Intramural Hematoma in Acute Aortic Syndrome

More Than One Variant of Dissection?

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Intramural hematoma (IMH) of the aorta came onto the clinical horizon with its recognition as a variant of aortic dissection and with the early diagnostic use of modern tomographic imaging modalities in the evaluation of acute thoracic pain syndromes. IMH, which is a precursor of dissection, cannot be reliably distinguished from classic dissection on clinical grounds and is considered to represent about 10% to 20% of all cases of acute aortic syndromes. Other syndromes include symptomatic aneurysm, penetrating ulcerations, and dissection.

Despite recent diagnostic advances, the complexities of aortic dissection remain the major challenge to cardiovascular medicine, with 30-day mortality rates of 27% for proximal and 29% for distal dissection even after surgery.1 In contrast to classic dissection, IMH is characterized by the absence of an aortic entry tear2–4 and the separation of media wall layers created by hematoma from rhexis of aortic vasa vasorum, which eventually leads to a secondary tear or to communication to the adventitial space. In both scenarios of progressive IMH, the evolution to overt dissection or even rupture may occur suddenly or be heralded by ongoing acute aortic syndrome. Unlike classic aortic dissection, IMH has no mechanisms of intramural decompression by a re-entry tear, but rather reveals intramural (intramedial) thickening or echolucent pockets of noncommunicating blood with potential for rupture or, at times, regression and resorption of the hematoma over time.4–9 Although the diagnostic implications of IMH continue to attract attention in the field of vascular medicine, predictions of both natural course and individual risk are far from reliable.9,10–15 Whereas our own data have always suggested that any IMH located in the ascending aorta should be considered a form of proximal dissection that should not be left without surgical repair,4 others have identified parameters, such as progressive maximal aortic wall thickness (increasing from 14±5 to 21±7 mm) and aortic diameter (increasing from 55±6 to 63±9 mm) as strong predictors of subsequent dissection with the need for surgical repair within days to weeks after the clinical presentation of acute aortic syndrome.10

In ancient Greece, Hippocratic physicians praised the skills of prognostication, which were usually used to foretell death. Some 2 millennia later, and after a brief period of decline, foretelling of death is enjoying a resurgence of interest in aortic disease. In this issue of Circulation, Ganaha et al16 unravel a new prognostic component for the individual risk assessment in 65 patients with evidence of IMH on tomographic imaging; in a retrospective analysis over 10 years, the presence of ulcer-like projections in areas of IMH was shown to identify a subset of patients at high risk.1 The authors found diagnostic evidence of penetrating atherosclerotic ulcers (PAU) in 34 patients with IMH preferentially (>90%) involving the descending aorta, whereas IMH without PAU was more frequently involving the ascending aorta. The authors claim both clinical symptoms and radiological findings, such as recurrent pain, pleural effusion, and both size and depth of an ulcer-like projection are predictive of disease progression.16 PAU is known to result from progressive erosion of atheromatose mural plaque penetrating the elastic lamina, separating media layers and setting the stage for adjacent intramural hematoma with either local or extensive progression.17,18 Symptomatic ulcers cause complications such as the formation of aortic aneurysm, pseudoaneurysm, and dissection, or even complete rupture.16–20 and are considered unpredictable.6,17,21 Thus, the observations by Ganaha et al16 that, in PAU, signs of clinical instability and evidence of progressive erosion or the combination of both are likely to herald disease progression, are logical and well documented as focal precipitation of a longstanding atherosclerotic process primarily in the descending aorta of elderly patients. Conversely, classic dissection or its preceding stage of intramural hematoma with no evidence of PAU are considered more prevalent in a middle-aged population. For both pathologies, however, pain of “acute aortic syndrome” is indicative of a dynamic element in the disease process and eventually of imminent events, and thus should focus our attention on diagnostic confirmation of progression with subsequent treatment by either surgical repair or interventional stent-graft placement.

Although the analysis by Ganaha et al16 identifies PAU as the common denominator of progression and adverse outcome, IMH without PAU may erratically suggest a stable disease course. A closer look, however, reveals that only 2 of 8 patients with IMH without PAU in the ascending aorta were alive at follow-up, while 2 died and 4 had undergone surgical repair;16 conversely, IMH of the descending aorta may indeed warrant watchful waiting, supporting a previous notion.3,4,5 Moreover, a recent meta-analysis on morbidity and mortality of both intramural hematoma and ulcerations of the aorta offers additional insight.8,17 The natural course of 168 cases of typical IMH

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without PAU led to overt aortic dissection in 25% of ascending aortic IMH and in 13% of descending IMH, ended in aortic rupture in 28% and 9%, respectively, or in stabilization in 28% and 76%, respectively; 30-day-mortality after surgery was 18% with repair of proximal IMH and 33% with surgery to distal IMH compared with 60% and 8% in-hospital mortality with medical treatment of proximal and distal IMH, respectively.

Patients with PAU tend to be older and have more risk factors and co-morbidities (hypertension: 85%; coronary artery disease: 61%; abdominal or thoracic aneurysm: 53%; chronic renal insufficiency: 31%; peripheral artery disease: 17%; and cerebrovascular accidents: 12%). Interestingly, the meta-analysis revealed an association of PAU with IMH in 73% and with acute aortic syndrome (chest and back pain) in 76%, with sudden onset in 68%. Only 16% were found with advanced calcification of a relatively short immobile dissecting flap. PAU of the ascending aorta led to dissection and rupture in 4 of 7 cases (57%) compared with 12% dissection and 5% death, respectively, in the descending aorta. The majority of medically managed patients with PAU in the ascending aorta died within 30 days of hospital admission, whereas 86% of patients with distal PAU had an uneventful long-term outcome without surgery, supporting the notion that distal (type B) PAU without clinical signs of instability should be followed by sequential imaging, whereas unstable distal PAU should undergo more aggressive treatment (possibly stent-graft placement); any proximal (type A) PAU should primarily be considered for surgical management.17 Thus, although PAU seems to be the common denominator of adverse events in the analysis by Ganaha et al,16 recent literature and current experience identified acute aortic syndrome, morphological evidence of progression (even in the absence of PAU), and, most importantly, the involvement of the ascending aorta as harbingers of adverse prognosis, with PAU considered a bystander with potential to transform from a quiescent state to progressive erosion. In this context, important questions remain unanswered, such as the fate of PAU without IMH, the temporal dynamics of IMH evolution, and the time course of regression of IMH.

Although retrospective in nature, the article by Ganaha et al16 is important and focuses on the reappraisal of combined analysis of clinical and morphological indices for prognostic assessment in patients with acute and chronic aortic disease. However, review of charts and images collected over 10 years of rapidly evolving radiographic imaging technology in an era of only sporadic awareness of acute aortic syndromes may limit firm conclusions. Moreover, the potential of more recent diagnostic tools, such as omniplane transesophageal ultrasound or 3D magnetic resonance angiography, has not been utilized, even though these methods are considered superior.22 Nevertheless, considering both the aging patient population in Western societies with prolonged survival despite hypertension and the better diagnostic strategies available to more patients, the cardiovascular community faces an increasing incidence of acute and chronic aortic problems that desperately need to be stratified. At this pivotal point in time, an elevated level of clinical awareness and the availability of modern imaging technology should trigger the interest in diagnosing and treating the complex of acute aortic syndrome, similar to previous efforts in acute coronary syndromes. Complete vascular staging in acute and chronic aortic diseases and uniform follow-up programs, as well as precise definitions of pathology, should be implemented into prospective registries of aortic diseases by a multidisciplinary team of physicians in an attempt to validate previous retrospective observations and to make the best use of evolving diagnostic and therapeutic strategies. Finally, we are in need of credible prognostic models that can support decisions for individual patient care independent of investigators, at different times, and in worldwide locations.

References

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