Atypical Presentation of Intimomedial Mucoid Degeneration: Ascending Aortic Aneurysm and Dissection in a Young White Man Mimicking Multifocal Pneumonia

Solomon Ocran, DO; Mukul Sharda, BS; Charles Haynes, MD; Pinky Jha, MD, MPH

ABSTRACT

Introduction: Intimomedial mucoid degeneration (IMMD) is a rare noninflammatory arteriopathy classically affecting young Black African women with hypertension.

Case Presentation: A 34-year-old White man with chronic tobacco use presented with progressive dyspnea initially attributed to multifocal pneumonia and pulmonary edema. Computed tomography revealed bilateral pulmonary infiltrates and a 6.2-cm ascending aortic aneurysm with a pericardial effusion. Echocardiography demonstrated severe aortic regurgitation. During hospitalization, he developed chest pain and asymmetric pulses; computed tomography angiography confirmed an acute arch dissection. Emergent valve-sparing aortic root replacement, total arch repair, and descending thoracic stent graft placement were performed. Histopathologic examination confirmed IMMD.

Discussion: This case expands the demographic profile of IMMD and illustrates its potential to mimic pulmonary infection, delaying recognition of underlying cardiovascular disease. The recent COVID-19 infection in this patient raises the theoretical possibility that viral-induced endothelial injury contributed to aortic wall degeneration, although this association remains speculative and warrants further investigation.

Conclusions: Clinicians should consider structural cardiovascular disease in patients with unexplained dyspnea or presumed pneumonia unresponsive to antibiotic therapy, particularly when cardiac murmurs are present or imaging reveals mediastinal abnormalities. Early recognition, prompt surgical intervention, and histopathologic confirmation are essential for favorable outcomes in patients with IMMD.

INTRODUCTION

Intimomedial mucoid degeneration (IMMD) is a rare, noninflammatory degenerative arteriopathy characterized by pathological accumulation of mucopolysaccharides within the intima and media, resulting in fragmentation of elastic fibers and weakening of the aortic wall.[1,2] IMMD is classically described in young to middle-aged Black African women with hypertension and most often involves the aorta and its major branches, predisposing affected patients to aneurysm formation, dissection, and valvular dysfunction.[1,3,4] Extra-aortic involvement, including the coronary, brachial, renal, and peripheral arteries, has been reported but remains uncommon.[2,5]

Diagnosis is most often established histologically, either at autopsy or from surgical specimens, and reveals mucin pools in the intima and media, elastic fiber disruption, and smooth muscle cell loss.[2,6] Preoperative imaging, such as transthoracic echocardiography (TTE), computed tomography (CT), and computed tomography angiography (CTA), can detect aneurysms, dissections, and valvular regurgitation but cannot confirm IMMD without tissue analysis.[4]

We describe an atypical case of an ascending aortic aneurysm with dissection and severe aortic regurgitation due to IMMD in a young White male who initially presented with presumed multifocal pneumonia and pulmonary edema shortly after COVID-19 infection. This case expands the demographic profile of IMMD and highlights its potential to mimic infectious pulmonary disease, leading to delayed recognition of underlying structural cardiovascular pathology.

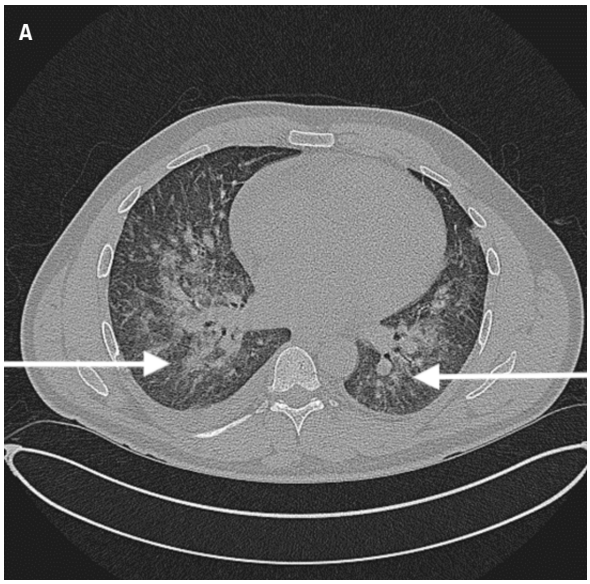
CASE PRESENTATION

A 34-year-old White man with a history of chronic tobacco use, attention-deficit/hyperactivity disorder, anxiety, and depression presented to our institution after transfer from an outside hospital with worsening shortness of breath initially attributed to multi-

Author affiliations: Medical College of Wisconsin, Milwaukee, Wisconsin (Ocran, Sharda, Haynes, Jha).

Corresponding author: Mukul Sharda, 9150 W Watertown Plank Rd, Milwaukee, WI 53226; email msharda@mcw.edu; ORCID ID 0009-0008-7711-386X

Figure 1. Computed Tomography of the Patient's Chest Demonstrating (A) Bilateral Pulmonary Infiltrates (arrows) Initially Interpreted as Multifocal Pneumonia and (B) Marked Dilation of the Ascending Aorta Consistent With Aneurysm (arrow)



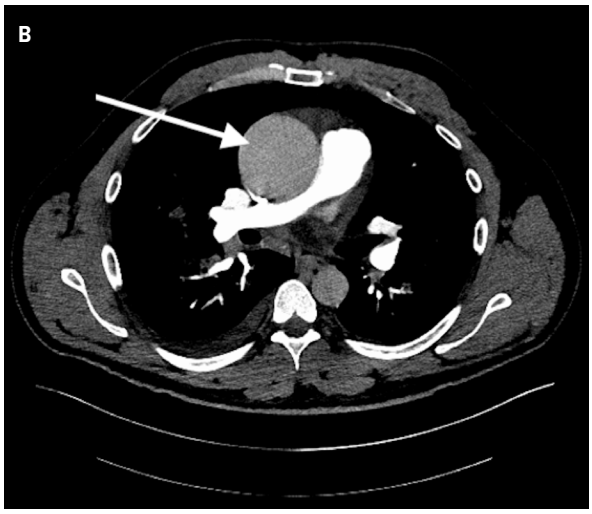
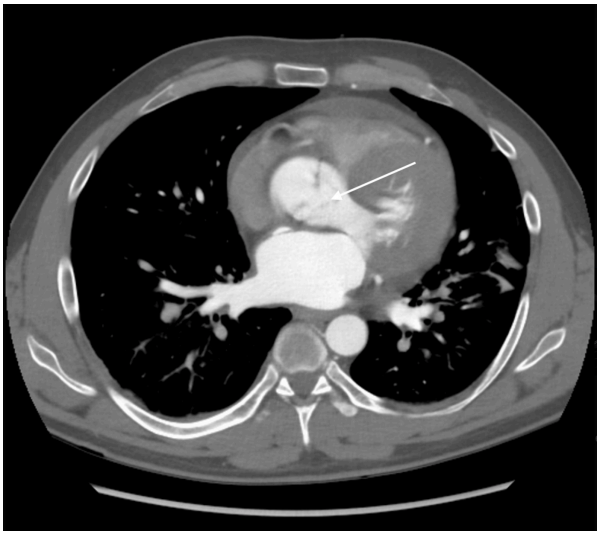


Figure 2. Computed Tomography Angiography Upon Admission Demonstrating an Intimal Flap Within the Aortic Arch Consistent With Acute Dissection (arrow)



focal pneumonia following a recent COVID-19 infection. Two weeks prior, he had been diagnosed with COVID-19 and subsequently developed suspected bacterial pneumonia, for which he was treated with doxycycline without symptom resolution.

At the outside facility, he reported pleuritic chest pain, orthopnea, cough, subjective fever and chills, and was hypoxemic. CT imaging revealed bilateral multifocal pneumonia and incidentally identified a 6.2-cm ascending aortic aneurysm with a small-to-moderate pericardial effusion (Figure 1). CT angiography ruled out pulmonary embolism. He was transferred to our tertiary center for further management and surgical evaluation.

Upon arrival, he was afebrile but required 4 L/min of supplemental oxygen via nasal cannula. Vital signs were notable for mild tachycardia (heart rate, 116 beats/min), oxygen saturation greater than 90%, and variable blood pressures. Physical examination revealed a loud diastolic murmur and bibasilar rhonchi; no signs of volume overload or Marfanoid features were noted. An extensive infectious workup, including testing for histoplasma, blastomycosis, syphilis, HIV, *Legionella*, *Streptococcus pneumoniae*, methicillin-resistant *Staphylococcus aureus* (MRSA), and respiratory viruses, was unrevealing. Empiric antibiotic therapy was narrowed to ceftriaxone on the recommendation of the infectious disease service.

Transthoracic echocardiogram revealed a left ventricular ejection fraction of 59%, a severely dilated ascending aorta, and severe aortic regurgitation. The N-terminal pro-B-type natriuretic peptide was markedly elevated at 8662 pg/mL, raising concern for acute decompensated heart failure. Given the bilateral infiltrates and elevated natriuretic peptide level, both pneumonia and pulmonary edema secondary to severe aortic regurgitation were considered. Empiric management for both conditions was initiated while awaiting further cardiac evaluation. Cardiology assumed primary management with plans for further cardiac evaluation, including left heart catheterization and cardiac magnetic resonance imaging.

On hospital day 6, the patient developed acute chest pain and asymmetric upper-limb pulses, with a diminished right radial pulse and a significant blood pressure differential between arms. Emergent CTA revealed a new dissection flap in the aortic arch (Figure 2), and he underwent emergent sternotomy with valve-sparing aortic root replacement (VSARR), total arch repair, and descending thoracic stent graft placement.

The postoperative course was notable for persistent pulmonary infiltrates and left lower lobe collapse, attributed to postopera-

tive atelectasis and residual inflammation, which improved with bilevel positive airway pressure support. He was gradually weaned to room air and discharged home in stable condition on postoperative day 12 with close follow-up arranged through adult congenital cardiology. Surgical pathology revealed focal myxoid (mucoid) degeneration within the intimo-medial layer of the aortic wall, consistent with IMMD.

DISCUSSION

IMMD is an uncommon cause of aortic aneurysm and dissection, with fewer than 100 cases described in the literature.[1,2] Its hallmark histopathologic feature is the accumulation of acid mucopolysaccharides within both the intima and media, accompanied by disruption of elastic lamellae and loss of smooth muscle cells.[2,6] This differs from cystic medial necrosis, which is confined to the media, and from cystic adventitial disease, which involves only the adventitia.[6]

While epidemiologic data suggest a predilection for young Black African women with hypertension,[1,3] cases have been reported in men, White individuals, and patients without hypertension.[4,5] The present case illustrates this atypical demographic profile. Clinically, IMMD can present with acute aortic syndromes, including aneurysm, dissection, and rupture, or with complications from valvular involvement, most commonly aortic regurgitation.[1,7] Our patient's presentation was unusual because his symptoms were initially attributed to multifocal pneumonia, likely reflecting pulmonary congestion from severe aortic regurgitation in the setting of a rapidly enlarging aneurysm rather than infection alone.

The temporal proximity between COVID-19 infection and presentation raises the theoretical possibility that SARS-CoV-2–induced endothelial dysfunction contributed to aortic wall weakening. COVID-19 is known to induce endothelial inflammation, cytokine release, and degradation of collagen and elastin fibers. Although IMMD is classically considered a noninflammatory process, viral endothelial stress could accelerate mucopolysaccharide accumulation or elastic fiber fragmentation in predisposed individuals. To our knowledge, no prior cases have linked IMMD directly to COVID-19 infection; however, this possibility warrants further investigation as delayed vascular complications of viral infections continue to be recognized.[8,9]

Diagnosis relies on correlation between imaging and histopathology. In our case, CT and CTA identified the ascending aortic aneurysm and subsequent dissection, while TTE confirmed severe aortic regurgitation. However, definitive diagnosis was made only after surgical excision and histopathologic examination, which demonstrated focal myxoid degeneration within the intimomedial layer.

Surgical management is the treatment of choice for symptomatic or complicated IMMD, given the high risk of rupture, dissection, and progression.[1,2] Surgical techniques depend on the location and extent of involvement and may include valve-sparing aortic root replacement, total arch repair, and adjunctive endovascular stenting.[3,5] Our patient underwent emergent valve-sparing aortic root replacement, arch repair, and descending thoracic stent grafting, with a favorable postoperative recovery.

An important perioperative consideration in IMMD is a tendency toward bleeding, reported in up to 14% of surgical cases and often attributed to primary fibrinolysis associated with the aneurysm wall.[1,5] Although our patient did not experience this complication, awareness of this potential risk and careful hemostatic management are warranted.

CONCLUSIONS

This case demonstrates that IMMD can occur in populations outside the classically described demographic and may present with symptoms that mimic respiratory infection. Clinicians should consider underlying structural cardiovascular disease in patients with unexplained dyspnea or presumed pneumonia that is unresponsive to antibiotics, particularly when cardiac murmurs are present or thoracic imaging reveals mediastinal abnormalities. Although causality cannot be established, recent viral infections such as COVID-19 may contribute to vascular injury in susceptible individuals. Early recognition, prompt surgical intervention, and histopathologic confirmation are essential for optimizing outcomes in patients with IMMD.

Financial disclosures: None declared.

Funding/support: None declared.

REFERENCES

1. Abdool-Carrim AT, Robbs JV, Kadwa AM, Kenoyer G, Cooper K. Aneurysms due to intimomedial mucoid degeneration. *Eur J Vasc Endovasc Surg.* 1996;11(3):324-329. doi:10.1016/S1078-5884(96)80079-7

2. Viljoen C, Szymanski P, Osman N, et al. Intimomedial mucoid degeneration causing aortic and renal artery aneurysms in a young adult. *Cardiovasc J Afr.* 2016;27(1):49-52. doi:10.5830/CVJA-2015-079

3. Tzavellas G, Skripochnik E, Landau D, Wain RA, Tassiopoulos AK. Intimomedial mucoid degeneration of the peripheral arteries. *J Vasc Surg Cases Innov Tech.* 2019;5(4):452-455. doi:10.1016/j.jvscit.2019.04.005

4. Raber MH, Meerwaldt R, van Det RJ. A young man with intimomedial mucoid degeneration of the brachial artery. *J Vasc Surg.* 2011;53(3):822-824. doi:10.1016/j.jvs.2010.10.107

5. Katz JR, West DL, Bui JT, Knuttinen G, Chejfec G, Owens CA. Endovascular treatment of intimomedial mucoid degeneration. *J Vasc Interv Radiol*. 2008;19(12):1765-1768. doi:10.1016/j.jvir.2008.08.019

6. Wiskott K, Genet P, Lobrinus JA, Fracasso T, Lardi C. Intimomedial mucoid arterial degeneration, a rare arterial disorder of forensic significance. *Forensic Sci Med Pathol.* 2019;15(4):591-594. doi:10.1007/s12024-019-00154-x

7. Agozzino L, de Vivo F, Falco A, de Luca L, Schinosa T, Cotrufo M. Surgical pathology of the aortic valve: gross and histological findings in 1120 excised valves. *Cardiovasc Pathol.* 1994;3(3):155-161. doi:10.1016/1054-8807(94)90024-8

8. Varga Z, Flammer AJ, Steiger P, et al. Endothelial cell infection and endotheliitis in COVID-19. *Lancet*. 2020;395(10234):1417-1418. doi: 10.1016/S0140-6736(20)30937-5.

9. Ackermann M, Verleden SE, Kuehnel M, et al. Pulmonary vascular endothelialitis, thrombosis, and angiogenesis in Covid-19. *N Engl J Med.* 2020;383(2):120-128. doi: 10.1056/NEJMoa2015432.

advancing the art & science of medicine in the midwest

WMJ

WMJ (ISSN 2379-3961) is published through a collaboration between The Medical College of Wisconsin and The University of Wisconsin School of Medicine and Public Health. The mission of *WMJ* is to provide an opportunity to publish original research, case reports, review articles, and essays about current medical and public health issues.

© 2026 Board of Regents of the University of Wisconsin System and The Medical College of Wisconsin, Inc.

Visit www.wmjonline.org to learn more.