A rare occurrence of myocarditis with acute heart failure as an extraintestinal manifestation of Crohn’s disease in a young male

Ferose MFW¹*, Ellepola KD¹, Bowattage S¹

Abstract

Crohn’s disease (CD) is an idiopathic inflammatory bowel disorder, which can be associated with various extraintestinal manifestations. Cardiac and pulmonary manifestations of CD are rare with only a few cases being reported. Myocarditis is an unusual and rare extraintestinal manifestation of CD. Only a few case reports associating CD with the above manifestation are found in the literature. Here, we report a case of an 18-year-old man diagnosed with CD for two years, on optimum treatment, presenting with the symptoms of myocarditis with acute heart failure, as extraintestinal manifestations in the absence of bowel symptoms, while being negative for infective and non infective aetiological screening. This case highlights the importance of knowing the rare extraintestinal manifestations of CD, which will ultimately guide the treating physicians.

Keywords: Crohn’s disease, myocarditis, acute heart failure

Introduction

Crohn’s disease (CD) is one of the major inflammatory bowel diseases, characterised by transmural inflammation, and can involve any portion of the gastrointestinal tract, from the oral cavity to the perianal area. Moreover, it can affect other systems causing extraintestinal manifestations. These manifestations are related to inflammatory disease activity and include musculoskeletal, ocular, dermatological, hepatobiliary, immunologic, haematological, renal, respiratory, and cardiac manifestations.(1) Although, certain extraintestinal manifestations are commonly associated with CD, cardiac and pulmonary involvement are deemed rare and unusual.

Myocarditis, complicated with acute heart failure in the absence of bowel symptoms, is one such rare manifestation of CD and is hardly found in the literature.(2)

Case presentation

An 18-year-old man, diagnosed with CD 2 years back, currently on maintenance therapy with oral azathioprine and tapering doses of oral prednisolone, presented to us with sudden onset shortness of breath at rest with mild bilateral ankle oedema for one-day.

He complained of sudden onset shortness of breath at rest, which had progressively worsened over time and had been associated with orthopnea and paroxysmal nocturnal dyspnœa. There was no
associated chest pain, palpitations, or autonomic symptoms. He denied fever, cough, or other respiratory symptoms.

He had also developed bilateral ankle oedema within the course of one day which had progressively worsened with time. There was no abdominal distention, facial swelling, jaundice, frothy urine, oliguria, or haematuria. He was clinically euthyroid and didn't give any history suggestive of an underlying connective tissue disorder. Interestingly, he didn't have any bowel symptoms. There was also no history of alcohol consumption or use of illicit drugs.

On examination, he was dyspnoeic with a respiratory rate of 28 breaths per minute. His Oxygen saturation on air was 72% and there were bilateral diffuse, fine, end inspiratory crepitations on auscultation. He was tachycardic with a pulse rate of 120 beats per minute and the blood pressure was 140/100 mmHg. The Jugular venous pressure was elevated. There were no murmurs. There was bilateral pitting ankle oedema. Abdominal examination was normal. There were no focal neurological deficits. Bilateral fundi didn't have chronic hypertensive changes or papilloedema. There were no features suggestive of connective tissue disorder.

His full blood count revealed a white blood cell count of 11200 cells/µL with neutrophil predominance. Haemoglobin was 11.6 g/dL and platelet count was 442 000 cells/µL. C reactive protein (CRP) was 38.4 mg/dL(normal 0-5) and erythrocyte sedimentation rate (ESR) was 59 mm/1ʰ hour(normal <20).

The electrocardiogram showed sinus tachycardia with nonspecific tall T waves and ST-T changes in the lateral leads (figure 1). Troponin I was significantly elevated with a level of 580 ng/mL (<0.03). 2D Echocardiogram revealed moderate to severe left ventricular dysfunction with an ejection fraction of 30% and global hypokinesia with mild pulmonary hypertension. Unfortunately, BNP levels were not available. The overall features were suggestive of myocarditis associated with heart failure.

The arterial blood gas analysis showed a PO2/FiO2 ratio of 200 and the chest x-ray showed bilateral diffuse patchy shadows more in the apical region suggestive of heart failure (Figure 2). Computed tomography pulmonary angiogram (CTPA) was performed to exclude pulmonary embolism which eventually revealed bilateral lower lobe patchy areas of consolidation and ground glass opacification compatible with acute respiratory distress syndrome (ARDS) without any evidence of thrombosis. However he did not fulfill the criteria of ARDS by definition.

Other basic investigations such as liver function tests, renal function tests, and serum electrolytes were normal. Screening for dengue, hepatitis B, hepatitis C, influenza A,B virus cytomegalovirus, Epstein Barr virus, and mycoplasma were negative. His thyroid-stimulating hormone level was also normal. Serum antinuclear factor(ANA) was marginally positive(1:80). Anti dsDNA and complement levels were negative.

The patient was initially resuscitated with high-flow oxygen via a facemask. Management for heart failure was commenced with diuretics and angiotensin-converting inhibitors. Beta blockers were introduced when he was much stable. He was ultimately managed for myocarditis with acute heart failure which is considered as an extraintestinal manifestation of CD. His maintenance dose of azathioprine was continued and a course of steroids was reintroduced at a higher dose. The patient was discharged after 12 days of hospital stay and the follow up-2D echocardiogram showed good biventricular function with an ejection fraction of 60%.
Inflammatory bowel disease (IBD) comprises two major disorders: ulcerative colitis (UC) and Crohn’s disease (CD). UC affects the colon and is characterised by inflammation of the mucosal layer. CD is characterised by transmural inflammation and may involve any portion of gastrointestinal tract, from the oral cavity to the perianal area. (3) Extraintestinal manifestations of CD are common and are seen in up to 25-30% of the population. However, myocarditis and acute respiratory distress syndrome (ARDS) are very rare manifestations and are seen in less than 1%. (2)

The cardiac manifestations of CD are pericarditis, pericardial effusion, myocarditis, endocarditis, arrhythmia and conduction anomalies. Crohn’s myocarditis can occur with or without acute flare up of the bowel disease activity and this can be complicated with heart failure, arrhythmia and death. (2,3)

The most common causative aetiologies of myocarditis can be categorised as infectious and non-infectious diseases. Infectious causes account for the majority and include viral (e.g.: hepatitis viruses, cytomegalovirus, human immunodeficiency virus, influenza A and B), bacterial (including tuberculosis), parasitic and fungal pathogens. (3)

The noninfectious causes of myocarditis include systemic diseases such as inflammatory bowel diseases, thyroid disorders, sarcoidosis, hypereosinophilia, rheumatoid arthritis, collagen-vascular diseases, hypersensitivity reactions, cardiotoxins and radiation exposure. (2,3) Interestingly, our patient demonstrated global hypokinesia with an ejection fraction (EF) of 30% in the 2D echocardiogram and negative blood cultures with negative screening for dengue, hepatitis B, hepatitis C, influenza A,B, cytomegalovirus, Epstein Barr virus and Mycoplasma serology.

There have been few documented cases of inflammatory bowel disease-associated myocarditis in America, Australia, and South Korea. However, there are no such reports from South Asia. One American study showed Crohn’s myocarditis involving inferior and septal hypokinesia with an EF of 30%. (3) A Korean study reported a young man with initial presentation of myocarditis associated with CD, with global hypokinesia and an EF of 38%. (1) An Australian study revealed myocarditis with an ejection fraction of 50% and endocarditis. (2)

Management of Crohn’s myocarditis in the absence of bowel symptoms is mainly symptomatic, involving the management of complications such as heart failure, arrhythmias and increasing the dose of steroids. (1-3) Our patient also received high dose of steroids after excluding the infective causes and showed a good clinical improvement.

ARDS is a complex response of the lung to direct and indirect insults. Numerous aetiological factors may cause this acute respiratory distress syndrome such as toxic inhalation, diffuse infection, sepsis, pancreatitis, etc. (4) Among patients with inflammatory bowel disease, up to 30%-50% have demonstrated pulmonary abnormalities according to available data such as bronchiolitis, bronchiectasis, organising pneumonia, alveolitis and acute respiratory distress syndrome. In addition, drug-induced lung injuries have also been reported in IBD patients. (5) However, ARDS occurring as an extraintestinal manifestation of CD is extremely rare.

**Discussion**

Inflammatory bowel disease (IBD) comprises two major disorders: ulcerative colitis (UC) and Crohn’s disease (CD). UC affects the colon and is characterised by inflammation of the mucosal layer. CD is characterised by transmural inflammation and may involve any portion of gastrointestinal tract, from the oral cavity to the perianal area. (3)

Extraintestinal manifestations of CD are common and are seen in up to 25-30% of the population. However, myocarditis and acute respiratory distress syndrome (ARDS) are very rare manifestations and are seen in less than 1%. (2)

The cardiac manifestations of CD are pericarditis, pericardial effusion, myocarditis, endocarditis, arrhythmia and conduction anomalies. Crohn’s myocarditis can occur with or without acute flare up of the bowel disease activity and this can be complicated with heart failure, arrhythmia and death. (2,3)

The most common causative aetiologies of myocarditis can be categorised as infectious and non-infectious diseases. Infectious causes account for the majority and include viral (e.g.: hepatitis viruses, cytomegalovirus, human immunodeficiency virus, influenza A and B), bacterial (including tuberculosis), parasitic and fungal pathogens. (3)

The noninfectious causes of myocarditis include...
Author details
¹National Hospital Kandy, Sri Lanka

References


Received: 27 May 2024 Accepted: 19 Jun 2024