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

E Feng Han Chiu
D Shih Hung Tsai
BC Cheng Hsuan Ho

Corresponding Author: Cheng Hsuan Ho, e-mail: erdoctorho@gmail.com

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

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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 hypoaesthesia 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      | Laminctomy 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.
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%
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 \(\geq 20 \text{ mm Hg}\) 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 repair 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 \[3,6\].

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.

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