Intramural Hematoma of the Ascending Aorta; Diagnosis, Management and Outcome

17

Martin Czerny and Jürg Schmidli

Abstract

The first description of IMH was given by Krukenberg in 1920 being defined at that time as aortic dissection without an intimal flap or tear that would lead to direct flow communication between the true and the false lumen. IMH was determined as to arise from hemorrhage of the vasa vasorum located within the medial layer of the aorta. The reason for the hemorrhage was thought to be the consequence of spontaneous rupture of the vasa vasorum. However, later on it was noted that hematoma formation within the aortic wall could also be the consequence of PAU.

Symptoms of IMH are very similar to those of acute aortic dissection and they may well be indistinguishable. Patients present with a history of chest and/or back pain and with a history of hypertension. Chest pain is associated with type A IMH and back pain is associated with type B IMH. Interestingly, patients with IMH report more intensive pain than patients with aortic dissection. In addition, patients with IMH are less likely to suffer from malperfusion syndrome. Finally, patients with IMH are in general one decade older than patients with classical acute aortic dissection.

The natural progression of IMH is unpredictable. While some cases of IMH progress to dissection, aneurysm or rupture, others remain stable, regress or completely resolve. Recent literature has identified several potential risk factors for progression including the presence of PAU, older age, an aortic diameter larger than 5 cm and wall thickness of the hematoma greater than 1 cm.

M. Czerny, MD, MBA (✉)
Department of Cardiovascular Surgery,
Inselspital Bern, Freiburgstrasse, Bern 3010,
Switzerland
e-mail: martin.czerny@insel.ch

J. Schmidli, MD
Department of Cardiovascular Surgery,
Inselspital, University Hospital Bern,
Freiburgstrasse, Bern 3010, Switzerland

R.S. Bonser et al. (eds.), *Controversies in Aortic Dissection and Aneurysmal Disease*,
DOI 10.1007/978-1-4471-5622-2_17,

The main objective of treatment of IMH is the prevention of aortic rupture as well to prevent the progression to classic aortic dissection. As type A IMH has a high and early risk of complication and death with medical treatment alone, surgery is usually indicated. Treatment of type B IMH is less straightforward as the prognosis is less uniform and predictable. A more conservative approach for uncomplicated type B IMH such as antihypertensive treatment and watchful monitoring is currently preferred as it appears to be a safer strategy. However, in some cases the disease might still progress despite optimal medical treatment.

Keywords
Intramural hematoma • Type A aortic dissection • Entry tear • TEVAR

Introduction

Intramural hematoma (IMH), penetrating atherosclerotic ulcer (PAU) and aortic dissection constitute the heterogenous group of acute aortic syndromes with the potential for transitions from one to the other [1]. Approximately 5–15 % of acute aortic syndromes are diagnosed as IMH [2]. In the biggest registry to date, the International Regsitry of Aortic Disscetion (IRAD), the incidence is quoted as 5.7 % [3] whereas the incidence in Asian countries is by far higher (Japan/Korea).

The first description of IMH was given by Krukenberg in 1920 being defined at that time as aortic dissection without an intimal flap or tear that would lead to direct flow communication between the true and the false lumen [4]. IMH was determined as to arise from hemorrhage of the vasa vasorum located within the medial layer of the aorta [5]. The reason for the hemorrhage was thought to be the consequence of spontaneous rupture of the vasa vasorum. However, later on it was noted that hematoma formation within the aortic wall could also be the consequence of PAU [6].

The Stanford classification of acute aortic dissection is also applicable and useful to classify IMH, meaning that type A IMH involves the ascending aorta and type B IMH involves the descending aorta, Another classification has been proposed by the European Society for Cardiology (ESC) in which IMH is classified into two distinct types. Type I IMH shows a smooth inner aortic lumen with the diameter usually less than 3.5 cm. Type II IMH has a rough inner surface due to severe aortic atherosclerosis with a dilated aortic diameter of more than 3.5 cm containing calcium deposits within the aortic wall [7].

Symptoms of IMH are very similar to those of acute aortic dissection and they may well be indistinguishable. Patients present with a history of chest and/or back pain and with a history of hypertension. Chest pain is associated with type A IMH and back pain is associated with type B IMH. Interestingly, patients with IMH report more intensive pain than patients with aortic dissection [8]. In addition, patients with IMH are less likely to suffer from malperfusion syndrome [9]. Finally, patients with IMH are in general one decade older than patients with classical acute aortic dissection [3].

The natural progression of IMH is unpredictable. While some cases of IMH progress to dissection, aneurysm or rupture, others remain stable, regress or completely resolve. Recent literature has identified several potential risk factors for progression including the presence of PAU, older age, an aortic diameter larger than 5 cm and wall thickness of the hematoma greater than 1 cm [8, 10].

The main objective of treatment of IMH is the prevention of aortic rupture as well the to prevent the progression to classic aortic dissection. As type A IMH has a high and early risk of complication and death with medical treatment alone,

surgery is usually indicated [5]. Treatment of type B IMH is less straightforward as the prognosis is less uniform and predictable [10, 11]. A more conservative approach for uncomplicated type B IMH such as antihypertensive treatment and watchful monitoring is currently preferred as it appears to be a safer strategy [12]. However, in some cases the disease might still progress despite optimal medical treatment [2].

Thoracic endovascular aortic repair (TEVAR) seems to be an excellent strategy in the treatment of the disease in particular when intimal disruptions are identified as aortic wall thickening is efficiently reduced by decompression [13].

This chapter should guide the reader to:

- Identify information regarding the natural history of acute type A IMH
- Identify imaging modalities to diagnose acute type A IMH
- Identify data regarding outcomes following medical treatment, surgical repair and TEVAR to treat acute type A IMH
- Identify information regarding recommendations of treatment of acute type A IMH

Natural History of Type A IMH

In contrast to type B IMH which has a natural history well comparable to the natural history of acute type B aortic dissection, there is broad consensus that the natural history of type A IMH is well comparable to the natural history of acute type A aortic dissection and therefore requires immediate surgical treatment. The combination of IMH and an ulcer like projection (ULP) or PAU are in particular indicative for an accelerated disease progression whereas the ones without ULP or PAU do have the potential to spontaneously regress [14].

There seems to be a difference in the natural course of the disease dependent on the geographical localization and ethnical origin whereas the natural course in Caucasians seems to be worse than in Asians [15] whereas it has to be stated that irrespective of geography and ethnics, the incidence of adverse clinical events in initially stable patients remains high.

Diagnosis of Type A IMH

Computed tomography represents the mainstay of diagnosis of acute type A IMH. To allow pulsation free visualization of the thoracic aorta up to the aortic root, a retrospective ECG-gating technique is recommended. Primarily, a crescentic or circular high-attenuation area along the aortic wall, seen without contrast enhancement on CT is regarded as diagnostic of IMH.

Afterwards our routine protocol for these examinations is performed as follows- CT angiograms are performed in the arterial phase during the intravenous administration of nonionic iodinated contrast material. A bolus triggering technique assesses the contrast medium transit time. A threshold of 150 HU (absolute) is used. In a biphasic fashion, 130 mL contrast agent is administered: the first 30 mL are injected at a flow rate of 6 mL/s followed by the second 100 mL administered at 5 mL/s. A saline flush of 40 mL is given after the contrast medium injection to optimize contrast utilization. With a post-threshold delay of 8 s, ECG-gated CT angiography of the entire aorta is performed using the following imaging parameters: a slice collimation of 64×0.625 and a pitch of 0.29 is used. The matrix size was 512×512. Images are reconstructed with a slice thickness of 1.4 and a slice increment of 1 mm as well as with a slice thickness of 3 and a slice increment of 2 mm without using the ECG-triggering ("untagged images"). Additionally, image redistribution according to the heart action is performed after scanning (retrospective ECG-triggering). Usually, images are triggered in a mid phase at 55 % of a R-R interval. If pulsation artefacts occur, other phases of the heart cycle are reconstructed. For every phase, two series are reconstructed (1.4/1 mm as well as 3/2 mm) [6].

By this approach a substantial number of patients presenting with the combination of IMH and PAU or ULP can be identified.

Transesophageal echocardiography (TEE) represents an excellent method to establish the diagnosis of IMH as well as to potentially detect small intimal tears especially in the descending thoracic aorta. However, it has to be taken into account that placing a TEE probe into the

esophagus in an awake patient, might abruptly increase intrathoracic pressure and may thereby expose individuals to rupture [16].

Outcomes Following Medical Treatment, Surgical Repair and TEVAR to Treat IMH

Medical Treatment

A recent Asian study described a series of patients with type A IMH as compared to classical type A aortic dissection in their institution. They excluded all patients with a visible PAU in the proximal thoracic aorta. One-hundred and one patients were analyzed where the primary indication for surgery was pericardial tamponade which was the case in 16 patients. The remaining ones underwent a watchful waiting strategy with serial close imaging examinations by echo and/or CTA. Initial medical treatment was by intravenous injection of a beta adrenergic receptor blocker accompanied by a long-lasting calcium channel blocker [17].

A substantial number of patients finally underwent surgery mostly due to the progression of type A IMH into classical aortic dissection. The mean duration between onset of symptoms and surgery was 27 days, interestingly the mortality rate was very low, as could be linked to the fact that the majority of these operations was then performed on an elective basis. Overall, among 85 stable type A IMH patients who received initial medical treatment, 31 (36.5 %) experienced adverse clinical events including development of classical type A aortic dissection (n = 25), delayed surgery (n = 25) or death (n = 6) within 6 months. In multivariate analysis, syncope during the initial event, hematoma thickness and aortic diameter turned out to be independent predictors for the development of adverse clinical events after initial medical treatment.

The best cutoff values for hematoma thickness and aortic diameter where 16 and 55 mm respectively. The event free survival rate was significantly different on whether a patient had neither, either or both risk factors [17]. Another recent study from Japan had similar conclusions with regard to cutoff values with regard to aortic diameter and increase in hematoma size [18].

Surgical Repair

The conceptual surgical approach in treating type A IMH is similar to the conceptual approach in treating type A aortic dissection consisting of ascending and hemiarch replacement as the lowest common denominator. This approach might be altered by the presence of an intimal tear, ULP or PAU in downstream aortic segments where the addition of a stent-graft or the application of a combined prosthesis (E-vita open®, Jotec GmbH, Hechingen, Germany) might add in achieving remodeling of the entire thoracic aorta. Surgical results regarding mortality in most series are well comparable to the ones after surgery for acute type A aortic dissection and range between 7 and 15 % in the current literature [19]. As in surgery for acute type A aortic dissection, current organ protection strategies are recommended such as selective antegrade cerebral perfusion during hypothermic circulatory arrest [20].

A very important message is that the after surgical treatment of type A IMH, freedom from aortic-related reintervention and freedom from any kind of aortic-related adverse event is extremely low. Despite serial follow-up examinations are recommended the probability for any kind of secondary intervention is low single digit [17].

TEVAR

There is clearly no place for TEVAR in the treatment of type A IMH without any signs of an intimal defect, ULP or PAU. However, if one of these distinct entities is detecting via advanced imaging modalities, closure of this defect might present an excellent strategy. Nevertheless, there is a big difference if this defect is located in the ascending aorta or in downstream aortic segments where retrograde extension of IMH has been causative for the involvement of the ascending aorta as these

cases might present a highly attractive scenario for TEVAR [6].

It is now known that- assumed an intimal defect of any kind can be detected- extension of IMH or also of classical aortic dissection has a retrograde component in the majority of cases [21]. Consequently, causative treatment of the underlying pathology is not necessarily destined to the entire extension of the disease but may well be treated by addressing this very primary intimal lesion irrespective of its site. By TEVAR, the defect can be closed and remodeling of the entire thoracic aorta can be achieved. It seems likely that the majority of such lesions can be located in the distal aortic arch at the concavity [6].

Recommendations of Treatment of Acute Type A IMH

Summarizing the mortality risk in patients with acute type A IMH is somewhat lower than in classical type A aortic dissection. In the very patients with additional ULP/PAU, tamponade or periaortic hematoma, emergency surgery is the treatment of choice. In addition, the high risk of clinical adverse events in stable patients with type A IMH without the previously mentioned signs favor early surgical repair. Finally, in a subgroup of patients where the mechanism of type A IMH can be clearly traced to a intimal lesion in the distal aortic arch or the descending aorta, TEVAR as less invasive but at least equally effective strategy, might be chosen.

References

1. Vilacosta I, Roman JA. Acute aortic syndrome. Heart. 2001;85:365–8.
2. Schlatter T, Auriol J, Marcheix B, Lebbadi M, Marachet MA, Dang-Tran KD, Tran M, Honton B, Gardette V, Rousseau H. Type B intramural hematoma of the aorta: evolution and prognostic value of intimal erosion. J Vasc Interv Radiol. 2011;22:533–41.
3. Evangelista A, Mukherjee D, Mehta R, O'Gara PT, Fattori R, Cooper JV, Smith DE, Oh JK, Hutchison S, Sechtem U, Isselbacher EM, Nienaber CA, Pape LA, Eagle KA, International Registry of Aortic Dissection (IRAD) Investigators. Acute intramural hematoma of the aorta: a mystery in evolution. Circulation. 2005; 111:1063–70.
4. Krukenberg E. Beitrage zur Frage des Aneurysma dissecans. Beitr Pathol Anat Allg Pathol. 1920;67: 329–51.
5. Reich DL, Sen S, Shinn JA, Svensson LG, Williams DM, American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines; American Association for Thoracic Surgery; American College of Radiology; American Stroke Association; Society of Cardiovascular Anesthesiologists; Society for Cardiovascular Angiography and Interventions; Society of Interventional Radiology; Society of Thoracic Surgeons; Society for Vascular Medicine. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease. A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. J Am Coll Cardiol. 2010;55:e27–129.
6. Grimm M, Loewe C, Gottardi R, Funovics M, Zimpfer D, Rodler S, Dorfmeister M, Schoder M, Lammer J, Aharinejad S, Czerny M. Novel insights into the mechanisms and treatment of intramural hematoma affecting the entire thoracic aorta. Ann Thorac Surg. 2008;86:453–6.
7. Erbel R, Alfonso F, Boileau C, Dirsch O, Eber B, Haverich A, Rakowski H, Struyven J, Radegran K, Sechtem U, Taylor J, Zollikofer C, Klein WW, Mulder B, Providencia LA, Task Force on Aortic Dissection, European Society of Cardiology. Diagnosis and management of aortic dissection. Eur Heart J. 2001;18: 1642–81.
8. Sundt T. Intramural hematoma and penetrating atherosclerotic ulcer of the aorta. Ann Thorac Surg. 2007;83:835–41.
9. Sundt T. Intramural hematoma and penetrating aortic ulcer. Curr Opin Cardiol. 2007;22:504–9.
10. Chao C, Walker G, Kalva S. Natural history and CT appearances of aortic intramural hematoma. Radiographics. 2009;29:791–804.
11. Li DL, Zhang HK, Cai YY, Jin W, Chen XD, Tian L, Li M. Acute type B aortic intramural hematoma: treatment strategy and the role of endovascular repair. J Endovasc Ther. 2010;17:617–21.
12. Dake M. Acute aortic intramural hematoma: current therapeutic strategy. Heart. 2004;90:375–8.
13. Monnin-Bares V, Thony F, Rodiere M, Bach V, Hacini R, Blin D, Ferretti G. Endovascular stent-graft management of aortic intramural hematomas. J Vasc Interv Radiol. 2009;20:713–21.
14. Kitai T, Kaji S, Yamamuro A, Tani T, Kinoshita M, Ehara N, Kobori A, Kim K, Kita T, Furukawa Y.

Detection of intimal defect by 64-row multidetector computed tomography in patients with acute aortic intramural hematoma. Circulation. 2011;124(11 Suppl):S174–8.
15. von Kodolitsch Y, Csösz SK, Koschyk DH, Schalwat I, Loose R, Karck M, Dieckmann C, Fattori R, Haverich A, Berger J, Meinertz T, Nienaber CA. Intramural hematoma of the aorta: predictors of progression to dissection and rupture. Circulation. 2003;107:1158–63.
16. Jánosi RA, Buck T, Erbel R, Eggebrecht H. Role of echocardiography in the diagnosis of acute aortic syndrome. Minerva Cardioangiol. 2010;58(3):409–20.
17. Song JK, Yim JH, Ahn JM, Kim DH, Kang JW, Lee TY, Song JM, Choo SJ, Kang DH, Chung CH, Lee JW, Lim TH. Outcomes of patients with acute type a aortic intramural hematoma. Circulation. 2009; 120(21):2046–52.
18. Kitai T, Kaji S, Yamamuro A, Tani T, Tamita K, Kinoshita M, Ehara N, Kobori A, Nasu M, Okada Y, Furukawa Y. Clinical outcomes of medical therapy and timely operation in initially diagnosed type a aortic intramural hematoma: a 20-year experience. Circulation. 2009;120(11 Suppl):S292–8.
19. Estrera AL, Safi HJ. Acute type A aortic dissection: surgical intervention for all: PRO. Cardiol Clin. 2010;28:317–23.
20. Czerny M, Krähenbühl E, Reineke D, Sodeck G, Englberger L, Weber A, Schmidli J, Kadner A, Erdoes G, Schoenhoff F, Jenni H, Stalder M, Carrel T. Mortality and neurologic injury after surgical repair with hypothermic circulatory arrest in acute and chronic proximal thoracic aortic pathology: effect of age on outcome. Circulation. 2011;124:1407–13.
21. Loewe C, Czerny M, Sodeck GH, Ta J, Schoder M, Funovics M, Dumfarth J, Ehrlich M, Grimm M, Lammer J. A new mechanism by which an acute type B aortic dissection is primarily complicated, becomes complicated, or remains uncomplicated. Ann Thorac Surg. 2012;93:1215–22.