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Case Report

Loeffler's endocarditis in a patient with a new diagnosed Churg-Strauss syndrome (CSS): A case report

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Abstract

Background: Loeffler's endocarditis is a rare disease, caused by endocardial involvement of esosinophils, which damages the heart and leads to endomyocardial fibrosis with consequent restrictive cardiomyopathy, mural thrombi or valvular dysfunction. The association between Loeffler's endocarditis and Churg-Strauss syndrome (CSS) was also reported. Abnormal elevation of peripheral eosinophil counts in a heart failure patient is a hint of disease. Though echocardiography or cardiac magnetic resonance imaging (MRI) facilitates diagnosis, endomyocardial biopsy is still the gold standard. Treatments include immunosuppressive agents, anticoagulant and guideline-directed medical therapy for heart failure.

Case presentation: A 59-year-old man presented with progressive dyspnea for one week and he was referred to our hospital for surgical treatment evaluation because valve destruction by infective endocarditis has been suspected at the local hospital. Echocardiography revealed biventricular mural thrombus and limited aortic valve opening caused by abutting thrombus. Moreover, eosinophilia, bronchial asthma, lung infiltration, acute kidney injury and positive perinuclear anti-neutrophil cytoplasmic antibody (p-ANCA) test implied Churg Strauss syndrome. Eosinophil infiltrate with fibrin thrombus was revealed by endomyocardial biopsy. The patient was diagnosed of cardiac involvement of CSS and recovered after immunosuppressive and anticoagulant treatments.

Conclusion: Loeffler's endocarditis should be suspected when physicians encounter restrictive cardiomyopathy accompanied by mural thrombus in a patient with eosinophilia. Prompt immunosuppressive and anticoagulant medication can bring the disease under control.

Keywords: Loeffler's endocarditis, Churg-Strauss syndrome, Hypereosinophilic syndrome

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Loeffler's endocarditis is a rare disease caused by cardiac involvement of eosinophils, which damages the heart and leads to endomyocardial fibrosis. Patients would have heart failure symptoms with abnormal elevation of peripheral eosinophil counts. The diagnosis can be facilitated by echocardiography, which shows restrictive pathophysiology, mural thrombus without akinetic or aneurysmal ventricular wall, valve motion restriction and ventricular chambers obliteration (1). Cardiac MRI also facilitates diagnosis, which can reveal diffuse endocardial hyper-enhancement and delineate left ventricle thrombus causing apical obliteration on a Gadolinium enhanced study (2). Though invasive, endomyocardial biopsy is still the gold standard of diagnosis. Loeffler's endocarditis can be a cardiac involvement in a patient with CSS, which is similar to hypereosinophilic syndrome (HES), both of which are multiple organ involvement with eosinophilia.

The presence of asthma, vasculitis and positive p-ANCA are characterized by CSS other than HES (3). Another study found cardiac involvement in patients with CSS is associated with the absence of ANCA and high eosinophil count (4). In this case report, we described a new diagnosed Churg Strauss syndrome patient with Loeffler's endocarditis and the patient recovered after immunosuppressive treatments.

Case presentation

A 59-year-old man was referred to our hospital for dyspnea on exertion for one week. He had been diagnosed with Streptococcus suis infectious endocarditis about 3 months ago, which was presented as fever initially, and one month antibiotics treatment was completed at local hospital. However, months later, he had mild fever again, along with new onset progressive dyspnea for one week, bilateral upper extremity numbness, weakness and mild cognitive impairment. He was referred to our hospital for a suspecting acute heart failure related to infectious endocarditis with valve destructing vegetation. He inhaled corticosteroid for 20 years for his underlying bronchial asthma with poor control. Upon admission, his clinical status was acute, ill looking and alert. Vital signs were: temperature 37°C, pulse rate 105 beats/minute, respiratory rate 19 cycles/min, blood pressure 113/82 mmHg and 91% saturation under ambient air. Physical examination showed regular heart beats, normal S1, increased P2, without S3, S4 and systolic murmur over right upper sternal border. All muscle power and sensory neurological test were documented within normal range even with subjective weakness and numbness.

Electrocardiography showed sinus tachycardia, interventricular conduction delay and left atrium enlargement. Chest x-ray showed cardiomegaly, left pleural effusion and confluent patchy infiltration over right and lower lung field. Blood laboratory examination were: white blood cell 12200/uL, the percentage of differential counts, neutrophil 64%, lymphocyte 10.4% and eosinophil 21.1%, hemoglobin 10.4 g/dL, and platelets 131,000/uL, erythrocyte sedimentation rate 55 mm/hr (0-15), N- terminal pro-brain natriuretic peptide (NT-proBNP) >35000 (<125), troponin I 0.105 ng/mL (<0.056), C reactive protein 4.36 mg/dL, serum glucose 132 mg/dL, ALT 20 U/L (5-50), and serum creatinine 3.60 mg/dL. Urinalysis showed microscopic hematuria without pyuria. Echocardiography was checked, which revealed biventricular mural thrombus, abutting thrombus restricting aortic valve and mitral valve motion with mean transaortic valve pressure gradient 33mmHg (fig. 1A-C), moderate aortic regurgitation and moderate to severe mitral regurgitation. Left ventricular systolic function was mildly impaired with left ventricular ejection fraction 45%, tissue Doppler showed reduced systolic and diastolic mitral annulus velocity (lateral S'= 4.64 cm/s, E'= 8cm/s and A'= 4.2 cm/s) and estimated right ventricular systolic pressure was 58 mmHg. Moderate aortic regurgitation and severe mitral valve regurgitation were also documented. Because of acute heart failure syndrome with valvular dysfunction, coronary angiography was arranged and excluded coronary artery stenosis. Cardiac hemodynamics showed right ventricular dip and plateau pressure tracing and prominent y descending, compatible with pathophysiology pattern of restrictive cardiomyopathy.

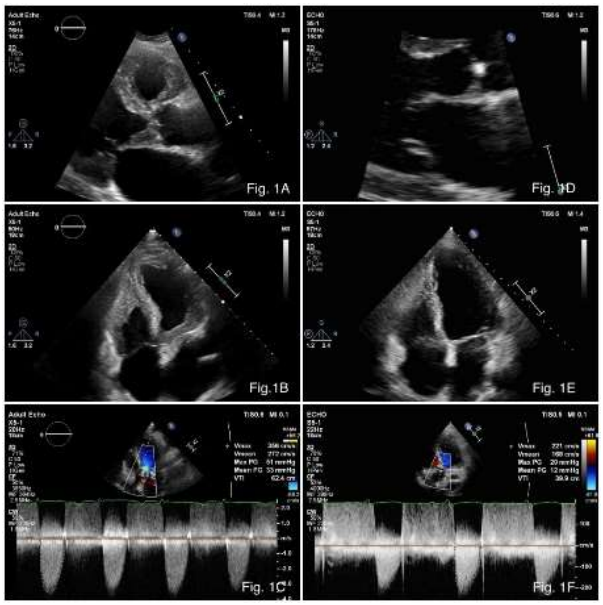

Fig. 1A and 1B: mural thrombus involving biventricular chambers and aortic valve.

Fig. 1D and 1E: Complete resolution of thrombus after months of anticoagulation and immunosuppressive treatment.

Fig. 1C and 1F: Aortic valve pressure gradient decreases after treatment.

Endomyocardial biopsy revealed eosinophil infiltrate with cardiac myocyte necrosis and fibrin thrombus (fig. 2A-C). Bone marrow biopsy was arranged, which showed eosinophil proliferation (fig.2D) and real time polymerase chain reaction showed negative result of the platelet-derived growth factor receptor-α (PDGFRA) gene fusing to a FIP1-like-1 (FIP1L1)

gene. Right patchy pulmonary infiltrate cannot be explained only by heart failure and typical pneumonia. Chest computer tomography was arranged, which showed right upper and lower lung ground glass opacity with patchy consolidation. Obstructive airway disease (the forced expiratory volume in the first seconds (FEV1) 46% and FEV1 to forced vital capacity (FVC) ratio 57% percentage of reference) was shown on pulmonary function test. Kidney sonography showed no hydronephrosis and kidney atrophy, with small renal stones in both kidneys.

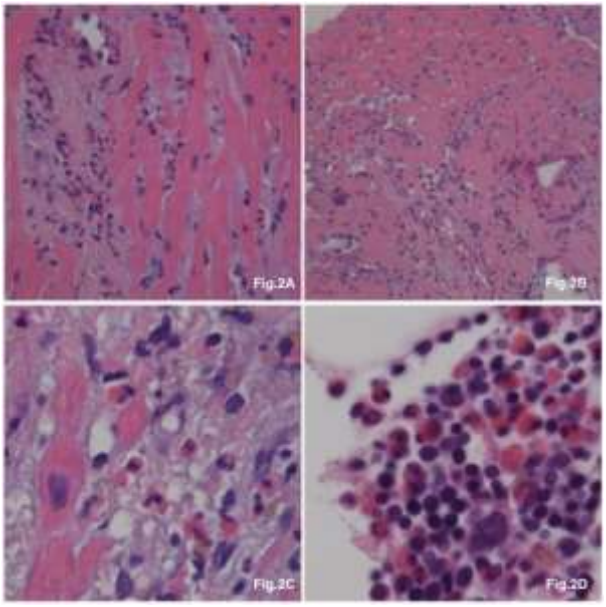

Fig. 2A: Eosinophil and lymphocyte infiltrates in cardiac muscle (hematoxylin and eosin stain, H&E stain, 400X).
Fig. 2B: A fibrin thrombus attach on endocardium (H&E stain, 200X).
Fig. 2C: The presence of myocyte necrosis (H&E stain, 1000X).
Fig. 2D: eosinophil proliferation within bone marrow (H&E stain, 1000X)

The finding of nerve conduction velocity and electromyogram of upper and lower extremities were compatible with sensory motor polyneuropathy and demyelinating type. Works-up of autoimmune disease and parasites infection showed negative stool ova and parasites examination, antinuclear antibody (ANA): negative (<1:80), cytoplasmic anti-neutrophil cytoplasmic antibody (c-ANCA): negative; p-ANCA: positive, anti-double stranded DNA (anti-dsDNA): negative. Because polyneuropathy, poorly-controlled asthma with airway obstruction, acute kidney injury and migratory pulmonary infiltrate, Loeffler's syndrome associated with Churg-Strauss syndrome was impressed. Intravenous methylprednisolone 40 mg four times a day and cyclophosphamide 600mg pulse therapy were initiated and warfarin 2.5 mg per day, keep international normalized ratio (INR) 2-3, after bridging Dalteparin 5000IU twice a day was used for mural thrombus. Dyspnea and general weakness subsided gradually, along with renal function improvement and resolution of left pleural effusion and pulmonary infiltrate. He was discharged after 3 weeks admission with oral prednisolone 5 mg and azathioprine 50mg twice a day. Eosinophilia remitted after immunosuppressive treatment and followed echocardiography months later showed mural thrombus disappeared completely and aortic valve motion recovered with little pressure gradient (fig. 1D-F).

Discussion

Loeffler's endocarditis is a cardiac manifestation of hypereosoinophilic syndrome, which can be divided into idiopathic, primary and secondary types based on the underlying disease (1). Churg Strauss syndrome can be considered as a combination of vasculitis and organ damaging eosinophilia. Diagnosis is made in the presence of four or more of six criteria, including asthma, eosinophilia over 10%, neuropathy, non-mixed pulmonary infiltrates, paranasal sinus abnormality and extravascular eosinophils (5). There are many overlaps of clinical presentation of HES and CSS, but the presence of bronchial asthma, polyneuropathy and positive p-ANCA test in the present patient are associated with CSS rather than HES (3).

Not only migratory pulmonary infiltrate but also acute kidney injury can be related to vasculitis. Though kidney or pulmonary biopsy is crucial to differential diagnosis, which can reveal granulomatous vasculitis, the patient hesitated about the procedure and prompt immunosuppressive treatments should be initiated. Successful immunosuppressive, anticoagulant and standard heart failure treatment for Loeffler's endocarditis associated with Churg Strauss syndrome were reported in the previous articles (6, 7). The patient also underwent bone marrow biopsy, which excluded myeloproliferative neoplasm and disclosed negative FIP1L1-PDGFR fusion gene, both of which were associated with primary hypereosinophilic syndrome (1). The patient had acute heart failure syndrome with aortic stenosis caused by

abutting thrombus restricting aortic valve motion, which was completely resolved after immunosuppressive and anticoagulant treatment. The pathology of endomyocardial biopsy of Loeffler's endocarditis in the present patient included endomyocardial inflammation with eosinophil infiltration and fibrin thrombus. No disclosed endomyocardial fibrosis may imply the response to immunosuppressive treatment. Aortic valve motion restriction and mural thrombus completely disappeared after immunosuppressive and anticoagulant treatment.

In conclusion, this case report describes a typical echocardiography finding of Loeffler's endocarditis, associated with Churg-Strauss syndrome with cardiac involvement. Eosinophil infiltrates within myocardial can be revealed in endomyocardial biopsy. The patient can recover completely after prompt immunosuppressive and anticoagulant treatments.

Author Statement

Authorship Responsibility: All authors have seen and approved the content of the submitted manuscript. The paper presents original work not previously published in similar form and not currently under consideration by another journal. If the paper contains material (data or information in any other form) that is the intellectual property and copyright of any person(s) other than the author(s), then permission of the copyright owner(s) to publish that material has been obtained, and is clearly identified and acknowledged in the text of the paper. The authors followed ethics guidelines.

Author Agreement

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