CASE REPORT

More than just muscle spasms: a rare presentation of aortic dissection

Ebisa Bekele, Deepthi Chaitanya Kagolanu, Martin Kim, Kent Stephenson

Nassau University Medical
Center, East Meadow,
New York, USA

Correspondence to

Dr Deepthi Chaitanya
Kagolanu, deepthi.lank@gmail.
com

Accepted 12 March 2017

SUMMARY

Acute aortic dissection is associated with significant morbidity and mortality, often from complications including aortic regurgitation, cardiac tamponade and myocardial infarction. Typical clinical presentation includes a sudden onset of severe chest pain, although this is not always consistent. Clinical signs and symptoms are diverse with an estimated 38% of cases being missed on initial evaluation. Primary neurological symptoms at presentation are rare but have been reported often to coexist with chest pain. We present a case of acute aortic dissection in which the initial presenting symptoms were predominantly neurological. Stanford type A dissection is a surgical emergency with a high burden of cardiovascular death; thus, aggressive identification and management is paramount. Our case re-emphasises the importance of having a higher index of suspicion and a keen clinical eye for atypical presentations of acute aortic dissection.

BACKGROUND

Aortic dissection is an uncommon disease with high mortality and morbidity. Early diagnosis with a high index of clinical suspicion and treatment is crucial for survival. The incidence of acute aortic dissection in the general population is estimated to be 2.6–3.5 per 100 000 person years.^{1 2} The mortality of acute aortic dissection is as high as 1% per hour initially.³ We present a case of aortic dissection with a predominantly neurological presentation. We have highlighted this atypical presentation in an attempt to emphasise the importance of having a strong clinical suspicion in such scenarios.

CASE PRESENTATION

A woman in her early 50s presented to the emergency room reporting of intermittent numbness, spasms and rigidity in both legs that started spontaneously about 4 hours prior to presentation. The patient denied upper extremity numbness, weakness, chest pain, back pain, shortness of breath and palpitations. Additionally, the patient reported of inability to void urine for 1 day prior to admission. Her medical history was significant for emphysema and she admitted to cigarette smoking. She denied family history at the time of presentation.

INVESTIGATIONS

On arrival, the patient's vital signs showed a blood pressure of 134/81 mm Hg and a pulse rate of 88 bpm with no discrepancy in the extremities. Cardiopulmonary examinations were unremarkable. Bilateral lower extremities sensory loss and

brisk reflexes were appreciated. There was bilateral lower limb rigidity with extension contracture of the feet and toes. There was mottling of the legs; however, the femoral, dorsalis pedis and posterior tibial artery pulses were intact.

While in the emergency room the patient developed tachycardia with her ECG showing atrial fibrillation with rapid ventricular response (pulse rate of 150 bpm). The basic metabolic panel and the electrolytes were within normal limits. The chest X-ray was unremarkable. The admission cardiac biomarkers showed a mildly raised troponin I of 0.72 ng/mL (normal range 0.00–0.045 ng/mL) and a disproportionate increase in creatine kinase (CK) of 921 U/L (normal range 26–192 U/L). After 8 hours, cardiac biomarkers were further elevated with troponin I level of 6.02 ng/mL and CK of 4409 mg/dL.

Given the elevated cardiac biomarkers, the patient was admitted to the coronary care unit for possible acute coronary syndrome (ACS). However, due to the acute neurological presenting symptoms, a decision was initially made to withhold dual antiplatelet therapy and systemic anticoagulation for atrial fibrillation until the aetiology of the primary neurological symptoms were elucidated. This decision was made by the cardiology resident and the attendant. The patient's symptoms remained the same and a bedside transthoracic two-dimensional echocardiogram (echo) was performed within hours which showed an echo dense linear density in the aortic root/ascending aorta highly suspicious for an aortic dissection with a possible fistula/shunt formation into the left atrium. An emergent chest, abdomen and pelvic CT angiogram was performed which showed an ascending aortic dissection extending down to the bilateral iliac arteries (figure 1). Transoesophageal echo also confirmed the diagnosis of type A aortic dissection with moderate aortic insufficiency and a fistula from the false lumen of the non-coronary sinus and the left atrium (figure 2). The patient then underwent an emergent operative procedure for repair of type A aortic dissection and repair of aortoleft atrial fistula.

A possible component of a mixed connective tissue disorder and/or metabolic myopathy was also initially considered. However, in light of a probable aortic dissection seen on the echo, an electromyography was not pursued immediately. Acute cord compression was also considered as a differential diagnosis but an acute aortic dissection was strongly suspected after the echocardiogram. Therefore, a CT scan or MRI of the brain and/or spine was not performed immediately.

CrossMark

To cite: Bekele E, Kagolanu DC, Kim M, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-218432

Figure 1 CT scan of the thorax showing the false lumen in the ascending aorta extending down into the descending aorta.

During a follow-up appointment after the surgery, she stated that her sisters might have had aneurysms but denied family history of sudden cardiac death.

DISCUSSION

The typical clinical manifestations of aortic dissection are severe chest, abdominal or back pain. Most patients with Stanford type A dissection (ascending aorta dissection) present with anterior chest pain, where those with type B (descending aorta dissection) present with abdominal and back pain.⁴

In some cases, typical symptoms reported above may be suggestive of myocardial infarction, pericarditis, new-onset arrhythmias or pneumothorax, which may coexist with neurological symptoms and in rare cases patients may actually present pain free.⁵

Our patient did not present with typical symptoms suggestive of an acute aortic dissection of either type A or B making it

more likely to be missed at the first evaluation. Other confounding factors at presentation were the presence of new-onset atrial fibrillation and positive cardiac biomarkers suggestive of an ACS. Given the neurological symptoms, treatment for ACS was withheld due to the clinical uncertainty and risks of antiplatelet therapy and systemic anticoagulation. A bedside echo was initially performed within hours into the presentation as it was faster and easier to arrange than MRI.

Our patient has recovered well after the emergent repair surgery; however, she continues to smoke cigarettes despite counselling.

A similar neurological presentation was recently seen in a 70-year-old patient with heaviness in bilateral lower limbs, flaccid paraplegia with urinary retention who was diagnosed with an acute aortic dissection.⁶ Kowalska-Brozda and Brozda⁷ presented a case of bilateral stroke in which the patient was eventually diagnosed with an underlying acute aortic dissection. Similar other cases have been reported recently, which further stress on the increased likelihood of missing an underlying lethal diagnosis of type B aortic dissection. In addition, coexisting diagnoses such as a myocardial infarction or a stroke requiring antithrombotic therapy may worsen the mortality risk.

Spinal cord ischaemia and ischaemic peripheral neuropathies are more common with distal aortic dissections. Based on clinical symptoms, spinal cord involvement has been reported in up to 10% of these cases.⁴ In type A dissection, spinal cord ischaemia occurs in up to 1% and ischaemic neuropathy in 11% of cases.⁸ Even though spinal involvement is rare, our case fits in to this scenario as she was found to have type A aortic dissection with involvement of the spinal cord which is more expected in descending aortic dissection.

Spinal cord involvement in patients with aortic dissections could be secondary to the occlusion of the intercostal arteries,

Figure 2 Transoesophageal echo showing a fistula between the aorta and left atrium.

the artery of Adamkiewicz, or the thoracic radicular arteries resulting in the various cord syndromes.^{9 10} Peripheral nerve involvement in acute aortic dissection is also very rare. It results from direct nerve ischaemia or due to compression of the nerves by the expanding false lumen. In addition to presenting as typical spinal cord syndromes, patients with spinal cord and/or peripheral nerve involvement can present with numbness, paraesthesia, pulse deficit and even paralysis.^{9 10} Our patient's neurological presentation is consistent with involvement of mainly spinal cord and possible peripheral nerves.

Aortic dissection is more commonly seen in men than in women. Women presenting with aortic dissection were generally older than the men (67 vs 60 years of age) and 50% of women with aortic dissection were 70 years of age or older.¹¹ Our patient was relatively younger; her age did not fit in to the existing understanding of gender-based epidemiology of the disease.

Aortic dissection is more common in patients with hypertension, connective tissue disorders, congenital aortic stenosis or a bicuspid aortic valve.¹² Our patient did not have any of the known potential predisposing conditions leading to dissection. Though smoking is not mainly associated as an aetiology of dissection, the role of smoking might have played an important factor in our patient. Although genetic defects have been known to cause thoracic aortic aneurysms and dissections, it is usually seen in families with other inherited syndromes, or in those with confirmed autosomal-dominant inheritance of familial aortic aneurysm and dissection.¹³ The lack of these syndromes

in our patient's family makes the presence of aneurysms in her sisters less likely to be a risk factor.

In the setting of an atypical age at presentation, female gender, no known initial risk factors and absence of a typical presentation made the diagnosis very challenging in our case. It is critical to have a high clinical suspicion for such presentations of aortic dissection, as they are more likely to be missed at initial evaluation.

Acknowledgements Samy Selim MD-Nassau University Medical Center.

Contributors EB is the first author. DCK is the second author and corresponding author. MK and KS are co-authors and editors.

Competing interests None declared.

Patient consent Not obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Clouse WD, Hallett JW, Schaff HV, *et al.* Acute aortic dissection: population-based incidence compared with degenerative aortic aneurysm rupture. *Mayo Clin Proc* 2004;79:176–80.
- 2 Meszaros I, Morocz J, Szilvi J, *et al.* Epidemiology and clinicopathology of aortic dissection: a population-based longitudinal study over 27 years. *Chest* 2000;117:1271–8.
- 3 Criado FJ. Aortic dissection: a 250-year perspective. *Tex Heart Inst J* 2011;38:694.
- 4 Hagan PG, Nienaber CA, Isselbacher EM, *et al.* The International Registry of Acute Aortic Dissection (IRAD): new insights into an old disease. *JAMA* 2000;283:897–903.
- 5 Spittell PC, Spittell JA, Joyce JW, *et al.* Clinical features and differential diagnosis of aortic dissection: experience with 236 cases (1980 through 1990). *Mayo Clin Proc* 1993;68:642–51.
- 6 Hdiji O, Bouzidi N, Damak M, *et al.* Acute aortic dissection presenting as painless paraplegia: a case report. *J Med Case Rep* 2016;10:1.
- 7 Kowalska-Brozda O, Brozda M. A patient with acute aortic dissection presenting with bilateral stroke—a rare experience. *Neurol Neurochir Pol* 2015;49:197–202.
- 8 Gaul C, Dietrich W, Friedrich I, *et al.* Neurological symptoms in type A aortic dissections. *Stroke* 2007;38:292–7.
- 9 Khan IA, Nair CK. Clinical, diagnostic, and management perspectives of aortic dissection. *Chest* 2002;122:311–28.
- 10 Gaul C, Dietrich W, Erbguth FJ. Neurological symptoms in aortic dissection: a challenge for neurologists. *Cerebrovasc Dis* 2008;26:1–8.
- 11 Nienaber CA, Fattori R, Mehta RH, *et al.* Gender-related differences in acute aortic dissection. *Circulation* 2004;109:3014–21.
- 12 Friedman WF, Silverman N. Congenital heart disease in infancy and childhood. *Heart Dis* 1997;6:1514.
- 13 De Backer J, Campens L, De Paepe A. Genes in thoracic aortic aneurysms/dissections—do they matter? *Ann Cardiothorac Surg* 2013;2:73.

Learning points

- ▶ Aortic dissection presenting with neurological symptoms is very rare, especially type A. It is a life-threatening medical condition that needs early diagnosis and prompt treatment.
- ▶ One has to have a very high index of clinical suspicion for aortic dissection even in the absence of typical clinical features.
- ▶ We recommend clinicians to have a low threshold to obtain early diagnostic imaging in such scenarios.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow