Atypical Presentation of Intramural Hematoma of the Ascending Aorta Using a Conservative Approach

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ABSTRACT

Aortic intramural hematoma is a severe disease, secondary to vasa vasorum bleeding and/or aortic micro-ulcers without connection with the true lumen of the vessel. Its occasional finding in the absence of symptoms or aortic trauma is quite rare. In this report, an asymptomatic female patient with involvement of the ascending aorta and transverse arch was maintained on clinical treatment, despite the prevailing recommendations for surgical repair in Stanford type A lesions. Relevant aspects of lesion characterization in different imaging tests, identification of high risk anatomical criteria and conduct are discussed. A satisfactory 7-month follow-up was observed.


Case Report

Since the original description of acute aortic syndromes,1 the growing diagnostic readiness and the availability of sophisticated imaging studies have allowed an early identification of patients with acute and typical aortic scenarios. In the case of intramural hematomas (IMH), as occurs in classic cases of dissection, the agility in diagnosis and the immediate treatment, especially for lesions classified as type A, are also crucial to success. A more widespread recognition of these diseases and the frequent use of advanced diagnostic methods can promote a clinical situation in which the image outweighs the importance or clinical significance of the disease itself. In this study, the case of an asymptomatic female patient presenting IMH of ascending aorta is described, with emphasis on diagnostic imaging and on medical management.

CASE REPORT

A female patient, aged 49, white, with a history of controlled hypertension and glucose intolerance, underwent preoperative evaluation for a hysterectomy for uterine myomatosis, during which an angiotomography of coronary arteries was requested, due to a family history of early coronary disease. In this procedure, performed on an outpatient basis, the presence of IMH of ascending aorta and transverse arch was observed as an incidental finding, occupying the entire circumference of the aorta, with a maximum thickness of 8 mm and discrete proximal progression to cervical vessels. Furthermore, a slight compression of the right coronary artery ostium was noticed (Figure 1).

The patient was referred for hospital admission and medical management. In a previous clinical history, she related an episode of prolonged fever that had occurred 16 years ago, accompanied by pleural effusion, when an investigation for systemic lupus erythematosus and tuberculosis (sic) was performed and subsequently discarded.

Since then, the patient was asymptomatic and practicing regular exercise (cycling); the woman denied...
previous episodes of chest pain or chest trauma at any time. Furthermore, no manifestation compatible with systemic inflammatory disease was perceived. The patient denied any family history of aortic disease or sudden death.

At the time of hospital admission, laboratory exams revealed metabolic profile, serum protein electrophoresis, and inflammatory activity profile (C-reactive protein, erythrocyte sedimentation rate, serum complement, rheumatoid factor, and antinuclear antibodies) with in normality; human immunodeficiency virus and syphilis serology were also negative. The hemogram revealed anemia (hemoglobin = 10.3 g/dL), which was considered compatible with her long-term hypermenorrhea. The electrocardiogram showed first-degree atioventricular block, and an echocardiogram revealed a mild thickening of aortic valve, with a mean gradient of 8 mmHg.

Considering that the patient was clinically stable, she was maintained under close observation in the hospital, and a first control with computerized tomography angiography (CT-A) of thoracic aorta was scheduled for the fifth day of hospitalization, in association with positron emission tomography (PET-CT), which demonstrated stability in aortic diameter (maximum diameter = 34 mm), predominance of the hematoma in posterolateral portions to the left of the ascending aorta, and with an apparently slight reduction of the hematoma, but with maintenance of a thickness of 8 mm. The pulmonary artery diameter was normal. In PET-CT, fluoro-deoxyglucose (FDG) uptake was observed in the portion corresponding to the highest volume of the hematoma (maximum standard uptake value – SUV max = 4.4), without abnormal uptake in other parts of the aorta or organs (Figure 2).

The patient was discharged, having been told to avoid contact sports or resistance exercises, and was prescribed beta blocker, angiotensin receptor blocker, and statin. At that time, a new control was scheduled to be held in 15 to 20 days, if the patient remained
asymptomatic. This third CT-Acontrol revealed hema-
toma stability.

After seven months, the patient remained asymptom-
atic and in regular use of the above-mentioned drugs
and a new CT scan of thoracic aorta revealed minimal
reduction in IMH thickness (7 mm) and maintenance
of aortic diameter (Figure 3).

DISCUSSION

Corresponding to about 6 to 10% of cases of acute
aortic syndrome, IMH of ascending aorta diagnosed dur-
ing the investigation of an episode of acute chest pain
is a severe disease with high mortality. The possibility
of progression to dissection or to frank rupture (20 to
50%) underlies the indication of immediate surgical
treatment for a great number of patients, particularly
those whose injuries are situated in the ascending aorta.
However, in recent years, several studies – especially in
the Asian population – have favored a strategy of close
observation for stable patients without signs of poor
prognosis, e.g., persistent pain, aortic diameter increased
on admission (> 48-55 mm), hematoma thickness > 11
mm, a concomitant presence of ulcer-like images (not
necessarily characteristic of a penetrating ulcer), or
hematoma progression. Thus, in stable patients, the
follow-up with imaging studies is a mandatory step in
the monitoring and in treatment indication.

Asymptomatic IMH is rarely described in the litera-
ture (two cases). One patient exhibited progression to
a penetrating ulcer and underwent surgical correction, and
another patient with vague symptoms underwent
an emergency thoracotomy, during which an aortitis ac-
compained by wall thickening was diagnosed, without
replacement of the aortic segment affected.

Despite the extent of IMH found in this case and
not withstanding its classification (Stanford type A),
the option in favor of a conservative management was
based on an absence of clinical manifestations of the
disease and on CT criteria for high risk, weighed against
the morbidity and mortality inherent to an extensive
aortic surgery.

In the case of an asymptomatic patient undergo-
ing CT or PET-CT for any indication (coronary heart
disease investigation, cancer monitoring, etc.) and in
whom IMH was detected, it is critical to be alert to
the possibility of differential diagnoses. In the present
case, images synchronized with the electrocardiogram
were requested in order to exclude artifacts, and the
possibility of aortitis was investigated, although, at first,
the patient’s clinical evaluation and laboratory tests
were not suggestive of this hypothesis.

The characteristic finding of IMH is the presence
of a thrombus (crescent-shaped, or occupying the en-
tire aortic circumference) in the vessel wall, without
communication with the vessel lumen, caused by vasa
vasorum rupture or by the occurrence of undetected
micro ruptures or micro ulcers. With multi-row detector
CT-A, the wall thickening with high attenuation (60-70
Hounsfield Units – HU), observed in the unenhanced
series, is clearly seen in the contrast-enhanced series
as a separate image, with no communication with the lumen. In MRI, additionally to the aforementioned anatomical aspect, a hyper-intense signal on T2-weighted sequences and an iso- or hyper-intense signal on T1-weighted sequences (black-blood), depending on the stage of evolution, are characteristic findings.

In cases of aortitis, the characteristic finding of CT-A in the unenhanced phase, as opposed to hematomata, is the low wall attenuation (< 40 HU). Another picture that leads to confusion is the presence of an intraluminal mural thrombus, often not distinguishable by CT studies. In case of persistent doubt, the differentiation may be achieved with magnetic resonance imaging (MRI), due to the presence of a hypo-intense or iso-intense signal.

While PET-CT is used predominantly in malignancy or inflammatory disease investigations, FDG uptake is also observed in cases of atherosclerosis and in the presence of a parietal thrombus. In the present case, PET-CT revealed a concentrated hyper-uptake area at the point of greatest volume and excluded uptake occurrence in other aortic portions or organs. The use of PET-CT in the prognostic assessment of patients, correlating the marked uptake with an increased risk of expansion and rupture, is still contradictory, but this may be a useful marker.

At this stage, no further investigation on myocardial ischemia was conducted, considering that the patient exhibited only mild compression in the right coronary

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Figure 3 – Multi-row detector computerized tomography angiography performed 7 months after treatment. Although the measured thickness of the hematoma is slightly smaller, with an apparent reduction in hematoma volume and maintenance of aortic diameters, the values are still within the range of variation of the method (8 and 7 mm, initial and final). Transverse sequences at pulmonary artery bifurcation (A) and aortic arc (B) levels. Reconstruction at the level of right coronary artery outflow tract (C) and longitudinal section of aortic arch and great vessels (D).
artery ostium, and also because the possibility of surgery was not excluded.

In the authors’ opinion, the clinical and imaging follow-up was an appropriate conduct in this case, avoiding a large surgical procedure, and also considering that the control follow-up revealed IMH stability.

The main stays of conservative treatment are the knowledge of relevant data pertaining to the diagnostic investigation by imaging studies and the association of these findings with the clinical assessment. The rapid development of these imaging methods and the low frequency of aortic diseases in the cardiology practice require a multidisciplinary participation. It is required that radiology services (or, when necessary, the physician) are accustomed to deliver reports that include the largest amount of relevant information for an initial risk classification, even if imperfect, and that inform the attenuation values observed in unenhanced series. An appropriate and sufficient clinical care strategy includes the classes of drugs that probably play a role in the reduction of aortic events\(^\text{10}\) (beta blockers, ACE inhibitors or angiotensin receptor blockers, and statins) and patient monitoring by imaging studies (preferably with nuclear magnetic resonance, to reduce exposure to X-ray). Considering that the pathophysiology of aortic diseases has only begun to be elucidated, it is possible to assume that silent episodes of intramural bleeding can occur as a mechanism of early dilation, or even of dissection. In this particular case, the possibility of progression to a late aortic dilation cannot be excluded.

CONFLICTS OF INTEREST
The authors declare no conflicts of interest.

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