Section: Radiodiagnosis

Original Research Article

PSEUDOCOARCTATION OF THE AORTA IN TURNER SYNDROME – A RARE MIMIC OF TRUE COARCTATION

Shikha Sood[1], Deepak Kumar[2], Kushagra Sharma[3], Kishitaa Rohilla[3], Harmandeep Kaur Sra[4], Rahul Paswan[2]

[1]Professor, Department of Radiodiagnosis, IGMC, Shimla, Himachal Pradesh, India.
[2]Junior Resident, Department of Radiodiagnosis, IGMC, Shimla, Himachal Pradesh, India.
[3]MBBS Student, MMU, Solan, Shimla, Himachal Pradesh, India.
[4]Consultant, Mercy St. Vincent Medical Center, Toledo, Ohio, USA.

Received : 18/06/2026
Received in revised form : 02/07/2026
Accepted : 16/07/2026

Keywords:
Pseudocoarctation, Turner syndrome, Coarctation of aorta, CT angiography, Congenital vascular anomaly.

Corresponding Author:
Dr. Deepak Kumar,
Email: deepakkrwork@gmail.com

DOI: 10.47009/jamp.2026.8.4.73

Source of Support: Nil,
Conflict of Interest: None declared

Int J Acad Med Pharm
2026; 8 (4); 420-422

ABSTRACT

Background: Pseudocoarctation of the aorta is an uncommon congenital defect marked by extension and kinking of the aortic arch, without substantial luminal obstruction or pressure gradient. It is very rarely associated with Turner syndrome, although true coarctation is often seen in these patients. Failure to differentiate pseudocoarctation from true coarctation can result in unnecessary surgical or endovascular interventions. The purpose is to illustrate an uncommon instance of pseudocoarctation of the aorta in a patient with Turner syndrome and emphasize the need of cross-sectional imaging to distinguish it from coarctation. **Materials and Methods:** A 14-year-old girl with a previous diagnosis of Turner syndrome presented with community acquired pneumonia. Transthoracic echocardiography and CT angiography were performed after identifying a suspected aortic arch abnormality. **Result:** Transthoracic echocardiogram showed bicuspid aortic valve without significant valvular dysfunction, preserved left ventricular systolic function (EF 58%), and a dilated tortuous aortic arch with diastolic spill and no measurable pressure gradient. CT angiography showed elongation and kinking of the aortic arch just distal to the left subclavian artery with preserved luminal caliber, absence of focal narrowing and absence of collateral circulation, confirming the diagnosis of pseudocoarctation of the aorta. **Conclusion:** The case highlights the role of cross-sectional imaging in differentiating pseudocoarctation from true coarctation in Turner syndrome. Accurate diagnosis can prevent unnecessary intervention, and ensure appropriate long-term surveillance due to the risk of aneurysm formation or aortic dissection.

INTRODUCTION

Pseudocoarctation of the aorta is an uncommon congenital defect marked by extension and kinking of the aortic arch, often occurring at the ligamentum arteriosum, without substantial luminal blockage or hemodynamic impairment. It is thought to be due to abnormal embryological development of the aortic arch and can closely mimic true coarctation of the aorta on initial evaluation. Pseudocoarctation, however, is not associated with a significant pressure gradient across the lesion or the development of collateral circulation as is true coarctation. Correct differentiation between pseudocoarctation and true coarctation is necessary for proper treatment.[1,2]

Turner syndrome is a chromosomal condition resulting from full or partial monosomy of the X chromosome. It occurred in 1 out of 2,500 female newborns. One of the most common congenital manifestations are cardiovascular abnormalities, including but not limited to bicuspid aortic valve, coarctation of the aorta and aortic root dilatation.[3] Up to one third of patients with Turner syndrome have coarctation of the aorta, and bicuspid aortic valve is reported in around one quarter of cases.[4] In contrast, pseudocoarctation of the aorta is exceedingly rare in this group.

The clinical presentation of pseudocoarctation is often nonspecific. In many cases the condition is discovered incidentally during imaging done for some unrelated indications.[5] CT angiography and magnetic resonance angiography are very important in confirming the diagnosis. They clearly demonstrate the characteristic elongation and kinking of the aortic arch while excluding features of true coarctation and its complications.[2]

We present an uncommon case of pseudocoarctation of the aorta in a 14-year-old female exhibiting phenotypic characteristics of Turner syndrome and possessing a bicuspid aortic valve. We emphasize the

ISSN (O): 2687-5365; ISSN (P): 2753-6556

importance of multimodality imaging to differentiate this uncommon entity from true coarctation and to avoid unnecessary intervention.

CASE REPORT

A 14-year-old girl was diagnosed with community-acquired pneumonia with a 3-day history of fever and cough. The patient had a pre-existing diagnosis of Turner syndrome. Physical examination revealed short stature, webbed neck, widely spaced nipples, and pectus excavatum. She had not attained menarche. Pubertal staging showed Tanner stage II breast development and Tanner stage I genitalia. Pelvic ultrasonography demonstrated a hypoplastic uterus and nonvisualization of the bilateral ovaries [Figure 1].

Figure 1: Clinical photograph demonstrating phenotypic features of Turner syndrome including short stature and characteristic body habitus.

Transthoracic echocardiography demonstrated a bicuspid aortic valve without significant stenosis or regurgitation. Left ventricular systolic function was preserved with an ejection fraction of 58%. The aortic arch appeared dilated and tortuous with evidence of diastolic spill. Although coarctation of the aorta was suspected, no measurable pressure gradient was identified across the suspected narrowing. Given these findings, CT angiography was performed for further evaluation.

CT angiography revealed elongation and kinking of the aortic arch distal to the origin of the left subclavian artery. The luminal caliber was preserved without focal narrowing or post-stenotic dilatation. There was no evidence of collateral circulation involving intercostal, internal mammary, vertebral, subclavian, or thyrocervical branches. These imaging findings established the diagnosis of pseudocoarctation of the aorta (Figure 2 and 3). The patient responded well to antibiotic therapy for pneumonia and was discharged with recommendations for endocrinological evaluation and periodic cardiovascular follow-up.

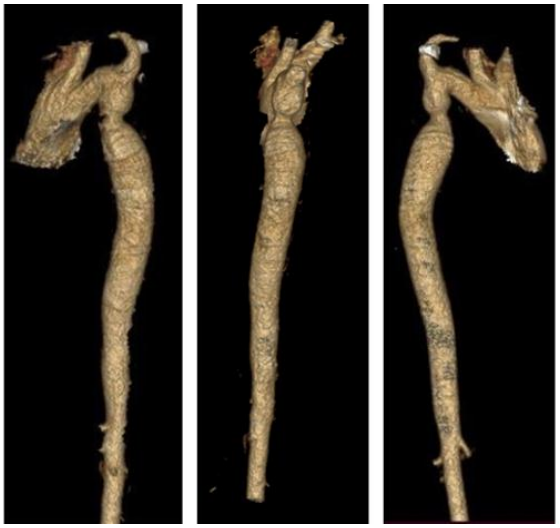

Figure 2: Elongation of the descending aorta with buckling at the level of the ligamentum arteriosum, distal to the origin of the left subclavian artery.

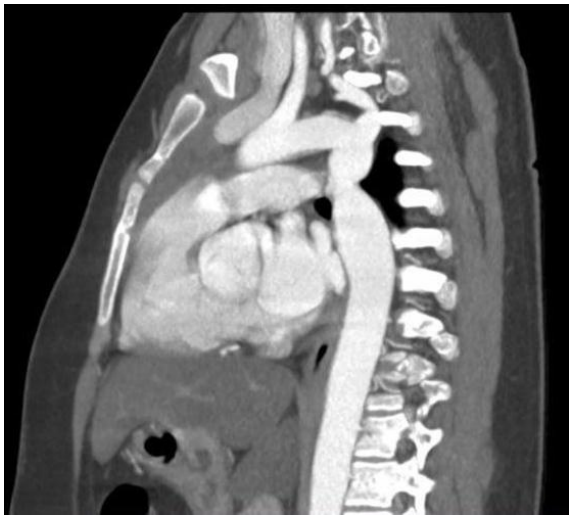

Figure 3: The aorta distal to the origin of the left subclavian artery demonstrates elongation and buckling with preserved luminal caliber. No evidence of dissection or collateral circulation is identified.

DISCUSSION

Pseudocoarctation of the aorta is an uncommon congenital defect characterized by extension and kinking of the aortic arch, often occurring near the location of the ligamentum arteriosum. Differentiation of pseudocoarctation from true coarctation still remains a diagnostic challenge as both entities belong to the same spectrum of aortic arch anomalies. However, pseudocoarctation can be differentiated with absence of significant blood pressure variation between upper and lower limbs,

ISSN (O): 2687-5365; ISSN (P): 2753-6556

absence of pressure gradient across the lesion and absence of collateral circulation.[1] The current case showed a dilated and tortuous aortic arch by transthoracic echocardiography without measurable pressure gradient. Computed tomographic angiography showed elongation and kinking of the aortic arch distal to the left subclavian artery with preserved luminal caliber and no collateral vessels. These findings correlated with the diagnostic criteria for pseudocoarctation and true coarctation was excluded.

This condition is rare, as shown by a systematic review published in 2014 in which only 18 reported cases of pseudocoarctation were identified over a period of 20 years. The most common presentation was hypertension, with or without unequal blood pressure in the extremities, followed by dyspnea and dysphagia, and the mean age at presentation was 43 years.[1] In contrast, our patient was a 14 years old girl with previously diagnosed Turner syndrome presenting with community acquired pneumonia and the vascular anomaly was an incidental finding during cardiovascular evaluation. This emphasizes the fact that pseudocoarctation may be asymptomatic and may be incidentally diagnosed during evaluation for other conditions.

Consistent with previous reports, CT angiography correctly demonstrated elongation and kinking of the aortic arch, while excluding collateral circulation, aneurysm formation and dissection. CT angiography and magnetic resonance angiography are considered essential imaging modalities to evaluate aortic morphology and related complications such as aneurysm or dissection.[1] In this case, the diagnosis was confirmed by CT angiography, which demonstrated elongation and buckling of the aortic arch distal to the left subclavian artery without focal narrowing, collateral vessels, aneurysm or dissection. Cardiac catheterization remains the gold standard for measuring pressure gradients across the lesion.[1] The diagnosis was made noninvasively in our case, given the absence of a measurable gradient on echocardiography and the characteristic CT findings.

The most frequent associations reported with pseudocoarctation are aortic valve abnormalities, especially unicuspid or bicuspid aortic valves.[6,7] Our patient also had bicuspid aortic valve without significant valvular dysfunction on echocardiography, supporting this previously recognized association. Furthermore, pseudocoarctation has been reported to occur with chromosomal abnormalities such as Turner syndrome, Noonan syndrome and Hurler syndrome and other congenital cardiovascular anomalies [6]. True coarctation of the aorta is a well-recognized cardiovascular manifestation of Turner syndrome, whereas pseudocoarctation is distinctly uncommon. Thus, the present case contributes to the scarce literature describing this rare association.

Management of pseudocoarctation is usually conservative as the disease has a benign clinical course.[8] Surgical and endovascular treatments have been described but the most accepted management is careful observation, reserving treatment for symptomatic patients or patients who develop complications such as aneurysm formation or dissection during follow-up.[8,9] Our patient had no evidence of hemodynamically significant obstruction, aneurysm or dissection and therefore conservative management with periodic cardiovascular surveillance was considered appropriate. Long term follow up is still important, especially in patients with Turner syndrome, given their increased risk of aortic complications.

CONCLUSION

Pseudocoarctation of the aorta is a rare but important differential diagnosis for patients with Turner syndrome and suspected coarctation. Multimodality imaging is a decisive confirmation of hemodynamic obstruction and collateral circulation. Proper identification prevents unnecessary intervention and allows for appropriate long-term surveillance for possible complications such as aneurysm or dissection.

REFERENCES

1. Asil, S., Geneş, M., Çelik, M., Yüksel, U. Ç., & Barçın, C. (2022). Pseudocoarctation of the aorta: A rare congenital aortic disease. Anatolian Journal of Cardiology, 26(1), 69–71. https://doi.org/10.5152/AnatolJCardiol.2021.934
2. Chou, Y. Y., Wang, C. J., Lin, C. H., Chung, H. T., & Lo, F. S. (2020). Association between cardiovascular anomalies and karyotypes in Turner syndrome patients in Taiwan: A local cohort study. Pediatrics & Neonatology, 61(2), 188–194. https://doi.org/10.1016/j.pedneo.2019.10.001
3. Khan, N., Farooqui, A., & Ishrat, R. (2024). Turner syndrome: Where are we? Orphanet Journal of Rare Diseases, 19(1), 314. https://doi.org/10.1186/s13023-024-03337-0
4. Nakai, H., Shun, M., & Tanaka, H. (2019). Successful treatment of hypoplastic aortic arch and pseudocoarctation of the aorta in an octogenarian. Interactive CardioVascular and Thoracic Surgery, 29(1), 150–151. https://doi.org/10.1093/icvts/ivy366
5. Sakurai, Y., Koide, M., Kunii, Y., Tateishi, M., Okugi, S., & Cao, Y. (2019). Severely kinked pseudocoarctation of the aorta with unicuspid aortic valve. The Annals of Thoracic Surgery, 107(6), e421. https://doi.org/10.1016/j.athoracsur.2019.01.027
6. Shafeeq, A., Nandakumaran, M., Prathapkumar, G., Ramesh, R., & Christudhas, A. (2024). Pseudocoarctation of the aorta: A rare incidental finding during a preoperative assessment. Cureus, 16(7), e65574. https://doi.org/10.7759/cureus.65574
7. Singh, S., Hakim, F. A., Sharma, A., Roy, R. R., Panse, P. M., Chandrasekaran, K., Alegria, J. R., & Mookadam, F. (2015). Hypoplasia, pseudocoarctation and coarctation of the aorta: A systematic review. Heart, Lung and Circulation, 24(2), 110–118. https://doi.org/10.1016/j.hlc.2014.08.006
8. Tung, R., & Siegel, R. J. (2007). Aortic pseudocoarctation associated with a stenotic congenitally bicuspid aortic valve. The American Journal of Cardiology, 100(1), 157–158. https://doi.org/10.1016/j.amjcard.2007.02.066
9. Zuo, Z. L., Tsauo, J. Y., Chen, M., & Feng, Y. (2017). Successful percutaneous stent implantation for isolated dismal transverse aortic arch kinking. Medicine, 96(10), e6089. https://doi.org/10.1097/MD.0000000000006089

ISSN (O): 2687-5365; ISSN (P): 2753-6556