IJC Heart & Vasculature 22 (2019) 154–155

Contents lists available at ScienceDirect

IJC Heart & Vasculature

journal homepage: http://www.journals.elsevier.com/ijc-heart-and-vasculature

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

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ARTICLE INFO

Article history:
Received 18 January 2019
Accepted 22 January 2019
Available online 8 February 2019

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, neurology and 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

Abbreviations: (AAS), acute aortic syndromes; (ACS), acute coronary syndrome; (CSFD), cerebrospinal fluid drainage; (CTA), computed tomography angiogram; (ECG), electrocardiogram; (ER), emergency room; (IMH), intramural hematoma; (MRI), magnetic resonance imaging; (SCI), spinal cord infarction; (TEVAR), thoracic endovascular aortic repair.

☆ All authors have read and contributed to this manuscript.
Declarations of interest: none
Institutional review board approval was not required for this report.

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https://doi.org/10.1016/j.ijcha.2019.01.006

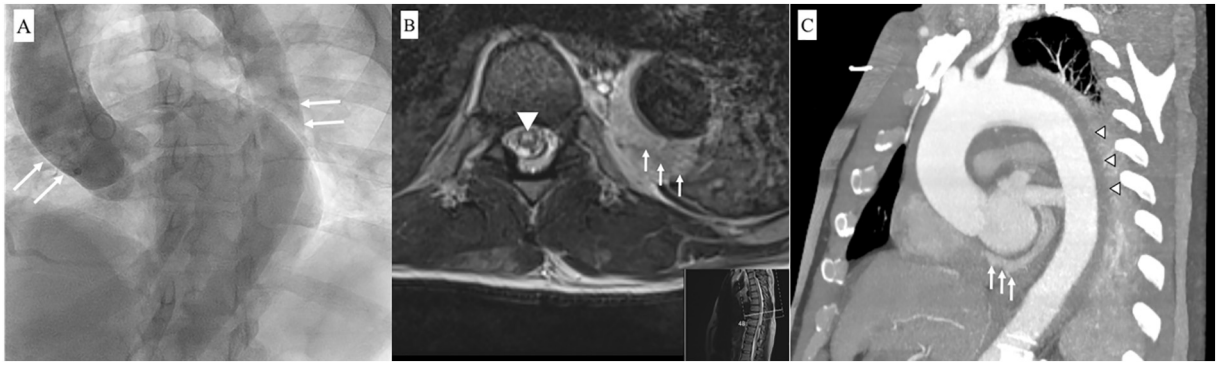

Fig. A. Aortography did not show any narrowing concerning for aortic dissection. Fig. B Whole spine magnetic resonance imaging showed increased T2 signal intensity and diffusion restriction predominantly involving the central gray matter of the spinal cord (white arrowhead). There was an abnormal enlargement of the descending aorta wall (white arrow). Fig. C Computed tomography angiogram demonstrated a small volume of pericardial effusion (white arrow) and intramural hematoma extending from ascending to the suprarenal aorta without evidence of great vessel occlusion, flap, extravasation of contrast, or hemothorax (white arrowhead).

Table 1
Former case reports of spinal cord infarction due to aortic intramural hematoma.

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.

often dramatic, masking the underlying vascular problems. This is a very common diagnostic dilemma especially in pain-free AAS. SCI due to IMH is so rare that we found only seven case reports in the last decade [3–9] [Table 1].

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 flap to seal the entry site of the dissection, resulting in thrombosis of the false lumen, and maintaining blood flow of the true lumen. However, the presence of a thrombosed false lumen precluded the application of endovascular strategies in our patient.

Acute aortic IMH masquerading as SCI is extremely rare and has a poor prognosis owing to the delayed diagnosis, short therapeutic window, and lack of consensus on management strategies. CSFD is one of the promising strategies for AAS-induced SCI and requires a prospective interventional study in the future.

Acknowledgements

None.

Funding sources

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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