Precipitous Resolution of Type-A Intramural Hematoma with Medical Management in a Patient with Metastatic Stage 4 Renal Cell Carcinoma

Camilo A. Velasquez, MD¹ Syed Usman Bin Mahmood, MBBS¹ Mohammad A. Zafar, MBBS¹
Adam J. Brownstein, BA¹ Ayman Saeyeldin, MD¹ Bulat A. Ziganshin, MD^{1,2} John A. Elefteriades, MD¹

¹Aortic Institute at Yale New Haven Hospital, Yale University School of Medicine, New Haven, Connecticut

²Department of Surgical Diseases 2, Kazan State Medical University, Kazan, Russia

Address for correspondence John A. Elefteriades, MD, Aortic Institute at Yale New Haven Hospital, Yale University School of Medicine, Clinic Building CB 317, 789 Howard Avenue, New Haven, CT 06519 (e-mail: john.elefteriades@yale.edu).

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Abstract

Keywords

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Intramural hematoma (IMH) is a variant form of aortic dissection characterized by involvement of the aortic media without the presence of an overt intimal flap. Surgical extirpation is the standard of care for type-A IMH in the Western world. However, a conservative approach with anti-impulse therapy has been advocated especially in Japan as a viable alternative. Here, we report a case of an elderly male patient with a history of metastatic stage 4 renal cell carcinoma who was treated with anti-impulse therapy for an acute type-A IMH. Blood pressure stabilization and continuous monitoring resulted in complete resolution of the IMH within 6 days. This report illustrates how immediate medical management in patients with acute type-A IMH who are not surgical candidates can alleviate the progression or even lead to regression as early as 1 week after initiating anti-impulse therapy.

Acute aortic syndrome (AAS) is a continuum of specific aortic diseases each of which portends an increased risk of morbidity and mortality.¹ Acute aortic dissection (AD), intramural hematoma (IMH), and penetrating aortic ulcer (PAU) all have a similar clinical presentation and can transform into one another, indicating a similar underlying pathophysiology.¹ AD is characterized by the presence of an intimal flap dividing the aortic lumen into a false and true lumen.² Despite being considered pathological variants of AD, IMH, and PAU are not associated with an intimal flap. In fact, an IMH is thought to arise from the rupture of the vasa vasorum in the media of the aortic wall and is referred to as a hematoma contained within the vessel wall.^{3,4}

IMH is second to AD in prevalence, accounting for 10 to 20% of all AAS,⁵ yet there are no clear guidelines regarding its optimal management. The Stanford classification for AD has

been the foundation for clinical decision-making in the treatment of IMH and PAU. Type-A IMH/PAU are managed surgically whereas type-B IMH/PAU are medically managed with conversion to surgery in the event of rapid aortic dilatation, impending rupture, and frank rupture.^{6,7} Nevertheless, this approach has been questioned by studies from Eastern countries. An unknown “Asian factor” has been invoked to explain the improved outcomes seen with medical management of type-A IMH in Japanese and Korean series.^{3,8} On the contrary, medical management has also been associated with an in-hospital mortality rate of 32%, questioning the effectiveness of an initial conservative therapy and reinforcing the concept that early surgical intervention may be warranted.⁹ An IMH has the potential to regress completely but also poses a very real risk of catastrophic complications, including aneurysm progression, frank AD,

and aortic rupture.⁵ It is this unpredictable natural progression of an IMH that confounds clinical decision-making.

We present the case of a 70-year-old man with a history of stage 4 renal cell carcinoma (RCC), who was admitted for management of an acute type-A IMH. Medical management resulted in resolution of the AAS within 6 days, as confirmed by serial imaging.

Case Report

We report the case of a 70-year-old male patient with a history of stage 4 RCC with metastasis to the adrenal glands, lymph nodes, lungs, and skin being treated with cabozantinib, an inhibitor of the tyrosine-protein kinase Met (cMET) and the vascular endothelial growth factor 2 (VEGFR2) used to reduce tumor growth, metastasis, and angiogenesis. He presented to the emergency department with complaints of pressure, such as lower back pain, abdominal pain, and bloating. Also, shortness of breath (SOB) and hypoxia were noted. As part of the workup for SOB, an echocardiogram showed a mild decrease in left ventricular ejection fraction (43%) and a dilated ascending aorta measuring 3.9 cm with a possible flap, raising concern for a dissection. Computed tomography angiography (CTA) scan of the chest revealed the presence of an IMH involving the aortic root, ascending aorta, aortic arch, descending thoracic aorta, and upper abdominal aorta. The maximal ascending aorta diameter was 4.3 cm at the level of the main pulmonary artery (►Fig. 1). The patient's RCC had been stable as per the Oncology Team, and they deemed surgical repair of the type-A IMH appropriate if indicated. The risk of progression to rupture and subsequent death without intervention was estimated to be 40%. Vascular surgery deemed the patient unsuitable for endovascular aortic repair of the descending IMH. However, the cardiac surgical team chose to manage the patient conservatively. Continuous monitoring and anti-impulse therapy were instituted in the cardiothoracic intensive care unit. Initially, hydralazine and nicardipine drips were

utilized to maintain systolic blood pressure below 120 mm Hg. Once this was achieved, the transition to oral labetalol and amlodipine was made. Follow-up CTA scan 6 days later demonstrated complete resolution of the acute IMH of the ascending aorta as well as a decrease in size from 4.3 to 3.3 cm with persistence of the descending IMH (►Fig. 2). Recommendation for continued medical therapy was made, and the patient was discharged home in satisfactory condition. He was seen in follow-up 2 months later with a normal-appearing ascending aorta and mild further enlargement of the descending aorta.

Discussion

IMH is described as a pathological variant of AD.⁷ It was first defined by Kruckenberg in 1920 as a dissection without an intimal flap.^{4,10} The pathophysiology of an IMH is attributed to the rupture of the vasa vasorum in the media of the aortic wall. However, this classical dogma for IMH formation has been recently challenged; the formation (or presence) of small intimal tears is now thought to be the inciting injury leading to IMH formation. Park et al¹¹ reported the presence of small intimal defects in 73% of patients who underwent surgery for type-A IMH. Uchida et al¹² confirmed the presence of an intimal tear in 78% of patients operated for an IMH, and they recommend the term thrombosed-type AD as an alternate term for IMH.

IMH has an unpredictable natural history. The clinical presentation is similar to that of any other AAS, with pain being the predominant symptom. Nevertheless, IMH tends to be present in older patients and has a female predominance in contrast to classical AD.⁴ As an IMH propagates, there is weakening of the aortic wall which may lead to aneurysmal dilation and as well as rupture, and fracture of the intima leading to overt dissection, a complication with an incidence of 12 to 47%.⁵ The presence or absence of spontaneous regression/resolution directly impacts the decision between surgical and medical treatment. Acute type-A IMH is treated surgically, whereas type-B IMH in the absence of complications is treated with anti-impulse therapy and strict follow-up.⁶ Conversely, this doctrine has been challenged by Asian studies that have shown good outcomes and resolution in medically treated patients with type-A IMH.^{2,13} Moizumi et al¹⁴ found that among the patients in their study, more than 50% were safely treated with medical therapy alone without the development of an overt complication. They postulated that the lack of flow in the thrombosed lumen decreases the risk of conversion to rupture.

Our patient presented with a symptomatic acute type-A IMH, and generally, these patients are treated surgically at our institution. However, the IMH developed in the context of a patient with stage 4 RCC with multiple metastases in the lung, adrenal glands, lymph nodes, and skin, findings which portend a poor prognosis with survival rates at 5 years of 8%.¹⁵ We felt aggressive surgical therapy was inappropriate. A medical management strategy with strict surveillance for life-threatening complications was adopted and proved fruitful.

Fig. 1 Chest computed tomography angiography at admission. Axial view demonstrating the presence of ascending intramural hematoma (arrow) extending to the descending thoracic aorta (arrowhead).

Fig. 2 Chest computed tomography angiography at admission showing the crescent type hematoma characteristic of Intramural hematoma (IMH) at admission and the resolution after 6 days of medical therapy, demonstrating interval resolution (arrows). The ascending IMH is resolved completely after 6 days of anti-impulse therapy. The descending IMH persists. (A) Axial view; (B) coronal view.

Table 1 A separate case, also a patient with RCC, with ascending IMH resolving in 24 h

Case report author	Case characteristics	IMH location	Time to resolution
Ohmi et al ¹⁶	Patient with RCC and two episodes of IMH. Second episode associated with left-sided hemothorax	First episode: Descending thoracic aorta Second episode: Ascending thoracic aorta	First episode: Undetermined Second episode: 24 h

Abbreviations: IMH, Intramural hematoma; RCC, renal cell carcinoma.

The uniqueness of this case lies in the complete regression of the ascending aortic IMH in a short period, thereby decreasing the risk of rupture or overt dissection by reducing the elevated shearing forces on the aortic wall. To the best of our knowledge, regression of an IMH has been described as early as 1 month,⁵ with one case report demonstrating rapid regression in less than 24 hours in another patient also with a history of RCC¹⁶ (► **Table 1**). Our group encourages surgical approach for patients with acute type-A IMH.⁴ However, this case highlights the role of medical management in a subset of patients in whom a surgical approach might not be best. This case also

demonstrates that adequate control of systolic blood pressure can engender regression of a type-A IMH as early as 6 days and thus reduce the risk of immediate catastrophic complications.

Conclusion

Anti-impulse therapy for acute type-A IMH is a valuable alternative for patients in whom the potential risk of an open surgical approach outweighs the benefits. Despite the Western consensus that type-A AAS should be managed surgically, autonomous regression is still one possible outcome.

Acknowledgment

None.

Conflict of Interest

None.

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