



Endovascular Repair for Primary Adult Coarctation of the Aorta Complicated with Acute Epidural Hematoma Leading to Paraplegia: A Case Report

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The incidence of neurovascular complications with paraplegia in patients of coarctation of the aorta (CoA) is very rare, and the prognosis of the outcome is poor. The traditional standardized management for the aortic coarctation is surgical repair, which can be associated with considerable intraoperative risk and postoperative morbidity. In this article, we present a case of diagnosis of the primary CoA complicated with acute epidural hematoma and paraplegia treated with endovascular repair with balloon angioplasty and stent placement successfully.

Key words: Coarctation of the aorta, endovascular management, case report, acute epidural hematoma, paraplegia

INTRODUCTION

Coarctation of the aorta (CoA) is one of the common congenital cardiovascular diseases. CoA can be diagnosed over a wide range of ages, degrees of severity, and with various presentations. Although the most common presenting age of aortic coarctation is 3–6 months, only a small proportion of adolescents and adults can be diagnosed with primary aortic coarctation as a first-time diagnosis.¹ In the undiagnosed and untreated older age group, primary adult aortic coarctation can be recognized as an incidental finding by a primary care physician who notices a heart murmur or decreased femoral pulses as part of a routine annual physical examination.²

Like the neonatal presentation, the standard treatment of adult aortic coarctation is surgical repair of the narrow segment. However, these adult patients have developed significant collateralization through intercostal arteries and are at increased surgical risk secondary to bleeding complications. In an effort to minimize the surgical risk and shorten procedural recovery times, a catheter-based approach could be an alternative treatment in adult aortic coarctation cases. Nevertheless, it has been successfully used in pediatric and adolescent patients, and little information exists regarding the endovascular repair in adults.

We report the case of a young adult who developed acute nontraumatic paraplegia. After a series of radiological examinations, CoA complicated with spontaneous spinal epidural hemorrhage was proved, and he was treated successfully.

CASE REPORT

A 21-year-old man who was a military soldier with a history of hypertension and mitral valve prolapse without regular outpatient department follow-up and the past medical treatment workup was due to the sudden onset shortness of breath. He had severe sharp pain from the neck to the lower back and experienced with weakness and numbness of his lower extremities.

In our emergent department, his blood pressure was 155/92 mmHg, and electrocardiogram revealed sinus tachycardia. He had an increased respiratory rate with oxygen saturation around 97%–99%. On physical examination, his consciousness was alert with a Glasgow Coma Scale score of 15. Although he had no petechia, ecchymosis, or contusion over the trunk or extremities, he had local tenderness from the

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cervical to the lumbar spinal region. He also had mid-systolic click murmur through heart auscultation, bilateral clear breathing sounds, and diminished pulse of the bilateral femoral artery, hypoesthesia below the T3 dermatome, poor joint position but no rigidity or spasticity. In addition, the tendon reflexes were absent below the T3 level; the initial presentation of paraparesis progressing to paraplegia with a muscle power score of 0 for the bilateral lower limbs and 5 for the bilateral upper limbs was observed, and a positive test of bilateral Babinski reflex was taken. Furthermore, the urine incontinence and loss of anal sphincter tone through digital examination were found.

Echocardiography revealed a normal chamber size, minimal mitral regurgitation, mild tricuspid regurgitation, mild aortic regurgitation, minimal pulmonary regurgitation, and normal left and right ventricle function. No aortic stenosis, bicuspid aortic valve, or ventricular septal defects were noted. Plain chest radiography showed bilateral upper rib notching and the characteristic 3 sign at the lateral contour of the aortic arch. Hemocoagulation tests, including protein C, protein S, and antithrombin III, were negative. Examination of the lumbar puncture was performed, and subarachnoid hemorrhage was excluded. Bacteria culture of the cerebrospinal fluid showed no growth. Computerized tomography scan of the chest with contrast medium enhancement demonstrated CoA with stenosis and aortic isthmus and several dilated collateral vessels in the bilateral upper intercostal regions [Figure 1]. No other congenital heart disease or aortic dissection was noted.

Computerized tomography scan of the brain without contrast enhancement showed normal gray and white matter attenuation without evidence of focal intracranial aneurysm formation. Magnetic resonance imaging of the thoracic spine with a gadolinium injection disclosed a fusiform vascular

lesion measuring 1.9 cm × 0.9 cm × 4.4 cm (length × width × height) with hemorrhage involving the anterior epidural space from the C7 to T2 level, causing compression and ischemic change of the spinal cord; it was more severe at the T1 and T2 levels [Figure 2].

We sufficiently explained the final cause of acutely developed paralysis to the patient and his family and then discussed the choice of treatment: conventional surgery or balloon angioplasty and stenting. The patient underwent surgery using percutaneous endovascular balloon angioplasty with Bard Conquest PTA balloon (Bard, Tempe, AZ, USA) and stent placement with a 30 mm × 140 mm Cook Zenith TX2 TAA endovascular graft (Cook, Bloomington, IN, USA).

Postoperative computerized tomography scan of the chest with contrast enhancement demonstrated one metal stent deployed in the narrowest location of the aortic coarctation, and normalized distal run-off was confirmed [Figure 3]. Postoperatively, his systolic blood pressure was within the normal range without antihypertensive medicine treatment, and a significant improvement in the upper to lower extremities systolic pressure gradient was noted; in addition, no delay in the brachial-femoral pulse was present, and pulsation of the bilateral dorsalis pedis was detectable. Since the patient had a relatively stable hemodynamic and postoperative condition for 1 year, he was referred to a special neurological and neurosurgery institute for further treatment of the acute epidural hematoma-related paraplegia.

DISCUSSION

CoA is a relatively common defect that accounts for 4%–6% of all congenital heart defects with a reported prevalence of



Figure 1: Preoperative chest computed tomography three-dimensional reconstruction revealing coarctation of aorta with stenosis, aortic isthmus, and several collateral vessels in the bilateral upper intercostal regions

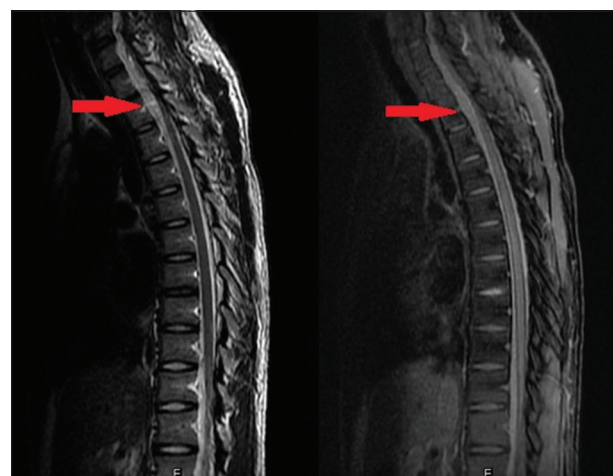


Figure 2: Magnetic resonance image with contrast of the thoracic spine (left: T2 frFSE; right: T1 FLAIR, sagittal view) showing a fusiform vascular lesion measuring 1.9 cm × 0.9 cm × 4.4 cm with hemorrhage in the epidural space from C7 to T2 level, causing compression and ischemic change in the spinal cord

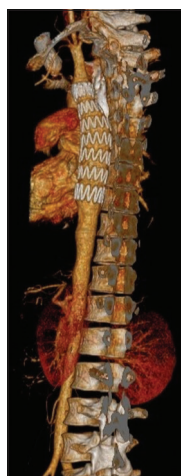


Figure 3: Postoperative chest computed tomography three-dimensional reconstruction revealing full expansion over the narrowest site of coarctation after balloon angioplasty and implantation with the self-expandable covered stent

about 4/10,000 live births.^{3,4} In delayed diagnosed adults, most patients are asymptomatic without developed overt congestive heart failure. Diagnosis is often made by incidental findings during radiological examinations for other medical problems. Historically, the classic clinical presentation is hypertension or a heart murmur. However, other symptoms include a headache, exertional claudication, and subtly diminished or delayed femoral pulses, which are not easily recognized by primary care physicians.^{5,6} In young individuals with systemic hypertension such as in our patient, careful assessment of the pulsation in the brachial-femoral delay or the gradient pressure between the upper arms and lower legs and further evaluation with color and continuous-wave Doppler echocardiography are necessary to avoid missing the diagnosis.

Development of collateral circulations connecting the pre- and post-stenotic parts with the thoracic aorta is one of the typical signs of coarctation.

In addition, collateral vessels may form aneurysmal dilatations due to muscle and elastic tissue deficiency on the background of longstanding arterial hypertension, which may cause these collateral vessels to rupture.^{2,7} To the best of our knowledge, the complication of acute nontraumatic epidural hemorrhage with paraplegia related to spinal cord compression, such as in our case, has been rarely reported. Except for neurological problems, the differential diagnosis of aortic coarctation should not be ruled out. Further physical and radiological examinations including checking the pressure gradient of the upper and lower extremities and performing computerized tomographic angiography are recommended to confirm the diagnosis. These adult patients with CoA have developed significant collateralization through intercostal arteries and are at increased surgical risk secondary to bleeding

complications. Because of this consideration and discussion with the neurosurgeon, prior to evacuate the spinal epidural hematoma, to restore the main blood stream in the thoracic aorta blocked by the coarctation, and to reduce the perfusion of intercostal collateral artery into the spinal epidural hematoma should be rational and have better clinical outcome.⁸⁻¹⁰

Standard management for CoA is surgical repair, which has proven to be an effective management for native CoA in neonates and infant stage but can be associated with considerable morbidity and operative risk. Over the past two decades, advances in operator experiences and technological evolvement of balloon and stent have improved the success rate and offered another safe and feasible solution for treatment of CoA. In patients with CoA who are at high surgical risk, balloon angioplasty offers a less invasive and equally effective method. Although balloon angioplasty has been successfully used in pediatric and adolescent patients with CoA, stent grafting is usually used for postoperative complications and has the unique property of high radial force to avoid elastic recoil to reduce the rate of reinterventions.¹¹⁻¹³ The most serious complication of angioplasty is rupture of the aorta, which although rare, may lead to severe hemothorax with circulatory collapse and death; however, the use of a stent graft at implantation has been proposed for reducing the risk of aortic wall complications such as aneurysm formation, aortic dissection, or vessel wall rupture.¹⁴⁻¹⁶ Therefore, we performed balloon angiography with stent-graft placement in our patient and treated CoA successfully. Balloon angioplasty with stent-graft implantation is another alternative choice for treating primary coarctation in adults.

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Conflicts of interest

There are no conflicts of interest.

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