otherwise healthy (with the exception of poorly controlled hypertension) patient. However, the diagnosis of type A IMH versus type A classic aortic dissection could not be unequivocally established. Additional contrast administration was deemed ill advised, as was a magnetic resonance imaging examination in the setting of suspicion for acute aortic dissection. The patient was therefore taken to the operating room for a TEE examination under general anesthesia. The type A IMH was confirmed and massive LV hypertrophy was noted, and it was thought that the creatinine elevation was likely secondary to hypertensive nephropathy.

Although concerns for abdominal malperfusion in our patient were sorted out quickly, the presence of bowel ischemia would have considerably complicated the clinical scenario, as well as increased morbidity and mortality. Therefore, in that setting addressing the gut malperfusion first may be warranted. Percutaneous fenestration procedures have become well established and, depending on the resources of the institution, may be a valuable option. The presence of a hybrid operating room would offer a particular advantage in this case because it would minimize patient transport and unnecessary delay in addressing the ascending aortic component. In the spectrum of a pathologic process of the ascending aorta, type A IMH is relatively straightforward, which may translate into shorter bypass and circulatory arrest times. Several reports of using less than deep hypothermia have been reported, and perhaps consideration of modifying the temperature threshold for circulatory arrest may be entertained, depending on the speed and experience of the surgeon, in an attempt to minimize bypass time for rewarming [7]. If the cardiac problem is addressed first, immediate fenestration after the open heart operation to reperfuse the bowel may be of benefit. The availability of a hybrid operating room would allow the second procedure to be done safely without transporting a potentially hemodynamically unstable patient to another part of the hospital. If percutaneous fenestration procedures are not available, careful abdominal examinations and the need for open fenestration or visceral bypass are necessary. Finally, bowel resection should be used as a last resort for the primary treatment of the gut ischemia. Careful serial abdominal examinations are warranted regardless, because the need for bowel resection may become evident even if the bowel was reperfused up front.

Our case report exemplifies some of the questions that may arise during the evaluation of patients with type A IMH in the setting of acute type B aortic dissection. Although the management of type B dissection is medical, the presence or absence of malperfusion may affect the treatment algorithm of the associated type A IMH. Conversely, depending on the characteristics of the descending dissection, surgical intervention on the ascending component may increase the risk of paraplegia. This case report emphasizes that the management of multilevel aortic disease is complex and needs to be carefully individualized.

References

Intercostal Artery Aneurysmosis
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True intercostal artery aneurysms have been reported to occur in conjunction with neurofibromatosis, coarctation of the aorta, and Kawasaki disease. However, there has not been a previous report of a patient with intercostal artery aneurysmosis and no known or diagnosed...
associated condition. We describe the first such patient and review the literature.


In 1979, Blaisdell described the first case of visceral artery aneurysmosis occurring in a patient with aneurysms of the splenic, superior mesenteric, hepatic, and gastroduodenal arteries [1]. There has not been a similar correlate described in the thoracic cavity. Based on known genetic and pathologic associations, all patients with true intercostal artery aneurysms also have coarctation of the aorta, neurofibromatosis, or Kawasaki disease [2–5]. In the absence of these conditions, patients may have a single isolated intercostal artery aneurysm, although these tend to actually be pseudoaneurysms from previous operations, thoracentesis, or trauma [6–9]. A patient with more than 1 true intercostal artery aneurysm in the absence of linked diseases has not been reported. We present the case of a 47-year-old woman who presented to the emergency department with chest pain and was found to have a ruptured 6-cm intercostal artery aneurysm and intercostal artery aneurysmosis.

A 47-year-old African-American woman with hypertension and chronic obstructive pulmonary disease presented to the emergency department with severe chest and back pain. With her history of hypertension, a computed tomographic scan was ordered to look for an aortic dissection. The scan revealed a ruptured 6-cm right intercostal artery aneurysm, and a total of 7 intercostal aneurysms, 3 in the right side of the chest and 4 in the left side (Fig 1A-D).

In the operating room, a right lateral thoracotomy was performed and the ruptured 6-cm aneurysm was excised, along with a second smaller unruptured 4-cm aneurysm, and the feeding arteries were ligated (Fig 1). A third aneurysm was identified in the right side of the chest that was 2 cm in diameter and clotted; it was not excised.

The patient was stabilized in the intensive care unit and several days later was taken to the interventional radiology department, where 3 of the 4 intercostal artery aneurysms in the left side of the chest were embolized (Fig 2A-D). No spinal cord complications occurred. The fourth aneurysm in the left side of the chest could not be embolized. The patient made an uneventful recovery.

The pathologic examination showed normal elastin and collagen content of the arterial wall and was unrevealing (Fig 3A-D). Fungal, aerobic, and anaerobic culture results were negative. Alpha-1 antitrypsin levels were normal. The intercostal nerves did not show neurofibromas. Workup for connective tissue disease was also negative. To exclude systemic lupus erythematosus, the following tests were performed with these results: antinuclear antibody determination was negative, rheumatoid factor determination was negative, anti-dsDNA determination was negative, and the erythrocyte sedimentation rate was normal. To exclude Ehlers-Danlos syndrome, the pathologist specifically examined the histologic slides for the elastin and collagen content in the aneurysm wall, which was normal. He also did not see any evidence of cystic medial necrosis or medial mucoid degeneration. In addition, because Ehlers-Danlos syndrome is an autosomal dominant genetic disease, the absence of this disease in her parents greatly decreased the likelihood of it...
being present. With no antecedent history of uterine hemorrhage, dermal pathologic process, or other previous vascular events, Ehlers-Danlos syndrome was further ruled out. The patient had no problems with wound healing. Specific testing for the genetic mutation COL3A1 was not performed. The patient was doing very well and had returned to work by her 6-month follow-up.

She returned to the emergency department 7 months after her initial operation with back pain, and a repeated computed tomographic scan of the chest showed that the
remaining fourth aneurysm in the left side of the chest had increased from 3.7 cm to 5.1 cm over 6 months (Fig 2C, 2D). Attempts at embolization were again unsuccessful. The patient was taken to the operating room where the aneurysm was excised (Fig 3A, 3B). She again made an uneventful recovery and was discharged.

Comment

This is the first case report to document intercostal artery aneurysmosis. The patient had no connective tissue or systemic vascular disease, except that she had undergone endovascular repair of an infrarenal abdominal aortic aneurysm 3 years earlier. Patients with more than 1 intercostal artery aneurysm have associated diseases such as neurofibromatosis, coarctation of the aorta, or Kawasaki’s disease [2–5]. Although we expected to find neurofibromas along the intercostal nerves, this was not the case. In addition, close review of the computed tomographic scan of her chest did not reveal an undiagnosed coarctation. Intercostal artery aneurysm formation and hemorrhage have been associated with systemic lupus erythematous and Ehlers-Danlos syndrome, but our patient had neither of these based on further testing [10, 11]. Because specific genetic testing for COL3A1 for vascular Ehlers-Danlos syndrome was not performed, there is a possibility that the patient carries these germline mutations. Testing for TGFBR1 and TGFBR2 for Loeys-Dietz syndrome was performed and was negative.

Interestingly, the aneurysms were along multiple different intercostal arteries, including the lower ones. With coarctation of the aorta, the aneurysms involve the upper intercostal arteries. There is a single case report in the literature of a mycotic intercostal artery pseudoaneurysm, but in our patient the fungal culture results were negative and she did not have any evidence of a systemic infection [12].

Endovascular management of these aneurysms has shown success in the small number of patients reported in the literature [2, 6, 8, 13–15]. We also had success using endovascular embolization to occlude flow into several of the aneurysms in our patient. Unfortunately, the feeding intercostal arteries tend to be elevated and stretched over the aneurysmal sac, which can make embolization difficult. Surgical repair was, and will remain, a safe alternative approach if embolization is not successful.

References


Cystic Atrioventricular Node Tumor Excision by Minimally Invasive Surgery

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Cystic tumor in the atrioventricular (AV) node region is a rare cardiac primary tumor that can lead to sudden death. Our patient was a 43-year-old woman who presented with dyspnea and a first-degree AV block seen on electrocardiography. Echocardiography revealed a cystic mass attached to the inferior portion of the interatrial septum. After surgical excision of the mass, placement of a permanent pacemaker was required for complete heart blockage. Histopathologic examination revealed the mass to be a cystic tumor of the AV node region. To our knowledge, this is the first report of this condition