A RARE CASE REPORT OF GRANULOMATOSIS WITH POLYANGIITIS PRESENTING WITH THROMBUS OF THE ASCENDING AORTA

DR IOANNIS TZANNINIS, DR AHMED ELSERWEY, DR MOHAMMED ELREFAI, DR ALICE MASON, DR JOHN RAWLINS

Introduction

Granulomatosis with polyangiitis (GPA) is a rare multisystem autoimmune ANCA positive vasculitis which in rare cases can affect the aorta causing aortitis, aneurysm and rupture.

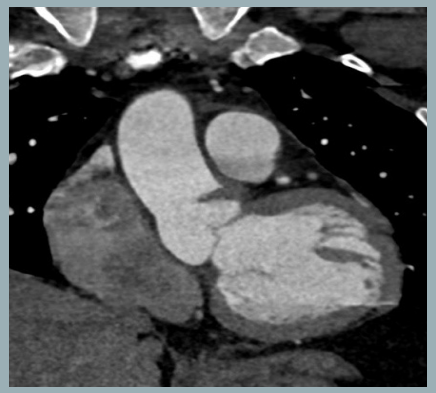
Case presentation

A 50-year old male presented with acute onset central chest pain, vomiting and haemoptysis preceded by arthritis of the right hand, ankle and knees. He was otherwise fit and well, non-smoker and without comorbidities.

Examination showed episcleritis and migratory arthritis without a rash. Electrocardiogram was consistent with atrial fibrillation and serial troponins had a rising trend (42 – 376 – 1300 ng/L). Repeated blood cultures were negative, but a vasculitis screen was positive for Proteinase-3.

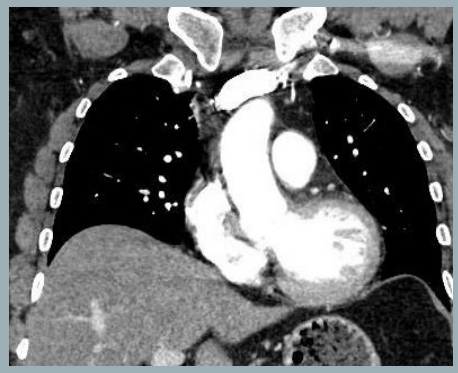
An initial high-resolution CT on admission was suggestive of right sided pulmonary haemorrhage.

A subsequent CT aorta 3 days later showed a new low-density filling defect in the aortic sinus, arising from the left/right coronary commissure and associated with minor thickening of the valve leaflets on that side. This was confirmed on an echocardiogram which showed no evidence of vegetation.



The impression was that the mass represented a thrombus rather than an infective vegetation considering its position and time of appearance. The patient was commenced on IV heparin infusion and IV methylprednisolone. Repeat CT aorta 3 days later showed reduction in volume of the low attenuation material. Overall, the appearances favoured thrombus which may had formed at a point of intimal inflammation or injury secondary to an underlying aortitis.

The patient was then switched to Warfarin and was discharged a reducing regimen of Prednisolone. He also received a 6-dose course of Cyclophosphamide as outpatient. Repeat CT scan 10 weeks later showed complete resolution of the lesion.



Discussion

GPA is a small vessel vasculitis mostly associated with PR-3 antibodies. In one fifth of cases MPO antibodies can also be present. PR-3 antibody positivity provides aid to the diagnosis and is also a marker of disease activity [1]. The disease is characterized by granulomatous inflammation and necrosis of small and medium vessels and most commonly affects the ears, nose and throat, the respiratory tract and the kidneys [2]. Cardiac manifestations of GPA are rare and can include pericarditis, cardiomyopathy, coronary arteritis, valvular lesions and conduction abnormalities [3]. Large vessels may also be affected, despite it being classified as a small vessel vasculitis. Specifically for the aorta, which was affected in our case, this can present with aneurysmal formation, dissection, rupture, regurgitation, or death. In those cases, a possible overlap between ANCA-associated vasculitis and large vessel vasculitis has been suggested [4].

Conclusion

We have presented a rare case of GPA presenting with thrombus of the ascending aorta that resolved following effective anticoagulation and immunosuppressive treatment.

References

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