ESC European Society of Cardiology

European Heart Journal - Case Reports (2019) **3**, 1–5
doi:10.1093/ehjcr/yty170

CASE REPORT
Congenital heart disease

A case report of mycotic pseudoaneurysm in childhood: an unusual complication of coarctation of the aorta

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Received 25 January 2018; accepted 23 December 2018; online publish-ahead-of-print 4 March 2019

Background We report on an unusual case of a 3 year-old girl with coarctation of the aorta complicated by mycotic pseudoaneurysm and infected with *Streptococcus pneumoniae*.

Case summary The only symptoms and signs were fever and weak femoral pulses. The echocardiography confirmed a localised isthmic's coarctation. In order to complete the evaluation, a CT scan was performed. This proved crucial in terms of the diagnosis and decision to perform emergency surgery. The diagnosis was confirmed surgically. An aortic rupture was contained by the parietal pleura. Bacteriological analysis of surgical specimens revealed bacterial DNA that tested positive for *Streptococcus pneumoniae*. The post-operative course was free from any cardiovascular or neurological complications after six weeks of antibiotic therapy.

Discussion Surgical repair of coarctation of the aorta is frequently performed in children. However, complicated forms are less common with a potentially fatal outcome. Indeed, there are no recommendations concerning the management and surgical timing of mycotic pseudoaneurysm. These rare complications should be kept in mind. Although short- and medium-term follow-up of these children is good, caution should be exercised with long-term follow-up because of complications in childhood and adulthood.

Keywords Case report • Paediatric coarctation of the aorta • Aortic rupture • Mycotic pseudoaneurysm • False aneurysm • Streptococcus pneumonia • Aortitis

Learning points

- Coarctation of the aorta may be complicated by mycotic pseudoaneurysm and *Streptococcus pneumoniae*—it is important to keep these rare complications in mind.
- The diagnosis of associated abscesses by transthoracic echocardiography is difficult or even impossible. The computed tomography scan was the key imaging tool for diagnosis and management when clinical history and echocardiography are unusual.
- Combined medical and surgical treatment for pseudoaneurysm of an aortic coarctation may be lifesaving.

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Handling Editor: Nikolaos Bonaros

Peer-reviewers: Cemil Izgi, Monika Arzanauskaite, and Esther Cambronero-Cortinas

Compliance Editor: Mohammed Akhtar

Supplementary Material Editor: Peregrine Green

Introduction

This case reports on an unusual case of coarctation of the aorta complicated by mycotic pseudoaneurysm infected by *Streptococcus pneumoniae* in a child. The co-existence of coarctation of the aorta and post-stenotic aneurysm is well known.[1–3] Endarteritis may develop beyond the coarctation area because of the turbulent flow of the blood stream directed against the aortic wall.[4] However, an infected pseudoaneurysm revealing coarctation of the aorta with a favourable outcome is exceptional.

Timeline

Fifteen days prior to presentation	No medical comorbidities. Deterioration in her general condition: fever, asthenia Possible diagnosis of atypical Kawasaki syndrome was considered
Upon presentation to our institution	Heart rate: 110 beats/minute, blood pressure in the right upper limb was 139/83 mmHg and 99/71 mmHg in the right lower limb. Moderate biological inflammatory syndrome (CRP was 74 mg/L). The white cell count was 24 000 giga/L with 85% of neutrophils. Transthoracic Echocardiography (TTE) demonstrated a coarctation of the aorta A cardiac CT-scan coarctation of the aorta complicated by pseudoaneurysm
Emergency surgical treatment of this pseudoaneurysm and coarctation	Microscopic examination of surgical specimens confirmed extra cardiac aortitis and bacteriological analysis revealed bacterial DNA (PCR RNA 16S), which tested positive for *Streptococcus pneumoniae*, fully sensitive to antibiotic therapy
After 13 days	CT scan did not detect any abnormality
After two months	End of antibiotic therapy
After a year	Last TTE: no complications

Case presentation

A 3-year-old girl with no medical comorbidities presented with deterioration in her general condition since 2 weeks (fever and asthenia). She was admitted to another hospital where a possible diagnosis of atypical Kawasaki syndrome was considered based on a prolonged fever of 39°C associated with cheilitis and cervical lymphadenopathy; therefore, the patient received human immunoglobulin and an anti-inflammatory dose of aspirin. Apyrexia was immediately achieved with no recurrence of febrile peaks after aspirin. Then, the patient was transferred to our institution. Her immunizations were up to date including the 7-valent pneumococcal conjugate vaccine (when she was 2 months, 4 months, and 11 months old).

The medical examination in our centre revealed a heart rate at 110 b.p.m., blood pressure in the right upper limb was 139/83 mmHg and 99/71 mmHg in the right lower limb. There was no fever, no cheilitis, and no cervical lymphadenopathy anymore. There was no evidence of heart failure. The femoral pulse was weak and cardiac auscultation revealed a discrete systolic murmur audible throughout the thorax but loudest in the third left intercostal space. Systolic functional murmur is frequently detected in paediatric populations, especially during bouts of fever. There was no radial femoral delay. There was a moderate biological inflammatory syndrome [CRP was 74 mg/L (0–6 mg/L)]. The white cell count was 24 000 G/L (7000–11 000 G/L) with 85% of neutrophils. Urine testing showed aseptic leucocyturia. Stomatological examination did not highlight any infectious focus. There was no evidence of immune deficiency.

Transthoracic echocardiography (TTE) demonstrated mild hypoplasia of the aortic arch associated with localized isthmic coarctation. The flow in the abdominal aorta was dampened. The aortic tract between the aortic arch and the coarctation was oddly configured and could not be clearly visualized on ultrasound examination (*Figure 1*). Transoesophageal echocardiography (TOE) was not performed because the transthoracic window is good in children. We did not see any vegetation or abscess in TTE technique and TOE technique is very difficult to implement in paediatric population.

Therefore, a cardiac computed tomography (CT) scan was performed to complete the evaluation. A Gothic-type aortic arch was observed. Aortic coarctation was located below the ductus arteriosus (*Figures 2*). The aortic wall in the descending thoracic aorta was very irregular and indicative of pseudoaneurysm. These findings were associated with low attenuation periaortic collection with enhanced periphery (Figure *3*). There was an arteria lusoria (aberrant right subclavian artery), the origin of which was occluded within the collection. Collaterals across the occluded segment were visualized.

Based on the imaging findings, the final diagnosis was coarctation of the aorta complicated by pseudoaneurysm and a thoracic aortic rupture contained by the compressive effect of the adjacent structures.

Consequently, the patient was prepared for emergency surgical treatment of this pseudoaneurysm and aortic coarctation.

The procedure was carried out by left thoracotomy at the fourth intercostal space under extra corporeal circulation. The CT diagnosis of contained rupture was surgically confirmed: aortic rupture was contained by the parietal pleura.

After careful dissection of the tissues near the aortic rupture, two aortic clamps were positioned before and after the rupture zone with no changes in cerebral and distal perfusion monitoring. Following double aortic cross-clamping, the pleura and the rearranged aortic tissues were incised and flattened. Surgical treatment included extensive debridement of all the rearranged tissues. The healthy part of the aortic wall was anatomically and macroscopically normal. There was no obvious sign of infection.

The aortic reconstruction was obtained by end-to-end anastomosis using the Crafoord technique.

The peripheral pulse was clearly perceived immediately after surgery. Intraoperative TOE did not reveal any vegetation.

Microscopic examination of surgical specimens confirmed extracardiac aortitis. A parietal inflammatory infiltrate with the formation of an aortic and periaortic abscess was also noted. Furthermore,

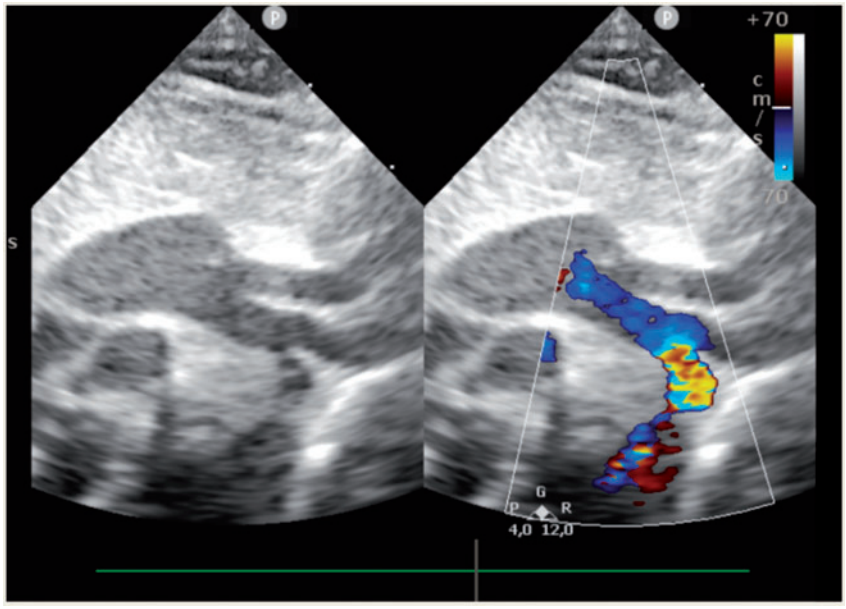

Figure 1 Transthoracic echocardiography demonstrating mild hypoplasia of the aortic arch associated with localized isthmic coarctation. The aortic tract between the aortic arch and the coarctation was oddly configured and could not be clearly visualized.

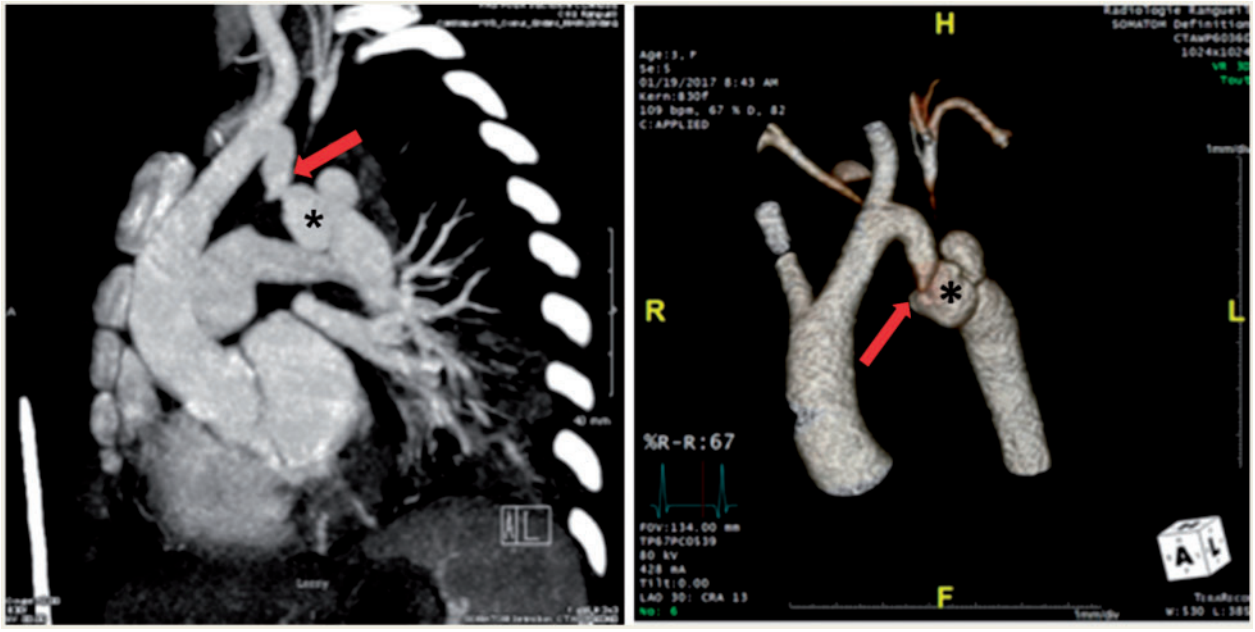

Figure 2 Cardiac computed tomography scan with three-dimensional reconstruction demonstrating coarctation of aorta (arrow) with the pseudoaneurysm (asterisk) below.

bacteriological analysis of the surgical specimens revealed bacterial DNA (PCR RNA 16S), which tested positive for *S. pneumoniae*. Surgical specimen and abscess cultures proved negative.

After initial empirical antibiotic therapy, treatment targeted Pneumococcal infection with amoxicillin followed by a switch to ceftriaxone for a total of 6 weeks.

The post-operative course was free from any cardiovascular or neurological complications. The biological inflammatory syndrome disappeared [CRP at 4 mg/L (0–6 mg/L)], and the white cell count was 8000 G/L (7000–11 000 G/L) with 3500 neutrophils (3500–6 000 G/L). A thoracic–abdominal–pelvic follow-up CT scan did not detect any vascular abnormality in the sub-diaphragmatic region.

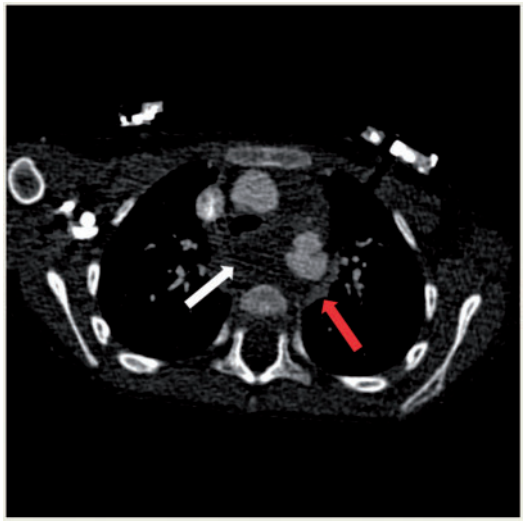

Figure 3 Cardiac computed tomography scan. Low attenuation peri-aortic collection (arrow) consistent with pseudoaneurysm contained by the compressive effect of the adjacent structures (tracheobronchial and oesophagus) with the enhanced periphery (arrow).

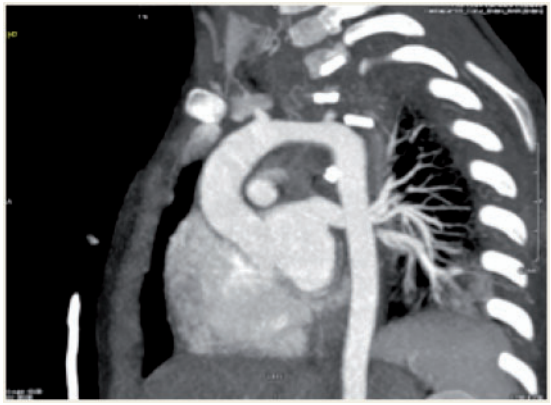

Figure 4 Computed tomography scan 15 days after surgery: the calibre of the aorta is regular without residual coarctation or aneurysm.

There was no evidence of septic emboli or secondary infectious sites of endocarditis and the aorta was well repaired (*Figure 4*).

The patient was able to return home 20 days after surgery. Antihypertensive drugs could be discontinued 4 months later and the last echocardiographic check-up performed after 1 year of follow-up was good.

Discussion

Children with endocarditis often present non-specific symptoms and are less likely to have classical peripheral vascular and immunological phenomena, vegetation and Osler's triad.[5,6] *Streptococcus pneumoniae* is detected in 3–7% of cases of endocarditis in children[6] and is thus a rare albeit often fatal infection. Choi and Mailman[5] found the mean age of presentation of pneumococcal endocarditis to be 4.1 years. There were no major diagnostic criteria of endocarditis in this case. No intracardiac abscess was highlighted on the echocardiogram or confirmed by histological examination. Microscopic examination of surgical specimens confirmed extracardiac aortitis. There was a discrepancy between clinical and surgical or anatomopathological findings. However, at least three minor criteria were present including predisposing heart condition, fever >38.0°C and vascular phenomena (mycotic pseudoaneurysm, which is a known complication of endocarditis). Therefore, the patient was treated for endocarditis for 6 weeks.

The clinical presentation was unusual. Kawasaki disease was initially suspected before imaging studies because of prolonged fever in a child coupled with moderate elevation of inflammation markers. The symptoms were crude with no typical pain. In addition, blood cultures were always negative and there was no sign of vegetation. Renal dysfunction is commonly associated with this condition,[7] but no immune complex glomerulonephritis or renal failure was observed.

To our knowledge, this is the first reported case of a 3-year-old child who survived[8] following mycotic pseudoaneurysm due to *S. pneumoniae* infection accompanied by coarctation of the aorta. *Streptococcus pneumoniae* has rarely been described in these situations.[6,9–11] Furthermore, the child was properly vaccinated against pneumococcus when she was 2 months, 4 months, and 11 months old, had no medical or surgical history and was not immunocompromised.

Surgery was successful without any complications or aortic allograft material. Following excision of the infected part of the aorta, antibiotic therapy was effective in eradicating the infection. The management of mycotic aneurysm is unclear. There are no strategies or recommendations regarding the timing of surgery (before or after antibiotics), aortic reconstruction (with or without material), and the duration of post-operative antibiotic therapy. Studies suggest that combined medical and surgical treatment decreases mortality.[5]

The CT scan was the key imaging tool for diagnosis and management in our case contrary to other case reports which used magnetic resonance imaging or TOE.[8,12,13] We retrospectively reviewed the CT scan, and we find that low attenuation periaortic collection with the enhanced periphery correspond to the periaortic abscess found macroscopically.

Patient perspective

Coarctation of the aorta is considered a complete repair. Although the short- and medium-term follow-up of these children is good, caution should still be exercised with long-term follow-up because of complications in adulthood.[14]

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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