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EDITORIAL COMMENT

Type A Intramural Hematoma

Watchful Waiting Might Sometimes Be an Option*

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Intramural hematoma (IMH) of the aorta is an acute aortic syndrome that is thought to be a variant of the more common "classic" aortic dissection (AoD). In classic AoD, there is at least 1 tear in the aortic intima that allows luminal blood at systemic pressure to penetrate the aortic wall and, in turn, split the aortic medial layer in two, producing true and false lumens separated by a dissection flap. Conversely, in IMH, hemorrhage into the aortic wall occurs in the absence of a grossly visible tear in the intima or ongoing communication between the aortic lumen and the hematoma. When first described, IMH was attributed to rupture of the vasa vasorum within the aortic wall. However, more recent evidence suggests that IMH results from very small tears in the intima (1,2) that result in transient blood flow from the aortic lumen into the aortic wall, where the blood then thromboses in the form of a hematoma. Importantly, whereas in classic AoD there is active communication between the true and false lumens, in IMH there is none. Indeed, some have argued IMH should be called "thrombosed-type acute aortic dissection," "aortic dissection with complete thrombosis," or "aortic dissection with completely thrombosed false lumen." But, regardless of the nomenclature, the thrombosis of blood within the aortic wall gives IMH a distinctly different appearance from classic AoD on imaging studies.

In classic AoD, it is well recognized that patients with type A dissection are at high risk of early death, most often due to rupture into the pericardial space, and therefore the standard of care has long been open surgical repair of the ascending aorta. However, the natural history of type A IMH has been a bit less certain. Indeed, it is known that IMH, whether it involves the ascending or descending aorta, can evolve in a number of different ways. According to an expert consensus report by Evangelista et al. (3), at 1 year, IMH is slightly more likely to stabilize or regress than to progress (59% vs. 49%); nevertheless, at 1 year, 27% do progress to an aneurysm, 25% to a localized dissection or ulcer-like projection, 5% to classic dissection, and 4% to rupture (3). Indeed, proximal IMHs can progress within hours to days to classic AoDs, which in turn can rupture and cause death. Indeed, in studies of Western populations with type A IMH (4), mortality for medical versus surgical management was 47% versus 24%, respectively, which is quite similar to rates of 58% versus 26%, for a contemporaneous series of classic type A AoD reported by the International Registry of Acute Aortic Dissection (5). Consequently, the American College of Cardiology Foundation/American Heart Association/American Association of Thoracic Surgery and the European Society of Cardiology guidelines recommend treating type A IMH with urgent ascending aortic repair, as they do for classic AoD (6,7).

Intriguingly, the clinical experience with type A IMH among populations in Japan and South Korea has been quite different. In 2009, Song et al. (8) reported a series of 357 consecutive patients with type A acute aortic syndromes, 256 (72%) of whom had AoD and 101 (28%) had IMH. Urgent surgery was performed in only 16 (16%) of those with IMH due to instability, whereas the other 85 patients with IMH were initially

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stable and thus managed medically. The overall hospital mortality of the patients with IMH was comparable to that of surgically treated patients with AoD (8% vs. 11%). Of the 85 patients with IMH who were managed medically, at 6 months, adverse clinical events occurred in 31 (37%), which included progression to AoD in 25, delayed surgery in 25, and death in 6. Predictors of adverse events were an initial aorta diameter of ≥55 mm and hematoma thickness of ≥16 mm.

In a similar report by Kitai et al. in 2009 (9), of 66 patients presenting with type A IMH, 16 (24%) underwent surgical repair because of instability, and the other 50 (76%) were managed medically. Among the latter, 15 (30%) demonstrated progression to classic AoD or an increase in hematoma size within 30 days and underwent late surgical repair. The 30-day mortality rate was 6% for emergency surgery versus 4% for medical management. In medically treated patients, an aortic diameter ≥50 mm was highly predicted progression of the ascending IMH.

Based on the preceding and similar evidence, the Japanese Circulation Society guidelines recommend medical rather than surgical management of uncomplicated type A IMH provided that there are no ulcer-like projections in the ascending aorta, the hematoma thickness is <11 mm, and the aortic diameter is <50 mm (10). A 2020 report from Japan followed those guidelines with favorable outcomes, but the authors consider their strategy more akin to "watch and wait" than simply medical management, highlighting the fact that they repeated CT angiography at days 1, 3, 7, and 14 following admission (11).

In this issue of *JACC: Case Reports*, Vlastos et al. (12) present a case of a patient with a subacute type A IMH. The diameter of the aortic root was 45 mm and ascending aorta 39 mm, and the maximal diameter of the hematoma of the ascending aorta was only 5 mm. Because the patient had presented 5 days after the acute onset of his pain, the decision was made by a multidisciplinary team to follow a strategy of optimal medical management with anti-impulse therapy and serial computed tomography angiography (CTA). The patient had a benign course, and at 28 days his CTA demonstrated a decrease in hematoma thickness.

What relevance does this case, and the broader Japanese and South Korean experience have, then, for clinicians in Europe and America? The preceding suggest that among patients presenting with type A IMH, there may indeed be a subgroup who are at lower risk for adverse events. Thus, although the large majority of our patients with type A IMH should still be managed with urgent aortic surgery, in selected cases it may not be unreasonable to consider the alternative strategy of medical management and watchful waiting, provided the ascending aortic diameter is ≤50 mm and the hematoma is ≤12 mm.

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