Acute liver failure and seizure: a case report of an unusual presentation of acute painless aortic dissection

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Background
Painless aortic dissection presenting with seizure and acute liver failure is uncommon. We described a case of early recognition leading to successful treatment of painless aortic dissection with atypical presentation.

Case summary
A young lady presented with generalized tonic-clonic seizures coupled with hepatic pattern of deranged liver function test. Examination revealed blood pressure of 99/75 mmHg and hepatic flap. Electrocardiography showed sinus tachycardia. Urgent bedside echocardiography showed preserved cardiac function without significant valvular pathology, but noted a moderate pericardial effusion. Abdominal Ultrasound excluded liver cirrhosis or biliary obstructions. Viral hepatitis serologies and anti-liver panel were negative. She was progressively hypotensive with concurrent acute liver failure and oliguric acute kidney injury. Despite no chest pain, her rising serum troponin and widened mediastinum prompted an urgent computed-tomography aortogram, which showed a 4.3 cm dilatation of ascending thoracic aorta with acute haemopericardium and cardiac tamponade. She was diagnosed with malperfusion syndrome from Stanford type A aortic dissection. She underwent emergent ascending aorta and aortic arch repair and dialysis. She experienced complete recovery in her kidney, liver, and neurological function post-operatively.

Discussion
Painless aortic dissection masquerade as acute liver failure is uncommon. We describe a successful early recognition of malperfusion syndrome from painless aortic dissection, thus providing window for timely, life-saving intervention. Clinical challenges in this case include: (i) atypical presentation of aortic dissection, (ii) worsening acute liver failure which could lead to unnecessary liver transplantation, and (iii) risk of contrast-induced nephropathy in the setting of acute renal failure.

Keywords
Malperfusion syndrome • Liver failure • Case report • Painless aortic dissection • Type A aortic dissection

Learning points
• Aortic dissection may present atypically with malperfusion syndrome such as neurological deficit and end-organ failure.
• Malperfusion syndrome with multi-organ failure should prompt the physician to consider aortic dissection as an important differential diagnosis.
Introduction

Aortic dissection is a medical emergency with high rates of mortality and morbidity.\(^1\) Although aortic dissection classically presents as sudden onset of chest or back pain, it may present atypically as end organs failure from malperfusion.\(^2\) Overall, the incidence of painless aortic dissection is 6.4%, with a higher prevalence among type A than type B dissections. As painless aortic dissection is associated with increased mortality, it is of clinical relevance to identify these patients promptly.\(^3\) In this case report, we describe an uncommon case of painless aortic dissection presented with acute liver failure and seizure.

Timeline

Case presentation

A 36-year-old female patient who was found unconscious with stiffness of all four limbs and urinary incontinence. There was no history of chest pain. Her only significant past medical history was recent gastroenteritis following consumption of shellfish 3 weeks previously while travelling to Vietnam. There was no family history of seizure or liver disease, in particular, Wilson's disease. There was no recent acetaminophen or excessive alcohol consumption. On examination, the patient’s blood pressure was 99/75 mmHg and her heart rate was 117 b.p.m. She was not in respiratory distress with a respiratory rate of 17 breaths per minute. Jugular venous pressure was not raised. There were no clinically detectable jaundice or Kayser–Fleischer rings. A hepatic flap was present upon presentation. On palpation of the abdomen, there was mild epigastric tenderness with no palpable organomegaly. Heart sounds were regular and no murmur was audible. Lungs were clear and there was no pedal oedema. Pupils were equal and reactive to light. Cranial nerves II to XII were intact, power, and sensation was full in all four limbs.

Laboratory evaluation revealed a liver function test (LFT) that showed a hepatic pattern of derangement [alanine aminotransferase (ALT) 388 U/L (normal: 10–55 U/L) and aspartate aminotransferase (AST) 378 U/L (normal: 10–45 U/L)], but normal serum bilirubin, alkaline phosphatase (ALP), international normalized ratio (INR), and prothrombin time (PT). Abdominal ultrasound excluded the presence of liver cirrhosis or biliary obstruction. Viral hepatitis serologies (A, B, C, EBV and CMV) and autoimmune screen were all negative. Computed tomography (CT) of the brain excluded intracranial bleeding or mass. However, a chest radiograph (CXR) showed a possible soft-tissue opacity over the right upper mediastinum. Electrocardiogram showed sinus tachycardia without significant ST-segment deviation (Figure 1). Urgent bedside echocardiography showed preserved cardiac function without significant valvular pathology. However, a moderate-sized pericardial effusion that did not demonstrate tamponade features was noted (Figure 2).

Differential diagnoses include drug induced liver injury, Wilson’s disease, epilepsy with rhabdomyolysis, ischaemic hepatitis from systemic hypoperfusion such as cardiac dysfunction or sepsis.

Within 12 h of her admission, she started to develop features of ischaemic hepatitis and acute liver failure [prolonged PT of 14.9 (normal: 9.5–11.5), worsening LFT with ALT 3430 U/L, AST 3000 U/L]. She also became progressively hypotensive with oliguric KDIGO grade 3 acute kidney injury (AKI), metabolic acidosis, and hyperkalaemia requiring medical treatment. Intravenous acetylcysteine was given in view of early hepatic encephalopathy and rapidly progressing acute liver failure.

While she had no chest pain, her uptrending serum troponin from 89 to 1089 to 1217 (normal: 0.29 ng/L) and widened mediastinum prompted an urgent CT aortogram despite her AKI that put her at risk of contrast-induced nephropathy. Computed tomography aortogram showed a 4.3 cm dilatation of the ascending thoracic aorta with...
Acute liver failure and seizure

**Figure 1** Electrocardiography of patient upon admission.

**Figure 2** Different views on bedside trans-thoracic echocardiogram showing pericardial effusion: (A) apical two-chamber view, (B) apical four-chamber view, (C) parasternal long axis, and (D) subcostal view. Green arrow denotes the pericardial effusion of ~2 cm on the different views.
an irregular ulceration along left lateral aspect of ascending thoracic aorta extending ~5 cm from level of bifurcation of pulmonary trunk to level of aortic arch. There was moderate haemopericardium, but no definite dissection flap and no extension to the aortic root or involvement of the aortic cusps or coronary arteries (Figure 3). The diagnosis of acute liver failure secondary to ischaemic hepatitis from Stanford type A acute aortic dissection became apparent.

She underwent an emergency ascending aorta and arch replacement with a Frozen Elephant Trunk Device (Thoraflex Hybrid Graft) placed into distal arch and proximally anastomosed to the left subclavian artery, left common carotid artery, and innominate artery. She also underwent temporary renal dialysis for 4 days. Her renal and liver function had completely resolved during her outpatient follow-up two months later.

**Discussion**

The clinical challenges of this case lie in: (i) atypical presentation of aortic dissection and (ii) worsening acute liver failure which could lead to unnecessary liver transplantation. It highlighted the importance of prompt identification of acute aortic dissection as the aetiology of fulminant acute liver failure. Both fulminant liver failure and aortic dissection carry high mortality. Timely diagnosis permits early intervention, avoids catastrophic complications, and mitigates the risk of liver transplantation.

In this patient, she did not have any risk factors for developing dissection such as poorly controlled hypertension, inflammatory diseases such as takayasu arteritis or congenital conditions like Marfan syndrome or Ehlers–Danlos syndrome. She was also a lifelong non-smoker and did not use any recreational drugs such as cocaine. Instead, she presented with features of end-organ malperfusion. Aortic dissection can result in malperfusion syndrome, which gives rise to ischaemic hepatitis and neurological deficits. Hence, it is crucial for doctors to have a high clinical index of suspicion when presented with mimics of other conditions (such as neurological deficit or visceral failure) as any delay could be catastrophic.

In addition, widened mediastinum on chest radiography should prompt the physician to consider aortic dissection as a differential diagnosis. Pooled data from 10 studies showed that the predictive sensitivity of a widened mediastinum and abnormal aortic contour is 64% and 71%, respectively. With the advent of CT imaging, CXR has been used as an initial screening rule-in tool but is not adequate for conclusive rule out. In our case, suspicion of aortic dissection was raised from the presence of an abnormal aortic contour.

**Figure 3** Different views on computed tomography aortogram showing features of aortic dissection with end-organ malperfusion. (A) An ulcerated ascending aorta dilatation up to 4.3 cm with unilateral pleural effusion. (B) A long segment of irregular, eccentric intramural haematoma along ascending aorta to aortic arch. (C) Hypoperfusion of liver; hypoenhancement of liver noted. AA, ascending aorta; AR, aortic arch; DA, descending aorta; PA, Pulmonary artery.


**Conclusion**

Seizures and acute liver failure are uncommon presentations for acute aortic dissection. Malperfusion syndrome with multi-organ failure should prompt the physician to consider aortic dissection as an important differential diagnosis. Timely diagnosis is crucial to avoid irreversible morbidity for the patient.

**Lead author biography**

Dr Yu-Jun Wong was actively involved in clinical research and had won the best oral presentation at GIHEP meeting 2018 for his studies on the cost-effectiveness of DAA for HCV in Singapore. He has first authorship in several peer-reviewed international journals. He currently pursues his Master of Clinical Investigator. His passion for education is admirable. He holds positions as teaching faculty in Gastroenterology, Internal medicine, post-graduate year-1 residency and adjunct posting-director of year-III undergraduate postings (National University of Singapore).

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

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**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

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**References**

1. Hagan PG, Nienaber CA, Isselbacher EM, Bruckman D, Karavite DJ, Russman PL et al. The International Registry of Acute Aortic Dissection (IRAD): new insights into an old disease. JAMA 2000;283:897–903.
2. Imamura H, Sekiguchi Y, Iwashita T, Dohgomori H, Mochizuki K, Aizawa K et al. Painless acute aortic dissection—diagnostic, prognostic and clinical implications. Circ J 2011;75:59–66.
3. Fatima S, Sharma K. Painless aortic dissection—diagnostic dilemma with fatal outcomes: what do we learn? J Investig Med High Impact Case Rep 2017;5:232470961772125.
4. Melvinsdottir IH, Lund SH, Agnarsson BA, Sigvaldason K, Gudbjartsson T, Geirsson A. The incidence and mortality of acute thoracic aortic dissection: results from a whole nation study. Eur J Cardiothorac Surg 2016;50:1111–1117.
5. Gawinecka J, Schönrath F, von Eckardstein A. Acute aortic dissection: pathogenesis, risk factors and diagnosis. Swiss Med Wkly 2017;147:w14489.
6. Crawford TC, Beaulieu RJ, Ehliert BA, Ratchford EV, Black JH 3rd. Malperfusion syndromes in aortic dissections. Vasc Med 2016;21:264–273.
7. Coyle S, Moriarty T, Melody L, Ryan D. Diagnostic testing in acute aortic dissection. Curr Emerg Hosp Med Rep 2014;2:97–103.
8. Nazerian P, Pivetta E, Veglia S, Cavigli E, Mueller C, de Matos Soeiro A, on behalf of the ADvised Investigators et al. Integrated use of conventional chest radiography cannot rule out acute aortic syndromes in emergency department patients at low clinical probability. Acad Emerg Med 2019;26:1255–1265.