

Atypical Aortic Coarctation Complicating Takayasu's Arteritis Presenting as Pyoderma Gangrenosum in A Young Boy: Case Report and Review of Literature

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Abstract

Case Report

Introduction: Atypical aortic coarctation is very rare, accounting for up to 2 % of aortic coarctation malformation. Several etiologies have been described and the most common is the Takayasu's arteritis. Skin manifestations of TA are very rare and an association with pyoderma gangrenosum has been reported. This case highlights the diagnosis and management challenges. **Case report:** A 10 years old boy with pyoderma gangrenosum was admitted for hypertensive emergency. TEE and CT scan further revealed an atypical aortic coarctation complicating Takayasu arteritis. The inflammatory storm was managed with immunosuppressive therapy. **Conclusion:** Atypical aortic coarctation is rare. Takayasu's disease is its main reported etiology and has poor prognosis. Early diagnosis and treatment are therefore crucial to prevent further damage to the blood vessel.

Keywords: Takayasu arteritis; pyoderma gangrenosum; atypical aortic coarctation; hypertension; children.

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INTRODUCTION

Coarctation of the aorta is typically a congenital narrowing of the aorta that occurs distal to the origin of the left subclavian artery at the aortic isthmus where the fetal ductus arteriosus inserts into the aorta. It accounts for 5-8% of all CHD. Atypical aortic coarctation is a rarer condition in which the stenosis is located far from the aortic isthmus and has several causes among which Takayasu's arteritis is the most common. Through this report and literature review we describe the diagnosis and management challenges in a 10 years old boy with history of Pyoderma gangrenosum who presented with secondary severe arterial hypertension revealing an atypical aortic coarctation complicating Takayasu's arthritis.

CASE REPORT

We describe a case of a 10 years old boy with history of pyoderma gangrenosum diagnosed at the age of 3 years old and treated with corticosteroids. He was

admitted in our cardiology department for hypertensive emergency.

Three weeks earlier, he developed an outbreak of his dermal pathology appearing as small papules on the right thigh which gradually increased in size complicated by painful central ulceration (Figure-1). He was first hospitalized in the dermatology unit for exacerbation of pyoderma gangrenosum, where he was treated with daily local care and oral prednisone 40mg per day. During that period, he accused atrocious headaches concomitant with very high blood pressure that required his transfer into our department for management and further screening. His clinical symptoms were frequent episodes of dizziness and shortness of breath three months earlier. Physical exam showed scarring plaques on the abdominal skin and both thighs, with ulceration of the right thigh. His HR was at 106 bpm. Blood pressure measured in the upper extremities was 240/130mm Hg in the right arm, 135/96 mm Hg in the left arm and in the lower extremities the BP was at 125/93 in the right limb and 120/92 in the left one. Right radial pulse was bounding, but the left radial arterial pulse was absent and both right and left femoral

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pulses were weak. Cardiac auscultation revealed a 3/6 pansystolic murmur radiating to the back, with enhancement of the second sound. He had no signs of Congestive heart failure.



Fig-1: Pyoderma gangrenosum with undermined lesion of the right thigh

The ECG showed a sinus rhythm with LV hypertrophy. Chest radiograph was normal. Results of complete blood count showed mild leukocytosis, normal hemoglobin and platelet counts, serum C-reactive protein (CRP) level of 35 mg/L and erythrocyte sedimentation rate (ESR) of 55mm/h.

His medical status was defined as hypertensive emergency and he was treated accordingly with intravenous calcium channel blocker associated with oral B blockers.

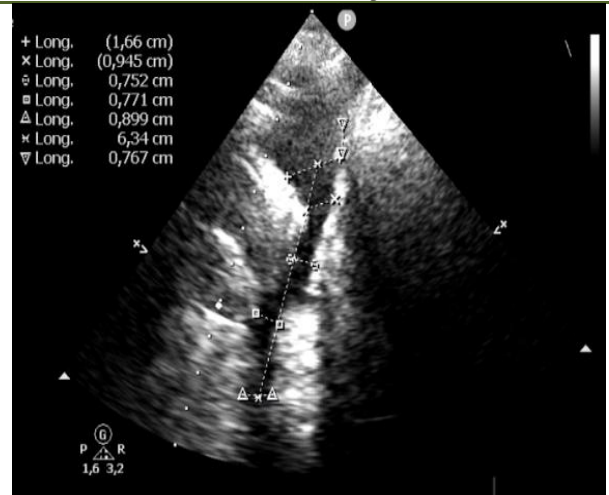


Fig-2: TTE suprasternal view showing diffuse narrowing of the descending aorta

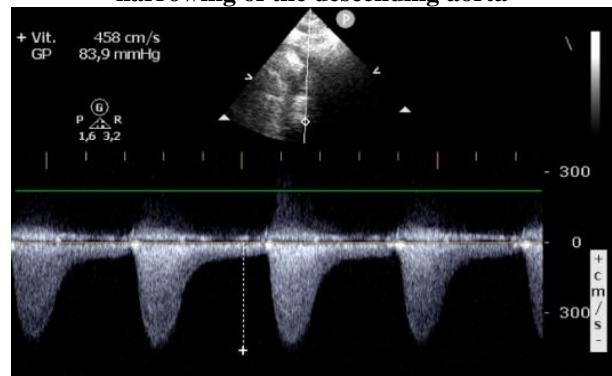


Fig-3: Continuous wave doppler in the descending aorta demonstrating a peak velocity of 4,6m/s comparable to a gradient of 84mmhg

Transthoracic echocardiography revealed a diffuse narrowing of the aorta at the isthmus segment prolonged 5 cm below and 1cm above the level of the left subclavian artery branch which was not visualized in the suprasternal view, the continuous wave doppler recorded a systolic-diastolic curve with a peak velocity of 4,6 m/s estimating a peak systolic gradient of 84mmhg, the diastolic antegrade flow didn't overcome 1,4 m/s, the LV function was preserved with left ventricular hypertrophy, no congenital heart abnormalities were associated (no bicuspid aortic valve , nor PDA or VSD) (Figure 2 and 3).

The CT angiography found a 5,4 mm thickened aortic wall, a diffuse and irregular lumen narrowing extending from the distal aortic arch into the descending aorta with left subclavian artery occlusion and left common carotid artery stenosis (Figure 4 and 5). The aorta in relationship with the renal arteries was normal, no collateral circulation was described.

According to the criteria of the American College of Rheumatology, alongside Ultrasound findings that excluded a congenital etiology given the anatomical aspect and the absence of associated heart

anomalies. The diagnosis retained was atypical aortic coarctation caused by Takayasu's arteritis

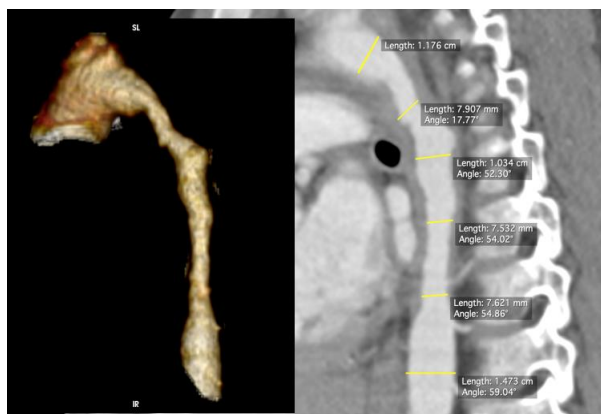


Fig-4: 2D and 3D CT angiography: oblique sagittal reconstruction showing long irregular stenosis of the descending aorta



Fig-5: CT angiography: 3D reconstructing showing the long-narrowed segment of the descending aorta

During his hospitalization, the patient was treated with oral prednisone 40mg per day, amlodipine 10 mg per day and a beta-blocker type atenolol 50 mg per day. His blood pressure was well controlled after 5 days dropping down to 130/84 mm Hg in the right arm and 113/83 mm Hg on the left side.

The management of this young boy who is in active phase of Takayasu's arteritis with diffuse inflammatory narrowing of the aorta involving its subclavian and common carotid branches, was established by a team of different specialties including dermatologists, vascular surgeons and interventional

cardiologists and it was decided to treat the inflammatory storm with immunosuppressive therapy and antiplatelet drug (aspirin 75 mg daily) at least for 6 months, the management of the aortic coarctation is to be discussed later depending on the course of the disease.

DISCUSSION

Takayasu arteritis (TA) is a chronic granulomatous large vessel vasculitis that predominantly affects the aorta, its main branches and the pulmonary arteries.¹ It affects predominantly females in their second and third decades.

Two stages of the disease have been described. In the early "prepulseless" stage, nonspecific symptoms such as fever, arthralgia, headaches, weight loss, can be present. In the later "pulseless" stage, narrowing of the arteries cause symptoms such as limb claudication, stroke, or congestive cardiac failure and characteristic clinical findings consisting of diminished or absent pulses, asymmetric blood pressures between right and left limbs, or vascular murmurs [1, 2].

In both stages, cutaneous manifestations of TA may be present. These are uncommon skin lesion, up to 2.8–28% including pyoderma gangrenosum, erythema nodosum and erythema induratum.

In our case despite the long history of pyoderma gangrenosum, the diagnosis of Takayasu's arteritis was overlooked until the later stage with appearance of clinical vascular signs.

The association of skin lesion like pyoderma gangrenosum and Takayasu's arteritis is very rare. It is a neutrophilic dermatosis characterized by recurrences and enlarging painful deep ulceration with a violaceous border, undermined edges and a necrotic base [3]. It most commonly involves lower extremities. Our patient in this current relapse was suffering of ulceration on the right thigh and presented multiples scarring plaques on the abdomen and the thighs.

Systemic inflammation may be detected by increased CRP levels or increased erythrocyte sedimentation rates but they lack sensitivity and specificity [3, 4].

Angiography used to be the most valuable tool for the diagnosis and classification of TA. More recently it has been replaced by computed tomography scanning and magnetic resonance angiography allowing early and easy diagnosis of vascular lesions.⁴ In our case the TTE showed diffuse narrowing of the descending aorta inconsistent with typical congenital aortic coarctation. Then CT angiogram revealed wall thickening of the aorta, its carotid and subclavian branches, making evidence of inflammation as the

underlining cause of this atypical coarctation of the aorta.

Despite the significant advance in the imaging modalities cited above, the diagnosis of TA remains challenging. The American College of Rheumatology (ACR) has therefore laid criteria including Age, clinical findings and arteriogram abnormalities for diagnosing TA (Table-1). A score of three or more has a sensitivity of 90.5% and specificity of 97.8% [2]. Our patient fulfilled four of six criteria for the diagnostic of Takayasu's arteritis.

Medical treatment of TA includes systemic corticosteroids and/or methotrexate with evidence of long-term angiographic improving in previous literature [4]. Corticosteroids are also the primary treatment of choice for PG. As an alternative therapy, immunosuppressive drugs, such as azathioprine, cyclophosphamide, cyclosporine, and tacrolimus, were also demonstrated to be effective, especially in the case of corticosteroid resistance [3]. The active phase of TA is often complicated by thrombosis in the affected vessels with stenotic lesions, long-term aspirin therapy is then recommended to prevent thrombus formation in vessels with endothelial damage [4].

Due to the severity of the lesions, our patient was treated associating corticosteroids and methotrexate, his blood pressure controlled with betablockers and amlodipine and acute thrombosis was prevented by aspirin.

Table-1: American College of Rheumatology 1990 criteria for classification of Takayasu's arteritis

Age of onset ≤ 40 years
Development of symptoms or signs related with Takayasu's arteritis ≤ 40 years
Claudication of extremities
Development and worsening of fatigue and discomfort in muscle in 1 or more extremity while in use, especially in upper limb
Decreased brachial artery pulse
Decreased pulsation in one or both brachial arteries
BP difference >10 mm Hg
Difference of systolic BP >10 mm Hg between arms
Bruit over subclavian artery or aorta
Bruit audible on auscultation of one or both subclavian arteries or abdominal aorta
Arteriogram abnormality
Arteriographic narrowing or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental

The effectiveness of percutaneous angioplasty or stent placement of typical aortic coarctation have been proven these past 10 years [5]. Atypical aortic

coarctation on the other hand, regardless of the etiology, are unlikely to be successfully treated with angioplasty alone because the fibrous nature of the arterial lesions, making the risk of restenosis higher [6]. In a series with 20 patients from Chile with arterial stenosis caused by Takayasu's arteritis, treated with angioplasty alone, only eight patients maintained patency at 5 years [7]. Nonetheless Tyagi and al reported from India the largest series of 36 patients with favorable initial and midterm outcome, especially in dilatating discrete-type aortic stenosis [8]. Similarly many surgical treatments like extra-anatomical bypass, aortic patch plasty, anatomic graft interposition, and stent-graft insertion procedures have been reported. Among those, an aorto-aortic bypass has been the most frequent choice, because of the difficulties in anatomic repair due to a longer segment of the affected aorta; In a series over 44 years of 33 patients with atypical aortic coarctation, surgery with an aorto-aortic bypass has shown a satisfactory long-term survival of 78% at 10 years [9]. In our case, given the limited data and the lack of consensus concerning the management of this atypical aortic disease, we settled with an optimal medical treatment with clinical surveillance and reevaluation of the vascular state in 6 months.

CONCLUSION

Atypical aortic coarctation is rare and Takayasu's disease is its main reported etiology. Uncommon Takayasu's cutaneous manifestations such as pyoderma gangrenosum must be well known for an early assessment of vascular involvement. The long-term prognosis depends on blood pressure control and the underlying disease, thus early diagnosis and treatment are crucial to control the inflammation and prevent further damage to the blood vessel. The choice between percutaneous treatment and surgery should be guided by the anatomical aspects and the response to the immunosuppressive treatment.

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Authors' contributions

Koné Guéladio Hassane, Bounsir Ayoub, Mehssani Zineb took care of the patient under Fellat Nadia and Lekehal Brahim supervision. Koné Guéladio Hassane wrote a manuscript draft. Koné Guéladio Hassane, Bounsir Ayoub, Mehssani Zineb and Fellat Nadia finalized the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Ethical approval was granted by the Mohammed V University Ethics Committee and informed consent was obtained from the patient.

Written informed consent was obtained from the patient's legal guardian(s) for publication of this case report and any accompanying images. A copy of

the written consent is available for review by the Editor-in-Chief of this journal.

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