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Original Research Paper

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A RARE CASE OF CRITICAL CORONARY SYNDROME WITH A SUSPICIOUS VASCULITIS

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ABSTRACT Eosinophilic granulomatosis with polyangiitis (formerly called Churg-Strauss syndrome) is a syndrome characterized by episodes of asthma, rhinosinusitis and peripheral eosinophilia in the blood. It is a disease of the small and medium sized vessels causing necrotizing vasculitis and peripheral eosinophilia with eosinophilic infiltration in various organs resulting in granuloma formation. Organs commonly involved include the lung and skin but symptoms can include kidney involvement resulting in rapidly progressive glomerulonephritis and mononeuritis multiplex. Cardiac involvement, particularly of the coronary artery can occur and is associated with higher mortality particularly in the elderly. Here we report an unusual presentation of this disease as a case of acute coronary syndrome with significant right ventricular strain pattern on ECG and echocardiogram.

KEYWORDS : Eosinophilic granulomatosis with polyangiitis,Churg-Strauss syndrome, Rhinosinusitis and peripheral eosinophilia, Glomerulonephritis, Mononeuritis

INTRODUCTION

Myocarditis due to involvement of the coronary micro vasculature, stenosis, aneurysm, or vascular thrombosis are the common manifestations.(1)

EGPA is defined as an eosinophil-rich and granulomatous inflammation, formerly known as "Churg–Strauss syndrome." (2)

American College of Rheumatology classification criteria and the former CHCC, the CHCC 2012 has reported for the first time that ANCA are found in EGPA, especially in patients with glomerulonephritis.(3)

Eosinophilic granulomatosis with polyangiitis incidence in Europe is 0.5–6.8 new cases/year per million populations, whereas its prevalence is 10.7–13 cases per million populations. (4)

Eosinophilic granulomatosis with polyangiitis is an HLA-associated disease (5).

It has been proven that it is associated with *HLA-DRB1*04* and **07* and with *HLA-DRB4* (6,7).

It has been also investigated the presence of single nucleotide polymorphisms (SNP) of the gene, which encodes interleukin (IL)-10, an important molecule for the activation of the Th-2 pathway; EGPA ANCA-negative subset has been associated with the IL10.2 haplotype of the IL-10 promoter gene, a condition, which leads to an increased production of IL-10 (8)

Cardiac vasculitis is recognized as a heterogeneous disease process with a wide spectrum of manifestations including pericarditis, myocarditis, valvular heart disease and less frequently, coronary artery vasculitis (CAV). (9)

Cardiac manifestations in the primary systemic vasculitides occur with variable frequency and can affect any of the cardiac layers.(10)

Case presentation

A 49 year old of male presented to our hospital with complaints of shortness of breath accompanied by three episodes of mild hemoptysis in the past 24 hours. He had also been experiencing paresthesia in his left leg and complained of a sensation of tingling and burning in both his legs for the past 15 days. On examination he was tachycardic, tachypneic and had a petechial rash present bilaterally on his lower limbs.

Peripheral blood work was normal except for eosinophilia of 29.2%. Pulmonary edema was initially suspected as the cause of his breathlessness and an ECG was done to rule out acute coronary syndrome. It showed a characteristic S1Q3T3 pattern with partial right bundle branch block and T wave inversions in V1,V2 and III [Fig 1]. D dimer was elevated. CT pulmonary angiography was done which was normal, ruling out pulmonary thromboembolism [Fig 2].

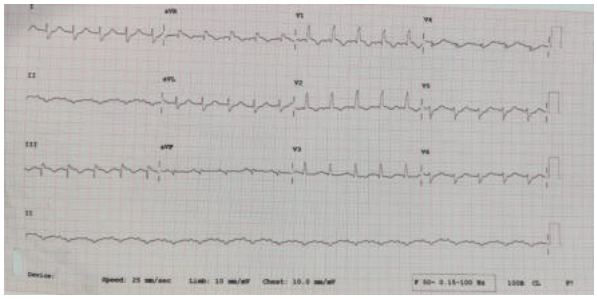
X ray showed bilateral peripheral wooly opacities unlike the classic butterfly pattern seen in pulmonary edema [Fig 3]. Echocardiogram was done which revealed right ventricular motion abnormality and a slightly reduced ejection fraction at 40%. Non-ST elevation myocardial infarction involving the right ventricle was suspected and later supported by elevated troponin I levels. He was started on dual antiplatelets immediately for the acute coronary syndrome.

On day 2, his hemoptysis increased and a repeat X ray revealed findings suggestive of diffuse alveolar hemorrhage [Fig 4]. A combination of acute coronary syndrome, hemoptysis with evidence of diffuse alveolar hemorrhage, recent onset of neuropathic pain in the legs, rash and eosinophilia in the blood prompted us to suspect Eosinophilic granulomatosis with polyangiitis.

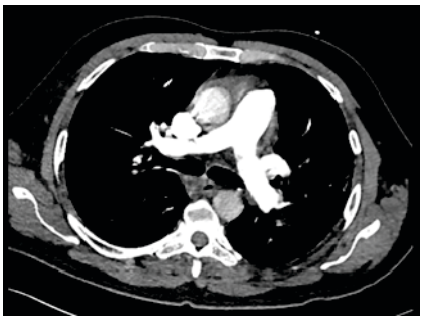
A nerve conduction study revealed motor axonal polyradiculopathy and p-ANCA was positive at 3+ [Fig 5]. MPO ELISA was positive further pointing towards a systemic vasculitis, possibly Eosinophilic granulomatosis with polyangiitis.

He was started on pulse therapy of Methylprednisolone along with Mycophenolate. His symptoms improved on day 5 and was shifted out of the ICU. He was discharged on day 8 and has been on follow up since recurrences can occur.

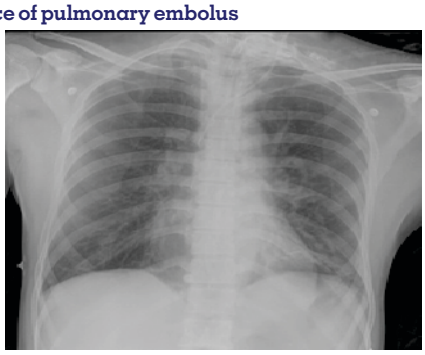
Images



Figure—1--ECG-Done at presentation



Figure—2--CT pulmonary angiography showing no evidence of pulmonary embolus



Figure—3--X-ray done at presentation

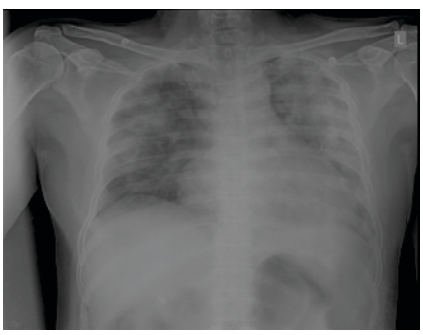


Fig 4: X ray done on day 2 showing diffuse alveolar hemorrhage

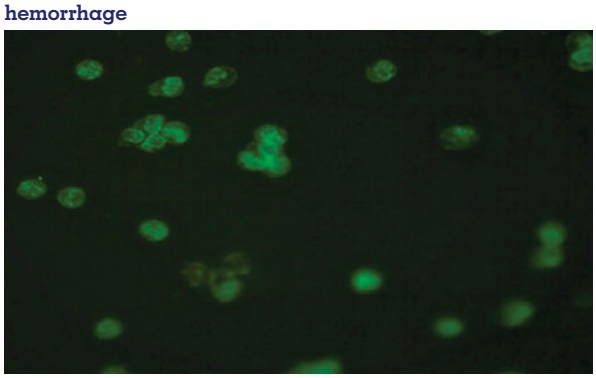


Fig 5: Image showing p-ANCA positivity

DISCUSSION

Eosinophilic granulomatosis with polyangiitis is multisystemic immune mediated vasculitis which in rare cases can cause serious complications including coronary artery involvement presenting as acute coronary syndrome or renal involvement with rapidly progressing glomerulonephritis. These are associated with worse outcomes. It is clinically divided into three stages, asthma and rhinosinusitis stage, eosinophilia stage, vasculitis phase. Presence of polyradiculopathy, coronary artery involvement points towards progression into the third stage of the disease. Cardiac involvement itself is a huge contributor to mortality in this disease. Approximately 50% of deaths in this vasculitis can be attributed to cardiac involvement (11)

Early involvement though difficult to detect, is very important. Treatment in the early stages of cardiac involvement is said to halt progression and reduce overall mortality (12)

A study done on 49 patients with Eosinophilic granulomatosis with polyangiitis revealed clinical cardiac involvement in 22 of them[3]. In patients with cardiac involvement, coronary vasculitis presenting as acute coronary syndrome is rare, as cardiomyopathy and pericardial effusion were the more common presentations (13)

Another important finding in our case was the MPO-ANCA positivity. This is important because previous case reports and studies have indicated a positive correlation of ANCA positivity with renal disease and a negative correlation with cardiac involvement (14)

Myopericarditis if present can cause chronic damage to the heart with replacement of cardiac tissue with fibrosis resulting in decline in cardiac function over time contributing significantly to cardiac morbidity in this disease. This further stresses on the importance of early detection of cardiac involvement. Current treatment guidelines highly recommend use of cyclophosphamide or rituximab in combination with systemic steroids for severe disease[5]. Cyclophosphamide is preferred in patients with cardiac involvement while rituximab is preferred in ANCA positive patients for achievement of remission (15)

Summary

Eosinophilic granulomatosis with polyangiitis is a disease of the small and medium sized vessels causing necrotizing vasculitis and peripheral eosinophilia with eosinophilic infiltration in various organs resulting in granuloma formation. Cardiac involvement particularly of the coronary artery is rare and is associated with higher mortality. Early detection is difficult but if done can lead to early initiation of treatment with steroids, which has shown to halt progression of the disease process and reduce mortality. In cases like ours where coronary artery is involved, they can present as acute coronary syndrome. We want to emphasize the need to keep vasculitis as a strong differential in patients presenting with acute coronary syndrome, particularly younger or middle aged patients with no risk factors for atherosclerosis and present with other symptoms suggestive of multisystemic involvement.

CONCLUSION

Through this case report we want to emphasize the importance of having an open approach to patients presenting with acute coronary syndrome. We want to highlight that this is particularly important in younger and middle-aged patients without significant risk factors for atherosclerotic disease like diabetes, hypertension, family history of early vascular events and smoking. Vasculitis though a rare cause of acute coronary syndrome must always be considered particularly in patients presenting with

complaints pointing towards multisystemic involvement or past history suggesting the same. We also want to point out the significant mortality benefit of early detection and appropriate treatment, particularly in cases with cardiac involvement.

Contribution of the author

Research concept- Dr.Vamsi Krishna Kaza
Research design- Dr.Vamsi Krishna Kaza
Data analysis and Interpretation- Dr.Vamsi Krishna Kaza
Materials-- Dr.Vamsi Krishna Kaza
Data collection- Dr.Vamsikrishna Kaza
Literature search- Raghavendra Rao,M.V
Critical review- Raghavendra Rao M.V
Supervision- Dr.K.Swarna Deepak
Article editing- Dr.K.Swarna Deepak
Final approval- Dr.K.Swarna Deeepak

REFERENCES

1. Andrea Gioffredi,Federica M,Elena Oliva and Carlo uzio,Eosinophilic granulomatosis with polyangiitis: an overview,Front. Immunol., 03 November 2014,Sec. Inflammation,Volume 5 - 2014 |
2. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 Revised international Chapel Hill consensus conference nomenclature of vasculitides. *Arthritis Rheum* (2013) **65**(1):1–11. doi: 10.1002/art.37715 CrossRef Full Text | Google Scholar
3. Vaglio A, Moosig F, Zwerina J. Churg-Strauss syndrome: update on pathophysiology and treatment. *Curr Opin Rheumatol* (2012) **24**(1):24–30. doi:10.1097/BOR.0b013e32834d85ce Pubmed Abstract | Pubmed Full Text | CrossRef Full Text | Google Scholar
4. Mouthon L, Dunogue B, Guillevin L. Diagnosis and classification of eosinophilic granulomatosis with polyangiitis (formerly named Churg-Strauss syndrome). *J Autoimmun* (2014) **48-49**:99–103. doi:10.1016/j.jaut.2014.01.018
5. Alberici F, Martorana D, Bonatti F, Gioffredi A, Lyons PA, Vaglio A. Genetics of ANCA-associated vasculitides: HLA and beyond. *Clin Exp Rheumatol* (2014) **32**(3 Suppl 82):90–7. Pubmed Abstract | Pubmed Full Text | Google Scholar
6. Wieczorek S, Hellmich B, Gross WL, Epplen JT. Associations of Churg-Strauss syndrome with the HLA-DRB1 locus, and relationship to the genetics of antineutrophil cytoplasmic antibody-associated vasculitides: comment on the article by Vaglio et al. *Arthritis Rheum* (2008) **58**(1):329–30. doi:10.1002/art.23209 CrossRef Full Text | Google Scholar
7. Vaglio A, Martorana D, Maggiore U, Grasselli C, Zanetti A, Pesci A, et al. HLA-DRB4 as a genetic risk factor for Churg-Strauss syndrome. *Arthritis Rheum* (2007) **56**(9):3159–66. doi:10.1002/art.22834 Pubmed Abstract | Pubmed Full Text | CrossRef Full Text | Google Scholar
8. Wieczorek S, Hellmich B, Arning L, Moosig F, Lamprecht P, Gross WL, et al. Functionally relevant variations of the interleukin-10 gene associated with antineutrophil cytoplasmic antibody-negative Churg-Strauss syndrome, but not with Wegener's granulomatosis. *Arthritis Rheum* (2008) **58**(6):1839–48. doi:10.1002/art.23496 Pubmed Abstract | Pubmed Full Text | CrossRef Full Text | Google Scholar
9. Shaun Khanna, Kartheek Garikapati, Daniel S. L. Goh, Kenneth Ch,Phillip Lo, Mohan V. Bhojaraja & Surjit Tarafdar, Coronary artery vasculitis: a review of current literature,*BMC Cardiovascular Disorders* ,**volume 21**, Article number: 7 (2021)
10. Miloslavsky E, Unizony S. The heart in vasculitis. *Rheum Dis Clin North Am* 2014;40:11-26.
11. Lanham JG, Elkon KB, Pusey CD, Hughes GR. Systemic vasculitis with asthma and eosinophilia: a clinical approach to the Churg-Strauss syndrome. Medicine (Baltimore). 1984 Mar;63(2):65-81. doi: 10.1097/00005792-198403000-00001. PMID: 6366453.
12. Guillevin L, Cohen P, Gayraud M, Lhote F, Jarrousse B, Casassus P. Churg-Strauss syndrome. Clinical study and long-term follow-up of 96 patients. Medicine (Baltimore). 1999 Jan;78(1):26-37. doi: 10.1097/00005792-199901000-00003. PMID: 9990352.
13. Neumann T, Manger B, Schmid M, Kroegel C, Hansch A, Kaiser WA, Reinhardt D, Wolf G, Hein G, Mall G, Schett G, Zwerina J. Cardiac involvement in Churg-Strauss syndrome: impact of endomyocarditis. Medicine (Baltimore). 2009 Jul;88(4):236-243. doi: 10.1097/MD.0b013e3181af35a5. PMID: 19593229.
14. Sinico RA, Di Toma L, Maggiore U, Bottero P, Radice A, Tosoni C, Grasselli C, Pavone L, Gregorini G, Monti S, Frassi M, Vecchio F, Corace C, Venegoni E, Buzio C. Prevalence and clinical significance of antineutrophil cytoplasmic antibodies in Churg-Strauss syndrome. Arthritis Rheum. 2005 Sep;52(9):2926-35. doi: 10.1002/art.21250. PMID: 16142760.
15. Chung SA, Langford CA, Maz M, Abril A, Gorelik M, Guyatt G, Archer AM, Conn DL, Full KA, Grayson PC, Ibarra MF, Imundo LF, Kim S, Merkel PA, Rhee RL, Seo P, Stone JH, Sule S, Sundel RP, Vitobaldi OI, Warner A, Byram K, Dua AB, Husainat N, James KE, Kalot MA, Lin YC, Springer JM, Turgunbaev M, Villa-Forte A, Turner AS, Mustafa RA. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis. Arthritis Care Res (Hoboken). 2021 Aug;73(8):1088-1105. doi: 10.1002/acr.24634. Epub 2021 Jul 8. PMID: 34235880.