Atypical complications of aortic intramural hematoma: Paraplegia resulting from spinal cord infarction

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Neurological complications of acute aortic syndromes (AAS) are common and occur in 17–40% of patients, but spinal cord infarction (SCI) is very rare and only seen in a few percent of all AAS patients [1]. Herein, we report a case of acute aortic intramural hematoma (IMH) extending from the ascending to suprarenal aorta complicated by SCI manifesting as bilateral paraplegia and paresthesia.

A 57-year-old man presented with worsening epigastric burning pain radiating to his upper back. On his arrival to the emergency room (ER), blood pressure was 120/95 mm Hg without significant difference between arms, heart rate was 62 beats/min, and there were no significant heart murmurs or lung crackles. He also complained of mild numbness in his bilateral lower extremities which started at the onset of epigastric pain a few days ago but denied progressive weakness or urinary retention. The electrocardiogram (ECG) showed ST-elevation in the precordial leads and the initial troponin was mildly elevated at 0.19 ng/ml (N: <0.03 ng/ml). Diagnostic coronary angiogram via right radial artery did not reveal obstructive coronary artery disease and the invasive aortography with runoff did not show any narrowing concerning for AAS [Fig. A].

Thirty minutes after the aortography, he acutely developed complete paraplegia, paresthesia, and areflexia in the bilateral lower extremities. Given concern for acute spinal cord infarction or compression, neurosurgery first recommended whole spine magnetic resonance imaging (MRI) to exclude compressive pathology. MRI showed increased T2 signal intensity and diffusion restriction predominantly involving the central gray matter of the spinal cord extending from the T4 to T11 level [Fig. B]. Incidentally, there was also an enlargement of the descending aorta with an abnormal signal intensity within its wall [Fig. B]. These findings suggested thoracic SCI and possible thoracic aortic aneurysm. His computed tomography angiogram (CTA) demonstrated a small volume of pericardial effusion and acute IMH extending from the ascending to suprarenal aorta without great vessel occlusion, flap, extravasation of contrast, or hemothorax [Fig. C]. Based on such incidental AAS evidence and normal anatomy in the coronary angiography, the initial ECG changes most likely resulted from functional coronary artery occlusion due to retrograde extension of AAS toward the ascending aorta.

Because the CTA already demonstrated a thrombosed false lumen without impending rupture or extravasation, thoracic endovascular aortic repair (TEVAR) was not indicated. By the time the CTA was obtained, it had already been several hours since the onset of acute paraplegia. Based on his imaging and progressive paraplegia, his SCI was considered to be in an irreversible phase and cerebrospinal fluid drainage (CSFD) was not indicated given poor procedural benefit. His mental status continued to worsen without developing other organ manifestations and he expired on the seventh day.

The patient’s autopsy identified an aortic dissection from the ascending to suprarenal aorta and there was 87 ml of blood in the pericardial cavity. However, there was no significant hemothorax or mediastinal hemorrhage and the exact site of entry tear was undetermined due to the extensive damage to the aorta. Based on these findings, his primary cause of death was suspected acute cardiac tamponade.

This case presented two challenges of diagnosing highly suspected acute coronary syndrome (ACS) and identifying SCI with unexpected AAS. Invasive angiography allows percutaneous intervention of the coronary arteries or branches of the aorta at the time of diagnosis [2]. However, as our case demonstrated, aortography can fail to diagnose IMH when there is a lack of luminal disruption. Thus, other modalities such as echocardiography, CTA, or MRI are required for diagnosis, but their time-consuming nature can jeopardize patients’ lives [2]. Therefore, clinicians must choose the best modality through a multidisciplinary discussion.

The frequency of paraplegia in all AAS patients is two to five percent [1]. Neurological symptoms at the onset of AAS are infrequent but are...
As Sandhu et al. reported, the severity of SCI can vary widely from mild weakness to paraplegia. However, predictors of chronological resolution or therapeutic strategies have not been established [1]. CSFD appears to be effective in postoperative SCIs after the descending thoracic aortic surgery via reduction of spinal cord canal pressure and improvement of spinal cord perfusion [10]. However, this intervention has never been evaluated for AAS before and the clinical impact of CSFD does not have an international consensus. Sandhu et al. reported that they did not experience remarkable improvements with CSFD and this result can be attributed to the fact that many of their patients suffered from prolonged SCI after the initial onset of AAS [1]. For these reasons, the CSFD strategy for AAS/IMH requires a prospective interventional study.

Thoracic endovascular aortic repair (TEVAR) has been used as a less invasive alternative to open surgery for the management of descending AAS. An increasing number of TEVARs are being performed and the in-patient mortality has dramatically decreased. The indications of TEVAR include: (a) evidence of end-organ malperfusion, (b) refractory pain in spite of adequate medical treatment, (c) rapidly expanding false lumen, (d) impending or frank rupture, and (e) aneurysmal dilation in the chronic phase [2]. The stent-graft is positioned to cover the intimal lumen, (d) impending or frank rupture, and (e) aneurysmal dilation in the chronic phase. The stent-graft is positioned to cover the intimal lumen, (d) impending or frank rupture, and (e) aneurysmal dilation in the chronic phase [2].

Table 1

| Reference/published year | Age/gender | Stanford type of dissection | Spontaneous neurological improvement | Additional treatment besides ICU admission | Outcome | Neurological recovery |
|--------------------------|------------|-----------------------------|-------------------------------------|------------------------------------------|---------|----------------------|
| 3/2010                   | 65/M       | B, prior ascending aorta replacement | None                               | CSFD                                     | Survive | Full                 |
| 4/2012                   | 49/M       | B                            | Yes                                 | None                                     | Survive | Full                 |
| 5/2013                   | 75/M       | A                            | None                                | CSFD ascending aorta replacement         | Survive | Partial              |
| 6/2014                   | 60/M       | B                            | None                                | NR                                       | Death   | NR                   |
| 7/2014                   | 49/F       | NR                           | NR                                  | NR                                       | Survive | Partial              |
| 8/2015                   | 64/F       | A                            | NR                                  | CSFD ascending aorta replacement         | Survive | Full                 |
| 9/2017                   | 69/M       | B                            | NR                                  | None                                     | Survive | Partial              |

Note: CSFD: cerebrospinal fluid drainage; ICU: intensive care unit; NR: not reported in detail.

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