CASE REPORT

A Case of Microscopic Polyangiitis Presenting As Recurrent Myocardial Infarction

ARSALAN NAEEM BUTT, KHURRAM KAZMI, MEHRIN FAROOQ, *MAIMOONA JAVAID*
Department of Medicine, Lahore Medical & Dental College/Ghurki Trust Teaching Hospital, Lahore
Correspondence to Dr. Arsalan Naeed Butt, Assistant Professor Email: arsi_naeem@yahoo.com Cell: 0333-7469379

SUMMARY

A young man presented with chest pain was found to have Myocardial Infarction (MI). Patient already had two episodes of MI in last 3 months. Examination showed findings consistent with Bilateral pleural effusion. Investigations showed hematuria, proteinuia along with deranged renal function tests and segmental wall motion abnormalities along with reduced ejection fraction on coronary angiography. Further investigations revealed positive MPO-ANCA. These features were suggestive of Microscopic polyangiitis, one of the renopulmonary syndrome. Patient showed dramatic improvement on steroid therapy with marked improvement in pleural effusion and renal function tests on follow up.
Keywords: MI, microscopic polyangiitis, pleural effusion

INTRODUCTION

Antineutrophil cytoplasmic antibody (ANCA) associated vasculitis include Wegener's granulomatosis (granulomatosis with polyangiitis), microscopic polyangiitis (MPA), the Churg-Strauss syndrome (CSS) and renal limited vasculitis[1,2] Microscopic polyangiitis is mostly seen in older adults, although it has been reported at all ages[3,4]. Patients typically present with constitutional symptoms including fever, malaise, anorexia, migratory arthralgias, and weight loss[3,5,6]. Patients with MPA may present with involvement of airways or pulmonary parenchyma causing hoarseness of voice, cough, dyspnea, wheezing, hemoptysis or pleuritic pain. These symptoms may be accompanied by signs of tracheal or subglottic stenosis, pulmonary consolidation and/or pleural effusion. Renal involvement is common in MPA[4,6,7]. The manifestations of the glomerulonephritis are similar to those in other causes of glomerulonephritis including i) asymptomatic hematuria with normal renal function. ii) Acute kidney injury with hematuria & cellular casts. iii) proteinuria of variable degree that is usually subnephrotic. iv) Rapidly progressive glomerulonephritis is common in this group of diseases

CASE REPORT

A 26 years old male presented to medical department with the complaint of recurrent chest pain for last 3 months. On presentation, he had retrosternal chest pain for last few hours which was moderate in intensity, aggravated on exertion and relieved partially by rest and sublingual nitrates. Patient had orthopnea, paroxysmal nocturnal dyspnea and NYHA grade II dyspnea but no history of cough, chest wheezing, decrease in urine output, blood in urine, froathing of urine, epistaxis, nasal crusting, pain in legs, paleness of complexion, tremors of hands or hot intolerance. Patient lost 15 kg weight in last 7 months, but no history of Tuberculosis or close contact with someone known to have tuberculosis. Patient denied joint pains, oral ulcers, photosensitivity, body rash or lumps in the body. Patient was admitted twice in a hospital in last 3 months and was found to have coronary artery disease. On examination, blood pressure was 100/70 mmHg, pulse 114/ minute, Respiratory rate 22 breaths/minute, temperature was within normal range, third heart sound was audible. Rest of the examination was within normal limits. Investigations revealed raised serum creatinine (2.9 mg/dl), hematuria (4+) and raised serum troponin I levels (0.59 ng/ml against reference range 0-0.1). ECG showed inverted T Waves in lead II,III and avf and Q waves in lead V2,V3 and V4. Echocardiography showed hypokinesia of distal interventricular septum, adjoining apex and lateral wall on segmental wall motion analysis with ejection fraction of 34%. Previous coronary angiography showed moderate stenosis in left anterior descending artery, anterior wall hypokinesia with 35% ejection fraction. Patient was managed conservatively and investigated for work up of recurrent Myocardial infarction at this young age. After 7 days of improvement in chest pain, patient started complaining of shortness of breath which was insidious in onset, aggravated on minimal exertion and on lying supine, associated with dry cough, but no history of new onset chest pain, wheezing or fever was reported. On examination, there was dull percussion note, decreased intensity of breath sounds and decreased vocal resonance in both infrascapular areas suggestive of bilateral pleural effusion. No new changes were present on ECG. Blood samples were sent to rule out new episode of MI which showed negative CK MB and troponin I. on the basis of this recent onset pleural effusion in the absence of new cardiac insult, patient was given prednisolone 60 mg/day. Investigations previously ordered revealed positive P-ANCA and negative renal angiogram. USG abdomen did not show any renal infarct. HBsAg was positive and PCR showed HBV Viral load of around 11,000 IU for which entacavir was started. Patient refused renal biopsy. Patient responded dramatically to steroids, shortness of breath and pleural effusion settled within 48 hrs of starting steroid therapy. Patient was labelled as a case of Microscopic polyangiitis and discharged on steroids, followed after 15 days with improvement in Shortness of breath & normalization of renal function tests.

Received on 12-12-2018
Accepted on 19-02-2019

DISCUSSION

The American College of Rheumatology (ACR) criteria was aimed at defining granulomatosis with polyangiitis before the broader separation of microscopic polyangiitis (MPA) from GPA[6]. The ACR proposed the following clinical criteria to distinguish patients of suspected wegners granulomatosis/MPA from those with other forms of vasculitis[8].

- Nasal or oral inflammation
- Abnormal chest radiograph showing nodules, fixed infiltrates, or cavities
- Abnormal urinary sediment (microscopic hematuria alone / hematuria + RBC casts)
- Granulomatous inflammation of an artery or perivascular area, seen on biopsy.

The presence of two or more of these four criteria showed a specificity of 92% and a sensitivity of 88% [8]. Since the ACR did not establish specific criteria for MPA, these criteria did **not** discriminate GPA from either MPA or non-vasculitic diseases that can mimic wegner's granulomatosis. Ear, nose, and throat (ENT) manifestations occur in patients with either GPA or MPA. However, they are much more common in patients with GPA (estimated frequency 90 percent versus 35 percent in MPA)[3,4,6].

European Medicines Agency (EMA) and Chapel Hill Consensus Conference (CHCC) criteria definitions for GPA[9]. included ANCA as one of the diagnostic criteria. A positive ANCA in a patient with surrogate markers for GPA allowed a diagnosis of GPA without a biopsy[9].

Approximately 70 percent of patients with microscopic polyangiitis (MPA) are ANCA positive[10]. In contrast to GPA, most ANCA-positive MPA patients have MPO-ANCA, with a few of them having PR3-ANCA. ANCA serologies can be used to distinguish Microscopic polyangiitis from classic polyarteritis nodosa (PAN). Classic PAN is not associated with antibodies to either PR3 or MPO, whereas three-fourths of patients with MPA are ANCA-positive. MPA may be distinguished from classic polyarteritis nodosa by the clinical presentation and ANCA. The cardinal features of classic polyarteritis nodosa include renal infarcts, renal artery stenosis, and visceral microaneurysms, which are not observed in MPA. Glomerulonephritis is observed in MPA but **not** in classic PAN[11].

REFERENCES

1. Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. Arthritis Rheum 1994; 37:187.
2. Hogan SL, Falk RJ, Chin H, et al. Predictors of relapse and treatment resistance in antineutrophil cytoplasmic antibody-associated small-vessel vasculitis. Ann Intern Med 2005; 143:621.
3. Jennette JC, Falk RJ. Small-vessel vasculitis. N Engl J Med 1997; 337:1512.
4. Seo P, Stone JH. The antineutrophil cytoplasmic antibody-associated vasculitides. Am J Med 2004; 117:39.
5. Falk RJ, Hogan S, Carey TS, Jennette JC. Clinical course of anti-neutrophil cytoplasmic autoantibody-associated glomerulonephritis and systemic vasculitis. The Glomerular Disease Collaborative Network. Ann Intern Med 1990; 113:656.
6. Hoffman GS, Kerr GS, Leavitt RY, et al. Wegener granulomatosis: an analysis of 158 patients. Ann Intern Med 1992; 116:488.
7. Fauci AS, Haynes BF, Katz P, Wolff SM. Wegener's granulomatosis: prospective clinical and therapeutic experience with 85 patients for 21 years. Ann Intern Med 1983; 98:76.
8. Leavitt RY, Fauci AS, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Wegener's granulomatosis. Arthritis Rheum 1990; 33:1101.
9. Watts R, Lane S, Hanslik T, et al. Development and validation of a consensus methodology for the classification of the ANCA-associated vasculitides and polyarteritisnodosa for epidemiological studies. Ann Rheum Dis 2007; 66:222.
10. Guillevin L, Durand-Gasselin B, Cevallos R, et al. Microscopic polyangiitis: clinical and laboratory findings in eighty-five patients. Arthritis Rheum 1999; 42:421.
11. Tervaert JW, Goldschmeding R, Elema JD, et al. Autoantibodies against myeloid lysosomal enzymes in crescentic glomerulonephritis. Kidney Int 1990; 37:799.