International Clinical and Medical Case Reports Journal

Salient Visionary Publications

Case Report (ISSN: 2832-5788)

IgG4-related Aortitis Mimicking an Ascending Aorta Intramural Haematoma: A Case Report

William J. Crawford[1], Ciro Amodio[2], Sarah Karrar[3], Anu Balan[4], Wael I. Awad[1*]

[1]Barts Heart Centre, St Bartholomew's Hospital, London, United Kingdom

[2]Department of Cardiac Surgery, University of Florida, United States of America

[3]Department of Rheumatology, The Royal London Hospital, London, United Kingdom

[4]Department of Radiology, St Bartholomew's Hospital, London, United Kingdom

Citation: *William J. Crawford, Ciro Amodio, Sarah Karrar, Anu Balan, Wael I. Awad. IgG4-related Aortitis Mimicking an Ascending Aorta Intramural Haematoma: A Case Report. Int Clinc Med Case Rep Jour. 2026;5(11):1-7.*

Received Date: 10 November, 2026; **Accepted Date:** 12 November, 2026; **Published Date:** 13 November, 2026

***Corresponding author:** Wael I. Awad, Barts Heart Centre, St Bartholomew's Hospital, London, United Kingdom

Copyright: © Wael I. Awad, Open Access 2026. This article, published in Int Clinc Med Case Rep Jour (ICMCRJ) (Attribution 4.0 International), as described by http://creativecommons.org/licenses/by/4.0/

ABSTRACT

Introduction: IgG4-related disease is a newly recognised autoimmune disease which affects almost all tissues, including the aorta. Here we present a rare case of IgG4-related aortitis with radiological features that mimicked an IMH.

Case Summary: A 54-year-old woman presented to the emergency department with exertional chest pain. CT imaging excluded pulmonary embolism, but showed an enlarged ascending thoracic aorta of 40 x 41mm and features of an Intramural Haematoma (IMH). Given these radiological findings and symptoms of chest pain, the patient underwent urgent surgery. Intraoperatively the aorta was found to be severely thickened and dilated, however there was no IMH present. The ascending aorta was replaced with an interposition graft under circulatory arrest. After an uneventful recovery, serological analysis displayed a raised serum IgG4, while histology demonstrated features in keeping with IgG4-related aortitis. The patient was subsequently started on prednisolone and methotrexate. Despite this, she was still experiencing some chest pain at her twelve-month post-operative follow-up. Fifteen months after her surgery an FDG PET-CT demonstrated ongoing aortic arch inflammation, and she was initiated on rituximab therapy.

Conclusion: IgG4-related aortitis can cause aortic dilatation and mimic acute aortic syndromes, such as an intramural haematoma. Definitive diagnosis of this condition is challenging and is typically made post-operatively. The diagnosis is made through histological confirmation, elevated serum IgG4 levels and typical radiological features. It may be possible to treat IgG4-related aortitis with immunosuppression only in the absence of any other indication for surgery.

Keywords: IgG4-related disease; IgG4-related aortitis; Aortitis; Intramural haematoma

INTRODUCTION

Aortitis is an overarching term referring to inflammation of the aorta of multiple aetiologies. It has a highly variable presentation, resulting in a significant diagnostic challenge. Systemic symptoms (such as fever, fatigue, or malaise) are often present. Depending on the segment of aortic involvement, patients can present with cerebrovascular accidents, acute coronary syndromes, occlusive disease of any of the abdominal organs (commonly the spleen, kidneys and gut), as well as peripheral arterial occlusive disease.[1] Patients with aortitis are at a higher risk of aortic aneurysm and acute aortic syndromes, and aortic inflammation is frequently only diagnosed post-operatively with histopathological examination after surgery.[2]

Aortitis is most commonly due to non-infective aetiologies.[2] The large vessel vasculitides such as giant cell arteritis and Takayasu arteritis are thought to be the most common non-infectious causes.[3] Recently, IgG4-related disease has emerged as a common cause of non-infectious aortitis. IgG4-related disease is an autoimmune condition first recognised in 2001 that can affect almost any organ.[4] It is characterised by chronic inflammation secondary to tissue infiltration of lymphocytes and IgG4-secreting plasma cells, resulting in a classical pattern of storiform fibrosis.[5,6] IgG4-related disease has a highly variable presentation, with the potential for single or multiple organ involvement. It now encompasses previously distinct conditions such as autoimmune pancreatitis, retroperitoneal fibrosis and tubulointerstitial nephritis.[4] Data regarding the incidence and population distribution of IgG4-related disease are scarce, however cohort studies have demonstrated a slight preponderance for middle-aged and older males.[7]

Here we present a case of IgG4-related aortitis of a dilated ascending aorta, with radiological features mimicking an intramural haematoma (IMH). This case highlights the difficulty in diagnosing the condition, and controversy in management approach.

CASE PRESENTATION

A 54-year-old woman presented to the emergency department with central, burning chest pain. The pain was exacerbated by exercise and associated with shortness-of-breath. Her medical history included stage I chronic kidney disease and hypertension, which was well controlled with Amlodipine and Ramipril. She did not complain of any arthralgia, fevers, night sweats, skin changes, Raynaud's phenomenon, or hair loss. She had no family history of rheumatological or connective tissue disease. On examination she had normal heart sounds and warm peripheries with a regular pulse. She did not have any rashes, ulcers, lymphadenopathy or organomegaly.

Electrocardiogram demonstrated a right bundle branch block, without any ischaemic changes. Blood tests demonstrated a normal troponin of 7ng/L (reference range 0-13ng/L), with an elevated d-dimer (0.98mg/L FEU, reference range <0.5mg/ml FEU). Computerised tomography (CT) pulmonary angiogram did not detect any pulmonary emboli, however it did show a dilated ascending aorta. Subsequent CT aortic angiogram confirmed a proximal ascending aorta of 40 x 41mm, with apparent wall thickening of 19mm, and normal calibre of the aortic root. The appearances were highly suggestive of IMH, indicated by focal, crescentic, high-attenuation

within the thick walled ascending aorta on non-contrast CT, with corresponding low attenuation on the arterial phase imaging. This suspected IMH commenced at the level of the aortic root and extended to the proximal aspect of the ascending aortic arch (Figure 1). CT coronary angiogram did not show any significant coronary disease, while transthoracic echocardiogram demonstrated no valvular pathology and preserved biventricular function. Given these findings she was referred for urgent surgery.

The patient underwent an ascending aorta replacement via median sternotomy. The ascending aorta appeared calcified and severely thickened, which was thought to be due to a chronic IMH. The excised aorta was sent for histological analysis, and a supracoronary 32mm Anteflow Dacron interposition graft was inserted with the distal anastomosis performed under circulatory arrest. The patient's post-operative recovery was uneventful, and she was discharged on post-operative day eight.

Histological analysis of the excised aorta demonstrated a severely thickened wall, with a maximum thickness of 18mm. There were no macroscopic features of an IMH. Immunohistochemistry showed large areas of adventitial fibrosis and inflammation, accompanied by copious IgG4-positive plasma cells. Infective causes of aortitis such as tuberculosis and fungal aortitis were excluded using Ziehl-Neelsen, Warthin–Starry, and Grocott's stains. Serological testing excluded common causes of infective aortitis including human immunodeficiency virus and syphilis.

The patient was subsequently reviewed in a rheumatology clinic with suspicion of IgG4-related disease. She was found to have an elevated serum IgG4 level of 161mg/dL (reference range 0 - 86mg/dL), which confirmed a diagnosis of IgG4-related aortitis. An FDG PET-CT performed nine months after her surgery demonstrated peri-aortic inflammation (likely due to ongoing aortitis or persistent post-operative inflammation), without pathological uptake in any other vasculature or organs (Figure 2). She was commenced on a weaning regime of prednisolone (starting at 20mg once-daily) and methotrexate 15mg once-weekly.

Five months post-operatively she was still experiencing mild chest pain similar to her initial presentation, without any features of extra-aortic IgG4-related disease. As repeat FDG PET-CT demonstrated persistent uptake in the wall of the aortic arch, she was initiated on rituximab therapy. After two years of rituximab there was reduction in inflammation of the native aorta.

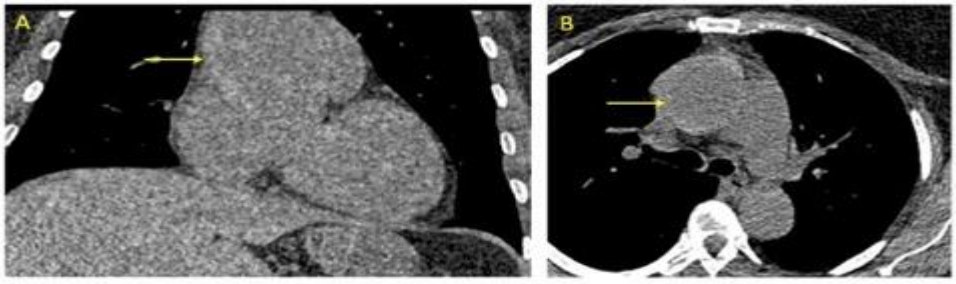


Figure 1: – Non-contrast Coronal (A) and Axial (B) CT reformats of ascending aorta. Yellow arrow shows hyperdense stripe applied to medial wall of proximal ascending aorta, corresponding to suspected intramural haematoma.

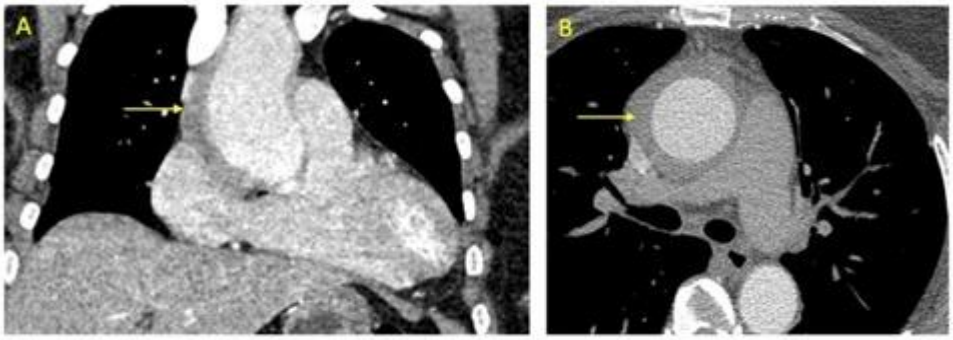


Figure 2: – Arterial phase Coronal (A) and Axial (B) CT reformats showing same region identified on pre-contrast study. Yellow arrow shows low attenuation suggestive of intramural haematoma.

DISCUSSION

We present a rare case of IgG4-related aortitis with a symptomatic ascending aortic aneurysm and false radiological signs of an IMH. This case demonstrates the challenge associated with diagnosing IgG4-related aortitis, and the management conundrum of whether urgent or emergent surgery should be offered for these patients.

IgG4-related disease was first recognised in 2001. Given that it can affect numerous organs, it has a highly variable presentation.[5,6] Key diagnostic criteria of IgG4-realted disease include clinical and radiological features, serological features (serum IgG4 >135mg/dL) and histological features (lymphoplasmacytic infiltration with IgG4-positive plasma cells). The presence of two of these criteria indicates possible or probable IgG4-related disease, while all three indicates a definite diagnosis.[6] International expert consensus strongly recommends that a histological analysis is performed to exclude other disease mimics (such as malignancy, or IMH as seen in this case).[8] As it is not feasible to obtain a pre-operative aortic biopsy, most cases of IgG4-related aortitis are only diagnosed after the patient has undergone aortic surgery.

There is increasing evidence that IgG4-related disease is a common cause of aortitis. In multiple retrospective analysis of patients with aortitis that underwent aortic surgery, IgG4-related disease was common alongside Takayasu's arteritis, Giant Cell arteritis and Rheumatoid aortitis.[9,10] IgG4-related aortitis also seems to cause a characteristic, severe aortic wall thickening.[9] These patients typically presented with chest pain (either acute or chronic), fevers, and one patient with neurological sequelae of IgG4-related disease. In similar reports of IgG4-related aortitis, the condition was diagnosed post-operatively via histological examination of excised aortic specimens.[11=15] Retrospective observational studies have also demonstrated that aortic aneurysms often occur secondary to IgG4-related disease, with approximately 1-3% of all aortic aneurysms meeting the diagnostic criteria for IgG4-related disease.[9,16] It has also been shown that aortic aneurysms with IgG4 positive cell infiltration are at a higher risk of dissection, which provides compelling evidence to proceed with surgical repair in these cases.[17]

In the absence of any surgical pathology such as a suspected acute aortic syndrome or aneurysm meeting the threshold for surgery, the management of IgG4-related aortitis involves immunosuppression with glucocorticoids. Due to its recent recognition, data regarding the natural history of IgG4-related aortitis is sparse, with no randomised clinical trial data. One prospective observational trial showed that glucocorticoids substantially decreased the thickness of periaortic/periarterial lesions in patients with IgG4-related disease, with a 50% reduction in wall thickness after approximately two months of treatment. There is however a distinct lack of data regarding medical management of aneurysmal IgG4-related aortitis, given concerns of an increased risk of rupture secondary to aortic inflammation and early recommendation for surgery.[18] There are also concerns that glucocorticoids fail to control IgG4-related inflammation when tapered to a lower dose, however second-line treatment with rituximab-induced B cell depletion and other steroid-sparing agents have shown to be efficacious.[4,8,19]

Although there is some evidence that non-aneurysmal IgG4-related aortitis can be effectively managed with immunosuppression, the main challenge remains diagnosing this condition before undergoing potentially unnecessary surgery. Clinicians should be suspicious of autoimmune causes of aortitis, keeping in mind that IgG4-related disease often does not cause systemic features.[20] Furthermore, IgG4-related disease is associated with lower inflammatory markers, when compared to other vasculitides such as Giant Cell Arteritis.[21,22] IgG4-related aortitis should be considered as a differential diagnosis when cross-sectional imaging is suggestive of an IMH, and measuring serum IgG4 levels can be helpful in making this diagnosis. Although PET-CT can demonstrate FDG uptake in IgG4-related disease, similar uptake is also observed with acute IMH.[23] Therefore histological analysis remains the gold standard to diagnose IgG4-related disease.[4]

CONCLUSION

This case highlights the challenge of diagnosing IgG4-related aortitis pre-operatively. IgG4-related disease is a common cause of non-infective aortitis and should be considered as a differential diagnosis to IMH. Although surgery is indicated in aneurysmal disease, there is some evidence that the significant aortic thickening often seen in IgG4-related aortitis is responsive to glucocorticoid and rituximab therapy. Robust randomised trial evidence may guide optimal therapy for these patients in the future.

REFERENCES

1. Shchetynska-Marinova T, Amendt K, Sadick M, Keese M, Sigl M. Aortitis - An Interdisciplinary Challenge. In Vivo. 2021;35(1):41–52.
2. Gornik HL, Creager MA. Aortitis. Circulation. 2008;117(23):3039–51.
3. Pugh D, Grayson P, Basu N, Dhaun N. Aortitis: recent advances, current concepts and future possibilities. Heart. 2021;107(20):1620 LP – 1629.
4. Maritati F, Peyronel F, Vaglio A. IgG4-related disease: a clinical perspective. Rheumatology (Oxford). 2020;59(Suppl 3):iii123–31.
5. Hamano H, Kawa S, Horiuchi A, Unno H, Furuya N, Akamatsu T, et al. High serum IgG4 concentrations in patients with sclerosing pancreatitis. N Engl J Med. 2001;344(10):732–8.

6. Umehara H, Okazaki K, Kawa S, Takahashi H, Goto H, Matsui S. The 2020 revised comprehensive diagnostic (RCD) criteria for IgG4-RD. Mod Rheumatol. 2021;31(3):529–33.
7. Al-Khalili OM, Erickson AR. IgG-4 Related Disease: An Introduction. Mo Med. 2018;115(3):253–6.
8. Khosroshahi A, Wallace ZS, Crowe JL, Akamizu T, Azumi A, Carruthers MN, et al. International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. Arthritis Rheumatol. 2015;67(7):1688–99.
9. Pérez-García CN, Olmos C, Vivas D, Ferrera C, García-Arribas D, Enríquez-Vázquez D, et al. IgG4-aortitis among thoracic aortic aneurysms. Heart. 2019;105(20):1583–1589.
10. Stone JH, Khosroshahi A, Deshpande V, Stone JR. IgG4-related systemic disease accounts for a significant proportion of thoracic lymphoplasmacytic aortitis cases. Arthritis Care Res (Hoboken). 2010;62(3):316–22.
11. Babur Güler G, Cantürk E, Güler E, Oran G, Demir GG, Akçevin A, et al. IgG4-related aortitis mimicking intramural hematoma. Anatol J Cardiol. 2016;16(9):728–9.
12. Shilagani C, Lansman S, Gilet A, Flusberg M. IgG4 aortitis of the ascending thoracic aorta: A case report and literature review. J Radiol Case Rep. 2021;15(5):1–9.
13. Leone A, Votano D, Amodio C, Fiaschini C, Berardi M, Pacini D. How to mimic a type A intramural hematoma: A case of IgG4-related aortitis associated with cerebral hemorrhage. J Card Surg [Internet]. 2022;37(1):252–4.
14. Byeon K, Han J, Kim JS, Kim WS, Choe YH, Lee EJ, et al. Immunoglobulin G4-Related Periaortitis Mimicking an Intramural Hematoma. ATS. 2011;92(4):1506–8.
15. Tay DZ, Goh PYT, Teo TK, Boey ML, Chachlani N, Wong PS. Immunoglobulin G4-related aortitis mimicking an intramural hematoma. Asian Cardiovasc Thorac Ann. 2015;23(9):1083–6.
16. Prucha M, Sedivy P, Stadler P, Zdrahal P, Prokopova P, Voska L, et al. Abdominal aortic aneurysm as an IgG4-related disease. Clin Exp Immunol. 2019;197(3):361–5.
17. Kajander H, Paavonen T, Valo T, Tarkka M. Immunoglobulin G4 – positive ascending thoracic aortitis may be prone to dissection. J Thorac Cardiovasc Surg. 2013;146(6):1449–55.
18. Mizushima I, Inoue D, Yamamoto M, Yamada K, Saeki T, Ubara Y, et al. Clinical course after corticosteroid therapy in IgG4-related aortitis/periaortitis and periarteritis: a retrospective multicenter study. Arthritis Res Ther. 2014;16(4):R156.
19. Lanzillotta M, Mancuso G, Della-Torre E. Advances in the diagnosis and management of IgG4 related disease. BMJ. 2020;369.
20. Legatowicz-Koprowska M. IgG4-related disease: why is it so important? Cent J Immunol. 2018;43(2):204–8.
21. Mackie SL, Dejaco C, Appenzeller S, Camellino D, Duftner C, Gonzalez-Chiappe S, et al. British Society for Rheumatology guideline on diagnosis and treatment of giant cell arteritis. Rheumatology. 2020;59(3):e1–23.
22. Wallace ZS, Deshpande V, Mattoo H, Mahajan VS, Kulikova M, Pillai S, et al. IgG4-Related Disease: Clinical and Laboratory Features in One Hundred Twenty-Five Patients. Arthritis Rheumatol (Hoboken, NJ). 2015;67(9):2466–75.

23. Hayashida T, Sueyoshi E, Sakamoto I, Uetani M, Chiba K. PET features of aortic diseases. AJR Am J Roentgenol. 2010;195(1):229–33.