

Magnetic resonance imaging-based diagnosis of aortitis preceding development of a thoracic aneurysm in a patient with giant cell arteritis: a case report

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Received 28 October 2021; first decision 20 December 2021; accepted 6 April 2022; online publish-ahead-of-print 12 April 2022

Background

Inflammatory manifestation in the aortic arch can be a complication of giant cell arteritis (GCA), potentially requiring surgical therapy in the case of aneurysmatic dilatation.

Case summary

We report the case of a 73-year-old female patient with GCA in whom a typical appearance of arteritis was visualized on magnetic resonance imaging of the superficial temporal arteries. Additionally, ectasia (4.7 cm) of the ascending aorta with a mural rim of increased contrast media uptake was detected at the time of the initial diagnosis, which is an indicator of aortitis. While the diameter had only minimally increased in a computed tomography angiography (CTA) examination after 8 months, a subsequent CTA revealed an increased diameter of 5.8 cm and maximum at the level of the ascending aorta another 22 months later, indicating urgent surgery to replace the ascending aorta.

Discussion

Magnetic resonance imaging can detect silent, generalized manifestations of GCA such as severe aortitis, which may possibly lead to aneurysmatic dilatation, urging closer follow-up imaging. Detection of the ongoing process and subsequent follow-up imaging protects patients by avoiding rupture.

Keywords

Giant cell arteritis • Aortitis • Ascending aorta aneurysm • MRI • Surgery • Case report

ESC Curriculum

2.3 Cardiac magnetic resonance • 7.5 Cardiac surgery • 9.1 Aortic disease

Learning points

- Despite the absence of clinical and serological signs of ongoing inflammation, an asymptomatic, life-threatening thoracic aortic aneurysm with persistent active vasculitis can develop in patients with giant cell arteritis (GCA).
- Findings of aortitis in patients with GCA should indicate long-term follow-up imaging to prevent the occult formation of an aortic aneurysm.

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Handling Editor: Tina Khan

Peer-reviewers: Matthew Williams and Andreas Mitsis

Compliance Editor: Alexander Tindale

Supplementary Material Editor: Tinka Julia van Trier

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Introduction

Giant cell arteritis (GCA) represents a primary large-vessel vasculitis occurring in elderly patients, predominantly in women. Rapid treatment with anti-inflammatory medication is considered the highest priority to prevent acute cranial complications such as ischaemic ophthalmic and cerebral events.¹ Extracranial manifestations in the aorta and the major aortic branches have been described in 30–80% of patients with GCA and can occur without specific symptoms or core clinical manifestations.² Data regarding increased risk of aortic aneurysm in patients with giant GCA range from a two-fold increase to being 17 times more likely (in the thoracic aorta) than in the normal population.^{3,4}

In addition to the clinical symptoms and inflammatory laboratory parameters, cranial GCA can be assessed by biopsy and imaging using ultrasound or magnetic resonance imaging (MRI). For extracranial large-vessel involvement, various imaging methods such as MRI, computed tomography (CT), or 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET)/CT are recommended.⁵ Studies using FDG-PET/CT revealed that patients with GCA with aortitis suffer from an increased risk of subsequent aneurysmatic dilatation.^{6,7} There are only limited data about the potential value of MRI in providing prognostic information about the risk of an aortic aneurysm and other aortic events.⁸

This case report illuminates the value of an MRI-based diagnosis of active thoracic aortitis for the awareness of the potential future development of a life-threatening aneurysm.

Timeline

June 2017	Initial diagnosis of GCA based on clinical and serological findings, supported by cranial MRI. Additional MRI finding of thoracic aortitis associated with mild ectasia (ascending aorta: 4.7 cm)
February 2018	Follow-up computed tomography: no relevant change: diameter of the ascending aorta: 4.8 cm
December 2019	Subsequent follow-up computed tomography: diameter of the ascending aorta: 5.8 cm. Patient is asymptomatic
January 2020	Replacement of the ascending aorta with an aortic graft with two bypass grafts towards the posterolateral myocardial wall

Case presentation

A female patient (73 years) presented with general signs of inflammation [C-reactive protein (CRP): 115 mg/L] and new onset of headaches. There were no thoracic symptoms. The MRI confirmed the clinical diagnosis of cranial GCA with typically increased contrast media uptake in the wall of the superficial temporal arteries

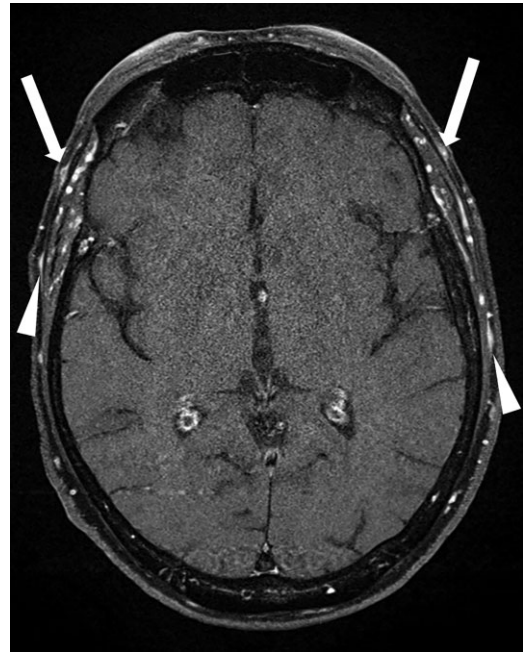


Figure 1 Cranial magnetic resonance imaging (high-resolution black-blood T1-weighted spin-echo sequence, fat-suppressed, contrast-enhanced) in 2017: increased signal at the frontal branches (arrows) and at the parietal branches (arrowheads) of the superficial temporal arteries as a sign of active giant cell arteritis.

(Figure 1). Additionally, acquired images of the thoracic aorta performed as a routine protocol in our clinic revealed ectasia of the ascending aorta (4.7 cm) with circumferential wall thickening accompanied by increased contrast media uptake, indicating aortitis (Figure 2). Under immunosuppressive therapy (prednisolone started with 100 mg daily with a subsequent long-term reduction to 0.5 mg daily), clinical symptoms and signs of inflammation subsided. Due to concerns about aortic ectasia with signs of active vasculitis in the initial MRI, annual follow-up was recommended. After 1 year, CT demonstrated a nearly stable aortic diameter of 4.8 cm (Figure 3A). There were no noticeable signs of inflammation in the CT, corresponding to clinical and serological regression (CRP: 2 mg/L). Surprisingly, subsequent CT follow-up indicated substantial progression with an increased aneurysmatic diameter up to 5.8 cm after 1.5 years (Figure 3B). There were still no significant serological signs of recurrent inflammation (CRP: 7 mg/L). Cardiac catheter examination as a routine procedure before replacing the ascending aorta indicated severe stenoses of the right coronary artery and in the left anterior descending artery. The patient was scheduled for aortic and bypass surgery. Although serological parameters were negative, histology of the ascending aorta revealed a fragmented Tunica media characterized by the presence of immune cell infiltrates, including macrophages and granulomas as a sign of active aortitis (Figure 4). According to the control MRI after 6 months, the patient's post-operative condition was normal and she did not present with any clinically remarkable signs of disease.

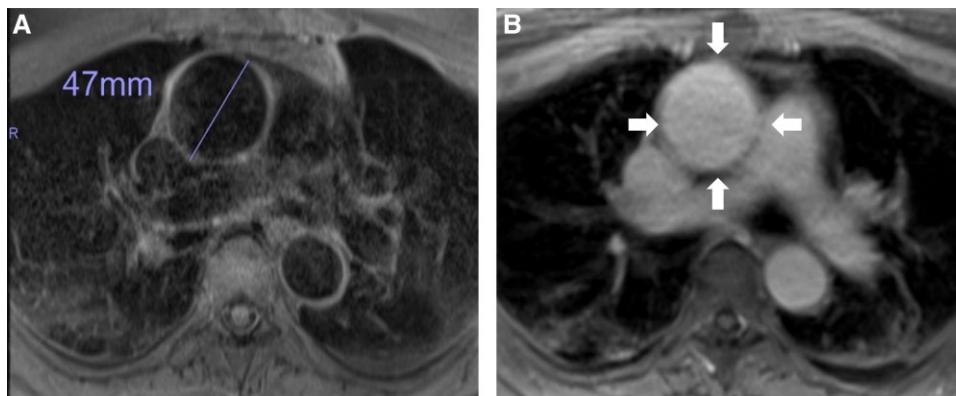


Figure 2 Thoracic magnetic resonance imaging in 2017: proton-weighted dark-blood sequence (A) and contrast-enhanced, T1-weighted, fat-suppressed volumetric interpolated breath-hold examination sequence (B). There is ectasia of the ascending aorta with a diameter of 4.7 cm (A), accompanied by a circumferential, moderately increased contrast media uptake in the thickened vessel wall (arrows), indicating aortitis.

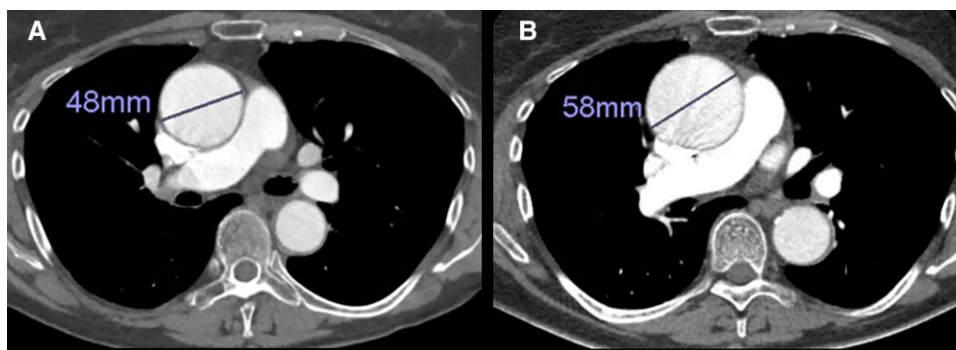


Figure 3 Contrast-enhanced computed tomography of the thoracic aorta: progressive diameter of the ascending aorta with transition from 4.8 cm (A, ectasia in 2018) to an aneurysm of 5.8 cm (B, 2019). In contrast to the more sensitive magnetic resonance imaging technique, no inflammation of the vessel wall can be determined on computed tomography at different time points.

Discussion

We report a case of granulomatous necrotizing aortitis with active immune cell infiltrations in an aneurysm of the ascending aorta. After an initial, uncomplicated course of GCA with signs of inflammation at the superficial temporal artery and the thoracic aorta, vasculitis was clinically in remission, and the diameter of the aorta was nearly stable during the first year (Figure 3A). Surprisingly, another CT in the following year demonstrated a marked increase in the ascending aorta diameter (Figure 3B), and the post-operative histological evaluation revealed active granulomatous vasculitis (Figure 4). In contrast, serological parameters and clinical findings were unremarkable.

The development of an aortic aneurysm several months or years after the primary diagnosis is known in particular from FDG-PET studies, in which progressive aortic dilatation was predominantly found in patients with initially active aortitis.^{6,7} Pathophysiologically, initial fragmentation of elastic fibres in the aortic wall as found in our patient (Figure 4) can be expected. The incidence of aortic structural damage (aneurysm or dilatation) was found to be maximal

within the first 5 years after diagnosis, developing continually over time and affecting up to 33.3% of individuals after long-term follow-up in a study of García-Martínez *et al.*⁸

Our case report points out that MRI findings of active aortitis in patients with GCA may also be found several years before clinically relevant aortic dilatation develops. Although patients with clinically symptomatic aortitis have been described to be at risk for a complicated disease course,⁹ our case demonstrates that the possible development of an aortic aneurysm should not be ignored in asymptomatic patients without serological signs of inflammation. Even though systematic follow-up imaging is performed emergency aortic surgery or interventional therapy might occur which is associated with a limited outcome compared to a planned procedure.¹⁰ However, it still remains unclear how follow-up should be performed. While monitoring is based on symptoms, clinical findings, and ESR/CRP levels, which is emphasized in the 2018 update of the EULAR recommendations for the management of large-vessel vasculitis, routine imaging to assess activity is not recommended in cases of clinical and biochemical remission but is suggested for long-term monitoring of dilatations and aneurysms. The best imaging

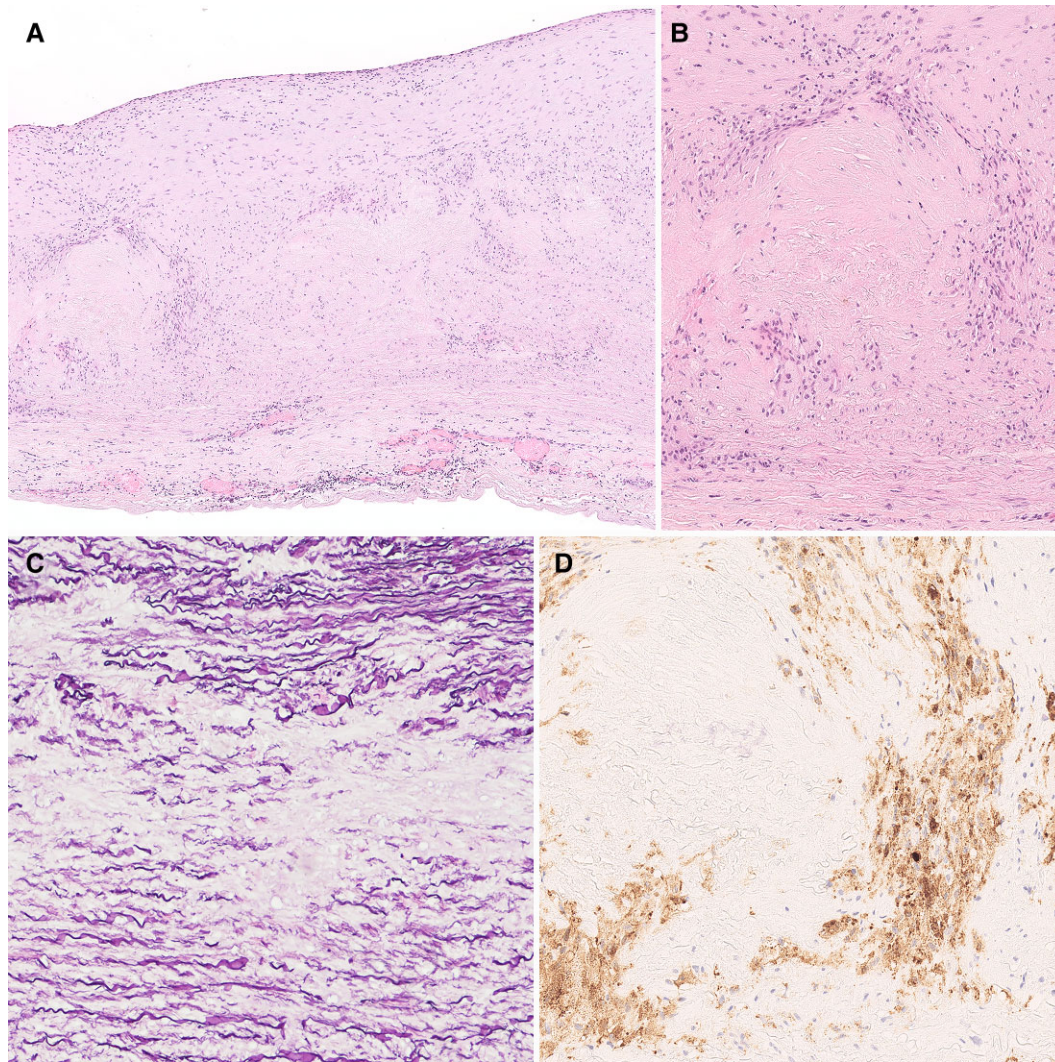


Figure 4 Histological assessment of the aortic resection specimen confirmed the presence of several granulomas (A and B) in the media of the aorta, leading to fragmentation of elastic fibres (C). Immunostaining confirmed the presence of CD68-positive histiocytes lining the granulomas. Haematoxylin and eosin (A and B), Elastic van Gieson staining (C), and anti-CD68 immunostaining. Original magnifications: 50-fold (A), 100-fold (B), and 200-fold (C and D).

method (MRI, CT, or FDG-PET/CT) should be chosen on an individual basis.^{5,11} Both FDG-PET/CT and MRI are valuable tools in the diagnosis of large-vessel vasculitis. Recently, it was reported that MRI better delineates disease extent and that PET is better suited to assess inflammatory vascular activity.¹² Nevertheless, MRI and PET were found to be unreliable for assessing large-vessel inflammation in patients with aortic arch syndrome in the course of GCA in the presence of pre-existing immunosuppressive therapy.¹³ Persistent mural inflammation or vascular re-modelling processes are presumed to cause these limitations.⁵ As our patient had also received immunosuppressive medication, we refrained from follow-up imaging by means of MRI or FDG-PET/CT. Instead, we applied CT, which also has limitations in assessing disease activity, but is more cost-effective than MRI and can reliably evaluate aortic diameter and vessel wall thickening.^{5,14} Radiation exposure must be taken

into account in CT and FDG-PET/CT, but this was not the most important concern in our 73-year-old patient. For exclusion of ongoing dilatation, ultrasound would represent a reasonable alternative in the abdominal aorta.⁵ Apart from initially identifying patients with large-vessel vasculitis, it is also still under discussion as to whether imaging of the aorta should be generally performed in patients with GCA or if screening should focus on sub-groups such as patients with extracranial symptoms or those with a severe inflammatory response associated with hypertension at the time of diagnosis of GCA.¹⁵

Conclusion

This case report demonstrates that a large thoracic aortic aneurysm with evidence of active vasculitis on biopsy can develop despite the

absence of clinical and serological signs of ongoing inflammation. The initial diagnosis of active aortitis as revealed by MRI should prompt long-term follow-up by appropriate imaging methods.

Lead author biography



Georg Lutter has a surgical professorship at the University Medical Center Schleswig-Holstein, Campus Kiel, Germany. He is Chair for Experimental Cardiac Surgery and Heart Valve Technologies at the Christian-Albrechts-University of Kiel, Germany. His main interests are innovative trans-catheter mitral, tricuspid, aortic, and tissue-engineered pulmonary valved stent technologies.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Acknowledgements

The authors are grateful for the support of Anna Wolf and the DZHK (German Center for Cardiovascular Research, Germany).

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: This study was financially supported by the German Centre for Cardiovascular Research (DZHK) and the Deutsche Forschungsgemeinschaft (DFG) within the funding programme 'Open Access publication'.

References

1. Soriano A, Muratore F, Pipitone N, Boiardi L, Cimino L, Salvarani C. Visual loss and other cranial ischaemic complications in giant cell arteritis. *Nat Rev Rheumatol* 2017; **13**:476–484.

2. de Boysson H, Liozon E, Espitia O, Daumas A, Vautier M, Lambert M, Parienti J-J, Granel B, Dumont A, Sultan A, Manrique A, Saadoun D, Ly KH, Agard C, Aouba A. Different patterns and specific outcomes of large-vessel involvements in giant cell arteritis. *J Autoimmun* 2019; **103**:102283.
3. Evans JM, O'Fallon WM, Hunder GG. Increased incidence of aortic aneurysm and dissection in giant cell (temporal) arteritis. A population-based study. *Ann Intern Med* 1995; **122**:502–507.
4. Robson JC, Kiran A, Maskell J, Hutchings A, Arden N, Dasgupta B, Hamilton W, Emin A, Culliford D, Luqmani RA. The relative risk of aortic aneurysm in patients with giant cell arteritis compared with the general population of the UK. *Ann Rheum Dis* 2015; **74**:129–135.
5. Dejaco C, Ramiro S, Duftner C, Besson FL, Bley TA, Blockmans D, Brouwer E, Cimmino MA, Clark E, Dasgupta B, Diamantopoulos AP, Direskeneli H, Iagnocco A, Klink T, Neill L, Ponte C, Salvarani C, Slart RHJA, Whitlock M, Schmidt WA. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice. *Ann Rheum Dis* 2018; **77**:636–643.
6. Blockmans D, Coudyzer W, Vanderschueren S, Stroobants S, Loeckx D, Heye S, De Ceuninck L, Marchal G, Bobbaers H. Relationship between fluorodeoxyglucose uptake in the large vessels and late aortic diameter in giant cell arteritis. *Rheumatology* 2008; **47**:1179–1184.
7. Muratore F, Crescentini F, Spaggiari L, Pazzola G, Casali M, Boiardi L, Pipitone N, Croci S, Galli E, Aldigeri R, Versari A, Salvarani C. Aortic dilatation in patients with large vessel vasculitis: a longitudinal case control study using PET/CT. *Semin Arthritis Rheum* 2019; **48**:1074–1082.
8. García-Martínez A, Arguis P, Prieto-González S, Espígol-Frigolé G, Alba MA, Butjosa M, Tavera-Bahillo I, Hernández-Rodríguez J, Cid MC. Prospective long term follow-up of a cohort of patients with giant cell arteritis screened for aortic structural damage (aneurysm or dilatation). *Ann Rheum Dis* 2014; **73**:1826–1832.
9. Espitia O, Blonz G, Urbanski G, Landron C, Connault J, Lavigne C, Roblot P, Maillot F, Audemard-Verger A, Artifoni M, Durant C, Guyomarch B, Hamidou M, Magnant J, Agard C, French Study Group for Large Vessel Vasculitis (GEFA). Symptomatic aortitis at giant cell arteritis diagnosis: a prognostic factor of aortic event. *Arthritis Res Ther* 2021; **23**:14.
10. Lau C, Leonard JR, Iannacone E, Gaudino M, Girardi LN. Surgery for acute presentation of thoracoabdominal aortic disease. *Semin Thorac Cardiovasc Surg* 2019; **31**:11–16.
11. Hellmich B, Agueda A, Monti S, Buttgerit F, de Boysson H, Brouwer E, Cassie R, Cid MC, Dasgupta B, Dejaco C, Hatemi G, Hollinger N, Mahr A, Mollan SP, Mukhtyar C, Ponte C, Salvarani C, Sivakumar R, Tian X, Tomasson G, Turesson C, Schmidt W, Villiger PM, Watts R, Young C, Luqmani RA. 2018 Update of the EULAR recommendations for the management of large vessel vasculitis. *Ann Rheum Dis* 2020; **79**:19–30.
12. Quinn KA, Ahlman MA, Malayeri AA, Marko J, Civelek AC, Rosenblum JS, Bagheri AA, Merkel PA, Novakovich E, Grayson PC. Comparison of magnetic resonance angiography and 18F-fluorodeoxyglucose positron emission tomography in large-vessel vasculitis. *Ann Rheum Dis* 2018; **77**:1165–1171.
13. Both M, Ahmadi-Simab K, Reuter M, Dourvos O, Fritzer E, Ullrich S, Gross WL, Heller M, Bahre M. MRI and FDG-PET in the assessment of inflammatory aortic arch syndrome in complicated courses of giant cell arteritis. *Ann Rheum Dis* 2008; **67**:1030–1033.
14. Lariviere D, Benali K, Coustet B, Pasi N, Hyafil F, Klein I, Chouchard M, Alexandra J-F, Goulenok T, Dossier A, Dieude P, Papo T, Sacre K. Positron emission tomography and computed tomography angiography for the diagnosis of giant cell arteritis: a real-life prospective study. *Medicine* 2016; **95**:e4146.
15. Mackie SL, Hensor EM, Morgan AW, Pease CT. Should I send my patient with previous giant cell arteritis for imaging of the thoracic aorta? A systematic literature review and meta-analysis. *Ann Rheum Dis* 2014; **73**:143–148.