

Peer Review File

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Reviewer A

This manuscript described GPA with cardiac involvement. However, I have identified several issues which I think should be addressed in the manuscript.

Comment 1: Authors describes coronary dilation as cardiac involvement caused by vasculitis. First, detail lesions should be clearly presented by cardiac echography, CT, MRI. And they should confirm association with vascular inflammation.

Reply 1: We have included the echocardiogram; however, we do not have CT angiography or MRI. In the text we include our cardiologist thoughts on relation to vascular inflammation in lines 153-160.

Changes in the text: Lines 153-160 Due to technical difficulty of EBUS, the pulmonologist was unable to obtain a lung biopsy, a recognized diagnostic gold standard, thus further complicating diagnosis. With ongoing rises in Troponin-T and BNP cardiology suspected coronary dilation attributable to vasculitis changes, though uncommon, due to rising suspicion on our differential for GPA and no other current explainable cause for ongoing dilation with repeat echocardiography revealing the right coronary artery size increasing to 4.4mm3.9mm and recommended daily aspirin therapy with plans for outpatient follow-up.

Comment 2: Was biopsy performed to detect necrotizing granulomatous lesions?

Reply 2: A sinus biopsy was unrevealing aside from necrosis, EBUS was technically difficult and lung biopsy was not able to be made.

Changes in the text lines 153-160

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Comment 3: Chest CT revealed cavity lesions typically found in GPA; however, Was it as considered as one of differential diagnosis?

Reply 3: Yes, this was in the differential, included with this is the figure for echocardiogram.

Changes in the text lines 127-130 with initial high suspicion for vasculitis vs a fungal

infection. Troponin T and BNP were both elevated prompting an echocardiogram which showed right coronary dilation Figure 2A-B with noted right coronary dilation measuring 3.9mm described as ectatic and diffusely dilated

Comment 4: Authors describe literature review, but detail method and the results are unclear. Is this article the first case to present coronary dilation as cardiac involvement in patients with GPA?

Reply 1: This is the first case to demonstrate coronary dilation, specifically in a pediatric patient, and significant coronary involvement as a cardiac manifestation. We did find and include a case report of a cardiac mass; however, no other pubmed cases of coronary dilation in GPA pediatric patients were found.

Changes in the Text line 179-182: In a thorough literature review via PubMed I was unable to find any specific cases regarding coronary dilation; however, we did find an interesting case of a cardiac mass in a 14-year-old woman with GPA (3) with most demonstrating variable cardiac involvement.

Reviewer B

Comment 1: this interesting report, the authors described a 15-year-old girl with granulomatosis with polyangiitis (GPA), presenting as sinusitis, pulmonary cavitory lesions and microscopic hematuria with the presence of ANCA-positive anti-PR3. In particular, there was a presentation of right coronary dilation by the echocardiography examination. Finally, this case was responsive to combined glucocorticoids/cyclophosphamide induction therapy.

Indeed, in GPA, pediatric-onset cases are responsible for less than 5% of patients, while they have rare cardiovascular involvement. Although it is worthy of publication, there is one issue needed to be clarified as follows.

The authors showed the typical chest x-ray/CT with pulmonary cavitations and sinus CT with marked sinusitis. Nevertheless, there were no available echocardiographic figures of dilated right coronary artery in their reported patient. Since they used “A challenging case of GPA with cardiac involvement” as their manuscript title, characteristic photographs to demonstrate the cardiac abnormalities are indispensable for this case report.

Reply 1: We have included the imaging

Changes in the text lines 127-130 with initial high suspicion for vasculitis vs a fungal infection. Troponin T and BNP were both elevated prompting an echocardiogram which showed right coronary dilation Figure 2A-B with noted right coronary dilation measuring 3.9mm described as ectatic and diffusely dilated