A case of left Haemothorax from contained rupture of Penetrating Atherosclerotic Ulcer

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Abstract

Penetrating Atherosclerotic Ulcer (PAU) is a rare clinical entity of Acute Aortic Syndrome (AAS). The rupture rate for PAU is almost ten-times that of aortic dissection but the clinical diagnosis of this condition can be challenging due to numerous other condition presenting with similar symptoms of “aortic pain”. However, with a unique prognosis, treatment strategy is distinct and prompt diagnosis with early specialist input is needed. We report a case of PAU presenting with collapse, chest and abdominal pain with a left Haemothorax.

Keywords Penetrating Atherosclerotic Ulcer; Acute Aortic Syndrome; TEVAR; Haemothorax
1. Introduction

Penetrating atherosclerotic ulcers (PAU) is part of the spectrum of acute aortic syndrome (AAS) alongside aortic dissection and intramural haematoma (IMH). (Committee et al., 2017) First described by Shennan et al in 1934 and further defined as a unique condition by Stanson et al. (Stanson et al., 1986) It is a condition where an atherosclerotic lesion ulcerates with subsequent penetration of the internal elastic lamina and media of the aortic wall. It commonly affects elderly patients with hypertension and only 2.3% - 7.6% of patients with AAS are due to PAU.(Committee et al., 2017) However, due to its high rupture rates with distinctive prognosis compared to other AAS entity, it has a different treatment strategy. (Chou et al., 2016; M. A. Coady et al., 1998; Tittle et al., 2002) Therefore, early accurate diagnosis of PAU is important. It can be clinically challenging to diagnose PAU as it shares presentation common to other AAS entity, high clinical suspicion has been advocated. (Michael A Coady et al., 2010; Gifford et al., 2016; Nathan et al., 2012) Although the natural history of PAU is better understood, there remain knowledge gaps about this condition and controversies regarding the timing of intervention especially in the asymptomatic group. (Cho et al., 2004; Chou et al., 2016; M. A. Coady et al., 1998; Gifford et al., 2016; Harris et al., 1994; Nathan et al., 2012)

Herein, we present a case of PAU with contained rupture in the descending thoracic aorta comparing with other case reports and review of literature regarding the natural history, investigation and management of this condition.
2. **Case presentation**

A 83 years old gentleman who is a non-smoker with a background of medication controlled hypertension and previous transient ischemic attack was brought to the emergency department in a district general hospital following an episode of collapse at home. He was complaining sudden onset severe “tearing” chest and abdominal pain associated with one episode of “coffee ground” vomitus. He was well prior to this presentation.

On examination, he was hypoxic and slightly hypotensive at 110/70 with a normal pulse. He had minimal air entry on the left chest, a soft but distended abdomen with no bowel sounds. Remaining physical examinations were unremarkable.

He had no ischemic changes on Electrocardiogram (ECG). Arterial blood gas suggested type 1 respiratory failure with metabolic acidosis. His Haemoglobin dropped from 94 g/L to 74g/L within hours of presentation and he had a raised cardiac enzyme. A chest x-ray (CXR) and abdominal x-ray showed a white out of the left-side of his lung (see figure 1) with no features of abdominal obstruction respectively.

A Computed Tomography Angiogram (CTA) of aorta was organised. This showed a contrast-filled focal protrusion (see figure 2 & 3) in the mid-descending thoracic aorta, in keeping with a PAU. Left haemothorax was confirmed. A small segmental pulmonary embolism was noted in the right lung.
Differential diagnoses are acute aortic syndrome, acute coronary syndrome, pneumonia, pulmonary embolism or upper gastrointestinal bleed.

Following resuscitation, the patient had a left-sided chest drain inserted which drained 1.5L dark red blood. The patient was transferred to our tertiary unit and underwent Thoracic Endovascular Aortic Repair (TEVAR). Intraoperative angiogram confirmed the position of the PAU at T8/9 (see figure 4). A 37 x 150 mm Conformable Gore ® TAG ® Thoracic Endoprosthesis (CTAG) device (W. L. Gore and Associates) was placed at T7 to mid T12 and a 4-5cm overlap proximally with 40 x 100 mm CTAG. Completion angiography confirmed exclusion of PAU (see Figure 5). There was no peri- or post-operative complication.

The patient made good recovery and was discharge with Rivaroxiban for treatment of incidental finding of pulmonary embolism 5 days post-operative.

3. Discussion

PAU is a rare yet potentially life-threatening condition with no known true incidence. (Committee et al., 2017) Although categorised under the umbrella term “Acute Aortic Syndrome (AAS)”, it is a distinct clinical entity with its own disease course and hence, management strategy. (Chou et al., 2016; M. A. Coady et al., 1998; Nathan et al., 2012) As presented in our case, the clinical diagnostic challenge is early accurate diagnosis of PAU. However, with high clinical index of suspicion, early appropriate imaging and prompt involvement of relevant specialist, this could be achieved.
Our patient reflects the described patient demographic of PAU in many studies. Commonly, they are elderly with hypertension. (Chou et al., 2016; M. A. Coady et al., 1998; Gifford et al., 2016; Nathan et al., 2012) In addition, other co-morbidities such as hyperlipidemia, ischemic heart disease, diabetes or chronic renal insufficiency may be present. In fact, majority of other case studies of patient with PAU were over the age of 65 years with hypertension. (Dalio, Dezotti, Ribeiro, Joviliano, & Piccinato, 2015; Kyaw, Sadiq, Chowdhury, Gholamrezaee, & Yoe, 2016; Samal, White, & Kot, 2001; Siegel, 2013; Soyama et al., 2015) Only in one case by Ando et al was the patient a 57 years old man with no known hypertension. (Ando, Minami, Muramoto, Narita, & Sakai, 1994) Meanwhile, some studies suggest that PAU has a female predominance. (Chou et al., 2016; Gifford et al., 2016)

The clinical presentation in our case of collapse with acute severe chest pain posed a diagnostic challenge. The symptoms could be classically termed as “aortic pain”. (Ahmad, Cheshire, & Hamady, 2006) But, severe chest pain is a common denominator for many other conditions such as acute coronary syndrome, pulmonary causes or other clinical entity of AAS. Patatas et al summarised that in symptomatic PAU, severe chest and or back pain is the most often complaint. (Patatas, Shrivastava, & Ettles, 2013) Also, acute pain may not always be the case. Soyama et al presented their patient with 11 days history of abdominal, left chest and back pain. (Soyama et al., 2015) Kyaw et al patient had 6 days of chest pain with similar symptom 3 weeks prior to diagnosis. (Kyaw et al., 2016)
The abnormal CXR in our patient was an additional sign to suggest a more sinister underlying cause to his presentation. Kazerooni et al showed that all patients with PAU in their series had an abnormal CXR in the form of enlargement of the descending thoracic aorta although they also described other findings such as widened mediastinum or pleural effusion. (Kazerooni, Bree, & Williams, 1992) Similarly, the CXR in the case described by Siegel et al and Ando et al found pleural effusion. (Ando et al., 1994; Siegel, 2013) In contrast, normal CXR was found in Soyama et al’s patient and on both admission in Kyaw et al’s patient. (Kyaw et al., 2016; Soyama et al., 2015)

In the acute setting, CT angiogram (CTA) has superseded all other imaging modalities such as Magnetic Resonance Imaging (MRI) or Transoesophageal Echocardiogram (TEE) or Angiography as an imaging modality of choice to diagnosis PAU. (Patatas et al., 2013) It is readily available, provide detailed anatomical information of the thoraco-abdominal aorta with two or three-dimensional reconstruction, less invasive and has a shorter examination time. On CTA, PAU is characterised as a contrast-filled outpouching of the affected artery sometimes described as “mushroom-like projection” in tangential view in the absence of a false lumen or dissection. (Hiratzka et al., 2010) In addition, there could be associated extensive aortic calcification and diffuse atherosclerosis.

The management strategy in our patient was resuscitation, optimisation and finally definitive treatment within the same admission. The management options here are medical management, early surgical intervention or delayed surgical intervention. In our case, although the patient’s symptoms were manageable with analgesia, the risk of expansion of rupture was deemed high
hence decision made for early surgical intervention. Chou et al demonstrated that early surgical management confers better survival outcome when compared to initial medical management. (Chou et al., 2016) Earlier studies recommended the same, although these conclusions were drawn from studies on symptomatic group. (M. A. Coady et al., 1998; Stanson et al., 1986; Tittle et al., 2002) Similarly, another case study managed their patient successfully with early surgical intervention. (Siegel, 2013)

In contrast, some studies showed that initial medical management and surveillance is an acceptable course for PAU particularly in asymptomatic patients. (Cho et al., 2004; Gifford et al., 2016; Harris et al., 1994) Kyaw et al successfully managed their patient with PAU medically. (Kyaw et al., 2016) However, not all cases managed medically were successful. Soyama et al’s patient had progression of PAU and symptoms recurrence requiring TEVAR. (Soyama et al., 2015) Samal et al’s patient did not survive following open surgery after failed medical management. (Samal et al., 2001) Meanwhile, Brittenden and colleagues performed TEVAR on 2 cases of symptomatic PAU six weeks following presentation with blood pressure control in the interim. (Brittenden, McBride, McInnes, Gillespie, & Bradbury, 1999) Available guidelines suggest endovascular repair for symptomatic or complicated PAU but the evidence is low and timing of intervention is still debatable. (Committee et al., 2017) In addition, endovascular repair in the form of TEVAR is now the preferred choice over open repair for PAU as it has better mortality rates and acceptable mid-term results. (D’Anoville, Ozdemir, Alric, Marty-Ané, & Canaud, 2016)

Current concept on PAU is that 90% of these are located in the descending thoracic aorta. (M. A. Coady et al., 1998) They can complicate into a localised dissection and intramural haematoma,
formation of pseudoaneurysm or rupture. The rupture rate ranges from 4.1% to 40%. (Chou et al., 2016; M. A. Coady et al., 1998; Nathan et al., 2012) Ganaha et al suggested that PAU with 20mm diameter or 10mm neck had a high risk of progression. (Ganaha et al., 2002) But Nathan et al did not see any correlation of disease progression with measurements. (Nathan et al., 2012) Gifford et al concluded that larger ulcers grow faster. (Gifford et al., 2016) Hence, there are still gaps of knowledge regarding why some PAU develop complications while others do not.

4. Conclusion

PAU is an increasingly recognised clinically entity of AAS that can be lethal without prompt accurate diagnosis and involvement of specialist for timely delivery of treatment. Clinicians should have a high clinical index of suspicion for all patients with risk factor presenting with “aortic pain”. Current research supports aggressive surveillance for asymptomatic PAU with a low threshold for surgical intervention for symptomatic PAU. The natural history and timing of intervention for PAU is an area for future research.

Declaration of competing interests

The author(s) declare that there is no competing interest.

Authors Contribution

(Sharon) Yen Ming Chan: literature review and writing of case report
James Matossian: preparation of radiological images
Bryce Renwick: Concept and editing of case report
Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanied radiological images.

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Figure 1: Erect chest x-ray showing white out of left lung

Figure 2: Computed Tomography Angiogram (CTA) of aorta showing A = contrast-filled outpouching wall of mid-descending thoracic aorta; B = Attenuation of pleural fluid in left chest suggesting left haemothorax; C = left intercostal drain in-situ.
Figure 3: Three-dimensional (3D) reconstruction of CTA of aortic arch and thoracic aorta demonstrating the focal projection in mid-descending thoracic aorta as labelled.

Figure 4: Intra-operative angiography showing contrast in the descending thoracic aorta. A = Conformable Gore ® TAG ® Thoracic Endoprosthesis (CTAG) device (W. L. Gore and Associates) over a guidewire; B = contrast-filled outpouching of aorta at T8/9 level.
Figure 5: Completion angiography showing exclusion of PAU with successful deployment of Conformable Gore® TAG® Thoracic Endoprosthesis (CTAG) device