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Atypical Presentation of Silent Aortic Coarctation in an Adolescent Female Presenting with Sudden-Onset Paraplegia

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ABSTRACT

Background: Aortic coarctation (CoA) is a prevalent congenital heart anomaly, accounting for 5-8% of congenital heart diseases and occurring at an incidence of 4 per 10,000 live births. While its presentation varies, ranging from cardiovascular collapse in neonates with severe CoA to chronic hypertension in adults, spinal neurovascular complications are rare. The formation of collateral arteries is a compensatory mechanism for obstructed aortic arch blood flow. Rarely, intraspinal collaterals and hypertrophied spinal arteries may contribute to compressive myelopathy.
Objective: This study aimed to report an atypical presentation of previously silent aortic coarctation manifesting as sudden-onset paraplegia due to spinal epidural hematoma from ruptured collateral vessels.
Methods: This case report detailed the clinical presentation, diagnostic workup, and management of a 13-years-old female. Diagnostic tools included spinal cord magnetic resonance imaging (MRI) with contrast and CT aortography.
Results: A 13-years-old female presented with sudden-onset bilateral lower limb symmetric paralysis. MRI revealed an abnormal intramedullary cord signal with a small epidural hematoma and multiple epidural vessels. CT aortography confirmed significant postductal aortic coarctation distal to the left subclavian artery, with extensive perivertebral and dilated intraspinal/paraspinal collateral arteries. One of these engorged collateral vessels ruptured, causing spinal cord compression and acute paraplegia. A Cheatham-Platinum stent was successfully implanted.
Conclusion: This case highlighted that previously undiagnosed aortic coarctation can present atypically with acute paraplegia due to spinal epidural hematoma from ruptured collateral vessels. Comprehensive vascular evaluation, including CT angiography, is crucial for accurate diagnosis and timely management in such rare presentations.
Keywords: Aortic coarctation, Paraplegia, Epidural hematoma, Collateral vessels, Spinal cord compression.

INTRODUCTION

Aortic coarctation (CoA) stands as one of the most prevalent congenital heart abnormalities (CHD), representing a significant component within the spectrum of congenital cardiac defects. This condition is responsible for an estimated 5% to 8% of all diagnosed CHD cases [1]. Epidemiologically, CoA manifests at an incidence rate of approximately 4 cases per 10,000 live births [2], underscoring its notable occurrence within the neonatal population.

Numerous variables, such as the severity of the CoA and the existence of concomitant cardiac diseases, especially those related to left-sided heart blockage, might affect the clinical presentation. Neonates may exhibit cardiovascular collapse when they have severe CoA, especially as their ducts close [3].

On the other end of the range, adults who exhibit symptoms of chronic secondary hypertension may be diagnosed with CoA [4]. When blood flow via the aortic arch is obstructed, collateral arteries form, enabling blood to move from high-pressure to low-pressure regions. The branches of the subclavian arteries above the obstruction are where collateral vessels most commonly arise, delivering blood to the tissues beneath the obstruction, in our case one of these aberrant vessels ruptured, and epidural hematoma was a major cause of spinal cord compression, leading to neurological deficits such as paraplegia and sensory loss [5].

Recent investigations have elucidated that the development of compressive myelopathy in a limited subset of patients with aortic coarctation can be attributed to distinct vascular abnormalities within the spinal canal. Specifically, these studies have demonstrated that the presence of intraspinal collaterals and a hypertrophied anterior spinal artery are anatomical factors contributing to this neurological complication in a small, though clinically significant, number of affected individuals [6]. This highlights an uncommon yet important mechanism of spinal cord compression in this patient population.

CASE REPORT

A 13-year-old female came at our Hospital's Emergency Room with bilateral lower limb symmetric paralysis that had developed suddenly, with grade 0 power, absent tendon reflexes and she had loss of pain and temperature sensation below the C7–T1 level. The patient had no additional contributing medical history, such as coagulopathy, fever, severe coughing fits, or any other long-term medical or surgical co-morbidities. In order to determine the reason of her paraplegia, she was referred for spinal cord magnetic resonance imaging (MRI) with contrast which revealed abnormal intramedullary cord signal with small epidural hematoma, along with multiple epidural vessels (figure 1). Vascular malformation was suspected and CT angiography was requested.

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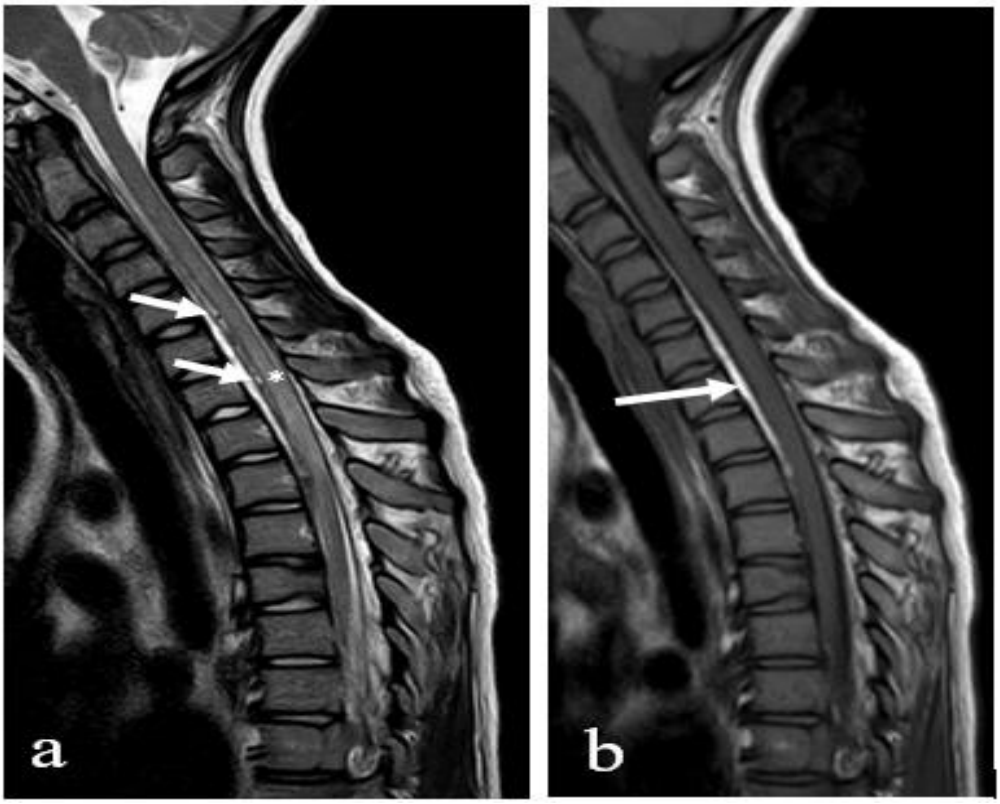

Figure (1): MRI of the cervicodorsal spine a) Sagittal T2 WI image showed increased cord signal (asterisk) likely congestive myelopathy with multiple engorge vessels (arrows) (b) Sagittal T1WI showed small epidural hematoma (long arrow) exhibiting bright signal.

Further assessment with CT aortography revealed significant postductal aortic coarctation distal to the origin of left subclavian artery associated with multiple peri-vertebral collateral with dilated intra-spinal and para-spinal arteries (figure 2). She was sent to the cardiovascular surgery unit for additional care. A Cheatham-Platinum (CP) stent 45 mm was manually crimped on Atlas balloon 16 mm/4cm and was implanted below the left subclavian.

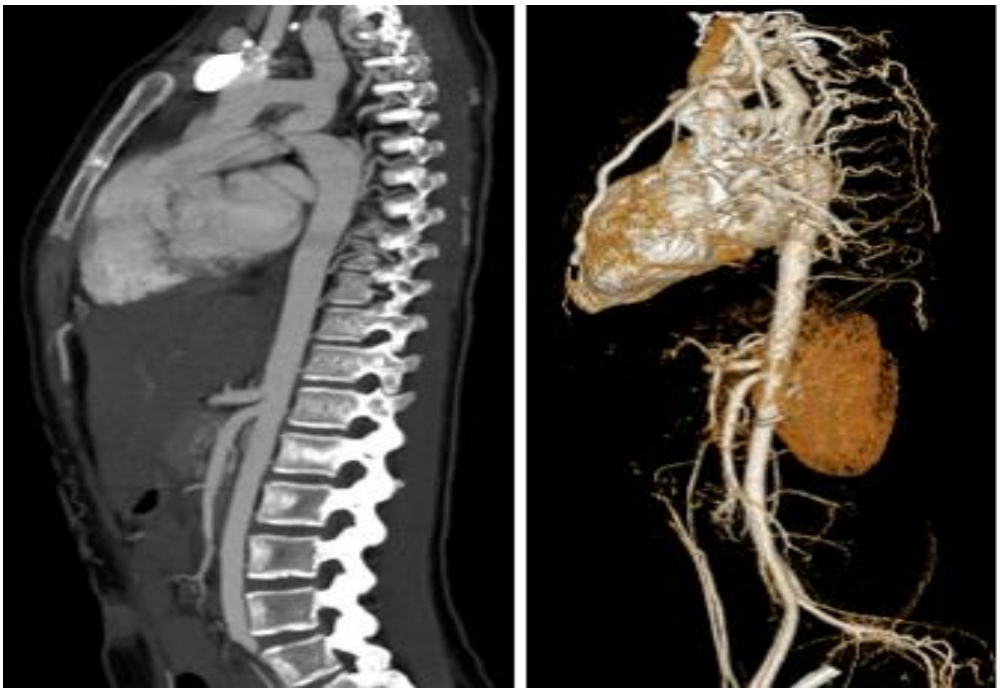

Figure (2): CT Aortography a) sagittal reformat, b 3D reconstruction showed aortic coarctation (short arrow) distal to the left subclavian artery with prominent internal mammary (long arrow), intercostal arteries, and multiple peri-vertebral collateral (double arrows).

DISCUSSION

Patients diagnosed with aortic coarctation demonstrate a significant susceptibility to a range of serious neurovascular complications. Among these, the development of intracranial hemorrhage stemming from an aneurysm is observed in approximately 12% of cases that remain untreated. Furthermore, intracranial subarachnoid hemorrhage represents a critical concern, accounting for a notable 7% of mortalities in patients afflicted with aortic coarctation [7]. In contrast to the relatively higher incidence of intracranial events, spinal hemorrhage as a complication of coarctation is an exceptionally rare occurrence, with only a limited number of isolated cases having been documented in the global medical literature [8]. In cases of post-ductal aortic coarctation, a critical physiological response involves the compensatory collateral shunting of blood. This typically occurs through the significant dilation of specific arterial networks, primarily the intercostal and internal mammary arteries [9]. This adaptive mechanism effectively serves to decompress the high-pressure proximal aortic segment, located before the coarctation, by rerouting blood flow to the lower-pressure distal segment, beyond the narrowed region. As the systemic pressure within the proximal aortic segment consequently increases due to the obstruction, certain pre-existing embryologic vessels, which might otherwise remain less prominent, undergo compensatory hypertrophy to further facilitate this vital collateral circulation.

In rare cases, the spinal cord vasculature may also enlarge and collaterals may form inside the spinal canal. These vessels have the potential to be hazardous when they are present, mostly because of the risk of bleeding from channel rupture, which is the scenario of our patient. Nevertheless, in very rare instances, these arteries may lead to spinal cord compression. This compression is most likely the result of continuous pulsing under pressure in a closed space, even though these arteries almost ever reach aneurysmal size [10].

Moreover, chronic myelopathy may arise from lympho-venous congestion brought on by elevated intraspinal canal pressure. Besides the more common mechanisms, other less frequent causes of primary compressive myelopathy in the context of aortic coarctation can include the dislodgement of microthrombi from dilated collateral vessels or the presence of a spinal artery aneurysm [11].

This case underscored the critical importance of comprehensive vascular evaluation in patients presenting with spinal epidural hematomas, particularly from an imaging standpoint. It cautions against premature diagnosis of spinal arteriovenous malformations without thorough investigation. In our patient, a previously asymptomatic aortic coarctation led to the development of markedly engorged collateral vessels, one of which ruptured, resulting in a small epidural hematoma evident on MRI. This hematoma caused spinal cord compression and acute paraplegia, serving as the first clinical indication of the underlying, silent coarctation. While MRI initially revealed spinal cord involvement, definitive diagnosis depended on CT angiography, which clearly delineated the aortic coarctation and abnormal collateral vessels. This case highlighted the essential role of CT angiography in evaluating sudden-onset paraplegia of suspected vascular origin—especially when epidural hematoma and collateral circulation are present—to accurately visualize the aorta and guide appropriate management.

CONCLUSION

We reported a rare case of acute paraplegia caused by spinal cord compression secondary to rupture of engorged collateral vessels, which developed as a consequence of previously undiagnosed aortic coarctation. In patients presenting with sudden lower limb weakness—especially without trauma—vascular etiologies, including silent coarctation with collateral formation, should be considered. Early recognition and appropriate management are essential to improve outcomes.

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