Radiology Case Reports 16 (2021) 2934–2937

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

"Undiagnosed aortic coarctation with 2 simultaneous acute aortic syndromes: Intramural hematoma and mycotic aneurysm"☆

Sowmiya Murali, MD[a,*], *Sherry S. Wang, MBBS*[b], *Michelle A. Grenier, MD*[a], *Jonathan W. Revels, DO*[a]

[a] *Department of Radiology, University of New Mexico, MSC 10 5530, 1, Albuquerque, NM 87131, USA*
[b] *Department of Radiology and Imaging Sciences, University of Utah, Salt Lake City, UT, USA*

ARTICLE INFO

Article history:
Received 23 June 2021
Revised 6 July 2021
Accepted 6 July 2021

Keywords:
Aortic coarctation
Intramural hematoma
Mycotic aneurysm
Computed tomography angiography

ABSTRACT

Acute aortic syndrome can be a fatal pathology if not diagnosed and managed early. Although acute aortic syndrome is more often a diagnosis of adulthood, it may occasionally afflict the pediatric patients. We herein present a case of a 5-year-old female that was discovered to have multiple acute and congenital aortic abnormalities after presenting to the emergency department with infectious symptoms and lower extremity pain. Acute aortic syndrome may not be a top differential consideration in children with acute chest pain; however, it is important to consider because delayed diagnosis and management can have fatal implications.

Introduction

Acute aortic syndrome is rare amongst the general pediatric population, with the exception of those with known congenital heart disease, connective tissues disorders, and/or trauma [1,2]. Similar to adults, early diagnosis and intervention is imperative to reduce the risk of patient mortality, which at baseline (onset of symptoms) can be 0.035% in those under 19-years-old and increase by 1-2% per hour after the onset of symptoms if left untreated [2]. Herein we present a case of a 5-year-old patient that was found to have acute and congenital aortic abnormalities after presenting to the emergency department with lower extremity pain: aortic coarctation, mycotic aortic aneurysm, and intramural hematoma.

Case report

During the height of the severe acute respiratory syndrome corona virus 2 (SARS-CoV-2, also known as COVID-19) pandemic, a 5-year-old female with no past medical history initially presented with 6 days of fever, body aches, chills, poor

☆ Competing interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author. S. Murali.
E-mail address: somurali1@salud.unm.edu (S. Murali).

https://doi.org/10.1016/j.radcr.2021.07.014

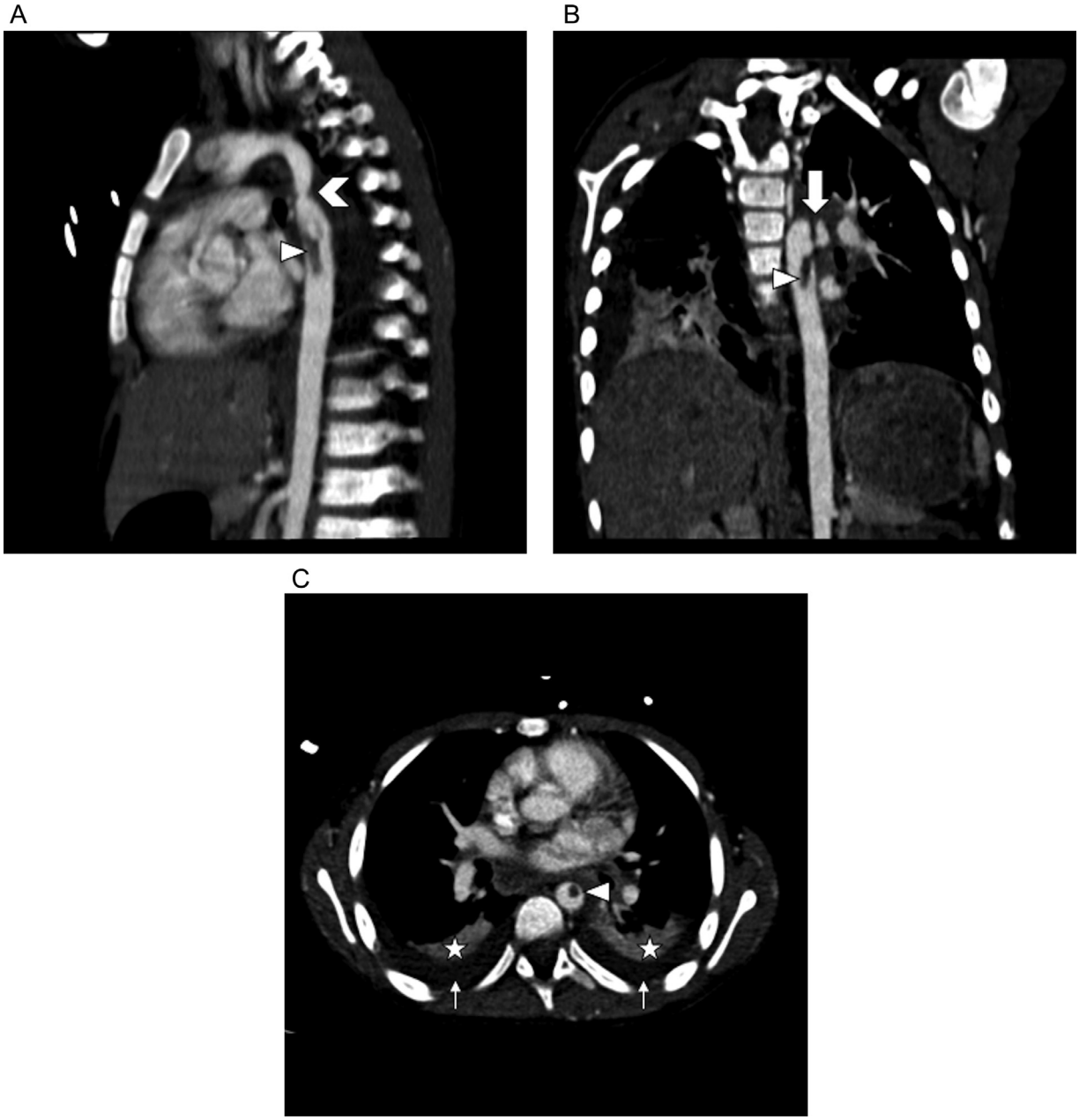

Fig. 1 – Non-cardiac gated contrast enhanced computed tomography angiogram of the aorta. (A) Sagittal oblique multiplanar reconstruction image of the thoracic aorta (candy cane view) demonstrating the post-ductal location of the aortic coarctation (chevron), as well as the intraluminal aortic thrombus (arrowhead). (B) Coronal MPR of the thoracic aorta demonstrates the aortic saccular outpouching along the left aspect of the proximal descending thoracic aortic segment (arrow) and adjacent aortic intraluminal thrombus (arrowhead). (C) Axial image of the mid-descending thoracic aorta demonstrating the aortic intraluminal thrombus (arrowhead), as well as mild volume pleural effusions (small arrows) and dependent lower lobe atelectasis (stars).

appetite, vomiting, left toe pain and swelling, and inability to bear weight on the left foot. Upon admission, she developed acute, severe chest pain and difficulty breathing. On physical examination, multiple splinter hemorrhages and petechiae were noted on her feet and toes. Her lower extremity pulses were difficult to obtain on physical examination, so a bilateral venous Doppler was ordered. No vascular thrombus was demonstrated on Doppler, but there was an incidental finding of bilateral monophasic arterial waveforms. This prompted further evaluation with an echocardiogram that revealed aortic coarctation.

Laboratory results demonstrated leukocytosis of 16.9, elevated C-reactive protein of 10.5, elevated D-dimer > 7650, and an elevated brain natriuretic peptide of 834. COVID-19 polymerase chain reaction testing was negative, as was COVID-19 antibody testing.

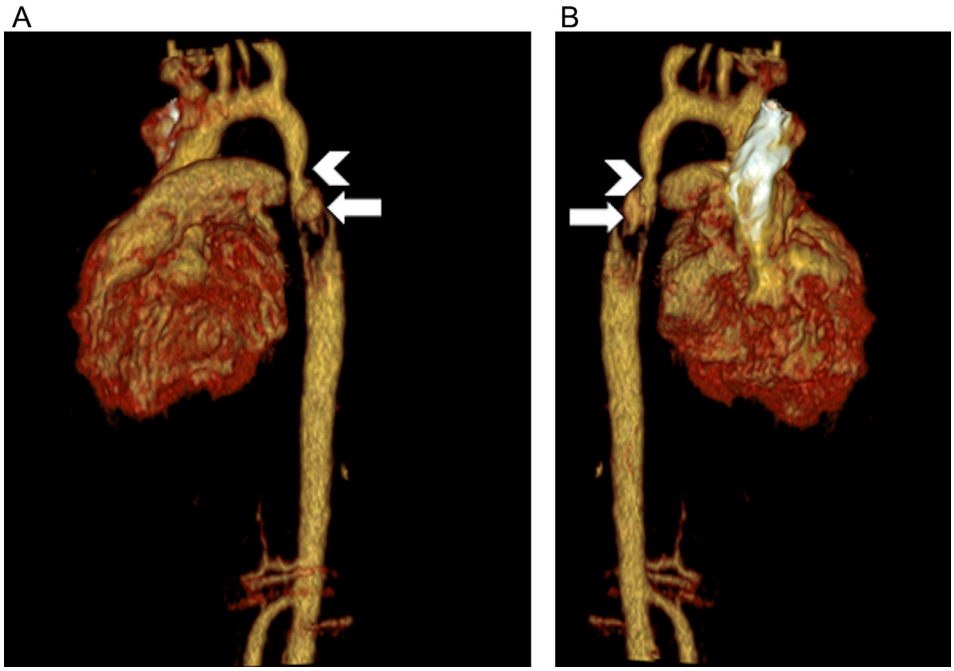

Fig. 2 – Three-dimensional volume rendered reconstruction of the aorta as viewed from the patient's left (A) and right (B) demonstrating the post-ductal location of the aortic coarctation (chevron), as well as the aortic saccular outpouching along the left aspect of the proximal descending thoracic aortic segment (arrow).

A contrast enhanced computed tomography angiogram of the aorta confirmed aortic coarctation with proximal descending thoracic aortic penetrating ulcer versus intramural hematoma with ulcer-like projection and associated intraluminal thrombus (Fig. 1 and Fig. 2).

The patient was transferred to an outside institution for surgical management. Intraoperatively, the patient was found to have a discrete aortic coarctation with isthmus hypoplasia. The post-stenotic descending aorta was dilated with a 3.0 cm mycotic pseudoaneurysm containing a large vegetation. There was also full thickness aortic wall penetration with a contained intramural hematoma. Resection of the aortic coarctation and mycotic aneurysm with interposition graft replacement was performed successfully. The patient tolerated the procedure with no complications. At 1 month follow-up, she is continuing antibiotics and is doing well.

Discussion

Aortic coarctation is a congenital abnormality diagnosed and repaired during childhood. In rare cases, the diagnosis is delayed until complications of the coarctation arise, such as hypertension [3]. We present a unique case of delayed presentation of aortic coarctations that was established after development of mycotic aneurysm and intramural hematoma. There have been reported cases of mycotic aneurysms in the setting of aortic coarctation, but to our knowledge there has not been a reported case of mycotic aneurysm and intramural hematoma in the setting of a native aortic coarctation, all being uncommon in children.

Mycotic aneurysms may develop when microorganisms inoculate a diseased aortic wall due to bacteremia or fungemia, which weakens the aortic wall thus leading to an aneurysm [4]. The incidence of aortic mycotic aneurysms is rare accounting for only 0.6% of all aortic aneurysms, rendering an even lower incidence with coexisting aortic coarctation [4]. Mycotic aneurysms can present with a non-specific constellation of symptoms, as in our case, which may delay diagnosis. They are associated with high mortality due to spontaneous rupture leading to fatal hemorrhage [5].

Intramural hematomas (IMH) occur due to spontaneous rupture of the vasa vasorum that can be caused by underlying hypertension and/or penetrating aortic ulcers [6]. IMH typically has the absence of an intimal tear resulting in a false lumen, which differs from an aortic dissection [7,8]. Although aortic coarctation can be a risk factor for aortic dissection, to our knowledge, there have been no reported cases of IMH with native aortic coarctation.

Our case is unique because 2 acute aortic syndromes coexisted within an undiagnosed aortic coarctation. We hypothesize that the patient's systemic infection may have been allowed for the formation of the aortic mycotic aneurysm due to her underlying coarctation that predisposed to disruption of the internal elastic lamina, intima, and media. This exposes the media to arterial flow, causing hemorrhage and creating an intramural hematoma, similar to the pathogenesis of a penetrating atherosclerotic aortic ulcer [6]. However, in this case infection is thought to be the primary culprit. The mycotic aneurysm was also likely to be a source of septic emboli

that led to the patient's lower extremity symptoms and findings: pain, splinter hemorrhages, etc.

Conclusion

Acute aortic syndrome is rare in children and may not be a top differential consideration in children with chest pain, especially if there are no known underlying aortic abnormalities, such as coarctation. Patients with aortic coarctation are at an increased risk of aortic wall complications both in the native setting, as well as in cases of prior repair. This is thought to be due to intrinsic histologic abnormalities of the aortic wall [9].

Patient consent

Patient and/or next of kin consent could not be obtained due to this being an older examination. All patient identifying information has been stripped from the images and movie files. Additionally, no patient identifying information is used in the case report.

IRB statement

No IRB approval was required for this manuscript.

REFERENCES

[1] Fikar CR, Fikar R. Aortic dissection in childhood and adolescence: an analysis of occurrences over a 10-year interval in New York State. Clin Cardiol 2009;32:E23–6.
[2] Eun LY, Cho DK, Cho YH, Byun KH. Aortic dissection and rupture in an 11-year-old child: A case report. J Cardiol Cases 2011;3:e46–9.
[3] Revels JW, Wang SS, Richards A. Bilateral renal artery parvus tardus: a sign of aortic coarctation in pediatric patients. Abdom Radiol (NY) 2021;46:841–3.
[4] Sörelius K, di Summa PG. On the diagnosis of mycotic aortic aneurysms. Clin Med Insights Cardiol 2018;12:1179546818759678.
[5] Ishizaka N, Sohmiya K, Miyamura M, Umeda T, Tsuji M, Katsumata T. Infected aortic aneurysm and inflammatory aortic aneurysm–in search of an optimal differential diagnosis. J Cardiol 2012;59:123–31.
[6] Macura KJ, Corl FM, Fishman EK, Bluemke DA. Pathogenesis in acute aortic syndromes: aortic dissection, intramural hematoma, and penetrating atherosclerotic aortic ulcer. AJR Am J Roentgenol 2003;181:309–16.
[7] Nienaber CA, von Kodolitsch Y, Petersen B, Loose R, Helmchen U, Haverich A, et al. Intramural hemorrhage of the thoracic aorta. Diagnostic and therapeutic implications. Circulation 1995;92:1465–72.
[8] Revels JW, Akaike G, Kicska G, Cham MD. Chinese Ring-Sword sign in intramural hematoma: feature of intramural blood pool. Abdom Radiol 2020;45:234–6.
[9] Oliver JM, Gallego P, Gonzalez A, Aroca A, Bret M, Mesa JM. Risk factors for aortic complications in adults with coarctation of the aorta. J Am Coll Cardiol 2004;44:1641–7.