

IMAGES

in PAEDIATRIC CARDIOLOGY

Tomar M, Parakh R, Bhan A. Mycotic aneurysm: a lesser known complication of coarctation repair in a child, managed by endovascular aneurysm repair. *Images Paediatr Cardiol* 2017;19(4):1-9.

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Abstract

We report a 12-year-old child diagnosed with mycotic aneurysm at the site of coarctation of aorta (CoA) repair. Surgical intervention, though very high risk, is the mainstay of management. In selected cases endovascular stenting is being reported in adults. Here we report the use of an endovascular stent in a child.

Keywords: Coarctation, mycotic aneurysm, endovascular aneurysm repair.

Introduction

Infectious thoracic aortitis and mycotic aneurysm (MA) represent two extremes of the same disease. MA usually develops when a preexisting aneurysm become infected and usually the site is treated or untreated coarctation of aorta (CoA).¹ MA represents 2.6 % of all aortic aneurysms, with the thoracic aorta the least common site of occurrence.²

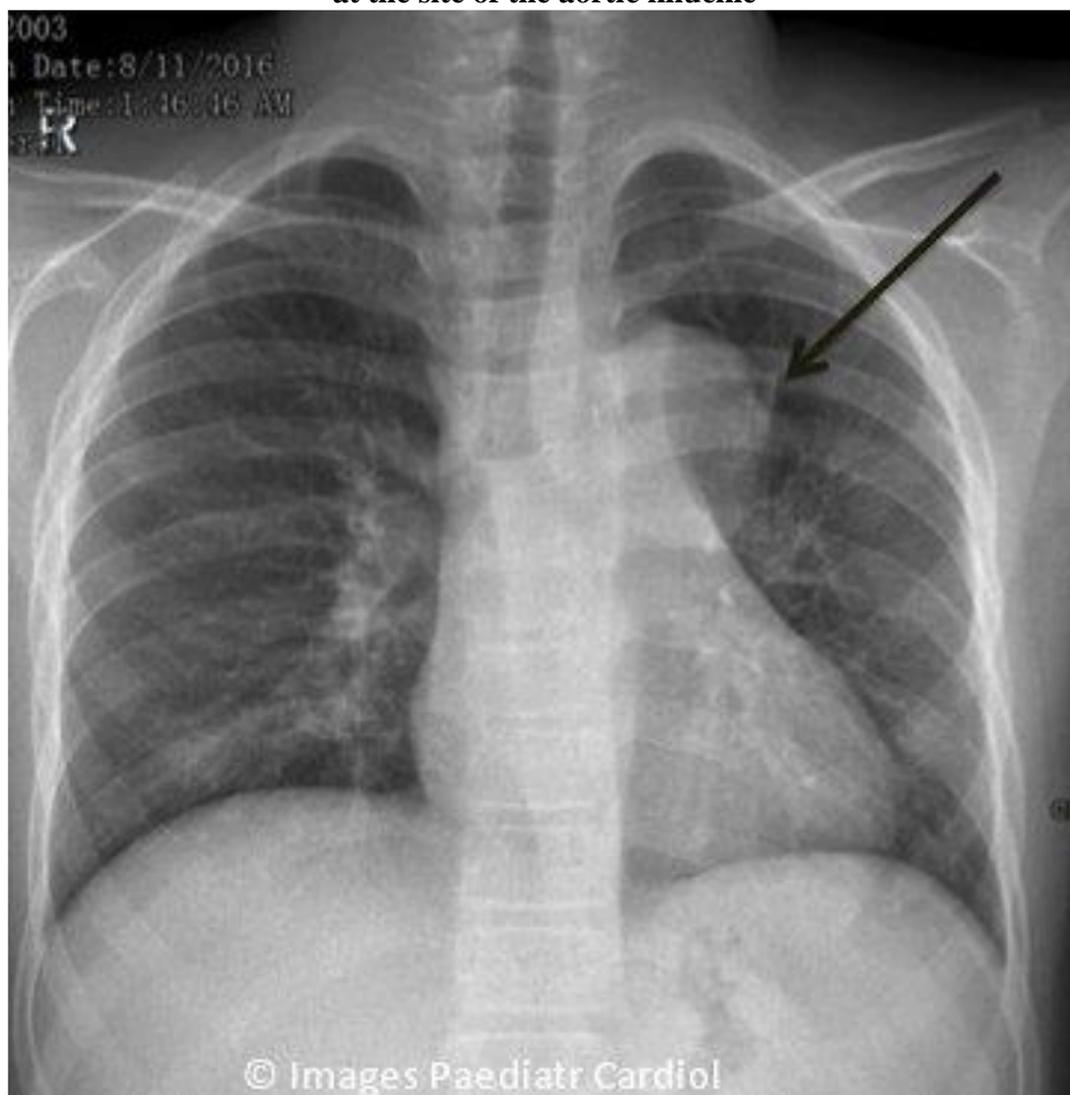
Unfortunately, the diagnosis of MA can be challenging and treatment carries significant risks and a high re-infection rates. Here we present the evaluation and management of a child with MA at the site of a previous CoA repair.

Case Report

We report a 12-year-old male child, post balloon dilatation of CoA at 3 years of age and followed by surgical intervention i.e. PTFE patch repair of CoA, ligation of ductus arteriosus and reimplantation of left subclavian artery to left carotid artery seven years later i.e. in 2014. The child was doing well till April 2016 when he had pain in the left hypochondrium and right leg followed by mild fever. There was no history of loss of appetite, weight loss, edema or any urinary complaint. The child was evaluated by a local physician. Work-up showed high leucocyte counts, high C-reactive protein and microscopic hematuria. Blood and urine cultures were sterile. He was treated as case of urinary tract infection with intravenous antibiotics (ceftriaxone and amikacin) for 14 days. One week after stopping the antibiotics, the child had recurrence of fever and was reevaluated. Repeat septic workup showed high inflammatory markers, numerous RBCs in urine while all cultures remained sterile. Chest X-ray frontal view showed a homogenous opacity of the left upper mediastinum at the site of aortic knuckle (figure 1).

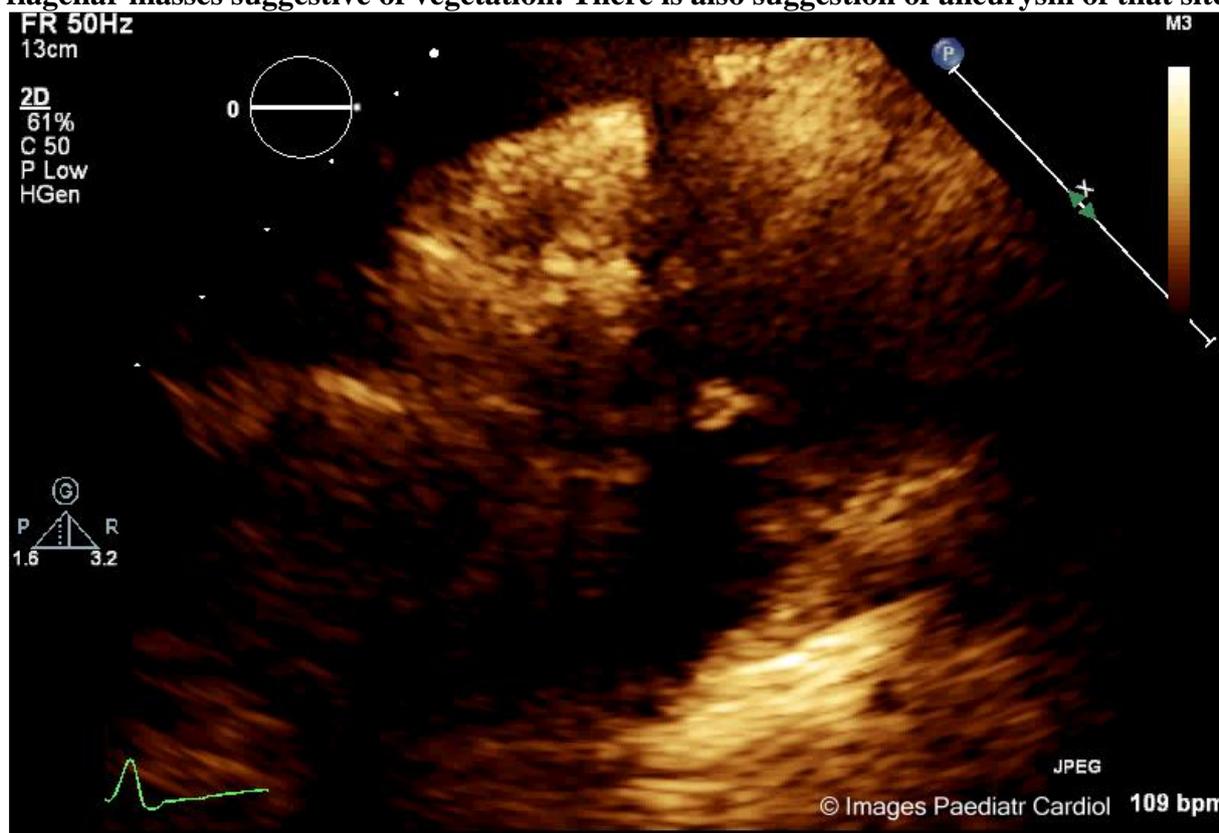
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Figure 1: Chest X-ray postero-anterior showing opacity in the left upper mediastinum at the site of the aortic knuckle



Echocardiography showed no evidence of intracardiac vegetation, no pericardial effusion and normal ventricular function. The suprasternal long axis view showed a well opened aortic arch, with no significant gradient but there was suggestion of aneurysmal dilatation of aorta. There were multiple echogenic flagellar masses at the site of the repair (figure 2).

Figure 2: Suprasternal long axis view of aortic arch showing a well opened arch with multiple flagellar masses suggestive of vegetation. There is also suggestion of aneurysm of that site.



CT angiography of the aorta (figure 3) showed multiple aneurysms at the site of surgical repair starting 5 mm distal to the left carotid artery and involving a length of 60 mm, mediastinal edema, enlarged mediastinal lymphnodes and multiple infarcts in the spleen. The child was treated as a case of mycotic aneurysm with intravenous antibiotics for 6 weeks.

He was planned for surgical intervention after completion of the antibiotic course. Repeat echocardiography, after completion of antibiotics showed no vegetations in the arch. Repeat CT angiography aorta showed no increase in number and size of aneurysms, the mediastinal edema had subsided but the mediastinal lymphnodes were still enlarged. In addition there was an aneurysm of the right profunda femoris artery. To rule out any active infection, positron emission tomography (PET) scan was also done.

With the diagnosis of MA, he was planned for intervention. Both options, surgical and nonsurgical i.e. endovascular aneurysm repair (EVAR) were discussed with the family and the parents agreed for endovascular stenting. After high risk consent the child was taken for EVAR.

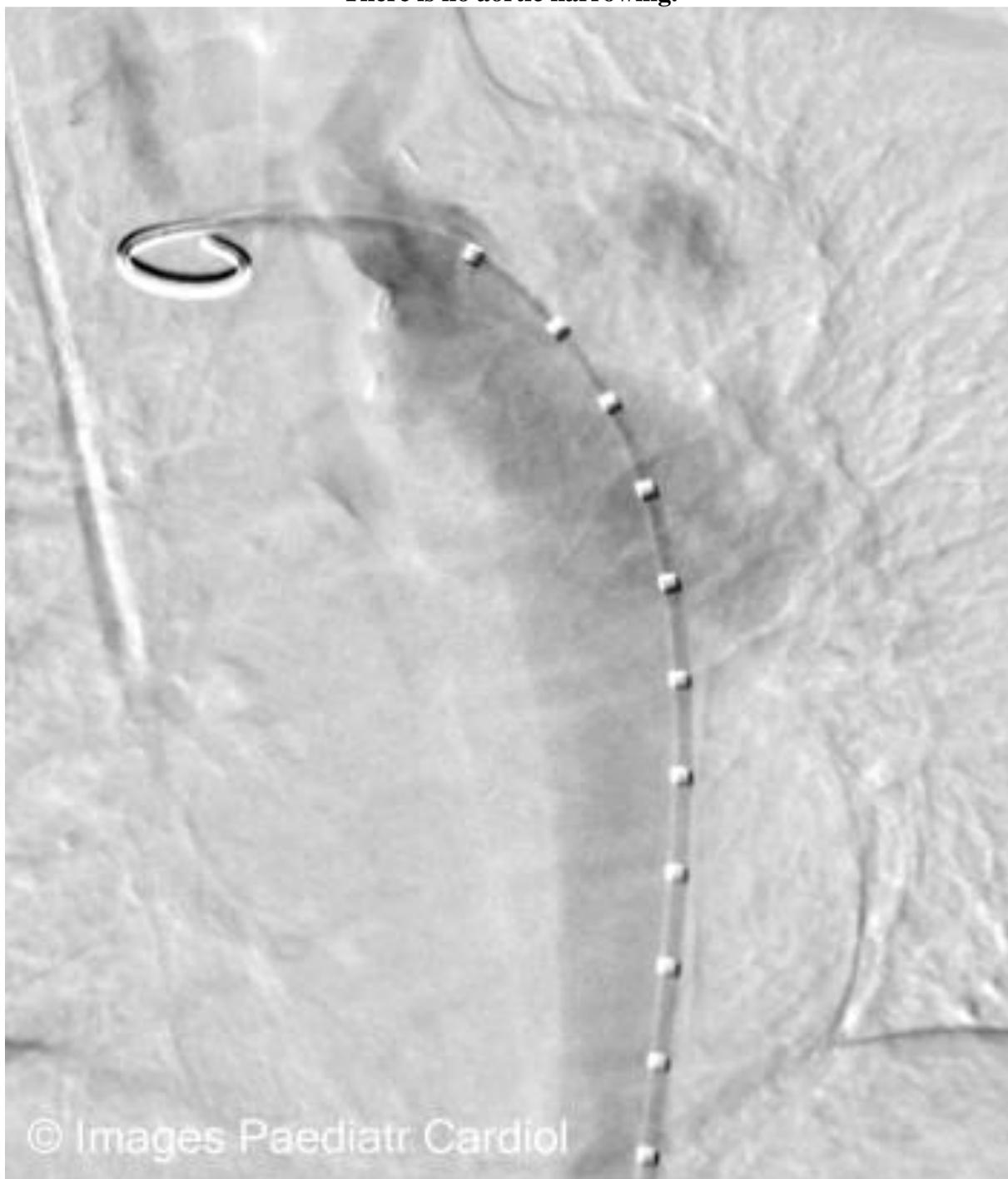
Figure 3: Three dimensional reconstruction of thoracic aorta with contrast angiography showing multiple aneurysms involving the site of coarctation of aorta repair



The procedure was done under general anesthesia with antibiotic cover. The right femoral artery lumen was small (5 mm) for a large sheath to be used for stenting. In addition there was small aneurysm involving the right profunda femoris artery. For these reasons, the procedure was planned from the right iliac artery. After profiling the aortic arch anatomy with from the left femoral artery (figure 4), the right iliac artery was exposed by incision in the iliac fossa. The aortic arch was crossed with the help of Multipurpose II catheter and an Amplatzer extrastiff wire (0.035”) was stabilized in the ascending aorta. Endurant Medtronic Limb Extension (16-20-93) was chosen to cover the length of diseased aortic segment and to exclude the aneurysms. Stent graft was deployed uneventfully under fluoroscopic guidance (figure 5). Post deployment angiography showed no restriction of flow to left carotid artery and to left subclavian artery, complete exclusion of aneurysms, and no dissection (figure 6). The pigtail catheter which was placed from the left femoral artery was removed over a Terumo wire as a precautionary measure.

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Figure 4: Aortic arch angiography showing multiple aneurysms of arch beyond left carotid artery. Left subclavian artery is being filled from left carotid artery. There is no aortic narrowing.



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Figure 5: Endurant Medtronic stent graft being deployed in diseased area under fluoroscopic guidance

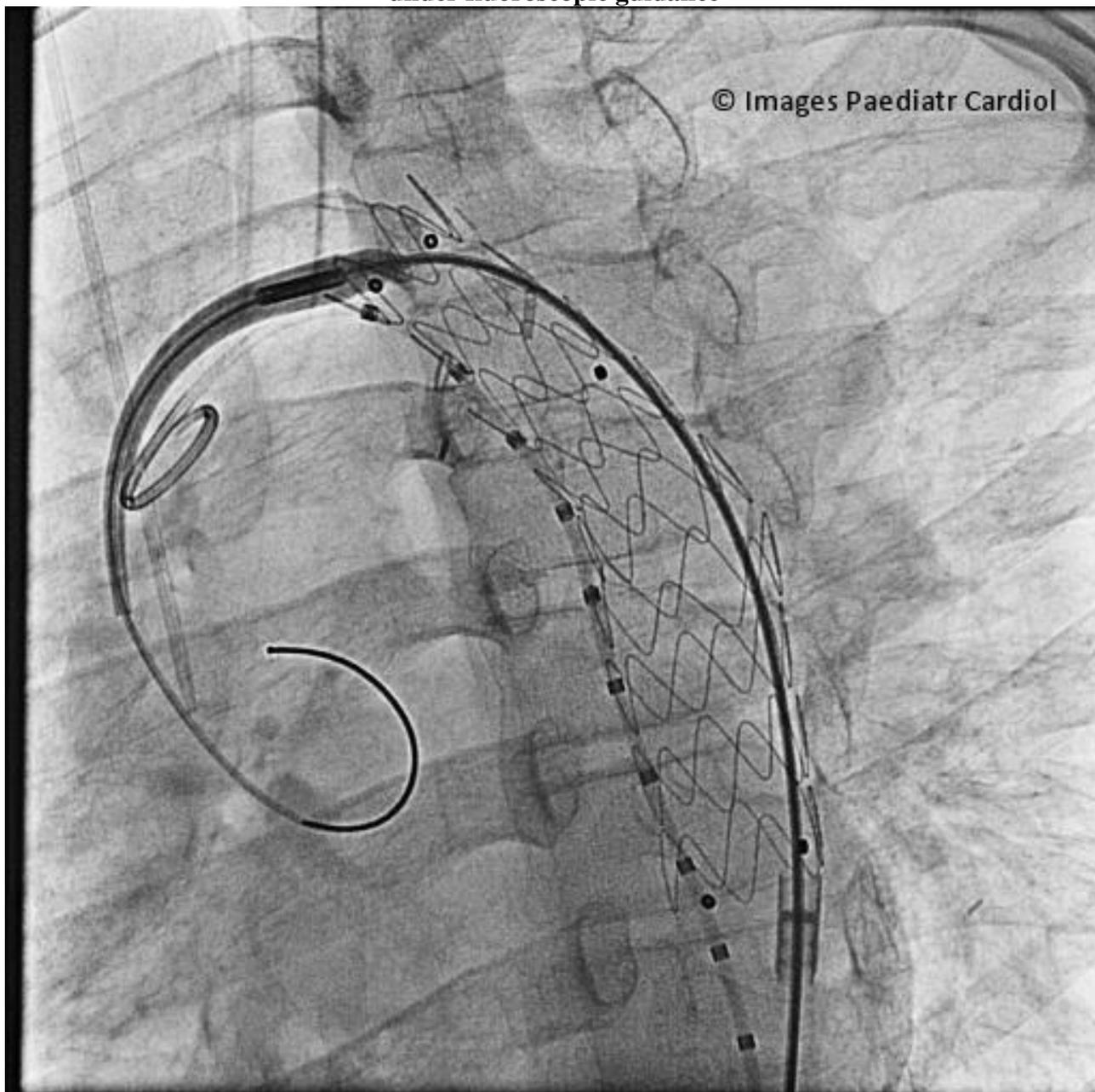


Figure 6: Aortic arch angiography post stent deployment showing well positioned stent graft with complete exclusion of aneurysms.



The post procedure antibiotic course was planned for 6 weeks and the family completed that under local pediatrician care. There was no recurrence of fever and inflammatory markers remained negative. CT angiography aorta 6 weeks and 6 months post procedure showed stent in situ, completely excluded aneurysms and no new lesion in the aorta.

Discussion

Aneurysms and pseudo-aneurysms have been described as late complications of CoA, repaired or un-repaired. Aneurysm formation at or near the site of CoA has been associated with both surgical and transcatheter relief of CoA.³ The formation of aneurysms at the site of coarctation repair appears to correlate to the time elapsed since the surgical correction and has a prevalence of ~5.4% post-coarctation repair.⁴ Perhaps because of this, the mean age of MA presentation is 40 years and the finding is uncommon in children. There is no gender predominance.²

Various microorganisms have been associated with MA, most commonly Staphylococcal, Enterococcus, Streptococcus, and Salmonella species. Infection by Mycobacterium tuberculosis, though rare, can occur due to extension of infection from nearby structures such as mediastinal lymph nodes and lung lesions.⁵ We were not able to isolate any organism in blood culture and workup for tuberculosis was also negative.

Clinical presentations and diagnosis: Clinical manifestations are nonspecific: fever (75%), thoracic pain (60%), abdominal pain (20%) and chills (16%) are common symptoms.² Mycotic aneurysm can cause compressive symptoms, such as dysphagia, dyspnea, hoarseness, cough and superior vena cava compression syndrome.² Possible complications such as thromboembolic phenomena and acute rupture will lead to emergency presentation. If left untreated, MA of aorta has a mortality rate close to 100% due to rupture of the lesion into the adjacent structures (such as the esophagus, bronchus or pleural cavity).¹

A high index of suspicion is necessary to promptly diagnose this entity, especially in those with a history of CoA repair. Our patient presented with fever and pain in the abdomen (left hypochondria) and right leg possibly due to embolic phenomena. He was initially treated as a case of urinary tract infection and as there was recurrence of symptoms. Detailed cardiac evaluation made the diagnosis of MA.

Echocardiography is a non-invasive and readily available diagnostic tool but in some cases it is not possible to make the diagnosis due to poor image quality. In our patient, we were able to demonstrate vegetation at the coarctation repair site with suspicion of aneurysm. Cardiac CT and MRI are more sensitive in the diagnosis of mycotic aneurysms.⁶ In our patient, CT angiography aorta was done to detail the diagnosis and showed a well-opened repaired arch with multiple aneurysms, splenic infarcts and also a mycotic aneurysm of right profunda femoris as a manifestation of septic embolism.

The appropriate treatment of mycotic pseudo aneurysms in a pediatric patient is not well defined. Some centers advocate for early surgery to prevent the risk of spontaneous rupture while acknowledging an increase in the risk for graft infection and insufficiency of the suture lines.⁶ Endovascular aneurysm repair has been advocated as an alternative in high risk surgical patients, though this does not allow removal of infected tissue, or assist in organism isolation.⁷ In some cases, EVAR is recommended as a bridge to open surgery.⁸ In cases not involving the aortic arch, the use of an extra-anatomic bypass (i.e. left subclavian artery to descending thoracic aorta) is preferred to avoid soiling of the new graft through the infected field. Our patient was hemodynamically stable and so was first treated appropriately with intravenous antibiotics for 6 weeks and as all inflammatory markers became negative, he was planned for intervention.

There is no standardization of antibiotic therapy or duration for mycotic aneurysm. Some authors suggest lifelong antibiotics⁹ while others suggest 6 weeks to 12 months of therapy.¹⁰ We used intravenous antibiotics for 6 weeks post procedure and followed with inflammatory markers and CT angiography.

Further follow up protocol is planned every 3 months with inflammatory markers for the 1st year and annually thereafter. Repeat CT angiography aorta has been planned at 6 months.

Conclusion

MA in children is a rare entity and requires high index of suspicion. Any patient under evaluation for pyrexia of unknown origin, especially with a previous history of coarctation intervention, should have MA ruled out. There is no role for medical management in mycotic aneurysm. Complete surgical excision of the infected aorta in combination with antibiotic coverage a (for at least 6-12

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weeks) is the mainstay of treatment. EVAR has been reported in adults with initial acceptable results but none in pediatric patient. Our patient underwent EVAR successfully although long-term device durability and infection free survival remain to be seen.

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