Unusual Case of Overt Aortic Dissection Mimicking Aortic Intramural Hematoma

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We report an interesting case in which overt aortic dissection mimicked two episodes of aortic intramural hematoma (IMH) (Stanford A, DeBakey I). This took place over the course of four days and had a major influence on the surgical treatment strategy. The first episode of IMH regressed completely within 15 hours after it was clinically diagnosed and verified using imaging techniques. The recurrence of IMH was detected three days thereafter, resulting in an urgent surgical intervention. Overt aortic dissection with evidence of an intimal tear was diagnosed intraoperatively.

Key words: 1. Aortic diseases  
2. Aortic dissection  
3. Aortic aneurysm

CASE REPORT

A 62-year-old woman presented to the emergency department of Central Hospital Bad Berka with nausea and an episode of acute chest pain. She had been previously diagnosed with a myxofibrosarcoma of the left forearm (April 2013), which had been treated surgically. At that time, the maximal diameter of her ascending aorta was 47 mm, and antihypertensive therapy with a calcium-channel blocker was initiated. A contrast-enhanced computed tomography (CT) scan upon admission to the emergency unit showed an eccentric hypodense thickening of the ascending aorta and aortic arch. The maximal thickness of the intramural hematoma (IMH) was 25 mm in the convexity of the ascending aorta and 10 mm in the aortic arch. No intimal tears or ulcer-like projections were detected. Moreover, no pleural effusion and only a small pericardial effusion were detected (Fig. 1A). Despite the presence of an intramural hematoma, the patient’s clinical symptoms did not recur, and she remained stable without progression of the pericardial effusion on echocardiographic follow-up. The IMH was interpreted as a stable and non-emergency finding, and the patient was treated medically.

Due to an episode of recurrent chest pain, the patient was referred to our cardiac surgical unit the next morning. As a part of a preoperative work-up in preparation for urgent surgery, another follow-up CT scan was conducted. The rationale behind this decision was to determine whether the status of the IMH had changed and to determine whether an intimal tear could be detected. To our surprise, significant regression of the aortic IMH was found, with only minor residual thickening in the aortic arch and virtually no detectable hematoma in the ascending aorta (Fig. 1B). Transesophageal echocardiography was then performed, demonstrating a complete disappearance of the IMH (Fig. 2). Moderate aortic valve re-

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Fig. 1. Transverse planes of the three consecutive preoperative CT scans. (A) The first CT scan at the time of hospital admission showing an eccentric hypodense IMH (arrow) involving the ascending aorta (1/23/2015). No evidence of pleural or pericardial effusion is present. (B) The second CT scan, showing complete regression of the IMH 15 hours after the initial CT scan. Note the increasing pleural effusion, especially at the right hemithorax. (C) The third CT scan, showing the recurrence of IMH 72 hours after the second CT scan as well as progressive right-sided pleural effusion. CT, computed tomography; IMH, intramural hematoma.

Fig. 2. Transoesophageal echocardiography conducted after the second computed tomography scan demonstrates no evidence of proximal aortic involvement.

gurgitation was detected, showing no obvious changes in comparison with previous echocardiographic findings. We decided to postpone surgery and to closely monitor the patient.

Although the patient remained asymptomatic, the recurrence of aortic IMH was diagnosed 72 hours after the second CT scan. We were surprised to find a recurrence of aortic IMH, especially since it predominantly involved the ascending aorta, with a maximal IMH thickness of 12 mm (Fig. 1C). Moreover, progressive pericardial effusion and pleural effusion were also detected. Due to the recurrence of type A IMH, in combination with the presence of aortic valve regurgitation and the increasing degree of pericardial effusion, the decision was made to undertake urgent surgery. Intraoperatively, an unanticipated intimal tear was identified in the noncoronary sinus at the level of the sinotubular junction (Fig. 3). Due to restriction of the left coronary cusp causing aortic regurgitation and moderate root dilatation (an aortic root diameter of 45 mm), root replacement (Bentall technique) using a bioprosthesis was conducted, as well as a partial aortic arch replacement. The patient’s postoperative course was uneventful and she patient was discharged on the eighth postoperative day. The unusual dynamics of this aortic IMH are demonstrated in the frontal reconstructions of consecutive CT scans (Fig. 4).

**DISCUSSION**

Aortic IMH has often been diagnosed as a variant of aortic dissection using modern imaging techniques. It may present with the clinical symptoms of aortic dissection, and may occasionally precede overt aortic dissection or rupture [1]. The clinical course of IMH has been reported to vary, ranging from progression to an overt aortic dissection to complete disappearance over the course of follow-up [2]. The serial changes of IMH during follow-up have yet to be established,
whereas its disappearance has been linked to a good long-term prognosis [3]. We report herein a patient in whom two episodes of aortic IMH (Stanford A, DeBakey I) occurred within four days, influencing the surgical treatment strategy.

Aortic IMH is a variant of overt dissection and may account for up to one of three to four cases of acute aortic syndrome [4]. IMH may completely resolve during follow-up, or it may progress to a classic aortic dissection [2]. In the largest case series, reported by Song et al. [5], the vast majority of the type A IMH patients (84%) were classified as being hemodynamically stable and hence were treated medically, resulting in a mortality rate of only 7.1%, whereas the International Registry of Aortic Dissection reported a mortality rate of medically treated type A IMH of as high as 33% [6].

Similarly to our patient, Ohmi et al. [7] reported a case in which an IMH exhibited rapid regression, affecting the descending aorta during the initial presentation and involving the ascending aorta one year later. On both occasions, the IMH regressed rapidly just hours after it was diagnosed. No aortic surgery was conducted due to the rapid and complete resolution of the IMH. The phenomenon of the regression and/or disappearance of an IMH is intriguing, and may explain to some extent the ongoing controversy with regard to
its optimal treatment. Nishigami et al. [3] claimed that the disappearance of an IMH suggests a good long-term prognosis. Although no significant changes were observed during the course of follow-up among their patients with an IMH that disappeared, they did not present guidelines regarding whether such cases of regressed IMHs should be frequently checked for malignant changes. Therefore, the results from this single study should be interpreted with caution and cannot be generalized to the entire spectrum of regressed IMHs. The rigid classification of IMH patients into ‘disappearance’ and ‘progressive’ groups might be suboptimal, as it may only represent different stages of ongoing aortic disease.

The clinical course of IMH is very similar to that of a possible overt aortic dissection, mainly because an IMH diagnosed using imaging techniques may precede an overt aortic dissection [1,2]. Although the resorption of an IMH may occur without specific treatment [3,7], a substantial number of medically treated patients may suffer sudden cardiac death during the course of follow-up. Moreover, data regarding the time course of IMH remain absent. Therefore, no consensus has emerged regarding the most appropriate surveillance strategy in asymptomatic patients with a known history of the regression of an aortic type A IMH. Nevertheless, as previously reported [5], new-onset pericardial and pleural effusion have been reported to be more common in IMH patients than in patients with an overt aortic dissection. Due to the fact that our patient presented with these two indirect signs of IMH-related complications and an initial IMH thickness of 25 mm, a rather aggressive treatment strategy was chosen in order to prevent the possibility of impending sudden death. The definite decision to perform surgical treatment in the present patient was made due to the recurrence of an IMH on the third CT scan, which demonstrated the malignant character of this case of acute aortic syndrome. Despite aortic imaging by means of serial CT scans and transesophageal echocardiography, the intimal tear and overt aortic dissection were only confirmed intraoperatively. Furthermore, it remains uncertain whether the intimal tear remained undetected or developed secondarily during the course of the IMH. Nevertheless, the majority of patients undergoing surgical treatment for acute type A IMH may present intraoperatively with an intimal tear [8]. The lack of a visible tear on the CT scan may contribute to the delay of surgical treatment. This possibility means that the appropriateness of aortic imaging as currently implemented should be questioned, and further raises the issue of whether other markers would lead to more accurate decision-making in the treatment of acute aortic syndrome.

**CONFLICT OF INTEREST**
No potential conflict of interest relevant to this article was reported.

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