Mycotic aneurysm in a child with history of coarctation of the aorta repair

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ABSTRACT

A mycotic aneurysm is a rare condition occasionally seen in patients with a history of prior cardiac or vascular surgery. Here we report the presentation of a mycotic aneurysm in a pediatric patient at the site of prior aortic coarctation repair. This patient's initial presentation suggested rheumatologic or oncologic disease, and after diagnosis he continued to show evidence of splenic, renal and vascular injury distal to the mycotic aneurysm site while being treated with antibiotics. We discuss the diagnosis, treatment and management of this condition.

Keywords: Coarctation, congenital heart disease, mycotic aneurysm

INTRODUCTION

A mycotic aneurysm is a condition caused by destruction of the arterial wall secondary to infection or the development of infection in a pre-existing aneurysm. Though mycotic aneurysms are rare, they occasionally present in patients with aortic coarctation, with or without a history of surgical repair. Unfortunately, identification can be challenging and treatment carries significant risks and a high re-infection rate. Here we present the evaluation, diagnosis and management of an 11-year-old male with a mycotic aneurysm at the site of a previous coarctation repair.

CASE REPORT

An 11-year-old male presented to the ER for evaluation of bilateral leg pain and fever. He had a history of a bicuspid aortic valve and coarctation of the aorta for which he underwent surgical repair with an end-to-end anastomosis at 3 weeks of age. He had been followed for residual stenosis across the coarctation site and mild hypertension.

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Two months prior to presentation he experienced chest discomfort followed by pain in the lower extremities and toes. He reported limping, unexplained bruises, high fever and a 40 lb unexplained weight loss. On examination he was obese, had a grade II/IV systolic murmur at the upper sternal border (consistent with prior records), tenderness on palpation of both knees and mild swelling of the lower extremities without pitting edema. He reported pain in his fingers with a hand grip and had bilateral leg rash with multiple small raised, red-blanching skin lesions extending up to the knee area. ESR and CRP were elevated; his white cell count was 16,000, had anemia with thrombocytopenia and a positive ASO titer.

Chest X-ray [Figure 1] suggested a mediastinal mass suspicious for malignancy. PET- computerized tomography (CT) scan showed a hypermetabolic subcarinal lymph node and bone marrow, splenic enlargement and increased soft tissue activity in the distal right leg suggestive of lymphoma. Bone marrow biopsy was consistent with chronic disease with no evidence of malignancy. A blood culture drawn on the second day of admission found Gram positive cocci in chains that were later identified as *Streptococcus viridans*. Limited echocardiogram showed stable cardiac function, no intracardiac vegetations, a bicuspid aortic valve and a slight residual stenosis (30 mmHg) at the site of prior coarctation repair.

Due to the aortic gradient and the clinical findings, a chest CT was performed [Figure 2a-c] which showed an aortic aneurysm at the site of the prior coarctation repair measuring approximately 4.4 × 4.2 cm. There were also multiple splenic lesions and focal cortical

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thinning of the left kidney felt to be due to embolic events from thrombus within the aneurysm. Subsequent complete echocardiogram showed an aneurysm at the superior descending aorta near the prior coarctation site [Figure 3]. With a second consecutive blood culture positive for *Streptococcus viridians*, imaging tests results and his clinical findings he was diagnosed with a mycotic aneurysm. He was started on cefepime, which was later changed to ceftriaxone and gentamicin as per sensitivity results.

Cardiothoracic surgery was consulted for consideration of surgical palliation after completion of the antibiotic course. Despite IV antibiotic therapy, he continued to have high fevers everyday and elevated inflammatory markers. Creatinine remained elevated suggesting continued renal injury. A chest/abdomen/pelvis CT was performed 4 weeks after the initiation of antibiotics to evaluate for progression of the aneurysm. This showed aortic pseudoaneurysm enlargement, an increasing number of splenic and renal embolic infarcts, and a small aneurysm within the splenic artery.

There were no new positive blood cultures since the initiation of antibiotics but given the progression of the aneurysm and the continued distal embolic damage, he was taken to the OR for surgical repair (in spite of fevers and elevated inflammatory markers) and cefepime was restarted for preoperative prophylaxis against *Staph aureus*.

A left lateral thoracotomy through the previous coarctation repair scar was performed and multiple adhesions from the lung to the descending aorta and pseudoaneurysm were found. Proximal and distal control of the aorta was attained on either side of the pseudoaneurysm, which was quite extensive, involving the area from the takeoff of the left subclavian artery through the mid-descending thoracic aorta. Patient was cannulated and placed on left heart bypass through the left superior pulmonary vein and the descending thoracic aorta. Clamps were placed on the aortic arch distal to the left common carotid artery, including the left subclavian artery and over the descending aorta. The pseudoaneurysm was then opened, revealing grossly inflammatory tissue of poor quality with no obvious purulence and a number of intercostal arteries arising from the area of the pseudoaneurysm, which were oversewn. The capsule of the pseudoaneurysm was thick and attached densely to the esophagus requiring debridement of abnormal tissue, mobilizing the capsule off of the esophagus and excising the entire capsule. In an area posteriorly where the abnormal tissue was unresectable due to its proximity to the esophagus, xenyl was applied and then neutralized with alcohol and copious irrigation. The underside of the arch was opened back to the base of the left common carotid artery, and

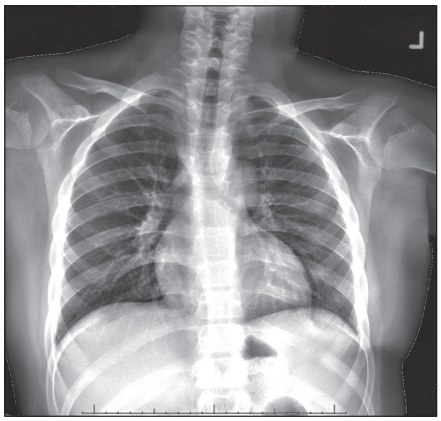

Figure 1: Chest X-rays on the day of presentation. Soft tissue density at the mediastinum measuring 3 × 3 cm

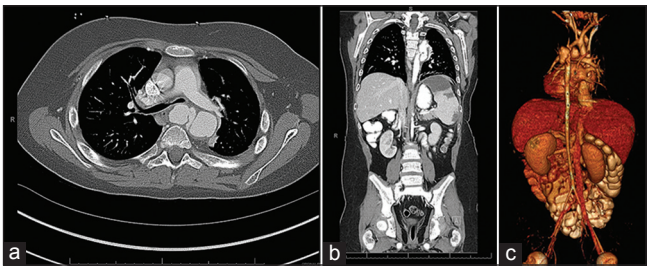

Figure 2: (a) Transverse view of chest CT with contrast, showing aneurysmal dilatation of the proximal descending aorta at the level of the origin of the left subclavian artery. Maximum dimensions of the multi-lobulated aneurysm are 4.4 cm in the transverse dimension and 4.2 cm in the craniocaudal dimension. No periaortic fluid collections. There is mass effect from the pseudoaneurysm with anterior displacement of the carina, (b) Coronal view of chest CT with contrast, showing aneurysmal dilation of the proximal descending aorta. Notice the different densities in the aneurysmal area, (c) 3-D reconstruction of chest CT with contrast, showing aneurysmal dilatation before surgery

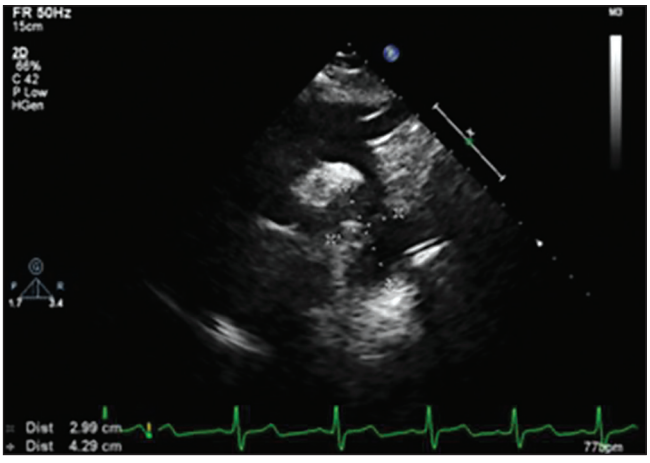

Figure 3: 2-D suprasternal long axis view of the aorta showing dimensions of the aneurysm

an 18 mm hemashield tube interposition graft was sewn to replace the excised segment of thoracic aorta. The total time on left heart bypass was 116 minutes, including 110 minutes of aortic cross clamp time. An intraoperative

biopsy of the aneurysm supported the diagnosis and a culture of the tissue showed one colony of gram positive rods and many white blood cells.

He was discharged after a postoperative period in which he showed significant anxiety and persistent pain of his legs and feet (normal imaging studies). Intravenous cefepime was continued at home for an additional six weeks. He had no recurrence of infection at one-year follow up and had normal inflammatory markers, cell counts and kidney function. CT scan at six months showed mild luminal narrowing of the thoracic aorta without signs of pseudoaneurysm, improvement of the splenic infarcts and scarring at the area of prior left kidney infarct. He continues to be treated for hypertension and his pain and cutaneous stigmata of endocarditis are now resolved.

DISCUSSION

The term "mycotic aneurysm" was initially used to describe aneurysms associated with bacterial invasion of the blood vessel wall by organisms other than syphilis[1] due to their appearance of "fresh fungus vegetations". Currently the term describes an aneurysm or pseudo-aneurysm,[2] caused by any infectious agent although the vast majority of them are caused by bacteria,[3] typically *Staphylococcus* and occasionally *Salmonella* species. If left untreated, a mycotic aneurysm associated with coarctation 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).[4]

In coarctation of the aorta there is proximal relative hypertension and distal turbulent, high velocity flow causing endothelial trauma predisposing to microbial colonization.[1] Aneurysms and pseudo-aneurysms have been described as late complications of coarctation of aorta that may develop in both repaired and un-repaired patients. In those undergoing repair, aneurysm formation at or near the site has been associated with both surgical and transcatheter relief of the coarctation.[5] 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.[6] Perhaps because of this, the finding is still uncommon in children.

Our patient had *Streptococcus viridans* confirmed by two blood cultures, which in addition to other streptococcal species and the HACEK organisms (*Hemophilus* species, *Actinobacillus actinomycetemcomitans*, *Cardiobacterium hominis*, *Eikenella corrodens*, and *Kingella* species) are somewhat more prevalent than other bacteria.[1] Though 50-75% of the patients have organisms isolated after early open repair of mycotic aneurysm,[7] we were not able to isolate the bacteria by culture of the surgical specimen.

A high index of suspicion is necessary to promptly diagnose this entity, especially in those with a history of coarctation repair. Physical examination and medical history are still the cornerstone of a clinical approach but appropriate selection of diagnostic modalities highly increases the likelihood of a successful outcome. Due to the variable presentation, as with our patient, with anemia, thrombocytopenia, fever and mediastinal mass it is not uncommon to entertain the possibility of malignancy. Echocardiography is a non-invasive and readily available diagnostic tool but in some cases (as with our first echocardiogram) it is not possible to make the diagnosis due to poor image quality. Cardiac CT and MRI are more sensitive in the diagnosis of mycotic aneurysms,[8] but may be less accessible. In our patient, chest CT and eventually echocardiography were fundamental to make the diagnosis.

The appropriate treatment of mycotic pseudoaneurysms 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.[8] Endovascular aneurysm repair (EVAR) 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. In some cases, EVAR is recommended as a bridge to open surgery.[7] 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; however, in this case, the pseudoaneurysm involved the base of the left subclavian artery, eliminating this potentially more attractive alternative. We weighed the risk of proceeding to surgery with elevated inflammatory markers and possible residual infection with the continued damage due to embolic events. Ideally, surgical reconstruction should be delayed until after serial negative cultures to prove sterility of the blood though there is no standardization of antibiotic therapy or duration. Some authors suggest lifelong antibiotics[9] while others suggest 6 weeks to 12 months of therapy.[10]

We followed our patient at 3, 6 and 12 months after discharge with inflammatory markers and at 6 months with chest CT, although some authors recommend more frequent imaging; every 3 months for the first year and annually thereafter.[10]

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