How to mimic a type A intramural hematoma: A case of IgG4-related aortitis associated with cerebral hemorrhage

Title page

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IgG4-related aortitis is an inflammatory condition of the aorta, characterized by aortic wall thickening and periaortic soft tissue involvement. Therefore, this condition can radiologically mimic an aortic intramural hematoma. We hereby report the case of an IgG4-related aortitis misdiagnosed as an intramural hematoma, associated with cerebral hemorrhage, possibly due to cerebral vascular system involvement.

Key words: IgG4-related aortitis, intramural hematoma, cerebral hemorrhage

Introduction

Aortic intramural hematoma (IMH) is a pathological process within the spectrum of acute aortic syndromes, associated with high mortality and morbidity. Diagnosis requires a high level of suspicion and it is confirmed by a computed tomography (CT)-angiogram of the aorta. Nonetheless, a CT scan suggestive of intramural hematoma can potentially hide a completely different pathology. Differential diagnosis includes IgG4-related aortitis, which is a rare inflammatory condition of the aorta, characterized by aortic wall thickening and periaortic soft tissue involvement, associated with high serum levels of IgG4 and non-specific onset symptoms [1]. We hereby report the case of an IgG4-related aortitis associated with cerebral hemorrhage, mimicking an aortic intramural hematoma.

Case report

A 61-year old man ~~of Lebanese origin~~ with new-onset double vision and paraesthesia to the right hand, right side of the face and left side of the tongue, lasting 24 hours, was admitted to our Emergency Department with a suspect of stroke. He did not complain of any pain whatsoever. A brain CT scan showed a small cerebral hemorrhage at the level of the left posterior pons, associated with a vascular lacuna at the level of the right anterior capsule and an irregular course of the left vertebral artery. An image suggestive for intramural hematoma was found at the level of the aortic arch and the origin of the innominate artery, therefore, due to this incidental finding, the scan was extended to the entire length of the aorta, showing a 57 mm ascending aorta aneurysm, with periaortic thickening, extended to the proximal arch and to the origin of the brachiocefalic trunk, suggestive of type A intramural hematoma. Due to the cerebral hemorrhage, decision was taken not to operate immediately and serial CT-scans showed a complete resolution of the cerebral lesion 11 days after the first episode and confirmed the intramural hematoma at the level of the aneurysmatic ascending aorta, with hemorrhagic suffusion of the periaortic adipous tissue (Figure 1). A new finding of contrast-enhancement of the proximal ascending aorta lesion was found with suspect flogistic features. The patient underwent ascending aorta and partial arch replacement with no post-operative issues. Intra-operative findings included an extensive thickening of the aortic wall, inflammatory involvement of the periaortic soft tissue and, as an incidental finding, also a bicuspid aortic valve. No trace of intramural hematoma was found. At the

post-operative CT-angiogram a right axillary colliquative lymphadenopathy was found. The patient was discharged at post-operative day 8. Pathological examination was available only after the discharge, revealing an IgG4-related aortitis with lymphocytes and plasma cells infiltration and severe chronic IgG4-related periaortitis. The patient was therefore referred to the Immunology Department for further treatment.

Comment

IgG4-related disease is a rare inflammatory condition characterized by high serum levels of IgG4 and infiltration of lymphocytes and IgG4-bearing plasma cells in the affected organs, mainly lacrimal and submandibular glands, pancreas and lymph nodes. Nonetheless, it can also involve the vascular system, generally targeting great and medium-sized vessels, mainly the aorta in 10-30% of the cases, showing as aortitis and periaortitis. 60-85% of the patients affected by IgG4-related vasculitis are males, usually older than 50 years, as in the case described [1,2].

A typical CT-finding in this type of presentation is aortic wall thickening and contrast-enhancement, along with periaortic soft tissue involvement. Due to these particular findings, IgG4-related aortitis can be easily misdiagnosed as an intramural hematoma. Therefore, although the incidence of this condition is low [3], IgG4-related aortitis has to be taken into account in the differential diagnosis of aortic intramural hematoma, as well as other rare conditions, which can mimic the disease radiologically and clinically, such as giant cell aortitis, Takayasu arteritis and even some neoplastic disease [4-6].

In the case hereby described, the first suspicion of inflammatory disease of the aorta was raised intra-operatively, since an extensive thickening of the aortic wall with a fibro-inflammatory periaortic reaction was found, in place of the expected intramural hematoma. The diagnosis of IgG4-related aortitis was confirmed at the histopathology examination only after the discharge of the patient. In light of this finding, looking back at the initial presentation, we can presume that the focal cerebral hemorrhage, causing visual disturbances and paraesthesia to the patient, might have been due to a blood pressure peak on a background of an inflammatory process involving cerebral vascular structures, as demonstrated by the vascular lacunae and the irregular course of the abovementioned cerebral vessels.

A possible immunological involvement of cerebral structures in IgG4-related disease has already been hypothesized [7].

IgG4-related aortitis can be treated conservatively with the use of glucocorticoids and/or other immunosuppressants and the treatment has to be undertaken urgently due to the risk of high enlargement rate and rupture [1]. Nonetheless, in this case the presence of an aortic aneurysm, associated with bicuspid aortic valve, implied in any case a mandatory surgical correction.

Conclusion

This is the first reported case of IgG4-related aortitis associated with cerebral vascular involvement. This type of inflammatory disease of the aorta can be radiologically misdiagnosed as an aortic intramural hematoma, implying different treatment options. Clinical features, which in this case were actually quite misleading due to the cerebral involvement, can help to spot the right diagnosis, optimizing the disease management.

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Figure legends

Figure 1. Pre-operative CT-angiogram showing aortic wall thickening and periaortic soft tissue soffusion