Aortic intramural hematoma: Importance of advanced MDCT imaging technique at diagnosis and follow-up.

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Learning objectives

The purpose of our educational exhibit is to:

1. Describe the classification, the main pathophysiology mechanisms and the clinical features of aortic intramural hematoma (IMH)
2. Illustrate a review of the main imaging findings of the disease with multidetector computed tomography (MDCT)
3. Describe the importance of advanced MDCT imaging technique for the diagnosis and assessment of natural history of IMH
Background

IMH accounts for 5-20% of cases of acute aortic syndrome together with aortic dissection (AD) and penetrating atherosclerotic ulcer (PAU) [1-5]. This pathology might evolve very dynamically in the short-term to regression, aortic enlargement, dissection, or even aortic rupture [1-5]. A huge advance in MDCT technology enables IMH-associated aortic lesions such as subtle intimal tears causing ulcer-like projections (ULPs) and aortic branch artery pseudoaneurysms (Ps) to be more frequently recognized. In this context, both the imaging quality and the deep knowledge of the main radiological manifestations of IMH are crucial.
Findings and procedure details

IMH Definition, Classification and Etiology

IMH is classically defined as a haematoma in the aortic wall that is distinguished from classic AD and haematoma secondary to PAU by the absence at imaging of any evident intimal tear [1-5]. The disease is classified in the same way as classic AD according to DeBakey or Stanford criteria [1-5] and it affects the descending aorta (Stanford type-B) more frequently, in almost 60% of cases [5].

Although the precise aetiology is still uncertain, the two most common pathogenetic theories are:

1. spontaneous rupture of the vasa vasorum in the media with intramural hemorrhage formation [6]; this is the most common theory reported in the literature;
2. microscopic (entry) tears in the aortic intima with false lumen thrombosis immediately thereafter, probably due to the absence of any re-entry (absence of outflow).

According to the last theory, in Japan, the term "thrombosed-type acute AD" has been preferably substituted for IMH [7]. A surgical study by Park et al. [8] showed that the prevalence of intimal tear in the ascending aorta or arch is high (73.0%) in the patients who were diagnosed as Stanford type A acute IMH. Therefore, in clinical practice IMH is defined as hemorrhage into the medial layer with thrombosed false lumen (absence of intramural longitudinal flow), regardless of small intimal tears [5].

Clinical Presentation and Natural History

The resulting hematoma may then propagate in an antegrade or a retrograde manner, producing symptoms that may be impossible to differentiate clinically from those of a classic AD [5]. Pain is characteristic of IMH, whereas malperfusion and pulse deficit are much less likely than with classic AD [5].

This pathology might evolve very dynamically in the short-term to [1-5]:

- Partial or complete resolution
- Remodelling:aortic aneurysm formation
- Rebleeding
- Development of an ulcer-like projection (ULP)
- Progression to AD
Rupture

According to the IRAD (International Registry of Acute Aortic Dissection), overall in-hospital mortality is similar to that of classic AD, and correlates with IMH location, being higher for type A IMH [5]: mortality for type A IMH compared to AD was 26.6% versus 26.5%; mortality for type B IMH was less but did not differ significantly (4.4% versus 11.1%) from classic AD. Type A IMH were managed mostly with surgery, whereas type B IMH were more frequently treated medically [5].

Predictors of disease progression [1-10].

- Involvement of the ascending aorta
- Maximum aortic diameter on initial CT scan (#5cm Ascending Aorta; >4 cm Descending Aorta)
- Progressive maximal aortic wall thickness
- Enlarging aortic diameter
- Persistent pain and/or hemodynamic instability
- Interval increase of associated pleural effusion
- Associated ULP

Role of MDCT Imaging

Typical features of IMH

At unenhanced MDCT[9]:

- IMH appears as hyperdense crescent-shaped wall thickening (usually >5 mm) due to intramural hemorrhage (Fig. 1 on page 8);

- displacement of sub-intimal calcification toward the aortic lumen may be observed (Fig. 1 on page 8);

- IMH tends to maintain a constant circumferential relationship with the aortic wall and not to spiral longitudinally (Fig. 1 on page 8, Fig. 2 on page 8);

- unlike mural thrombus, IMH shows smooth interior border (Fig. 1 on page 8).

At enhanced MDCT IMH[9]:

- IMH does not show contrast-enhancement (Fig. 1 on page 8).
Ulcer-like Projections (ULPs)

ULPs represent subtle sites of intimal disruption that may be observed at initial MDCT study or may develop during imaging follow-up (Fig. 3 on page 9, Fig. 4 on page 10, Fig. 5 on page 11, Fig. 6 on page 12, Fig. 7 on page 13, Fig. 8 on page 14):

- Prevalence of ULPs at imaging (initial ULPs, newly developed ULPs or both): 15-78% [7-8, 11-15]. Studies using CT scan with slices thickness >=2.5 mm (range: 2.5-5 mm) reported the lowest prevalence (15-33%) [11, 15], whereas recent studies using latest MDCT technology with para-millimetric thickness (range: 1-1.5 mm) reported the highest prevalence of ULPs (71-78%) [7, 12].

- Mean time between the initial event and the first appearance of ULPs: 17.8 days - 2.4 months, mainly within 30 days from the onset [17, 11, 13, 15];

- Progression (aortic enlargement, dissection, rupture) of ULPs: 31-70% of cases [7-8, 11-15]; it is higher for ULPs developed in the ascending aorta and decreases moving to the proximal descending aorta [7-8, 11-15] (Fig. 6 on page 12, Fig. 7 on page 13, Fig. 8 on page 14).

Aortic Branch Artery Pseudoaneurysm (Ps) in the context of IMH

- The pathogenesis of these lesions entails injury to the branch artery origin during the propagation of a dissecting hematoma[16-17]

- Ps are intramural collections of contrast material at the level of a branch artery, from which the aortic branch itself originates, associated with a small communication in the intimal-medial flap (between the true and false lumen of the IMH), corresponding to the branch artery origin (Fig. 9 on page 15).

- Ps may extend both along the aortic circumference and in a craniocaudal direction

- Ps more frequently involve intercostal, bronchial or lumbar arteries, in contrast to visceral vessels[16-17].

- The temporal presentation and sequential changes in size varied significantly. Most Ps were associated with complete or incomplete resorption or stability (86-95%)[16-18].

- In general, Ps appear to bear a relatively benign clinical course[16-19], although in some circumstances they need to be treated with endovascular embolization[20].
Importance of Advanced MDCT Imaging Technique

The above observations suggest that the detection (imaging prevalence) of ULPs is strictly dependent on the MDCT technology applied, e.g. many subtle and tiny ULPs may be overlooked at initial imaging study. Furthermore, using sub-optimal slice thickness (>\(=2.5\) mm), many Ps may be confused with ULPs and vice versa; this may negatively affect the imaging prognostication and the management of the disease in the individual patient with acute IMH.

Therefore, advanced MDCT imaging technique with ECG-gating for a motion-artifact-free evaluation of the ascending aorta using sub-para-millimetric slice thickness (0.7-1.2 mm) is recommended in the acute phase evaluation and during the follow-up. Especially for type A IMH, that often requires surgical intervention, coronary assessment is very important for a correct treatment planning.

An evaluation based on MPR reconstruction images is also essential for a detailed, precise and complete examination of the vessels. MPR images, which include curved planar reformation (CPR) and sagittal, coronal, and oblique reformations, are typically isotropic tomographic images (Fig. 10 on page 16, Fig. 11 on page 17, Fig. 12 on page 18).

\textit{CPR MPR} are obtained from a cutting plane parallel to the vessel and passing through its center, thus showing the anatomical details of the chosen vessel in one plane. The images of the vessel can be angled and rotated in all directions and planes, therefore highlighting some details that could be lost with an axial evaluation only. On the reconstructed images the real dimensions of the vessel and of the hematoma perpendicular to the longitudinal aortic axis can be easily measured and orthogonal images of the selected level are shown.

With \textit{virtual angioscopy}, internal vessels are seen as if a virtual endoscope is penetrating the body and viewing the vessel from the inside thus facilitating the detection of small ULPs. This allows an interactive fly-through animation with arbitrary positioning within the vessel (Fig. 13 on page 19 - Movie). In particular, it can be useful to assess the morphology of the ULPs (transversal vs. longitudinal tear respect to the major aortic axis) (Fig. 14 on page 20).
A. IMH at Unenhanced MDCT: hyperdense crescent-shaped wall thickening (arrows). Note the displacement of sub-intimal calcification (arrowhead).

B. IMH does not show contrast-enhancement.

Differential Diagnosis:
IMH vs. thrombosed FL: tends to maintain a constant circumferential relationship with the aortic wall and not to spiral longitudinally

IMH vs. mural thrombus: smooth interior border

Fig. 1

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Fig. 2

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Fig. 3

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Fig. 4

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Fig. 5

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Fig. 7

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Fig. 8

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Fig. 9: Pseudoaneurysms of the intercostal arteries at D8 and D9 level. A small pleural effusion can be seen on the left (asterisk).

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Fig. 10

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Fig. 11

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Type B IMH → 6-Month Follow up

Type B IMH- same case as in Fig. 10-11. At imaging FU, complete resorption of IMH with aneurysm evolution at the isthmus level (A) and development of an ULP at the proximal descending aorta (B)

Fig. 12

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**Fig. 13:** Virtual angioscopy: a fly-through animation from the proximal descending aorta to the ascending aorta (the aortic valve leaflets can be seen at the proximal extremity) demonstrating two intimal tears (see also fig. 14).

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Fig. 14

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Conclusion

Because a dynamic change in morphology or rapid evolution of IMH might be frequently observed, close clinical and imaging follow-up is mandatory. MDCT play a key role in assessing IMH and IMH-associated lesions (ULPs and Ps). Advanced MDCT imaging is fundamental for the diagnosis, assessment of natural course and of the acute and chronic complications during follow-up.
Personal information

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