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## Case Report

# Successful medical treatment of aortic intramural hematoma (Stanford type B) in a patient with aberrant right subclavian artery: A case report <sup>☆</sup>

Pasan Witharana<sup>a,c,\*</sup>, Mohamed Sherif<sup>a,b</sup>, Govind Chetty<sup>a</sup>, Douglas Turner<sup>a</sup>

<sup>a</sup> Northern General Hospital, Herries Rd, Sheffield, S5 7AU, UK

<sup>b</sup> Leeds General Infirmary, Leeds, LS1 3EX, UK

<sup>c</sup> Imperial College London, London, SW7 2BX, UK

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## ABSTRACT

Intramural Hematoma (IMH) forms part of the acute aortic syndrome, aortic dissection, and penetrating aortic ulcer. It is a life-threatening aortic disease that warrants prompt diagnosis and management. Like aortic dissections, it is classified using the Stanford classification system as type A (proximal to the origin of the left subclavian artery) and type B (distal to the origin of the left subclavian artery). Patients with type A IMH is generally managed surgically, and uncomplicated type B IMH is managed medically. The right subclavian artery arises typically from the brachiocephalic trunk. Aberrant right subclavian arteries (ARSA) are rare and derive directly from the aortic arch distal to the left subclavian artery. In this case report, a 73-year-old female presented with right-sided chest pain and shortness of breath. On examination, her heart rate was 100 bpm and blood Pressure was 185/85 and her ECG showed sinus rhythm. Following a CT scan, she was found to have a type B Aortic IMH with an ARSA. She was medically managed with vigorous blood pressure control. After a period of intravenous blood pressure treatment, she was treated with oral medication. Her subsequent CT scan showed that the hematoma was stable. She was followed up with MRI scanning 1 year later, which showed complete healing of the aorta with no changes in diameter. This case illustrates the importance of strict blood pressure management and follow-up imaging in patients presenting with type B IMH. It is important to monitor these patients regularly and where blood pressure control alone is not sufficient, further intervention may be required. Even though the complete resolution may be achieved as in this case, these patients will need to be kept under surveillance with repeated scans to monitor for any changes.

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\* Corresponding author.

E-mail address: [p.witharana@nhs.net](mailto:p.witharana@nhs.net) (P. Witharana).

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## Background

Aortic intramural hematoma (IMH) is described as a hemorrhage within the medial layer of the aortic wall with no detectable blood flow or initial flap in the vessel wall [1]. Rarely, patients with an aortic IMH may have an aberrant subclavian artery. The right subclavian artery arises typically from the brachiocephalic trunk. Aberrant right subclavian arteries (ARSA) are rare and found in 0.4%-1.8% of the population and derive directly from the aortic arch distal to the left subclavian artery and not from the brachiocephalic artery [2]. In this case report, we present the case of type B intramural hematoma in a patient with an ARSA and the importance of blood pressure management.

## Case report

A 73-year-old female presented after a week of having a right total hip replacement with severe epigastric pain radiating to the back associated with shortness of breath and right-sided pleuritic chest pain.

The patient's past medical history included hypertension, hypercholesterolemia, and a right total hip replacement. She was on simvastatin, bisoprolol and had no known allergies. She was independent, mobile with a stick after the hip replacement, lived with her husband, was a non-smoker, and drank moderately.

On presentation to the local emergency department, the patient had a respiratory rate of 22, Oxygen saturation of 97% on air, heart rate of 100 bpm, blood pressure (BP) of 185/85 mmHg, and a temperature of 37.3 degrees. The patient was alert and orientated, and her Glasgow Coma Scale (GCS) was 15/15. Examination revealed a general tenderness across the epigastric area of the abdomen. However, a respiratory, and cardiovascular examination was unremarkable. She was well perfused peripherally with a capillary refill time of (CRT) < 2 seconds and good peripheral pulses.

As part of the investigation, the electrocardiogram (ECG) showed a sinus rhythm with a rate of 100 and no ischemic changes. The patient also had a CT pulmonary angiogram (CTPA), which showed that she had an aberrant right subclavian artery and a type B aortic intramural hematoma (IMH). The IMH was extending from the aberrant right subclavian artery to the bifurcation of the aorta, measuring 8-9 mm in diameter. Figures 1 and 2 show the sagittal and cross-sectional views of the pathology. There was an ulcer at the aberrant right SCA (Fig. 3). After a discussion between the vascular, interventional radiologist, and the cardiothoracic team, the patient was treated conservatively with strict blood pressure control. She was started on a Labetalol infusion at a rate of 20 mg/ h. Systolic BP was titrated strictly between 100 and 120 mmHg with close monitoring in the progressive care unit for 72 hours.

The patient remained stable with a well-controlled BP and the CT scan was repeated in 72 hours which showed that the hematoma size was stable and not increased in size. The patient was converted to oral blood pressure control medica-

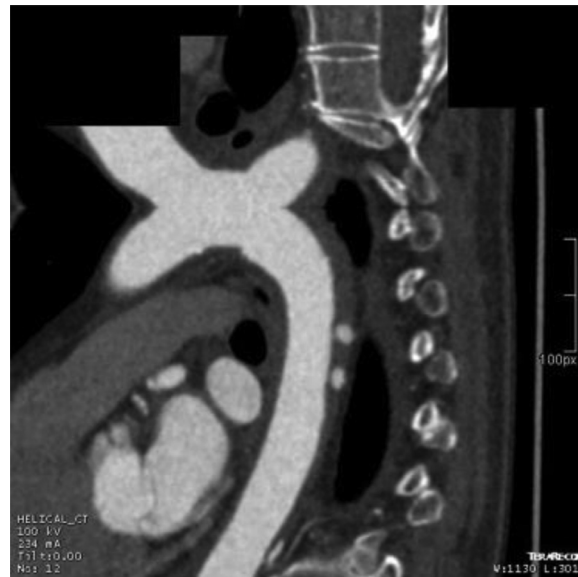


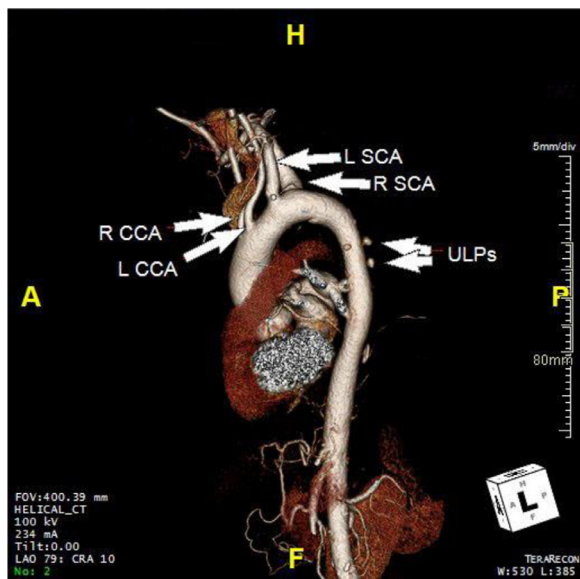
Fig. 1 – Sagittal view CT image of the pathology.



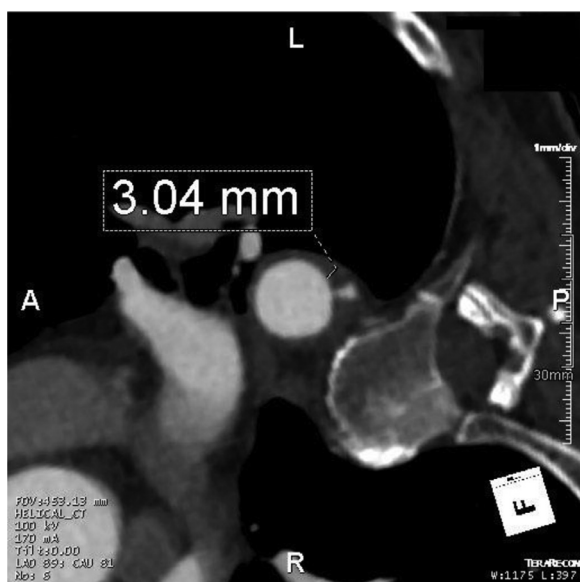
Fig. 2 – Cross-sectional view CT image.

tion including Bisoprolol 5 mg and Ramipril 2.5 mg and had another repeat CT scan before discharge, which showed no change in the size of the hematoma nor the aorta.

The patient was followed as an outpatient with another CT scan 4 weeks later, showing that type B hematoma has healed with no change in the aortic size (Fig. 4). Another repeat MRI scan a year later confirmed the complete healing of the intramural hematoma (Fig. 5). The volume-rendered (VR) MR aortogram images showed the complete resolution of ULPs as shown in Figure 6. The aortic MDT team decided to discharge the patient with no further follow-up.



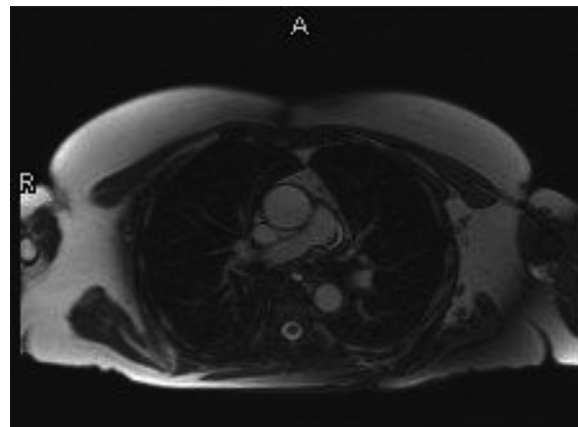
**Fig. 3 – Reconstructed Image showing ulcer-like projections (ULPs).**



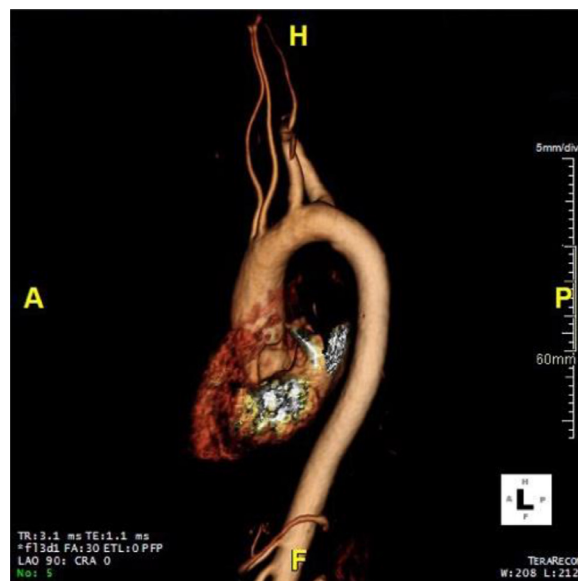
**Fig. 4 – Cross-sectional view CT image showing healing of the hematoma.**

## Discussion

The most common symptom in patients with IMH in more than 90% of the cases is a sudden onset of severe anterior or posterior chest pain that may radiate to the back of the neck. IMH accounts for 4%-11% of acute aortic syndrome cases in the United States and Europe, whereas it is 32% in Asia [1]. Type A, IMHs involve the ascending aorta and the aortic arch up to the left subclavian artery, while type B IMHs occur distal to this. In contrast, to type A IMH, type B IMH seems to have a better prognosis than type B aortic dissections [3].



**Fig. 5 – MRI showing healed hematoma.**



**Fig. 6 – MRA VR reconstructed images showing complete resolution of ULPs.**

The average aortic wall thickness size is less than 3 mm, whereas, in IMH, the aortic wall diameter is more than 5 mm. Most (90%) cases of IMHs are spontaneous, while some are traumatic. One suggestion is that IMH occurs due to the spontaneous rupture of vasa-vasorum in the medial layer due to a combination of wall stress, fragile vessels, chronic hypertension, and inflammation [1].

Almost 90% of type B, IMH cases are medically managed, whereas less than 10% are surgically managed. The initial defects of the aortic wall are referred to as ulcer-like projections (ULP), and the presence of these with a communicating orifice of more than 3 mm in size warrants serial imaging and close follow-up as these may indicate that the IMH is progressing. These patients may benefit from a TEVAR or surgical repair [1].

IMH may lead to acute aortic dissection in 3%-14% of the cases of type B and 88% of type A IMHs. Acute IMH can be life-

threatening and require urgent medical therapy to reduce the risk of progression to aortic dissection or rupture [4].

Patients with complicated type B IMH (aortic rupture, impending aortic rupture, or concerns about clinical deterioration) will have endovascular repair with TEVAR. Uncomplicated type B IMH are managed medically with strict blood pressure (systolic below 120), heart rate control, and pain management. These patients are closely monitored with CT scanning (repeated with worsening or new-onset pain or concerns about end-organ ischemia). Those who are not responding to blood pressure medication despite being on multiple agents, intractable pain, or worsening IMH thickness are considered for TEVAR [5].

Beta-blockers consist the mainstay of treatment in the medical management of type B IMH as it controls both the heart rate and the blood pressure. Other medications that may be used include calcium channel blockers or sodium nitroprusside. However, these should be used together with beta-blockers to prevent reflex tachycardia. Lifelong angiotensin-converting enzyme inhibitors or angiotensin receptor blockers may promote longer-term aortic remodeling as well as control blood pressure [6].

Type B IMHs are defined as acute if they are within 14 days of symptom onset. The mortality rate of open repair of type B IMHs in the acute phase has been reported to be as high as 16%. A multi-center study involving 41 patients with type B IMH of whom 19 received the best medical therapy (BMT), showed that 47% of these patients had progression of the aortic lesion [7].

Among the many different types of aortic arch anomalies, ARSA is the most frequently encountered. As they are asymptomatic, these are often diagnosed as incidental radiological findings. In some rare cases, patients may present with compressive symptoms on the trachea and esophagus. They will usually have dysphagia of solid food [8].

Contrast CT scans are beneficial in diagnosing IMH and confirming the presence of intima-media flap in aortic dissection, differentiating IMH from a penetrating aortic ulcer, or visualize ULP and intramural blood pool (IBP). IBP is another type of intimal defect seen in some patients with IMH. Although limited benefits in the acute setting, MRI remains a gold standard imaging modality in the follow-up phase. Small IMHs may be missed on CT scanning and could be picked up by MRI. During the early course of IMH formation, hematoma shows an iso-intensity on T1 imaging and hyper-intensity on T2. After the first couple of days, oxyhemoglobin gets converted to methemoglobin, which causes a hyper-intense signal in both T1 and T2 modalities. This makes MRI an excellent choice to help assess the age of the IMH [1].

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## Conclusion

This case illustrates the importance of strict blood pressure management and follow-up imaging in patients presenting with type B IMH. It is important to monitor these patients regularly and where blood pressure control alone is not sufficient, further intervention may be required. Even though the complete resolution may be achieved as in this case, these patients will need to be kept under surveillance with repeated scans to monitor for any changes.

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## Patient Consent

Written informed consent was obtained from the patient, prior to the publication of this article.

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