CASE REPORT

A Massive Aortic Intramural Haematoma — an Aortic Emergency

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ABSTRACT

Aortic intramural haematoma (AIH) is a rarely recognized disease characterized by a sudden haemorrhage into aortic media in the absence of any intimal tear. The clinical evolution and mortality rates of AIH are similar to those of acute aortic dissection. However, in the acute clinical care of patients presenting with chest pain of aortic origin, it is important to differentiate intramural haematoma from aortic dissection. A case of an elderly patient with an intramural hematoma (IMH), which progressed to very large dimensions and involved the entire aortic wall, resulting in fatal complications is presented here.

Keywords: aortic hematoma, intramural, aortic syndrome

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INTRODUCTION

Aortic intramural haematoma (AIH) represents a specific type of aortic disease which is considered to be a severe cardiovascular emergency. The symptomatology, the risk factors (hypertension, pregnancy), and the position of the lesion, usually at the level of the ascending aorta, aortic arch or both in type A, or at the level of descending aorta in type B, are similar to those associated with aortic dissection. Due to this similarity, 13 to 27% of cases diagnosed as having aortic dissection are in fact intramural haematomas (IMH). However, in contrast to an aortic aneurysm, in which a laceration of the aortic intima and the inner layer of the aortic media form a rupture that allows blood to enter and split the aortic media creating an intimal flap, in IMH both the rupture and the intimal flap are absent.

The outcome for patients with IMH is very poor in the absence of prompt treatment, and the appropriate management of IMH in an emergency department is essential. Treatment procedure depends mainly on the location of the hematoma, and surgical intervention is usually required in cases of proximal localization, whereas aggressive medical therapy appears justified in cases of descending aorta involvement. However, clear guidelines for treatment of this disease have yet to be established. According to published data, the progression to classic aortic dissection occurs in 28 to 74% of cases.¹ and aortic rupture in 20 to 45% of cases.² Each of these complications can be life-threatening and any signs of...
rapid deterioration in any of these complications or development of a penetrating aortic ulcer, indicate urgent surgical repair.

Factors which increase the risk of deterioration are age >70 years, a maximum haematoma diameter >10 mm or an aortic diameter >50 mm. Total resolution has been described in 5 to 20% of AIH cases.3

CASE PRESENTATION

An 84-year-old woman with a history of untreated essential hypertension presented at the emergency unit with prolonged chest pain in the inter–scapulovertebral region. This had started 5 to 7 days before, progressing in intensity and associated with shortness of breath. The patient had a medical history of scoliosis and reported a syncopal episode two weeks before, which did not require any specific treatment.

The informed consent was obtained from the patient for publication of this case report and any accompanying images. The publication of this case was accepted by the Ethics Committee of the hospital.

A general examination uncovered no abnormal findings, except an elevated blood pressure of 190/100 mmHg, there being no difference between measurements from the left or right arm, and an increased heart rate of 110 beats per minute. Her oxygen saturation was 98%, and both lungs were clear on auscultation. Laboratory results indicated values within normal limits for all biochemical parameters, including specific cardiac biomarkers (cTnI, D-Dimers, NT-proBNP). Results of an electrocardiogram showed normal sinus rhythm without any significant changes of the ST segment or T waves. Echocardiography did not reveal any wall motion abnormality, thus excluding an acute coronary syndrome, or any sign of intimal flap in the aortic lumen, thus excluding an aortic dissection. As the diagnosis was unclear at this stage, the patient underwent unenhanced computed tomography examination, which revealed a low attenuation hallow starting immediately below the origin of the left subclavian artery and extending down to the right external iliac artery, producing a luminal stenosis of 50% at that level. The maximum diameter of the aortic lumen was 9 mm at the level of the aortic arch. Parietal calcifications were visualized in the center of the aorta, without any capitation of contrast substance, visceral involvement or intimal flap (Figure 1, Figure 2). The patient was diagnosed as having an intramural haematoma and hypertensive spurt and was treated with intravenous nitroglycerin and beta-blockers. At 72 hours after the onset of symptoms, subsequent to medication for high blood pressure, the patient still presented with recurrent chest pain.

Repeated 64-multislice detector computed tomography was performed, which confirmed the presence of a

![Figure 1](image1.png)  **Figure 1.** Multislice Angio CT of the aorta, showing intraluminal low attenuation represented by an intramural haematoma

![Figure 2](image2.png)  **Figure 2.** 3D reconstruction of the aortic wall with an intramural haematoma
giant intramural haematoma involving the entire aortic wall, in addition to a jagged edged mushroom-like aortic wall lesion, positioned 7 mm from the sino-tubular junction on the ventral part of the ascending aorta. No local calcifications were present and there was minimal pleural effusion (Figure 3). The final diagnosis was a massive type A intramural haematoma with an ulcer-like protrusion in the proximal part of the ascending aorta.

The patient refused surgical intervention and was discharged after two days having responded well to adjusted antihypertensive treatment. However, the patient died suddenly two days after discharge from the hospital.

**DISCUSSION**

IMH has been described in 5 to 20% of patients who presented to the emergency department with symptoms and signs suggestive of acute aortic syndrome, mostly with severe hypertension, but without a history of trauma.

Based on the location of the lesion, two different types of IMH can be distinguished, according to the Stanford classification: type A is located proximally, involving the ascending aorta and type B is located distally, without involvement of the ascending aorta. Most IMH are located in the descending aorta, while the ascending aorta is more frequently involved in cases of aortic dissection. One third of type A IMH develop a penetrating aortic ulcer within the first three months of follow-up, probably resulting from a new intimal tear rupture, and 20–45% of them progress to saccular dilatation in the first six months of follow-up.\(^2\)\(^3\) Other described complications are aortic dissection in 28 to 47% of cases,\(^4\) pleural effusion, cardiac tamponade or aortic valve insufficiency. In 10% of cases there is a spontaneous and complete absorption of the haematoma.\(^3\)

As in any type of aortic disease, the clinical presentation is atypical and imaging has a crucial role in the early diagnosis and initiation of an appropriate treatment strategy, including the risk assessment in the follow-up period.\(^3\) Currently non-invasive investigations such as transoesophageal or transthoracic echocardiography have been replaced with 64-multislice detector computed tomography or magnetic resonance imaging (MRI) for diagnosis of IMH.\(^6\)

In asymptomatic patients, treatment is directed towards reduction of the workload of the left ventricular ejection, while in symptomatic patients, presenting at acute care units, the medical therapy targets heart rate and blood pressure reduction, together with prompt analgesia for reduction of catecholamine induced tachycardia and hypertension. Usually such measures have been already initiated in the emergency department. The target systolic pressure is 100 to 130 mmHg, and target heart rate is 60 to 80 bpm. First line medication includes beta-blockers in combination with vasodilator therapy such as sodium nitroprusside. In patients with asthma, bradycardia, heart failure or severe lung disease, who have the potential of intolerance to beta-blockers, short-acting esmolol, intravenous verapamil or diltiazem represent reasonable alternatives. As these cases are almost always associated with a diffuse atherosclerotic process, irrespective of the location of IMH, statin therapy should be initiated according to current guidelines (Class IIa, Level of Evidence: C).\(^3\)

In addition to medical treatment, patients with type A IMH should undergo surgical intervention because of the high risk of rupture or the development of dissection.\(^7\) Surgical intervention can be delayed in patients clinically stable at two to three days after medical stabilization. Alternative non-surgical techniques include stent-graft implantation when anatomical conditions allow this procedure, or in selected cases, a hybrid approach using both surgical and endovascular techniques. Surgical treatment of a penetrating aortic ulcer involves aortic reconstruction with the removal of the affected segment of the aorta and placement of a graft. However, a clear indication for urgent or delayed surgical management of ascending aorta IMH, versus medical treatment alone has not yet been established. Delayed surgery repair therapy within three
days from the onset of symptoms does not change the instantaneous risk of atypical dissection episode, but an increased risk of conversion up to and beyond eight days was noted.\(^3\)

Comparing the morbidity and mortality rates after medical and surgical treatment, the results showed a 30-day mortality of 18% associated with surgical repair, and 33% associated with surgery to a distal IMH compared to 60% and 8% mortality rate respectively, attendant on medical treatment of type A and type B IMH.\(^8\)

According to current recommendations, type B IMH should be treated with beta-blockers and other vasodilator drugs in order to control the blood pressure, the target values for blood pressure being set to less than 140/90 mmHg in patients without diabetes or less than 130/80 mmHg in patients with diabetes or chronic renal disease. This regime for type B IMH is associated with superior outcomes compared to its use in type A IMH or to aortic dissection.\(^9,10\) Treatment should be followed using imaging techniques.\(^11\) The predictors for regression of the acute phase of type B IMH are represented by an aortic diameter less than 40 mm and a thickness less than 10 mm,\(^12\) while the presence of a penetrating atherosclerotic ulcer, or a high level of C-reactive protein have been correlated with a significant risk for further deterioration of a type B IMH lesion.

In stable patients with IMH, but without penetrating aortic ulcer who respond to medical treatment, the intervention of endovascular or surgical reconstruction can be delayed.\(^3\) Symptomatic ulcers associated with recurrent chest pain are more prone to develop a dissection or rupture of the vessel. In these patients, endovascular stent grafting is emerging as an attractive therapeutic modality.

Isolated IMH, without the presence of any intimal tear, are considered relatively stable lesions,\(^13\) resulting in total regression in a significant number of patients, without any changes in aorta size or morphology. However, this condition is not without the development of serious complications and medically managed patients require to be closely monitored for at least two weeks and a routine long-term follow-up regime put in place.

In cases of IMH associated with penetrating aortic ulcer (PAU), several predictors of potential deterioration have been described, such as, sustained or recurrent pain (\(p <0.0001\)), increasing pleural effusion (\(p = 0.0003\)), enlarged aortic diameter, greater than 4.8 cm, progressive maximum aortic wall thickness greater than 11 mm, maximum PAU diameter >20 mm (\(p = 0.004\)), and maximum PAU depth >10 mm (\(p = 0.003\)). Contrary to these findings, Patel \textit{et al.} demonstrated that neither aortic nor PAU size-related criteria may be helpful in predicting PAU progression.\(^14\) Nevertheless, urgent surgical intervention should be considered in these cases, even in the absence of a clear indication for surgical intervention.

The present reported case demonstrated that in a patient with an extended IMH, accompanied by the presence of clear signs that the disease may progress, a conservative approach, in the absence of surgical intervention, can lead to death despite an encouraging and favorable response to medical treatment. In the present case, the patient refused surgery despite recurrent chest pain and despite clear signs of progression of the aortic hematoma as evidenced by serial CT examinations. The IMH clearly progressed over several days, reaching a massive size and involving the entire aortic wall. The patient felt well after several days of administration of intravenous nitroglycerin and blood pressure lowering agents, but this did not prevent the adverse progression of the disease, resulting in the death of the patient.

CONCLUSION

IMH is a serious condition that can be life-threatening. The optimal treatment strategy for type A intramural hematomas depends on the characteristics of each individual case. As the present case showed, a patient with IMH and apparently stable clinical status can rapidly progress to severe deterioration of the clinical parameters and death, due to the enlargement of the hematoma. Computed tomographic imaging plays a crucial role in assessing patients with an intramural hematomas, being superior to any other available non-invasive method.

CONFLICT OF INTEREST

None declared.

REFERENCES


