events. To the best of our knowledge, this was the first case of granulomatosis with polyangiitis with both NBTE and myocarditis, which was diagnosed before life threatening embolic events.

Conclusions

Herein, we reported an unusual patient who presented with dyspnea. The ECG changes suspected us to myocarditis. We performed TTE which showed LV systolic dysfunction and a mass on the anterior mitral valve leaflet, suggestive for NBTE. Our diagnosis was confirmed with TEE and computed tomography scanning. Our case emphasizes the need for searching for cardiac involvement in all autoimmune and connective tissue diseases by echocardiography to prevent life threatening complications.

References