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Newly diagnosed patient with eosinophilic granulomatosis with polyangiitis (Churg-Strauss) in the cardiac ward - A case report

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Abstract

Background: Churg-Strauss syndrome is a disorder marked by blood vessel inflammation. It is a multisystem disorder characterized by asthma, prominent peripheral blood eosinophilia, and vasculitis. This condition is also known as eosinophilic granulomatosis with polyangiitis.

Case Summary: We presented here a 52-yr old male who has been diagnosed with Churg Strauss disease while he was admitted to the cardiac ward as a case of acute coronary syndrome. Heis known to have bronchial asthma, diabetes, and hypertension, and he has a history of coronary angiography10 years ago. He presented to the emergency department with complaints of shortness of breath, and orthopnea with progressive lower limb swelling over the last 2 weeks. Associated with ischemic-type chest pain for the last 2 days. Depending on the history, presenting symptoms, blood investigations, and the histopathology of the gallbladder biopsy the diagnosis of Churg-Strauss syndrome was confirmed.

Conclusion: we reported a confusing case of asthma, progressive dyspnea, weight loss, and bilateral lower limb ulcers finally diagnosed as CSS with cardiac involvement. In any patient with refractory asthma, we should not neglect the diagnosis of CSS. Effective treatment methods can improve the prognosis if diagnosed in time.

Abbreviations: ANCA = antineutrophil cytoplasmic antibodies, CPR = C-reactive protein, CSS = Churg–Strauss syndrome,

Keywords: Churg–Strauss syndrome, acute coronary syndrome, histopathology.

Introduction

Churg–Strauss syndrome (CSS) is an eosinophil-rich necrotizing vasculitis of small-to-medium blood vessels that affects many organs including cardiac, pulmonary, renal, nervous, and vascular systems.[1] Symptomatic

cardiovascular involvement occurs in as much as 27% to 47% of CSS cases and can present with eosinophilic vasculitis, pericarditis, pericardial effusion, valvular heart disease, cardiomyopathy, acute myocardial infarction, myocarditis, and acute heart failure. [2–4] Although uncommon, cardiac involvement is a major cause of morbidity and mortality in this disorder. Here we report a newly diagnosed case of CSS presenting with acute coronary syndrome in the cardiac ward. We reviewed the literature on CSS with cardiac involvement.

Case report

52 yrs. old male presented to the emergency department with complaints of shortness of breath, and orthopnea with progressive lower limb swelling over the last 2 weeks. Associated with ischemic-type chest pain for the last 2 days. He reported 35 kg weight loss over the last 5 months associated with night sweats.

Regarding his past medical history, he is known to have bronchial asthma, diabetes, hypertension, history of coronary angiography10 yrs. ago.

In addition, he reported recurrent joint pain and skin rashes, chronic dyspepsia, and dysphagia. He is on omeprazole 40 mg od, aspirin 81md od, Lipitor 40 mg od, Lasix 40 mg od, salbutamol 2 puff sod, Atrovent 2puffs od, Ferrol 1tab od, perindopril 5mg od and fuciderm topical bid.

The patient was admitted to the medical cardiac ward as a case of no segment elevation myocardial infarction for medical treatment and possible cardiac catheterization. On general examination, he was pale, cachexic, with a dry mouth, and multiple aphthous ulcers. Lower limb examinations: tenderness at the knees and elbows, bilateral lower limb ulcers, and skin rashes. His vital signs: BP: 150/90, PR: 92, Temp: 36.9, SPO2: 96%.

During his hospital stay, he developed acute stridor for which an ENT consultation was done and revealed acute epiglottitis.

The patient had received iv dexamethasone with good improvement. A Pan CT scan and chest x-ray were done and revealed a huge hiatus hernia with gastric herniation. In addition, there was a calculator gallbladder.

The surgical team decided on urgent hiatal hernia repair with mesh + cholecystectomy+ fundoplication. The sam ple of the gallbladder was sent for histopathology.

Cardiac catheterization wasn't done due to recent surgery. Given his history of bronchial asthma, ischemic heart disease, GERD, weight loss, night sweats, significant anemia, multiple joint pain, and skin rashes, the medical team advised investigations for Autoimmune diseases, tumor markers, chronic infections including AAFB sputum C/S, ulcers swab C/S.

Investigations results and progression during hospital stay

- Serum troponin on admission:0.361 then 0.440
- HB: 8.5 on admission and before the surgery, so he was transfused with 2 units of RBCs, 6 units of FFP, and 6 units of the platelet.
- Serum Albumin was 25.
- ESR: 50. CRP: 168.
- CBC showed marked EOSINOPHILIA:1.9.
- Echocardiography: EF: 40%
- Autoimmune studies: C3, C4, ANA, D2DNA, tumor markers ALL were NEGATIVE.
- AAFB: NEGATIVE.
- Leg ulcer swab showed: coagulase -ve.

Pt was discharged in good condition on ant-ischemic, anti-failure medications and to follow his

regular medications at home. Cardiac catheterization was deferred due to recent surgery. The biopsy result was still bending.

Four months later, he had been admitted to the intensive care unit with severe shortness of breath. His Spo2 was 88% on room air, and a diagnosis of Asthma exacerbation Vs decompensated heart failure was entertained.

During this admission cardiac catheterization was done and showed no obstructive coronary-artery disease. Echocardiography revealed an ejection fraction of 40% with no interval changes.

Microscopic examination of the gallbladder after cholecystectomy shows

Multifocal vascular lesions with perivascular erosive eosinophilic infiltration and fibrinoid necrosis of small to medium-sized arteries. considering the patient's prior history of bronchial asthma, multiple skin rashes, and peripheral eosinophilia, Churg - Strauss syndrome is highly suggested.

This result was agreed upon by two doctor's consultant histopathologist.

Later on, this patient has been on follow-up with the rheumatologist and during his hospital stay, he received 6 cycles of iv Cyclophosphamide. Then he started on Azathioprine 50 then 150 then 200 mg. Also, he received Mepolizumab 300 mg (1^{st} dose only). A tracheostomy was done to this given recurrent attacks of acute obstructing epiglottitis.

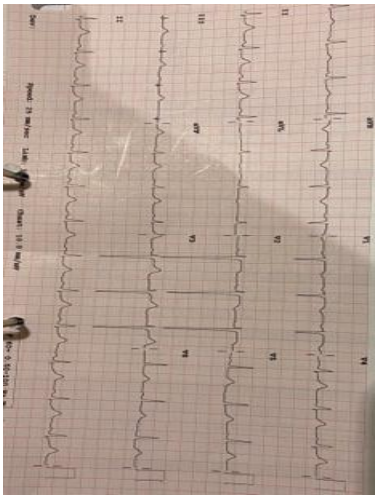

Figure 1

This is the ECG of the pt. upon admission.

This is the chest radiograph during hospitalization showing a large hiatus hernia

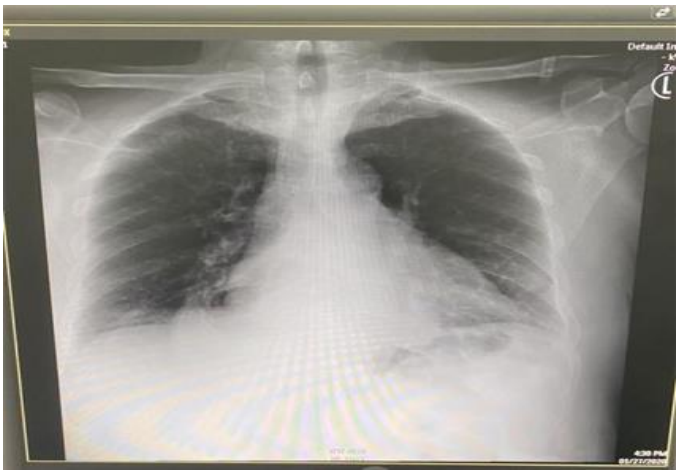

Figure 2

Discussion

CSS has an extremely low incidence, ranging from 0.5 to 6.8 new cases per million patients per year, and prevalence from 10.7 to 13 per million adults, varying by location and the diagnostic criteria applied. The mean age at diagnosis is 48 years,[5] and both sexes are affected equally. We have little data on the race distribution. The diagnostic criteria of CSS require the presence of any 4 or more of the following: asthma, eosinophilia >10%, neuropathy, pulmonary infiltrates, paranasal sinus abnormality, or extravascular

eosinophils.[6] Our patient had had asthma for 4 years, and the eosinophil proportion was >10% of the white cell count. Furthermore, the bilateral lower limbs numbness and ulcers represented neuropathy. Finally, the gallbladder biopsy revealed Multifocal vascular lesions with perivascular erosive eosinophilic infiltration and fibrinoid necrosis of small to medium-sized arteries. In addition to acute the chronic epiglottis. Thus, our case met the diagnosis of CSS. On admission to the cardiac ward, our patient felt sudden onset dyspnea and stridor, an ENT consultation was done and revealed a diagnosis of acute epiglottitis. Our patient received iv steroids with good improvement in symptoms. Cardiac catheterization revealed non-obstructive coronary artery disease. Echocardiography showed no problems with cardiac structure and no valvular vegetation. To explore the causes of continuous eosinophilia, we performed laboratory examinations. For parasite infection, our patient had no recent contact, and the sputum culture did not reveal relevant microbial pathogens. Furthermore, a consultant physician suggested an autoimmune screening as well.

Thus, we could exclude rheumatological diseases. Glomerular filtration rate and urea and urine testing were normal, which helped rule out interstitial nephropathy. The pathophysiology of CSS can be divided into 3 stages. The prodromal stage is characterized by asthma and atopic disease and can last a few years, even as long as 30 years, according to the report. In the next stage, eosinophils infiltrate into tissues such as the lungs or myocardium. The final stage is usually the diagnosis and when necrotizing vasculitis appears.[7] The pathogenesis of CSS is not detailed thoroughly. Genetics, environment, and interactions all play important roles. Several inducing factors, such as infection, drugs, and especially leukotriene receptor antagonists (e.g., montelukast) [8] and vaccinations, have been found responsible for the onset of CSS. Immune functional disturbance and dysregulated release of cytokines have been suggested to be associated with eosinophilic disorders.[9] CSS patients usually have asthma, whereas leukotriene receptor antagonists may be involved in the onset of CSS.[10] Patients are positive for erythrocyte sedimentation rate and CRP level, indicating an inflammation response, whereas CSS patients with cardiac involvement are usually ANCA-negative. CSS usually responds quickly to immunosuppressive therapy, associated with rather a good prognosis. Corticosteroids are the first-line therapy, resulting in remission and improved survival. When recurrences are frequent or associated with a serious form of necrotizing vasculitis in organs such as the gastrointestinal tract or the heart, the use of cyclophosphamide is recommended. Such favorable outcomes might not apply to patients with organ system involvement that indicates a poor prognosis. The French Vasculitis Study Group has recently revised 5 prognostic factors, the so-called 5-factor score (FFS). The new FFS comprises 4 factors that indicate poor prognosis (age >65 years, cardiac involvement, gastrointestinal manifestations, and renal impairment characterized by serum creatinine level >150mmol/ L) and 1 factor that indicates a better outcome (the absence of ear, nose and throat manifestations). According to the revised FFS, the presence of 0, 1, or 2 factors associated with CSS represents necrotizing vasculitis of medium and small vessels, typically characterized by asthma and hyper eosinophilia. Multiple systems can be involved in CSS, which easily misleads doctors to other diseases. We conclude that (1) the younger age of CSS, the greater the

occurrence rate of complicating cardiac disease and the poorer [11] Qiao and Gao Medicine (2016) 95:51 Medicine 4 prognosis; (2) patients with cardiac involvement usually have a history of severe asthma; (3) markedly increased eosinophil count suggests a potential diagnosis of CSS (when the count increases to 20% of white blood cell counts or 8.1109 /L, eosinophils start to infiltrate into myocardium); and (4) negative ANCA status is associated with heart disease in CSS.

Conclusion

In summary, we report a confusing case of asthma, progressive dyspnea, weight loss, and bilateral lower limb ulcers finally diagnosed as CSS with cardiac involvement. Symptoms improved and eosinophil count was sharply reduced with oral steroids. In any patient with refractory asthma, we should not neglect the diagnosis of CSS. Effective treatment methods can improve the prognosis if diagnosed in time.

Consent for publication: Informed consent was taken from the patient to publish this case report in a medical journal.

Ethical consideration: Permission from the ethical committee of the institute was taken.

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11. Qiao and Gao Medicine (2016) 95:51 Medicine 4 prognosis.