HK J Paediatr (new series) 2022;**27**:188-191

Case Report

A Rare Complication of Aortic Coarctation: Aneurysm and Aortoesophageal Fistula

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Abstract ***Introduction:*** Rupture of an aortic aneurysm and aortoesophageal fistula formation is a rare but serious complication of aortic coarctation. ***Case Presentation:*** A 13-year-old girl was admitted with a minor haematemesis in whom computed tomography examination showed aortic coarctation and a poststenotic aneurysm surrounded by a haematoma and right aberrant subclavian artery. An endoscopic examination revealed bleeding from a small orifice at the posterior wall of proximal esophagus which is suggestive of an aortoesophageal fistula. Resection of coarctation and aneurysmatic segment, graft replacement and closure of aortoesophageal fistula were performed with no complication. ***Conclusions:*** As major complications such as aortic aneurysm and aortoesophageal fistula are rarely reported in children with previously undiagnosed aortic coarctation, this entity must be kept in mind for the patients who present with vomiting even small amounts of blood. Also the X-ray findings of aortic coarctation and its' complications must be aware by the clinicians and radiologists.

Key words *Aneurysm; Aortic coarctation; Aortoesophageal fistula; Haematemesis; Rupture*

Introduction

Coarctation of the aorta is a congenital focal narrowing of the aortic lumen which accounts for 4% of all congenital heart disease. Serious complications of aortic coarctation such as aneurysm may be especially seen in previously undiagnosed or treated elderly patients.[1] Rupture of coarctation related aneurysm which can lead to aortoesophageal fistula and massive haematemesis is rarely reported.

In this report, we present computed tomography (CT) findings of a coarctation case that was complicated with haematemesis secondary to rupture of aneurysm and aortoesophageal fistula.

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Received July 17, 2020

Case Presentation

A 13-year-old girl, was admitted to the paediatric emergency department with a complaint of vomiting bright red blood. Her parents reported that she vomited about half a cup of blood 20 minutes before admission. Her medical history was unremarkable. A physical examination of oropharynx revealed fresh blood in pharynx but no any bleeding source. In the emergency room, she developed two vomiting episodes with a total of 10-15 cc fresh blood. Her heart rate was 124 beats per minute and blood pressure was 150/90 mm Hg at right upper extremity. On laboratory examination, haemoglobin level was 13 g/dl. A PA chest roentgenogram showed tracheal deviation and a figure of 3 sign in upper mediastinum (Figure 1). An urgent contrast-enhanced CT and endoscopy were planned. CT examination showed a coarctation of descending aorta distal to left subclavian artery. The coarctation segment has 8 mm caudocranial length and the widest diameter of the lumen was 9 mm in most stenotic point while the diameter of the prestenotic segment was 31 mm and poststenotic segment was 23 mm. A saccular aneursym

with a sizes of 12x11x10 mm, arising from the posteromedial wall of the descending aorta which was 13 mm distal to the coarctation site was demonstrated. The aneurysm was surrounded by a 32x31x28 mm haematoma which was compressing and markedly narrowing esophagus. Also a mild right- sided tracheal deviation secondary to the compression of haematoma was seen. An aberrant right subclavien artery which arising from just proximal to stenotic segment was demonstrated (Figures 2-5). During endoscopy, bleeding from a small orifice in the posterior wall of proximal esophagus was detected. Also at this site a marked extrinsic compression which narrowing the lumen was noted. She was afebrile (37°C) at admission. Besides there was no growth on blood culture which excluded an infectious process. With radiological and endoscopic findings, a diagnosis of an aortoesophageal fistula associated with a saccular aneurysm related to aortic coarctation was made and urgent resection of coarctation and aneurysmatic segment, graft replacement and also direct closure of the esophageal fistula were performed. She was discharged on the 15th postoperative day without

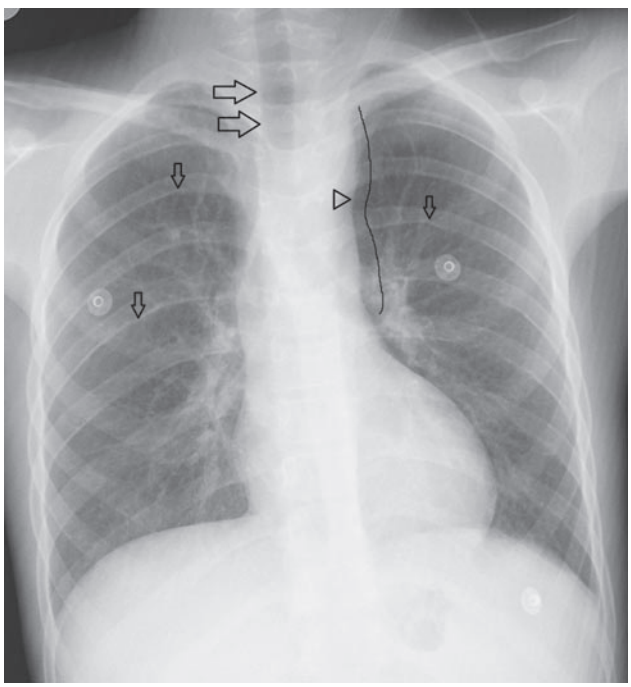

Figure 1 A PA chest X-ray shows stenotic segment of aorta (arrowhead) which forms 'figure of 3' sign (lines). Rib notching secondary to formation of intercostal collaterals is seen (short arrows). Tracheal deviation to the right side caused by the compression of haematoma is also shown (long arrows).

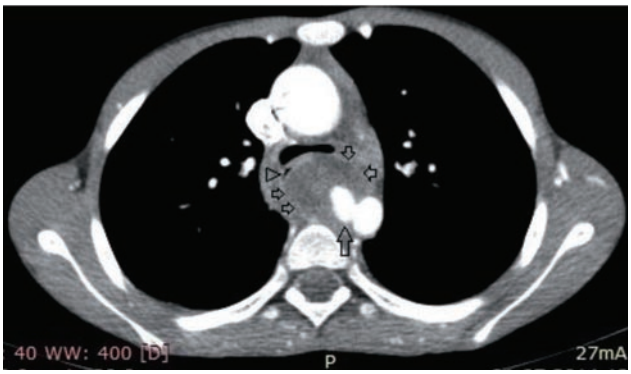

Figure 2 An axial contrast-enhanced CT angiography image demonstrates a saccular aneurysm (long arrow) which is surrounded by a large haematoma (short arrows). Haematoma caused marked compression, and right deviation of esophagus. Marked narrowing of the lumen of esophagus is seen (arrowhead). Also a mild compression to trachea is shown. Intercostal-paraspinal collaterals and dilated internal thoracic arteries are seen.

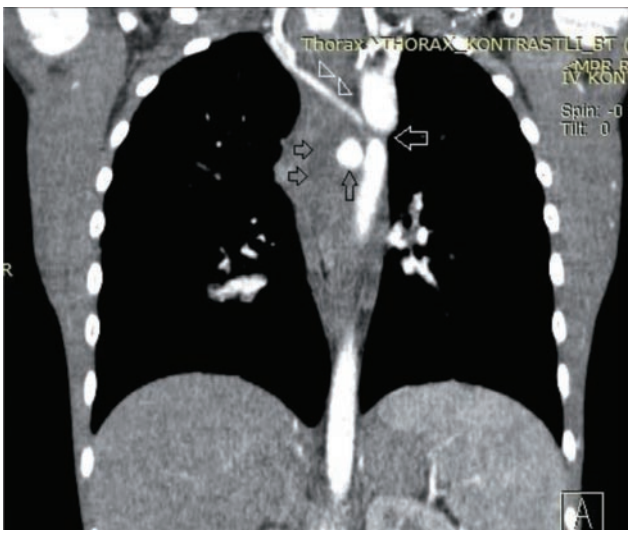

Figure 3 A coronal reformatted CT image shows coarctation site (white arrow) and an aberrant right subclavian artery (white arrowheads) which is originated from the prestenotic segment of aorta. Saccular aneurysm (black long arrow) and the haematoma (black short arrows) are also seen.

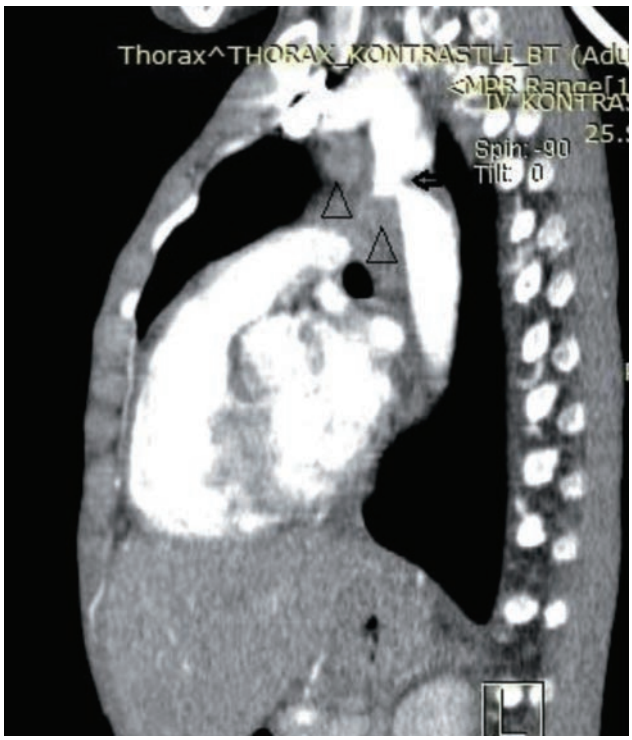

Figure 4 A sagittal reformatted CT image shows an aortic coarctation (arrow) and haematoma (arrowheads).

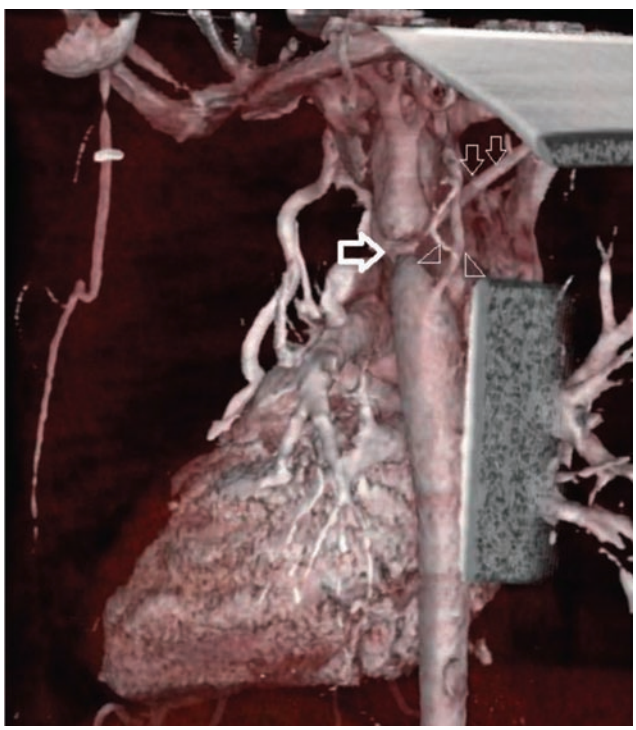

Figure 5 The 3D-volume rendering image shows aortic coarctation (long arrow) and the aneurysm (arrowheads).Also an aberrant right subclavian artery originates from prestenotic

complication. There was no evidence of recurrence of aneurysm or residual stenosis on follow-up at 6 and 12 months. An endoscopy on follow-up at 6 months showed normal esophagus lumen with no stenosis or stricture at the site of closure.

Discussion

Post-stenotic aortic aneurysm is a common and serious complication of aortic coarctation which occurs in 17% of unrepaired coarctation.[2] Preventza et al, reviewed a total of 943 patients in whom coarctation repair was performed in their institute from 1962 to 2011. Among these 943 patients aortic aneurysm was detected in 55 ones (5.8%) (mean age 39.9+-11.3 years (11-74). In 42 (76.4%) of these patients the aneurysm was located in descending aorta. Also in 25 patients (45.5%) coarctation was associated with bicuspid aortic valve, in 7 patients (12.7%) with aortic valve dysfunction, in 3 ones (5.4%) with ductus arteriosus, in 3 ones (5.4%) with ventricular septal defect, in one patient (1.8%) with foramen ovale, in one patient (1.8%) with partial anomalous pulmonary venous return, in one patient (1.8%) with Turner syndrome and in one patient with right aortic arch. There was no patient with coarctation related aneurysm and right aberrant subclavian artery.[1]

Postcoarctation turbulent blood flow which leads to endothelial trauma and subsequent colonsation of microorganisms at the site of damaged endothelium is the suggested mechanism for aneurysm formation in patients with aortic coarctation.[3]

In literature, there are a few case reports about haematemesis due to aortic aneurysm and aortoesophageal fistula associated with aortic coarctation. Burns et al, reported an 11-year-old boy with previously undiagnosed coarctation of the aorta who presented with massive haematemesis due to a pseudoaneurysm and aortoesophageal fistula.[4] McKell reported a 14 years old male with aortic coarctation presenting with massive haematemesis.[5] Sheiko and Hoffenberg reported a 15 years old male associated with Congenital Aortic Coarctation with Superinfected Aneurysm and Aortoesophageal Fistula who presented with massive haematemesis.[6] Grant et al, reported massive haematemesis in an 11-year-old male with undiagnosed aortic coarctation and mycotic aneurysm.[7] Krieves et al, reported massive haematemesis associated with aortoesophageal fistula and coarctation of the aorta in a 15-year-old child.[3]

When compared with previous cases, our patient presented with a relatively mild haematemesis. Also, her haemoglobin levels were within normal limits (13 g/dl). This may be due to the compression of marked perianeurysmal haematoma to the esophagus and also to aortoesophageal fistula. As similarly with reports above, our case was previously undiagnosed, early diagnosis of aortic coarctation is very essential to prevent serious complications such as cardiac failure, aortic rupture, endocarditis, aortic aneurysm and intracranial haemorrhage.[3] A careful and detailed examination of chest roentgenograms may be very helpful when rib notching detected which occurs secondary to dilated and tortiosed intercostal vessels.

Inferior rib notching occurs secondary to various conditions such as coarctation by the dilated and tortious intercostal arteries which subsequently erode the inferior margins of the ribs, to by-pass the stenotic segment and supply poststenotic descending aortic segments. It is suggested that if bilateral rib notching present, the coarctation must be distal to the origin of both subclavian arteries. Unilateral right rib notching is seen when coarctation lies distal to the brachiocephalic trunk, but proximal to the origin of the left subclavian artery or there may be a right-sided aortic arch with aberrant left subclavian artery distal to coarctation. Also if there is unilateral left rib notching it is associated with aberrant right subclavian artery arising after the coarctation.[8] In our case bilateral inferior rib notching is present because right aberrant subclavian artery arising just proksimal to aortic coarctation.

Also 'figure 3 sign' is an important radiological finding which is formed by stenotic segment (Coarctation site) in middle and the cranial (prestenotic) dilated segment of aortic arch and caudal (poststenotic) dilated segment of descending aorta. In our patient, right-sided tracheal deviation was suggestive of a mediastinal lesion. When the lack of evidence of pneumothorax, atelectasis, consolidation, or fibrosis in lung zones on chest roentgenograms, further radiological examinations such as contrast-enhanced computed tomography must be performed to rule out mediastinal lesions.

To the best of our knowledge this is the first aortic coarctation case with right aberrant subclavian artery which was complicated with aortic aneurysm and aortoesophageal fistula. Contrary to previous reports, this case was haemodynamically stable and presented with vomiting relatively small amounts of blood. Aortic coarctation and its' major complications must be kept in mind for the children who present with minor haematemesis. Also, the chest roentgenogram findings of aortic coarctation and its' complications must be well-known by the clinicians and radiologists.

Declaration of Interest

There are no conflicts of interest and no funding in this report.

Acknowledgement

An informed written consent was obtained from the parents of the patient.

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