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Aortic dissection of the young subject: think of Takayasu's arteritis!

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Abstract

Introduction: Takayashu vasculitis is a chronic inflammatory disease of unknown etiology affecting large and medium caliber arteries. Its clinical presentation is very polymorphic and is responsible for a delay in diagnosis.

The aorta and its main branches are the most affected with aneurysmal involvement in the majority of cases. Nevertheless, aortic dissection is an extremely rare complication, often diagnosed during the occlusive phase. It requires a multidisciplinary management and its prognosis depends on the evolutionary complications.

Case report: We report the case of a young man admitted for management of intermittent claudication of the right lower limb in connection with a type B aortic dissection and occlusion of the right primitive iliac artery.

Conclusion: Aortic dissection is a rare manifestation of Takayashu's aortopathy, the evolution of which can be life-threatening in case of delayed diagnosis.

Key words

Lower limb claudication, Aortic dissection, Takayashu arteritis

Introduction

Takayasu's arteritis (TA) is a non-specific inflammatory disease affecting the aorta and its branches. Its clinical presentation is polymorphic due to the heterogeneity of the affected vessels, making diagnosis

generally difficult. It is responsible for lesions such as occlusions, stenoses or aneurysms of the affected arteries. (1) Aortic dissection is an extremely rare complication, particularly in men (2).
We report the case of a young man admitted for management of a claudication of the lower limb in connection with an aortic dissection revealing takayasu aortopathy.

Clinical case

This was a 40-year-old patient, a former smoker, admitted for a claudication of the right lower limb evolving for 6 months, with no known cardiovascular history or systemic disease. On physical examination, the heart rate was regular at 90 beats per minute without any additional heart murmur, the blood pressure was 220/110mmhg. The rest of the examination was unremarkable. The electrocardiogram showed a regular sinus rhythm without signs of cavitary hypertrophy. The C-reactive protein (CRP) was 10mg. Transthoracic echocardiography was unremarkable. The thoracoabdominal angiogram showed a type B aortic dissection over the entire descending and abdominal aorta with occlusion of the right iliac artery (Figure 1). Our patient was referred to the department of internal medicine for further management. The main differential diagnosis evoked in front of this clinical presentation was Horton's disease. An immunological workup was negative. According to the criteria of the American College of Rheumatology, the diagnosis of Takayasu disease was retained. The patient was put on corticosteroid therapy and optimal antihypertensive treatment with a good clinical evolution.

Discussion

First described in 1908 by the Japanese ophthalmologist Mikito Takayasu, TA is a chronic inflammatory vasculitis of unknown etiology involving large and medium caliber vessels, mainly the aorta and its main branches and the pulmonary arteries. (3) The aorta is involved in 60% of cases, but aortic dissection remains an exceptional complication of this disease. Rare cases have been reported in the literature. (4-5)

TA has been reported extensively in Asia, more specifically in Japan, South America and Africa. Its prevalence is still poorly known throughout the world. However, it is estimated to be between 1.2 and 3.6 cases per million inhabitants per year. (4)
TA frequently affects young women but can be seen at any age (2, 4, 5). However, its occurrence after the age of 40 is not uncommon (4).

Anatomically, TA is characterized by panarteritis with lymphocytic and macrophagic infiltration of the adventitia and media of the arterial wall. Recently, histologic evidence is no longer a diagnostic criterion for this pathology. (4, 6)

The clinical presentation of TA is very variable depending on the evolutionary phase of the pathology, which explains the delay in diagnosis. TA is characterized by 2 phases: a pre-occlusive phase generally asymptomatic or associated with non-specific general signs and an occlusive phase characterized by vascular symptoms depending on the degree of inflammation of the arteries (stenoses or occlusions). These manifestations may or may not be associated with an inflammatory syndrome. (4)
Our patient presented with claudication of the lower limbs and low back pain, reflecting the occlusive phase of the disease, with a Stanford type B aortic dissection on angioscan with occlusion of the right primitive iliac artery. The main differential diagnosis evoked in our patient was that of Horton's disease characterized by temporoparietal headaches, jaw claudication, positive temporal biopsy and ocular involvement. The absence of these signs ruled out this diagnosis in our subject.

In a Japanese study reported by Wu et al in 2017, the majority of aortic involvement was type B. (5)

The American College of Rheumatology has proposed a classification based on a set of clinical and imaging arguments to establish the diagnosis, which remains a real challenge for practitioners. (7) Our patient presented 4 of the 6 criteria of the ACR in favor of Takayasu arteritis.

The treatment of aortic dissection associated with Takayasu's arteritis is based on a combined management.
Corticosteroid therapy at a dose of 0.7 to 1 mg/kg/day is the treatment of choice for this vasculitis. In case of non-remission, immunosuppressive therapy is necessary. In addition, 60% of patients report clinical and biological improvement with resolution of inflammatory involvement of the aorta, stenoses and aneurysms.

The surgical management of type B aortic dissection is complex, with a high operative and postoperative morbidity. It should be reserved for a minority of patients after stabilization of the disease. (8) Interventional treatment of type B aortic dissection has resulted in better outcomes than vascular surgery.
It is important to note that corticosteroid therapy must be maintained postoperatively. (5)

The prognosis of aortic dissection in Takayasu disease is poor and depends on progressive complications. (2, 5)

Conclusion

Aortic dissection in Takayasu disease is a very rare condition requiring multidisciplinary management. Appropriate treatment, including antihypertensive therapy and corticosteroid therapy, can help control the extension of the dissection. Complicated aortic dissections should be actively treated with endovascular stenting or surgery.

Competing interests

The authors declare no competing interest.

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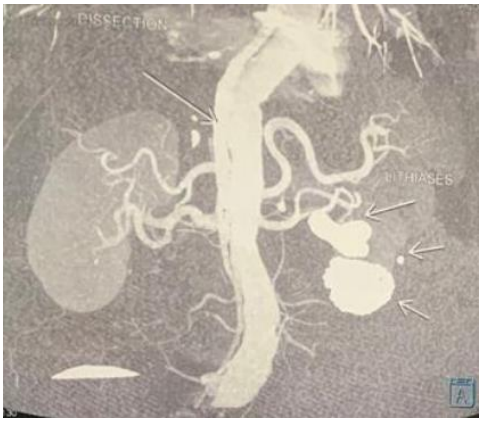
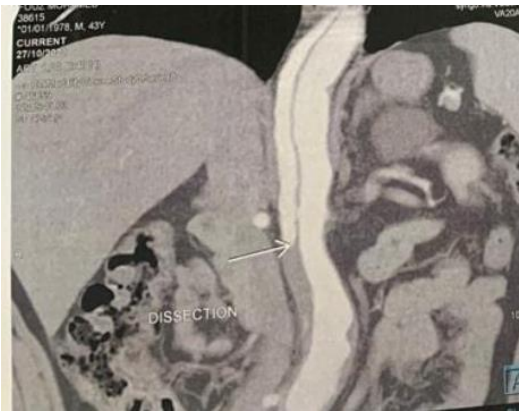
None

Figures





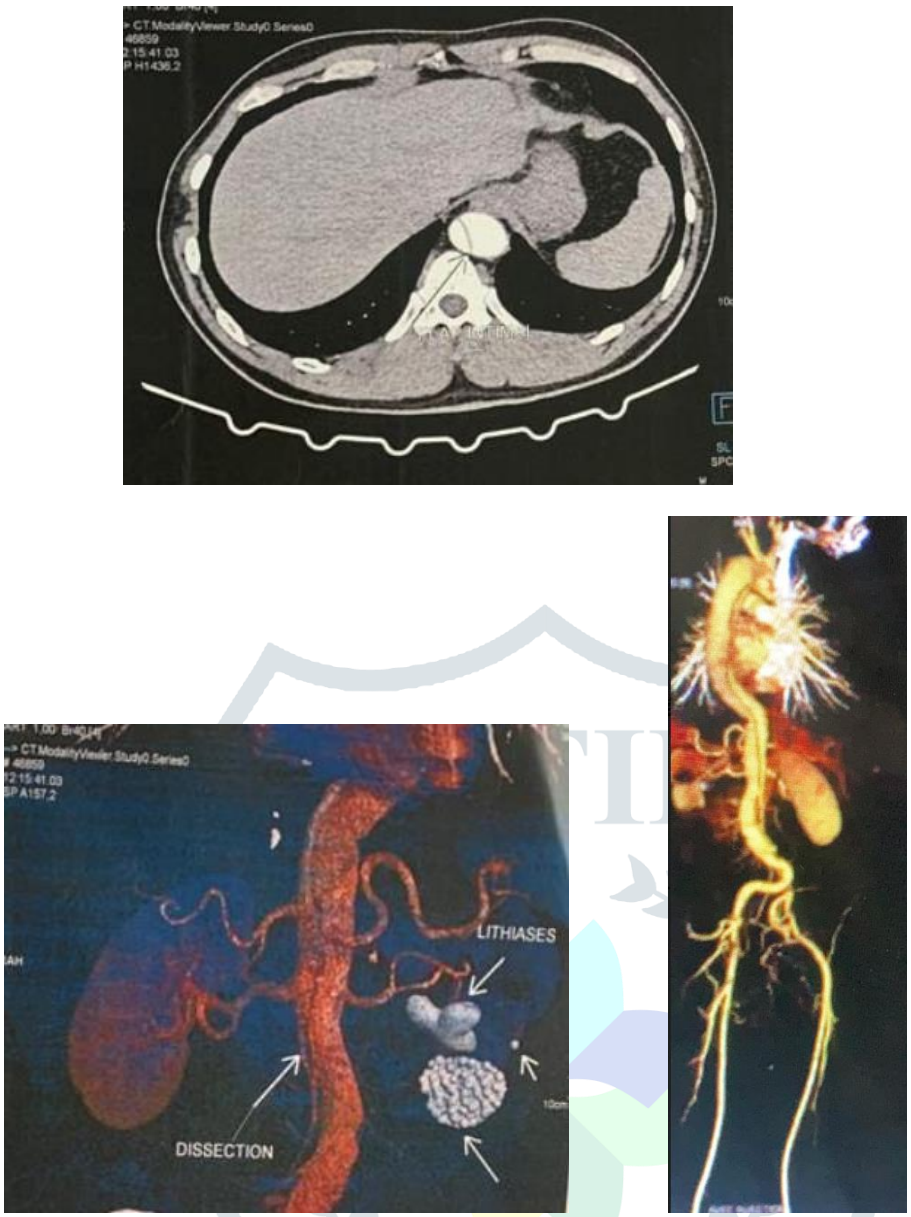


Figure 1: CT scan and thoraco-abdominal CT angiography showing type B aortic dissection with occlusion of the right common iliac artery.

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