Case report

Masquerade presentation of acute type B aortic dissection as isolated acute limb ischaemia treated with endovascular fenestration with angioplasty: A case report

A.M. Omar Mohamed Ozaal*, Thanoj Fernando

Vascular and Transplant Surgical Unit I, National Hospital of Sri Lanka, Colombo, Sri Lanka

ARTICLE INFO

Keywords:
- Acute type B aortic dissection
- Acute limb ischaemia
- Malperfusion syndrome
- Endovascular fenestration
- Angioplasty
- Case report

ABSTRACT

Introduction and importance: Masquerade presentation of acute type B aortic dissections (TBAD) as acute limb ischaemia (ALI) is rare. Holistic clinical assessment, preferably with the help of scoring systems and timely computer tomographic angiogram (CTA), is needed for early diagnosis. Acute TBAD and its complications are increasingly treated with endovascular therapies.

Case presentation: A 21-year-old male with poorly controlled essential hypertension was admitted with prominent clinical features of ALI. No clinical pointers of a TBAD were present. Doppler ultrasound revealed an arterial occlusive pattern, and an urgent surgical embolectomy was performed. On failure to retrieve any thrombi, a CTA was performed, and diagnosis of TBAD complicated with ALI was made. The limb was revascularised with guidewire directed aortic fenestration with angioplasty. TBAD was managed conservatively.

Clinical discussion: We report a case of acute TBAD presented as isolated ALI, which was initially diagnosed and treated as an ALI unrelated to aortic dissection. TBAD with typical or atypical clinical features presented with ALI as a malperfusion syndrome is not uncommon. However, masquerade presentations of TBAD as ALI are rare in the literature. Endovascular fenestration with or without stenting has fewer neurological complications and long-term mortality than thoracic endovascular aortic repair (TEVAR). Moreover, they become convenient in resource-poor settings without dedicated aortic centres.

Conclusion: Masquerade presentation of TBAD should be recognised in the differential diagnosis of ALI. Timely CTA would prevent unnecessary interventions and help diagnose TBAD complicated with ALI. Despite their availability, outcomes will depend on proper patient selection for endovascular, surgical, and TEVAR options.

1. Introduction

Acute aortic dissection is the most common life-threatening acute aortic syndrome (AAS). It occurs in 3 to 5 patients per 100,000 per year, and two-thirds of them are men with a mean age of 63 years [1]. Risk factors for aortic dissection are hypertension, atherosclerosis, known aortic aneurysm, previous cardiac surgery and Marfan syndrome [2]. According to the International Registry of Acute Aortic Dissection (IRAD), 33% of aortic dissections are type B. Type B aortic dissection (TBAD) commonly presents with back pain than chest pain, and 80.9% of them would have hypertension [2]. Complicated acute TBAD is characterised by malperfusion syndromes such as mesenteric ischaemia, acute renal failure, lower limb ischaemia, heart failure, major neurological deficit, and spinal cord ischaemia [3].

Acute limb ischaemia (ALI) occurs in 9.7% of acute TBAD in the IRAD series. Reports of masquerade presentation of TBAD as ALI are scarce in the literature. It is significantly associated with acute renal failure, acute mesenteric ischaemia and death [2]. Endovascular therapy is commonly performed over surgical bypass for TBAD complicated with lower limb ischaemia [2-4].

We report a case of acute TBAD in a 21-year-old hypertensive young male presented as an isolated ALI without any clinical cues of aortic dissection. The limb was revascularised successfully with guidewire directed aortic fenestration and angioplasty due to the unavailability of

Abbreviations: TBAD, Type B aortic dissection; ALI, Acute limb ischaemia; CTA, Computer tomographic angiogram; AAS, Acute aortic syndrome; DSA, Digital subtraction angiography; DUS, Doppler ultrasound; TEVAR, Thoracic endovascular aortic repair.

* Corresponding author.
E-mail address: ozaalnew@gmail.com (A.M.O.M. Ozaal).

https://doi.org/10.1016/j.ijscr.2022.106857
Received 23 January 2022; Received in revised form 4 February 2022; Accepted 22 February 2022
Available online 25 February 2022
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thoracic endovascular aortic repair (TEVAR). This work is reported in line with SCARE criteria [5].

2. Presentation of case

A 21-year-old male presented to the emergency unit with a history of sudden onset left lower limb pain for 5 h, without chest or back pain. He had clinical features of acute limb ischaemia such as numbness, coldness, diminished toe movements, and absent left femoral and distal pulses. His contralateral limb was normal. There was no past or family history of aortic syndromes, connective tissue disorders or sudden cardiac deaths. On admission, blood pressure and pulse rates were 240/150 mmHg and 90 bpm. Abdominal examination was unremarkable. He did not reveal the history of essential hypertension at the time of presentation which was diagnosed six months back after excluding secondary causes. He did not take regular anti-hypertensives.

All laboratory tests, including renal function and cardiac enzymes, were normal. An electrocardiogram showed a left ventricular strain pattern with no dynamic changes. Doppler ultrasound (DUS) showed monophasic flow in the left common femoral artery with no distal arterial waveforms.

The initial working diagnosis was isolated left-sided ALI in Rutherford stage IIb based on the clinical features and DUS findings. An initial computer tomographic angiogram (CTA) was not performed due to the need for immediate revascularisation and the institutional practice because of limited facilities and financial constraints.

He was commenced with opioid analgesics, Glyceryl trinitrate (GTN) and unfractionated heparin (UFH). The vascular surgical team has performed an urgent surgical embolectomy under local anaesthesia. Intra-operatively, the Fogarty catheter was passed via transverse arteriotomy on the left common femoral artery but failed to retrieve any thrombi proximally and distally. Moreover, a significant inflow reduction was noted. The negative embolectomy raised the suspicion of aortic dissection or an alternative pathology. An urgent aortic and lower limb CTA was performed, revealing a TBAD spanning from the left subclavian artery to aortic bifurcation without contrast extravasation (Fig. 1).

A complete thrombus extended into the left common iliac artery to its bifurcation through the maintained distal flow (Fig. 2). The celiac trunk originated from a false lumen with contrast flow. Superior and inferior mesenteric and bilateral renal arteries originated from true lumen. Right lower limb arteries were patent.

Once systolic and mean arterial blood pressures were maintained less than 120 mmHg and 90 mmHg, respectively, with GTN and labetalol infusion, intravenous drugs were switched to oral anti-hypertensives. Left lower limb malperfusion was treated with guidewire-directed aortic fenestration and angioplasty on day four since admission by the intervention radiology team at our institution (Fig. 3). The delay is due to the limited resources and UFH was continued till the procedure.

The procedure was performed under intermittent pulsed fluoroscopic screening. The left common and external iliac arterial retrograde puncture was done with an 18-gauge puncture needle. Seldinger technique was utilised to insert a 7-French gauge arterial sheath. Negotiation was done into the lower aorta with multiple catheter/guidewire combinations, and fenestration of dissected aortic membrane was achieved. Gradual dilatation of the lower aorta, common and external iliac arteries was performed with a 10 mm × 80 mm angioplasty balloon.

Post-procedure digital subtraction angiography (DSA) showed satisfactory dilatation of the lower aorta, common and external iliac arteries with a good distal flow (Fig. 4). He was given a short course of Enoxaparin to dissolve any distal thrombi if present. Aortic dissection was managed conservatively. There were no adverse events encountered in the therapeutic intervention the patient underwent. A repeat CTA was performed two months from the presentation, which showed a short segment narrowing in the false lumen of the distal aorta. Still, normal run-off of contrast was seen in the left common iliac artery and its branches (Fig. 5). The patient did not report any symptoms or signs of left lower limb ischaemia on review.

![Fig. 1. CT angiogram (axial view) following embolectomy A) type B aortic dissection with a spared arch of aorta and its branches [red arrow shows the true and false lumen] B) bilateral intact renal arteries C) aortic bifurcation [red arrow shows occluded false lumen with patent collateral to left lower limb] D) bilateral common iliac arteries [red arrow shows left artery with occluding thrombus]. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)](image-url)
3. Discussion

Malperfusion syndrome contributes to 30% of acute TBAD complications [2]. Lower extremity malperfusion was reported in 5 to 12% of TBAD and ranged from diminished pulse to acute limb-threatening ischaemia, where the latter was the commonest [4]. TBAD presenting with ALI is commonly associated with more dreaded complications than our case, where no end-organ dysfunctions were identified. TBAD occurs commonly in males of sixth and seventh decades of life [2]. Furthermore, apart from bilateral ALI in a 25-year-old male with graft related aortic dissection, no cases of TBAD or TBAD with ALI were found in the literature to occur in a male less than 25 years of age [6].

Most of the cardinal features of ALI were present in this case, along with DUS findings. American Heart Association (AHA) guidelines proposed a sensitive risk assessment tool to rule out aortic dissection based on three groups of information, namely predisposing conditions, pain features, and clinical examination. This scoring system considered the number of the information groups involved, from 0 (none) to 3. The presence of 0, 1, 2, or 3 groups of information increases the pre-test probability of an AAS [7]. The absence of these high probability features (Score-0) and the presence of most cardinal features and DUS findings of ALI contributed to the initial working diagnosis of isolated left ALI rather than aortic dissection complicated with ALI.

The decision for surgical embolectomy was derived as he was diagnosed with Rutherford IIB stage ALI which needs immediate revascularisation [8]. The latest European society for vascular surgery (ESVS) guidelines accept the usage of DSA, CTA, DUS and Magnetic resonance angiography to diagnose ALI. CTA is widely used as it is accurate and evaluates entire vascular tree and adjacent structures [8].

Liu et al. reported a similar case of TBAD presenting with isolated ALI of the right leg, which was initially missed but later diagnosed with CTA. He also stated the failure of surgical embolectomy for ALI, which raised the suspicion of an aortic dissection. Aortic dissection was managed medically, but endovascular or surgical options done for the patient were not mentioned [9]. Apart from Liu et al., other cases of TABD presenting with ALI reported in the literature had clinical pointers for the TBAD, such as chest or back pain to some extent [4,10–12].

According to the latest ESVS guidelines, acute TBAD management aims to restore perfusion to vital organs and prevent progression of dissection and aortic rupture [13]. Treatment comprises analgesics, antihypertensives (intravenously in an acute phase) and multi specialistic surgical or endovascular options. IRAD series indicates endovascular therapies (93%) are more commonly performed in ALI in TBAD than surgeries (7%) [2].

TEVAR is indicated for complicated TBAD patients, which showed benefits over the open repair and medical treatment in terms of early mortality [13]. It is directed to cover the entry tear and thereby reduce the false lumen perfusion where the visceral vessel patency is maintained in 97% of cases. However, TEVAR has higher neurological complications and long-term mortality in TBAD complicated with visceral malperfusion [11]. Fenestration of intimal flap with or without stenting is beneficial in these patients.

The mechanism of blood flow limitation via branch vessels in aortic dissection is either by static or dynamic obstruction. Our case had dynamic obstruction at the left common iliac artery on CTA. A dynamic obstruction can be managed by increasing false lumen outflow with fenestration of intimal flap. In contrast, a static obstruction or ostial disruption should be treated by stenting of malperfused branch vessel [12].

We opted for guidewire directed endovascular fenestration with balloon angioplasty in our case due to the unavailability of TEVAR, lack of signs of rupture, and dynamic obstruction to the left common iliac artery. Post-procedure DSA showed adequate visualisation of distal vasculature with enhancement and drainage of contrast. This precluded us from stenting the branch vessel in addition to the consideration of stent-related complications in young male patients [11]. In the recent case series, endovascular options had been performed for more than half of the cases of acute TBAD with ALI [4]. In our case, the reported procedure-related complications such as gastrointestinal ischaemia, reperfusion injury, and acute renal failure were not encountered [4].
Fig. 3. Digital subtraction angiography (DSA) and balloon dilatation A) pre-angioplasty film showing filling defect beyond left common iliac artery B) guidewire directed fenestration C) 6 mm × 80 mm balloon dilatation D) 10 mm × 80 mm balloon dilatation.

Fig. 4. Post angioplasty DSA A), B) & C) shows the successful revascularisation without a stent.
4. Conclusion

ALI in an emergency department should be assessed holistically, bearing in mind that it can present exclusively or as part of a masqueraded aortic syndrome. There are established scoring systems for ruling out aortic dissection. Timely CTA will help to diagnose aortic dissection and its complications. Moreover, it prevents unnecessary interventions and helps to streamline treatment options.

Endovascular fenestration alone or with stenting is a recognised option for ALI in acute TBAD, especially in resource-poor settings. It can also be combined with aortic stenting and involves a lower mortality rate than TEVAR and surgical options.

Ethical approval

N/a.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Availability of data

N/a.

Source of funding

None.

CRediT authorship contribution statement

AMOMO wrote this paper. TF reviewed and proofread the content. Both the authors read and approved the final manuscript.

Registration of research studies

N/A.
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