

CASE REPORT

Where's the Flap?

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ABSTRACT

We present a case of aortic intramural hematoma (IMH) in an elderly woman who presented with sudden onset of mid-scapular pain. The patient underwent a series of multimodality imaging studies before the diagnosis of IMH was evident by cardiovascular magnetic resonance, which is rapidly becoming the gold standard in the evaluation of acute aortic disease due to its high spatial resolution and ability to characterize tissue composition. Early diagnosis and prompt treatment is critical in improving the outcome of patients with IMH.

CASE REPORT

A 66-year-old female with a history of hypertension and tobacco use presented to her local emergency room after waking at 2 am with sudden, severe, pressure-like pain, located in the mid-scapular area of her back. Her associated symptoms included nausea, vomiting, left arm pain, and numbness.

Upon presentation, her heart rate was 40 bpm, and systolic blood pressure was 70 mmHg, with symmetric upper and lower extremity pulses. The electrocardiogram (ECG) demonstrated third degree atrioventricular block with 1 mm ST elevation in leads V1 and V2 and diffuse ST depression in the inferior-lateral leads. A subsequent ECG showed new inferior ST elevation, with resolution of the anterior ST elevation. Her initial cardiac enzymes were normal but later became elevated with a CK 376 U/L, CK-MB 40 ng/mL and Troponin T 1.2 ng/mL (normal ranges; CK = 20–200 U/L, CK-MB = 0–9 ng/mL, Troponin T = 0.0–0.1 ng/mL). The chest radiograph demon-

strated a widened mediastinum, and an MRI scan of her chest done at the outside hospital (details of which are not available to us) revealed a diffusely thickened (8–15 mm) aortic wall extending from the sinus of Valsalva to the level of the iliac arteries. There was no dissection flap evident or penetrating ulcer appreciated.

As the diagnosis was still unclear (the differential diagnosis included intramural hematoma, aortic dissection and vasculitis), she was transferred to our hospital for further management. She immediately underwent transesophageal echocardiogram (TEE), confirming the diffuse aortic wall thickening and the absence of an intimal dissection flap. MRI (Fig. 1), and CT scans were repeated at our institution. The MRI scan began with HASTE images in all three planes through out the thorax as “scout” images. Cine true fisp images were then obtained at the ascending aorta, descending aorta, aortic valve, and the left ventricle. TSE, T₁ and T₂ weighted images were obtained at the descending and ascending aorta which revealed marked thickening of the aortic wall that was circumferential in nature. A 3D contrast angiogram of the thoracic aorta was then obtained using the HASTE images as a reference. This revealed the penetrating ulcer in the descending aorta and did not reveal a dissecting flap. These MRI findings were consistent with the diagnosis of intramural hematoma (IMH) and a penetrating ulcer. Coronary angiography demonstrated a calcified and aneurismal left main coronary artery but no significant obstructive disease in any of the coronary vessels.

The following day, she underwent endovascular stenting of her penetrating ulcer with 2 overlapping nitinol stents and was discharged home 2 days later. A CT scan at one month follow-up

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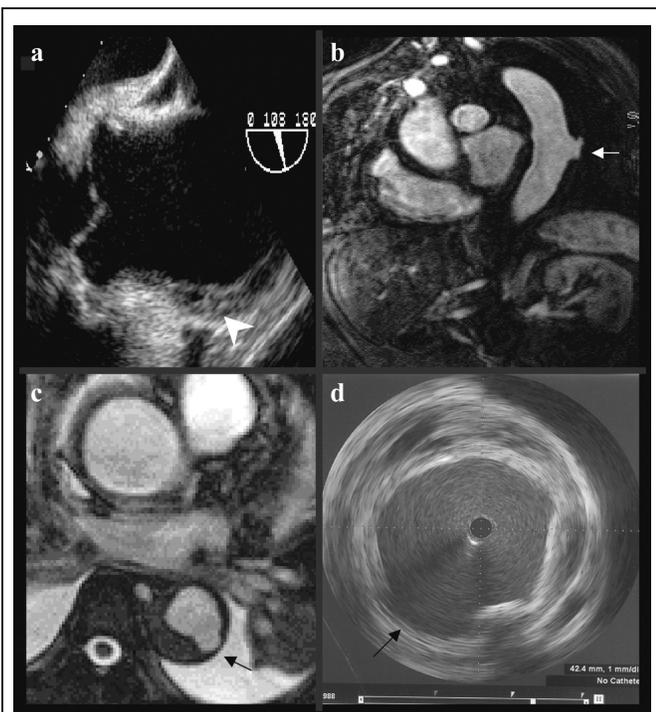


Figure 1. The ascending aorta by TEE showing thickened wall, identified by the arrowhead (A); contrast MRI of the descending aorta revealing extravasation into the penetrating ulcer as identified by the small arrow (B, C); and IVUS at the site of the penetrating ulcer as shown by the small arrow (D).

(the pictures of which are not available) showed dramatic resolution of the IMH.

Unfortunately, 2 months later, she returned to our hospital with severe back pain and was found to have a large dissection extending from the left subclavian artery to the proximal portion of the aortic stent. During surgical correction of the dissection, the dissection extended retrograde back to the aortic valve, leading to surgical replacement of the aorta and the aortic valve.

DISCUSSION

This patient presented with symptoms indicative of an acute aortic dissection. However, the absence of a dissection plane and the marked, diffuse thickening of the aortic wall led to the diagnostic confusion and delay in treatment. The recognition of aortic IMH in patients presenting with symptoms of aortic dissection has increased over the last decade owing primarily to improved imaging capabilities (1–5). There is still considerable controversy over the etiological basis and relationship of dissection and IMH. Clinical studies have indicated that IMH is a precursor of aortic dissection and a significant number of these cases progress to overt dissection (6, 7). The absence of continuous direct flow communication between true and false lumen

through an intimal tear is the fundamental pathologic feature that differentiates IMH from aortic dissection. The most common diagnostic criteria used for IMH include regional aortic wall thickness >7 mm without a dissection flap or symptoms of severe chest or back pain associated with CT findings consistent with IMH.

If acute aortic dissection is suspected, time is of the essence due to the high mortality rate in the initial hours of a type A dissection. Therefore, either CT scan or TEE is recommended as it is faster to obtain. However, if a patient is beyond the acute setting or the other imaging modalities are ambiguous, an MRI is an excellent diagnostic tool. CT scan has its own limitations including need for contrast and exposure to radiation. TEE has challenges of visualizing the aortic arch.

Depending on the acuity of the symptoms and the associated cause of the IMH, differing rates of mortality (0–89%) have been published in the literature for medical versus surgical management (1–2). Specifically, patients with an IMH and a coexisting penetrating atherosclerotic ulcer have been shown to have a higher rate of progression requiring surgery than those without a penetrating ulcer (4). Endovascular stent-graft repair has shown promise as a possible therapy in patients with penetrating atherosclerotic ulcers with or without an IMH (5). However, there are no data to help determine whether aortic stenting or surgical repair should be the therapy of choice and limited data exists on the intermediate and long-term efficacy of aortic stenting for IMH.

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