Asian Journal of Cardiology Research

Volume 7, Issue 1, Page 52-57, 2024; Article no.AJCR.116093

A Rare Case of Aortic and Cardiac Thrombosis Revealing a Horton's Disease: Clinical Presentation, Diagnostic Challenges and Therapeutic Considerations

Sami Belkouchia [a*], Zineb Amine [a], Zaid Ammouri [a], Meryem Haboub [a] and Rachida Habbal [a]

[a] Cardiology Department, IBN ROCHD University Hospital, Casablanca, Morocco.

Authors' contributions

This work was carried out in collaboration among all authors. Author SM designed the study, performed the statistical analysis, wrote the protocol, and wrote the first draft of the manuscript. Authors ZA and ZA managed the literature searches, the rest of authors did the scientific supervision as a professor and chief department. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/116093

Case Report

Received: 11/02/2024
Accepted: 15/04/2024
Published: 23/04/2024

ABSTRACT

Horton's Disease, also known as Giant cell arteritis is a disease of the blood vessels predominantly affecting medium and large-sized arteries mainly the branches of the aorta and its main vessels. In these patients, there is an increased cardiovascular risk for the development of both arterial and venous thromboembolism. In this case report we discussed a rare case of aortic and cardiac thrombosis associated with Horton's disease which is quite unusual. Early diagnosis is mandatory as the prognosis of this disease mainly depends on timely management. Horton's disease should be suspected in people aged above 55 years as this disease is unlikely and rare in people aged

**Corresponding author: Email: sami.belkouchia@gmail.com;*

less than 55 years and females are more affected by this disease (men to women ratio is 2:1). This rare case highlights the importance of considering unusual and infrequent associations of this disease and accordingly tailoring the treatment strategies for optimal patient outcomes. decrease medical as well as financial burden, hence improving the management of cirrhotic patients. These predictors, however, need further work to validate reliability.

Keywords: Horton's disease; thrombosis; cardiology; autoimmune; giant cell arteritis.

1. INTRODUCTION

Horton's disease commonly known as giant cell arteritis or temporal arteritis is an autoimmune inflammatory disease involving large and medium-sized arteries with well-developed wall layers and adventitial vasa vasorum [1]. It is mainly characterized by the formation of clusters of inflammatory cells (granulomas) in the injured vessels. The vascular beds that are more commonly affected mainly include the branches of the external carotid artery (e.g. temporal and occipital arteries), the ophthalmic, vertebral, distal subclavian, and axillary arteries, and the thoracic aorta [2].

It is the most common adulthood vasculitis and affects 1 in 15000 people per year. It most commonly occurs in people above 50 years of age, being most common among those in their 70s. Females are more commonly affected than males (women to men ratio is 2:1) [3].

Here we report the observation of a 67-year-old female patient in whom atypical thoracic pain and exertional dyspnea revealed Horton's disease while evaluating the patient in the suspicion of any cardiac disease. It came out to be an incidental finding along with aortic and cardiac thrombosis. This case report looks into an intriguing and rare association between Horton's disease and both aortic and cardiac thrombosis, highlighting the intricate association between inflammatory processes and thrombotic events within the cardiovascular system. This study aims to shed some light on the fact that Horton's disease although rare should be suspected in patients with aortic or cardiac thrombosis to prevent the risk of serious vascular complications which may occur as a result of diagnostic and therapeutic delay.

Hence clinicians must provide timely and effective management strategies for patients presenting with these co-existing conditions.

2. CASE PRESENTATION

We present the case of a 67-year-old female patient with no significant medical history, admitted for acute atypical thoracic pain radiating to the back, accompanied by exertional dyspnea. Clinical examination revealed no particularities, with the electrocardiogram showing repolarization disturbances characterized by negative T waves in the anteroseptal region. Transthoracic echocardiography revealed a non-dilated, non-hypertrophied left ventricle with global hypokinesia and a left intraventricular mass adherent to the apical and inferoseptal walls measuring 28.47mm. Filling pressures were normal, with mild mitral regurgitation and no signs of pulmonary hypertension or pericardial effusion.

The patient underwent thoracic scanning revealing a thrombus in the descending aorta and splenic infarction due to splenic artery occlusion in its distal portion. Cardiac MRI confirmed the thrombotic origin of the intracardiac mass. Inflammatory markers were elevated, with a CRP of 200 and twice the normal fibrinogen levels, while troponins were elevated at 100. A PET scan, conducted in consultation with internists, revealed multifocal large vessel vasculitis affecting the descending aorta, right humeral artery, subclavian arteries, abdominal aorta, iliac arteries, and femoral arteries.

The diagnosis of Horton's disease was established based on clinical and biological signs indicative of inflammation, as well as imaging findings from CT and PET scans revealing medium-sized artery vasculitis. Temporal artery biopsy could not be performed due to the significant risk of hemorrhage associated with antiplatelet and anticoagulant therapy. However, it was substituted according to protocol with Doppler ultrasound of the temporal artery, which demonstrated signs of vasculitis and edema (Halo sign), characteristic of Horton's disease.

Therapeutic management included anti-platelet therapy, anticoagulation, ACE inhibitor, beta blocker, statin, and corticosteroid therapy with close cardiologic and internist monitoring.

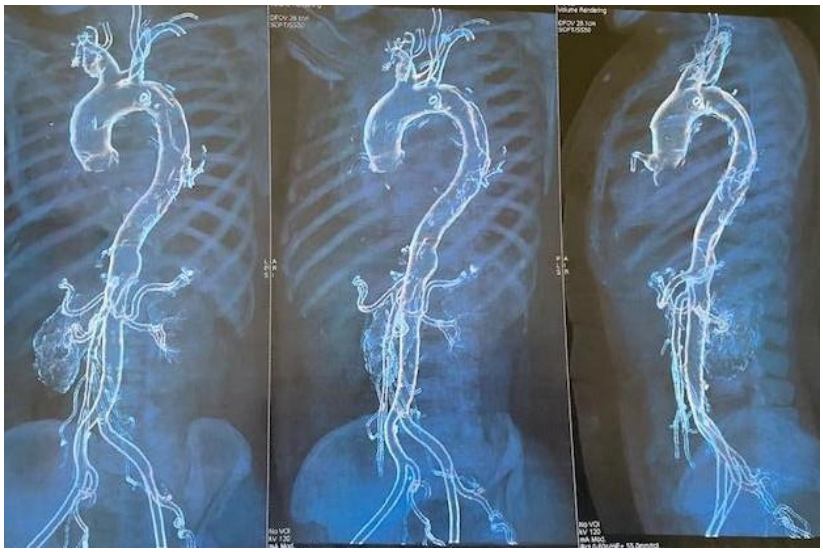

Fig. 1. Scan image showing disease

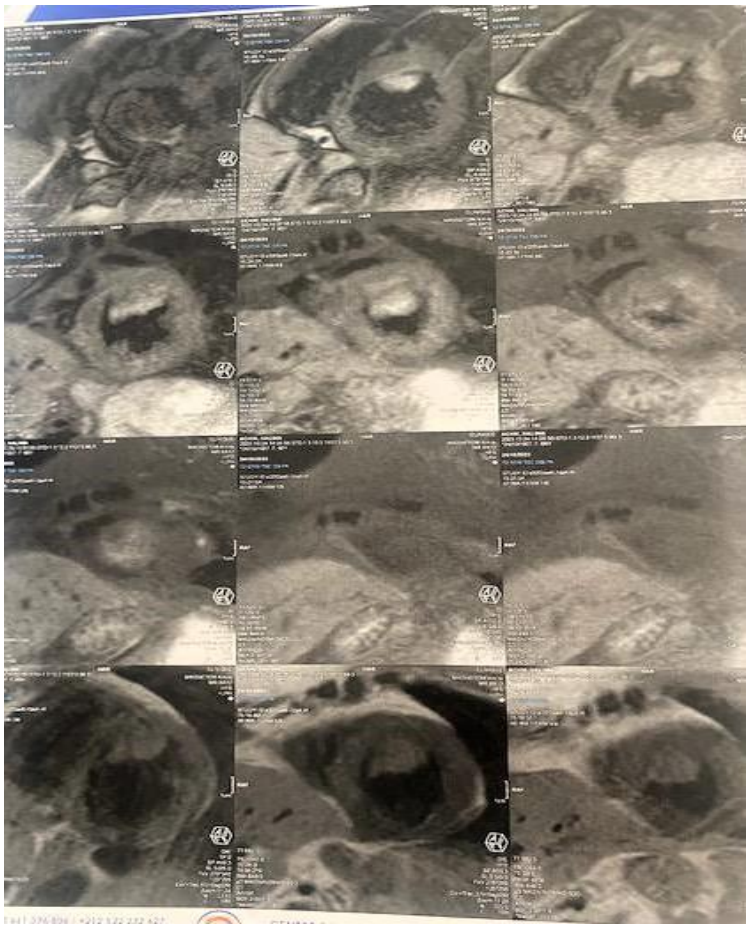

Fig. 2. MRI cardiac thrombosis

3. DISCUSSION

As previously discussed, giant cell arteritis is an autoimmune vasculitis that primarily involves medium and large-sized vessels. It is predominantly observed in elderly patients that preferentially involves the external carotid artery and its branches. Although the involvement of

cranial arteries is typical of this condition [4,5] in recent decades with the advancement in imaging techniques, the involvement of the aorta and its main branches is increasingly recognized [6-8].

Like in this case report we reported a case of an elderly female who presented with atypical thoracic pain accompanied by dyspnea leading to the suspicion of cardiovascular disease which ultimately after undergoing diagnostic evaluation via imaging modalities unrevealed the diagnosis of Horton's disease along with aortic and cardiac thrombosis which is undoubtedly quite rare.

The clinical manifestations are divided into those caused by the involvement of cranial vessels, those due to inflammation of large vessels (aorta and branches of the aorta), and systemic inflammatory signs [9].

Symptoms due to cranial vascular involvement include headache, jaw claudication pain on chewing), scalp tenderness, loss of vision, abnormalities of the temporal arteries, pain, nodules, and absence of pus. Bitemporal accentuated headaches refractory to analgesia occur in about 3 quarters of patients [10].

Symptoms due to the involvement of great vessels like the aorta and its branches include claudication of extremities (especially the arm).

Similarly, systemic inflammatory involvement is accompanied by several non-specific symptoms such as exhaustion, fever, night sweats, and weight loss [9].

Risk factors for developing Giant cell arteritis/ Horton's disease include individuals older than 50 years of age and females are twice more affected than males. The aging process remains the strongest risk factor for clinically evident vasculitis [11]. Epidemiological studies have described lower body mass index and low fasting blood glucose levels many years before disease onset [12,13]. The strongest genetic risk factor has been implicated in the HLA region, with particular HLA-DRB1*04 alleles conferring the risk of developing polymyalgia rheumatica (PMR) as well as giant cell arteritis (GCA) [14-16]. The aorta and its main branches are affected in 40% to 80% of cases, even though they do not induce any specific symptoms [5].

Horton's disease/ GCA is diagnosed based on a combination of symptoms, clinical findings, laboratory findings, and findings on diagnostic imaging [10,17,18]. However temporal artery biopsy remains the gold standard for diagnosis [18]. The American College of Rheumatology has established a criteria that is still used in clinical practice to classify the disease. The diagnosis of GCA is highly probable in the presence of at least 3 of the following 5 criteria:

1) age > 50 years;
2) an erythrocyte sedimentation rate (ESR) > 50 mm (and, by extension, high inflammatory parameters such as increased C-reactive protein (CRP));
3) recent onset of headaches;
4) abnormalities of the temporal artery; and
5) a positive temporal artery biopsy (TAB) showing transmural inflammation with the presence of giant cells. These criteria do not take into consideration the presence of clinical or radiological large-vessel involvement (LVI) [5].

In our case 2 the above-mentioned criteria are met; 1) the age of our patient was 67 years old and 2) inflammatory markers were elevated including a CRP of 200 and twice the normal fibrinogen level although there was no history of recent onset of headaches and no abnormalities of the temporal artery.

The consensus among the collegiate body was to refrain from proceeding with the temporal biopsy, in light of the significant hemorrhagic risk associated with this intervention. This decision was underpinned by the fact that the patient had already been administered tritherapy, consisting of dual antiplatelet therapy in conjunction with therapeutic anticoagulation.

Temporal artery ultrasound, utilizing Color Doppler, has the potential to identify vessel wall edema, manifesting as a halo, particularly when conducted by skilled practitioners, thus serving as a potential alternative to temporal artery biopsy for diagnosing giant cell arteritis [19]. It is recommended that ultrasound of the temporal artery be performed prior to initiating treatment or within a 5-day window, as corticosteroids can diminish test sensitivity. Notably, this technique offers numerous advantages, including its noninvasiveness, absence of radiation exposure, and capability to visualize other cranial vessels. Nevertheless, the diagnostic efficacy of temporal artery ultrasound hinges significantly on the proficiency of the ultrasound operator and the quality of equipment employed.

However over the past few decades because of the wider use of imaging methods such as computed tomography angiography (CTA),

magnetic resonance angiography (MRA) of the aorta or positron emission tomography with computed tomography (PET/CT) the proportion of GCA patient described with LVI has increased [5]. In our case, our patient underwent thoracic angioscanning which revealed a thrombus in the descending aorta.

The cardiac MRI played a crucial role in the diagnostic process as it enabled the confirmation of the intracardiac mass's nature as a thrombus. Simultaneously, it facilitated the exclusion of the initially hypothesized diagnosis of autoimmune myocarditis, which was considered based on the clinical and biological context. In addition, a PET scan was also conducted which showed multifocal large vessel vasculitis involving the descending aorta, right humeral artery, subclavian arteries, abdominal aorta, iliac arteries, and femoral arteries.

However, in GCA it is important to keep in mind aortitis because it may present at the initial stage of the disease, to diagnose delayed complications of GCA it is recommended by some authors to carry out annual chest radiographic and transthoracic echocardiogram to detect signs suggesting a thoracic aortic aneurysm in these patients [20]. As mentioned in our case report we also conducted transthoracic echocardiography of our patient which showed a non-dilated, non-hypertrophied left ventricle with global hypokinesia and a left intraventricular mass adherent to apical and infero-septal walls measuring 28.47mm.

The mainstay of treatment includes, high dose corticosteroids, and additional cytotoxic drugs, antitumor necrosis factor monoclonal antibody, and antiplatelet aggregation therapy may be used. The main goal of the treatment is the prevention of ischemic damage [18]. In our case the therapeutic management of our patient includes corticosteroid therapy, antiplatelet therapy, anticoagulation, ACE inhibitor, beta blocker, and statin which showed impressive regression of aortic and cardiac thrombosis within a period of three months.

4. CONCLUSION

This case report highlights the significance of considering and keeping in mind that cardiac and aortic thrombosis may present concomitantly in a patient diagnosed with Horton's disease and vice versa. This will help in preventing any diagnostic delay and sequential complications, hence improving the outcome of the patient by offering prompt treatment. However further treatment trials and research are still needed to develop more sensitive and specific diagnostic approaches and new corticosteroid-sparing treatment strategies.

SUPPLEMENTARY METARIALS

Supplementary metarials available in this link: https://journalajcr.com/media/2024AJCR116093.mp4

CONSENT

All authors declare that ‘written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Weyand CM, Goronzy JJ. Clinical practice. Giant-cell arteritis and polymyalgia rheumatica. The New England Journal of Medicine. 2014;371(1):50-7.
2. Weyand CM, Liao YJ, Goronzy JJ. The immunopathology of giant cell arteritis: Diagnostic and therapeutic implications. Journal of Neuro-ophthalmology: The Official Journal of the North American Neuro-Ophthalmology Society. 2012;32(3): 259-65.
3. Gomułka K, Radzik-Zając J, Zaleska-Dorobisz U, Panaszek B. Horton's disease: Still an important medical problem in elderly patients: A review and case report. Postepy Dermatologii I alergologii. 2017; 34(5):510-3.
4. Muratore F, Crescentini F, Spaggiari L, Pazzola G, Casali M, Boiardi L et al. Aortic dilatation in patients with large vessel vasculitis: A longitudinal case-control study using PET/CT. Seminars in Arthritis and Rheumatism. 2019;48(6):1074-82.
5. de Boysson H, Daumas A, Vautier M, Parienti J-J, Liozon E, Lambert M et al. Large-vessel involvement and aortic

dilation in giant-cell arteritis. A multicenter study of 549 patients. Autoimmunity Reviews. 2018;17(4):391-8.
6. Salvarani C, Soriano A, Muratore F, Shoenfeld Y, Blockmans D. Is PET/CT essential in the diagnosis and follow-up of temporal arteritis? Autoimmunity Reviews. 2017;16(11):1125-30.
7. Maleszewski JJ, Younge BR, Fritzlen JT, Hunder GG, Goronzy JJ, Warrington KJ et al. Clinical and pathological evolution of giant cell arteritis: A prospective study of follow-up temporal artery biopsies in 40 treated patients. Modern Pathology. 2017; 30(6):788-96.
8. Stone JR, Bruneval P, Angelini A, Bartoloni G, Basso C, Batoroeva L et al. Consensus statement on surgical pathology of the aorta from the Society for Cardiovascular Pathology and the Association for European Cardiovascular Pathology: I. Inflammatory diseases. Cardiovascular Pathology. 2015;24(5):267-78.
9. Ness T, Bley TA, Schmidt WA, Lamprecht P. The diagnosis and treatment of giant cell arteritis. Deutsches Arzteblatt international. 2013;110(21):376-85;quiz 86.
10. Vaith P, Warnatz K. [Clinical and serological findings of giant-cell arteritis]. Zeitschrift fur Rheumatologie. 2009;68(2): 124-31.
11. Weyand CM, Goronzy JJ. Immunology of giant cell arteritis. Circulation research. 2023;132(2):238-50.
12. Jakobsson K, Jacobsson L, Warrington K, Matteson EL, Liang K, Melander O et al. Body mass index and the risk of giant cell arteritis: Results from a prospective study. Rheumatology (Oxford, England). 2015; 54(3):433-40.
13. Wadström K, Jacobsson L, Mohammad AJ, Warrington KJ, Matteson EL, Turesson C. Negative associations for fasting blood glucose, cholesterol, and triglyceride levels with the development of giant cell arteritis. Rheumatology (Oxford, England). 2020; 59(11):3229-36.
14. Weyand CM, Hicok KC, Hunder GG, Goronzy JJ. The HLA-DRB1 locus is a genetic component in giant cell arteritis. Mapping of a disease-linked sequence motif to the antigen binding site of the HLA-DR molecule. The Journal of Clinical Investigation. 1992;90(6):2355-61.
15. Weyand CM, Hunder NN, Hicok KC, Hunder GG, Goronzy JJ. HLA-DRB1 alleles in polymyalgia rheumatica, giant cell arteritis, and rheumatoid arthritis. Arthritis and Rheumatism. 1994;37(4):514-20.
16. Carmona FD, Mackie SL, Martín JE, Taylor JC, Vaglio A, Eyre S et al. A large-scale genetic analysis reveals a strong contribution of the HLA class II region to giant cell arteritis susceptibility. American Journal of Human Genetics. 2015;96(4): 565-80.
17. Ness T, Auw-Hädrich C, Schmidt D. [Temporal arteritis (giant cell arteritis). Clinical picture, histology, and treatment]. Der Ophthalmologe: Zeitschrift der Deutschen Ophthalmologischen Gesellschaft. 2006;103(4):296-301.
18. Kale N, Eggenberger E. Diagnosis and management of giant cell arteritis: A review. Current Opinion in Ophthalmology. 2010;21(6):417-22.
19. Chrysidis S, Døhn UM, Terslev L et al: Diagnostic accuracy of vascular ultrasound in patients with suspected giant cell arteritis (EUREKA): A prospective, multicentre, non-interventional, cohort study. The Lancet Rheumatology. 2021; 3(12):e865-e873. DOI:org/10.1016/S2665-9913(21)00246-0
20. Nuenninghoff DM, Hunder GG, Christianson TJ, McClelland RL, Matteson EL. Incidence and predictors of large-artery complication (aortic aneurysm, aortic dissection, and/or large-artery stenosis) in patients with giant cell arteritis: A population-based study over 50 years. Arthritis and Rheumatism. 2003;48(12): 3522-31.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/116093