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Chest Pain and Sudden-Onset Paraplegia at the Emergency Department: An Uncommon Presentation

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Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 45
Final Diagnosis: Acute coarctation with spinal epidural hemorrhage
Symptoms: Chest pain with bilateral lower limbs paraplegia
Medication: —
Clinical Procedure: Percutaneous transluminal angioplasty and thoracic endovascular repair followed by bilateral hemilaminectomy
Specialty: Surgery

Objective: Rare disease





Background: Coarctation of the aorta is characterized by narrowing of the descending aorta. The narrowing typically is at the isthmus, the segment just distal to the left subclavian artery. Adults with undiagnosed aortic coarctation are asymptomatic or may present with nonspecific hypertension. We present a case that highlights the uncommon complication of aortic coarctation with spinal compression syndrome.

Case Report: A 45-year-old male presented to the emergency department (ED) with acute-onset chest pain; he experienced urinary incontinence and bilateral lower limb weakness during his ED visit. Chest CT showed coarctation of the aorta and MRI of the spine showed an epidural nodular lesion. He received emergency aortic stent placement surgery, followed by successful hematoma removal and was discharged with residual lower-extremity paraplegia.

Conclusions: Chest pain with lower limb paraplegia presentation should consider aortic coarctation complicated with spinal hemorrhage as a possible cause.

MeSH Keywords: Aortic Coarctation • Chest Pain • Hematoma, Epidural, Spinal

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/903503>

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Background

Chest pain is a common chief complaint of patients visiting the Emergency Department (ED); however, the diagnosis of aortic coarctation is rare and not usually considered a common etiology of spinal hemorrhage leading to paraplegia. Adults with undiagnosed aortic coarctation are asymptomatic or may present with nonspecific hypertension. Aortic coarctation with secondary intraspinal epidural hemorrhage is extremely uncommon. A PubMed literature search revealed only two cases of spinal epidural hemorrhage and three cases of spinal subarachnoid hemorrhage associated with aortic coarctation (Table 1) [1–5]. We describe a case of chest pain followed by paraplegia at the time of ED presentation. Computed tomography (CT) and magnetic resonance imaging (MRI) of the spine played a critical role in the diagnosis.

Case Report

A 45-year-old male presented to the ED with acute-onset chest tightness since the previous night. The patient had a medical history of hypertension, for which he took regular medication. He stated that he had not started any new medications and denied any occurrence of chest trauma. On examination, he was afebrile with a heart rate of 80 beats/minute and blood pressure of 202/186 mm Hg; a higher systolic blood pressure was observed over the upper limbs than over the lower limbs. He was in mild distress due to pain with moderate chest tenderness. The results for complete blood cell counts, hemocoagulation, and biochemistry tests were normal. Electrocardiography showed normal sinus rhythm and left ventricular hypertrophy.

Chest radiography showed no significant abnormalities. CT showed a marked narrowing of the aortic arch with several engorged intercostal arteries (Figure 1), suggestive of coarctation of the aorta (Figure 2). During the CT, the patient experienced urinary incontinence and sudden onset of bilateral lower limb weakness. Lower-extremity motor strength was 2/5 bilaterally. MRI of the spine showed an epidural nodular lesion, approximately 6.2 cm in length and 7 mm in thickness, located in the anterior aspect of the spinal canal, at the levels of C6–T2. An epidural hematoma was suspected (Figure 3).

The patient underwent emergency percutaneous transluminal angioplasty with balloon dilation and thoracic endovascular repair with stent placement in the descending aortic coarctation (Figures 4, 5), followed by hematoma removal at C6–T3. Two weeks after aortic stent coarctoplasty, the patient's hypoesthesia improved with gradual recovery of muscle strength in both lower limbs. The patient was ultimately discharged to a rehabilitation facility with residual lower-extremity paraplegia. One month later, he was able to ambulate with the assistance of a walker with near-normal motor function in both lower extremities.

Discussion

Our patient presented to the ED with severe chest pain. In general, the suspicion of a possible aortic dissection diagnosis is usually raised by the ED physician based on a patient's clinical presentation [6,7]. Based on our patient's normal cardiac biomarkers and electrocardiography showing normal sinus rhythm without clear changes of ischemia, we initially

Table 1. Reports of aortic coarctation causing spinal hemorrhage in worldwide literature.

Year	Age (years)	Gender	Onset time of paraplegia	Complication	Spinal level	Treatment	Outcome	Reference
1967	44	Female	N/A	Spinal SAH	T7-T10	Laminectomy only	Neurologic defect	[4]
1973	40	Male	N/A	Spinal SAH	C6–C7	Conservative treatment	Death due to second infection	[3]
2001	20	Male	N/A	Spinal EDH	C1–T7	Surgical implantation of an oval Gore-Tex patch	No neurologic defect	[1]
2014	45	Male	N/A	Spinal SAH	C2–T4	Conservative treatment	Neurologic defect	[5]
2016	21	Male	N/A	Spinal EDH	C7–T2	Conservative treatment	Neurologic defect	[2]
2016	45	Male	6 hours	Spinal EDH	C6–T3	Aortic stent coarctoplasty → laminectomy	Near normal motor function Paresis below T4 dermatome	Present case

SAH – subarachnoid hemorrhage; EDH – epidural hemorrhage; N/A – not available.



Figure 1. Computed tomography angiogram coronal view demonstrated several engorged intercostal arteries and mammary arteries.



Figure 2. Computed tomography angiogram sagittal view demonstrated aortic coarctation with stenosis over the aortic isthmus.

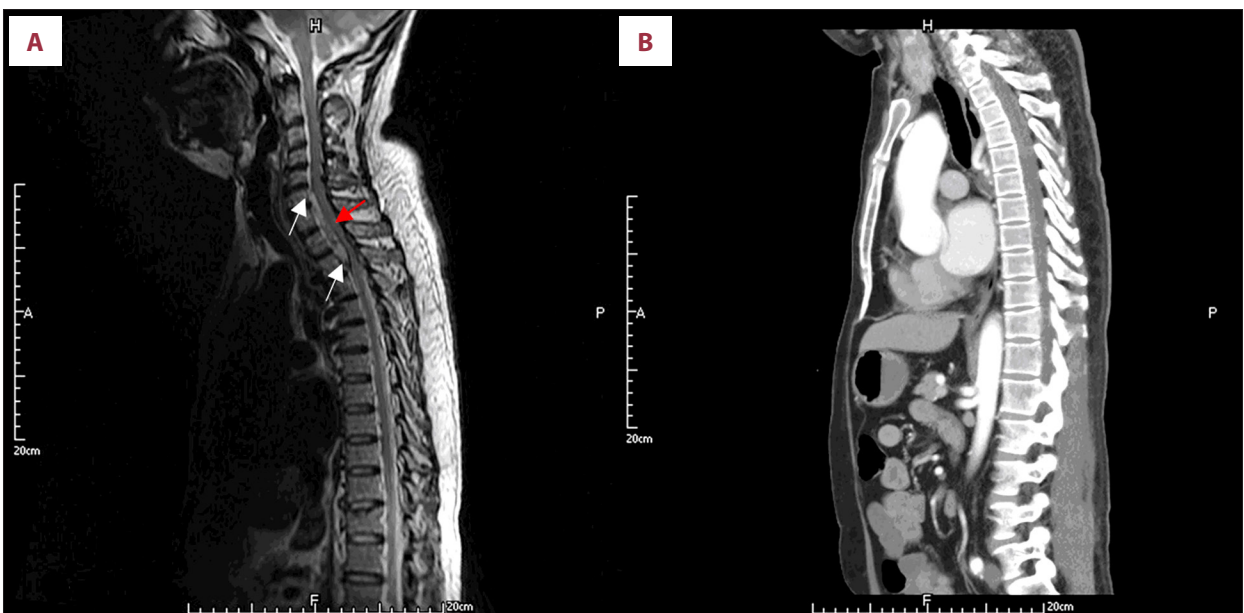


Figure 3. (A) A T2 phase sagittal view showing a fusiform lesion involving the anterior epidural space from C6 to T2 level (white arrow) and the squeezed medullary cord (red arrow). (B) Computed tomography angiogram sagittal view without obvious fusiform lesion over the same spinal level.

performed a chest computed tomography angiography (CTA) to rule out this critical condition as well as rule out other potential etiologies [7]. The chest CT showed a narrowing of the aortic isthmus without dissection or intramural hematoma (Figure 2). Once the patient's condition worsened, with the

sudden onset of lower limb weakness, we suspected a complicated aortic coarctation.

The proportion of aortic coarctation in men is slightly higher than in women, and overall accounts for approximately 5–8%

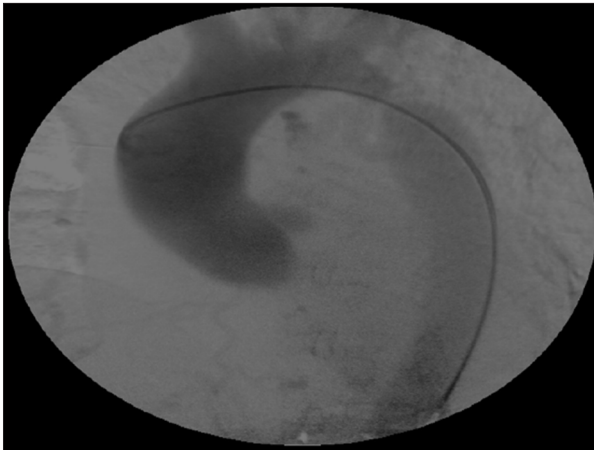


Figure 4. Percutaneous transluminal angioplasty to descending aortic coarctation.

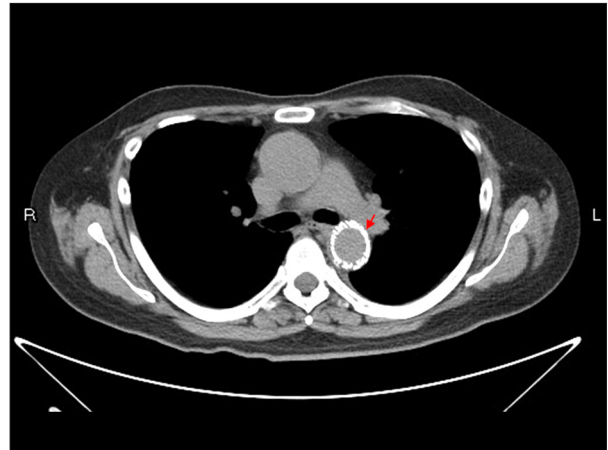


Figure 5. Computed tomography angiogram axial view after transluminal angioplasty to descending aorta.

of all patients with congenital cardiovascular diseases [8,9]. The mean survival of patients with untreated aortic coarctation is 35 years, with a 75% mortality rate by the age of 46 years [10]. The indications for thoracic endovascular repair of an aortic coarctation include hypertension, a peak instantaneous pressure gradient across the coarctation ≥ 20 mm Hg or imaging evidence of collateral circulation [10,11]. Aortic coarctation can lead to the mechanical obstruction of blood flow by the narrowed aorta, and left ventricular afterload increases with systolic blood pressure elevation in the upper extremities, and low or unobtainable blood pressure in the lower extremities. In the case of long-term arterial hypertension, the collateral vessels of the collateral circulation that connects between the prestenotic and poststenotic thoracic aorta may develop aneurysmal dilatations because of deficiencies in muscle and elastic tissues [2,12]. Aortic coarctation with abnormally dilated spinal arteries or collateral vessels may increase the risk of spontaneous hemorrhage, including intracranial subarachnoid hemorrhage [2,3,12]. However, spinal hemorrhage is infrequently described in the literature. Our literature search revealed only five prior cases of aortic coarctation with spinal hemorrhage. The most reported complications are intracranial aneurysm, occurring in approximately 12% of untreated patients, and intracranial subarachnoid hemorrhage, accounting for 7% of deaths in such cases [3,6].

References:

1. Zizka J, Elias P, Michl A et al: Extensive spinal epidural hematoma: A rare complication of aortic coarctation. *Eur Radiol*, 2001; 11: 1254–58
2. Yi-Da T, Chia-Ching H, Wen-IL, Sy-Jou C: Paraplegia caused by aortic coarctation complicated with spinal epidural hemorrhage. *Am J Emerg Med*, 2016; 34: 680.e1–2
3. Banna MM, Rose PG, Pearce GW: Coarctation of the aorta as a cause of spinal subarachnoid hemorrhage. Case report. *J Neurosurg*, 1973; 39: 761–63
4. Watsan AB: Spinal subarachnoid hemorrhage in patient with coarctation of aorta. *Br Med J*, 1967; 4: 278–79
5. Chang-Bum P, Dae-Jean J, Min-Ki K, Sang-Hyun K: Paraplegia due to acute aortic coarctation and occlusion. *J Korean Neurosurg*, 2014; 55: 156–59
6. Domenicucci M, Ramieri A, Paolini S et al: Spinal subarachnoid hematomas: Our experience and literature review. *Acta Neurochir (Wien)*, 2005; 147: 741–50 (discussion 750)
7. Aram B, Stefania LB, Krishan P et al: An emotional stress as a trigger for reverse Takotsubo cardiomyopathy: A case report and literature review. *Am J Case Rep*, 2016; 17: 137–42
8. Swan L, Wilson N, Houston AB et al: The long-term management of the patient with an aortic coarctation repair. *Eur Heart J*, 1998; 19: 382–86

Our patient presented with epidural spinal hematoma, which led to the sudden onset of acute and severe back pain. The sudden onset of motor and sensory deficits was observed long after chest tightness presentation. Immediate surgical repair was indicated for aortic coarctation. Cardiac catheterization can widen a narrowed aorta from the inside by balloon dilation or stent placement [13]. Our patient underwent surgical repair, balloon dilation, and stent placement simultaneously. He underwent surgery immediately, resulting in a successful recovery of his muscle strength two weeks after the event. Very few similar cases of aortic coarctation complicated with spinal hemorrhage have been reported, and even fewer cases have reported successful immediate emergency surgery.

Conclusions

We report here on an uncommon presentation of aortic coarctation causing spinal hematoma. This case suggests the need to increase awareness among emergency physicians who may be faced with a similar case of atypical paraplegia. The chest CT and spine MRI were critical for the timely diagnosis and management of our patients with atypical presentation.

9. Talner CN: Report of the New England Regional Infant Cardiac Program, by Donald C. Fyler, MD, *Pediatrics*, 1980; 65(Suppl): 375–461. *Pediatrics*, 1998; 102: 258–59
10. Warnes CA, Williams RG, Bashore TM et al: ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). *Circulation*, 2008; 118: e714–833
11. Attenhofer Jost CH, Schaff HV, Connolly HM et al: Spectrum of reoperations after repair of aortic coarctation: Importance of an individualized approach because of coexistent cardiovascular disease. *Mayo Clin Proc*, 2002; 77: 646–53
12. Neji H, Attia M, Zaouali S et al: Mid-arch aortic coarctation: An uncommon presentation. *J Cardiovasc Comput Tomogr*, 2017 [Epub ahead of print]
13. O'Brien P, Marshall AC: Coarctation of the aorta. *Circulation*, 2015; 131: e363–65