

Case Report

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Juvenile Takayasu's disease: A new case report

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Abstract

Takayasu's Arteritis (TA) is a rare granulomatous vasculitis affecting large vessels, mainly the aorta and its branches as well as the pulmonary arteries. Although it occurs most often in young women of childbearing age, it is also considered among the main causes of aortic occlusive disease and pediatric renovascular hypertension. The diagnosis of Takayasu's arteritis during childhood remains difficult due to non-specific symptoms and biomarkers, a source of delayed diagnosis and vascular complications. We report a new case of Takayasu's disease diagnosed in a 26-year-old woman at a very late stage of the disease with clinical symptoms that evolve from childhood, marked by systemic signs and a major biological inflammatory syndrome, the disease is revealed ten years later by vascular complications attributed to occlusive arteritis.

Keywords: Takayasu; Pediatric.

Introduction

Takayasu's Arteritis (TA) is a rare inflammatory vasculitis involving large vessels with an unclear etiology, which most often affects women of childbearing age, usually under the age of 40, with a peak in the third decade [1-4]. It is often known as pulseless disease characterized by damage to the medium and large arteries as well as their branches. However, cases with juvenile onset have been rarely reported; this could be explained by the non-specific nature of the clinical signs and biological markers [5], a source of frequent delay in diagnosis. The disease is thus revealed by its own complications, namely staged arterial stenoses, a cause of significant morbidity and mortality and an alteration in the quality of life.

We report a new case of early-onset TA, victim of a diagnostic delay of more than 10 years, and treated at the stage of very advanced lesions.

Case report

It is a 26 years old woman, in whom TA was diagnosed at a very late stage of the disease, stage V of the angio-graphic classification [6] (Table 1). Clinical symptoms dates back to

childhood marked by recurrent episodes of fever, asthenia, arthromyalgia and headache; our patient has been the subject of multiple medical exams and no diagnosis was retained. Ten years later, faced to vascular symptoms such as carotidodynia, claudication of the four limbs and abdominal pain, a specialized clinical expertise revealed a decreased pulses and blood pressure in the four limbs and a bruit over carotid artery.

Doppler ultrasound and angio-CT of the aorta showed diffuse thickening of the pulmonary artery and along the thoracic aorta and its branches, as well as severe narrowing of the common carotid arteries (degrees of stenosis estimated around 95% on the left and 70% on the right), of the brachiocephalic arterial trunk, of the two sub-clavian arteries and of the left vertebral artery. It was also found a circumferential thickening of the abdominal aorta above and below the kidney with a 90% narrowing, associated with a thickening of the celiac trunk and the superior mesenteric artery. A diffuse parietal thickening of the pulmonary artery and along the thoracic aorta as well as its branches, responsible for a narrowing of the common carotid arteries (estimated at 95% on the left and 70% on the right), the brachiocephalic arterial trunk, the two subclavian.

Table 1: New angiographic classification of TA [6].

Type	Vessel involvement
Type I	Branches from the aortic arch
Type IIa	Ascending aorta, aortic arch and its branches
Type IIb	Ascending aorta, aortic arch and its branches, thoracic descending aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Combined features of type IIb and IV

Arteries and the left vertebral artery (estimated at 50%). It was also objectified a circumferential thickening of the above and below renal portions of the abdominal aorta with a 90% narrowing, associated with a thickening of the celiac trunk and the superior mesenteric artery. All of these elements were associated with an accelerated ESR of 80 mm, an elevated CRP of 68 mg/l and an inflammatory anemia. These abnormalities strongly suggested an active TA, based on the Ishikawa diagnostic criteria modified by Sharma in 1996 with 3 major criteria and several minor criteria [7,8]. Our patient is treated with a high dose of corticosteroids and Methotrexate. Remission is obtained after 08 months of treatment, no vascular complication (renovascular hypertension or renal failure) has been observed.

Discussion

TA is the most common cause of inflammatory granulomatosis of large vessels and the third most common cause of vasculitis in the pediatric population affecting the aorta, its branches and the pulmonary artery [9]. Inflammation of the vessel wall can lead to thickening, stenosis and thrombus formation; aneurysms and dissections may also be observed [10]. Clinical symptoms result from systemic inflammation and organ dysfunction secondary to ischemia. The diagnosis is based essentially on the analysis of clinical criteria and angiographic abnormalities.

The clinical presentation of the infantile form is most often non specific, contributing to an important delayed diagnosis and therapeutic, which differs from that of adults by the frequency of systemic manifestations (fever, asthenia, weight loss, abdominal pain and myalgia) and the importance of the biological inflammation. High blood pressure is reported to be the most common symptom in these two age populations [2,11]. Studies carried out in children are not numerous, Brunner and al [12] published a meta-analysis (nine studies and 241 patients) which noted the clinical symptoms and particularities of TA in children and adolescents, the therapeutic possibilities and its prognostic factors. Juvenile TA has been described at any age, even in 6-month-old infants [12], with an average age at disease onset estimated at 12 years [13]. Its exact incidence is not known, although most studies in Europe and North America estimated an overall incidence between 1 and 2.6/1.000.000 population/year [2,14]. Some authors have found a double incidence peak: one at 10-15 years and a second at 20-24 years [15,16]. with a clear female predominance in different regions: Mexico [17], Korea [18], USA [4], Turkey [19], India [11] and South Africa [20]. However, this female preponderance is lower than that of adults [13]. Topographically, adults tend to have a lesion of the aortic arch and its branches, while children tend

to have a lesion of the thoracic and abdominal aorta [21]. This disease is considered the most common cause of renovascular hypertension in Asian children [22].

An extended period ranging from 2 to 11 years can elapse between the onset of symptoms and the diagnosis of the disease, and this delay can be four times longer than that of adult patients [1-3]. Up to 25% of children are diagnosed in a late stage of the disease with symptoms resulting from irreversible vascular damage [13]. However, this latency period is less pronounced in the Indian and Turkish cohorts (less than one year) [11,19,23].

Arterial hypertension, the main clinical feature (56 to 100% of children according to ethnic origin, with a higher prevalence among Asians), is mainly linked to renal artery stenosis. Its association with other signs of low perfusion such as pulse deficits and arterial bruits are found in more than 60% of children at the time of diagnosis, highlighting the need for a thorough physical examination, especially in a child with unexplained systemic inflammation. The prevalence of cardiovascular complications such as cardiomegaly, ischemic heart disease, heart failure and valvular regurgitation is estimated to be between 5 and 27% [4,13], but coronary stenosis is reported in only 11% of pediatric population. Neurological impairment including headache, dizziness, seizures.

Finally, juvenile TA seems to have a more severe clinical presentation, a worse prognosis than that of adults and a mortality rate that can reach 50% at 5 years [24]. The extent of vascular involvement and the severity of hypertension are the main determinants of morbidity and mortality in most series [1]. The evolution of juvenile TA seems very variable, about 50% of patients relapse or develop a vascular complication within 10 years of diagnosis. Silent course of the disease and involvement of the thoracic aorta are associated with the risk of vascular complications [25,26].

Conclusion

Early-onset TM, rare and potentially serious, is a source of significant morbidity and mortality. The clinical presentation can be highly variable and often non specific, contributing to diagnostic and treatment delays. Early identification of patients with poor prognostic factors could help prevent serious complications.

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