EJCRIM
European Journal
of Case Reports
in Internal Medicine

PROLONGED FOLLOW-UP OF A CASE OF TAKAYASU'S ARTERITIS PRESENTING WITH SPASTIC PARAPLEGIA SECONDARY TO THORACIC COMPRESSIVE MYELOPATHY

Matthew Spiteri, Victoria Farrugia, Michela Frendo

Department of Internal Medicine, Mater Dei Hospital, Msida, Malta

Corresponding author's e-mail: victoria.farrugia.3@gov.mt

Received: 18/12/2025
Accepted: 12/01/2026
Published: 03/02/2026

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: Written informed consent was obtained from the patient for publication of this report and accompanying images.
This article is licensed under a Commons Attribution Non-Commercial 4.0 License

How to cite this article: Spiteri M, Farrugia V, Frendo M. Prolonged follow-up of a case of Takayasu's arteritis presenting with spastic paraplegia secondary to thoracic compressive myelopathy. *EJCRIM* 2026;**13**:doi:10.12890/2026_006089

ABSTRACT

This is the case of a 63-year-old woman who presented with a few weeks' history of progressive bilateral lower limb weakness and spastic paraparesis. Urgent imaging revealed spinal cord compression caused by dural thickening between the T2 and T9 levels, as well as an incidental finding of ongoing aortitis. The impression of a large-vessel vasculitis with concurrent thoracic pachymeningitis was given. The patient underwent an urgent thoracic laminectomy and tissue samples were sent for culture and histology. Tissue cultures later identified Enterobacter and Pseudomonas species, introducing further diagnostic complexity. Histology showed findings consistent with a non-specific chronic inflammatory process. An autoimmune and vasculitic screen revealed nothing of note and IgG4 levels were within normal range. A diagnosis of Takayasu's arteritis with thoracic compressive myelopathy was made. Long-term follow-up was essential to the patient's management following her diagnosis of Takayasu's arteritis. Ongoing surveillance was required to assess physical recovery after prolonged immunosuppression and rehabilitation, as well as to recognise and detect delayed complications. These were related both to the vasculitis and to her prolonged corticosteroid therapy, despite an initial favourable response to the multidisciplinary treatment.

KEYWORDS

Takayasu's arteritis, spastic paraplegia, thoracic myelopathy, aortitis, immunosuppression

LEARNING POINTS

- Spastic paraplegia is an uncommon and potentially misleading presentation of Takayasu's arteritis. The resulting dural thickening may initially mimic primary spinal pathology.
- The presence of concurrent large-vessel inflammation is a crucial diagnostic clue; positron emission tomography/computed tomography scans and magnetic resonance imaging are invaluable in confirming vasculitis and monitoring treatment response.
- Long-term follow-up is essential, as it reveals new and significant complications related both to Takayasu's arteritis and to its management.

EFIM
European Federation of Internal Medicine

Publisher
smc media srl

INTRODUCTION

Takayasu's arteritis (TA) is a chronic, granulomatous large-vessel vasculitis, that typically causes symptoms due to occlusive thromboarthropathy[1]. It predominantly occurs in young women and affects the aorta and its main branches. Vessel inflammation leads to wall thickening, fibrosis, stenosis and thrombus formation[1]. TA can present with neurologic manifestations due to ischaemia such as visual loss, transient ischaemic attack and stroke[1,2]. Compressive thoracic myelopathy is a rare neurological manifestation of TA, seen in only a handful of cases[3,4].

CASE DESCRIPTION

A 63-year-old woman presented with a four-month history of progressive bilateral lower limb weakness, initially asymmetrical affecting the right leg more severely. She described associated sensory disturbance in both feet and an unsteady gait. She denied any history of back pain, urinary incontinence or retention, and bowel incontinence. She also denied any joint pains, morning stiffness or rashes. Her only medical condition at the point of admission was hypertension.

In the weeks prior to presentation the patient became rapidly dependent on a walking stick for mobility where beforehand she was achieving this independently. Neurological examination at the emergency department revealed spastic paraparesis; there was loss of power affecting most of the lower limb muscle groups. She had brisk reflexes and a positive Babinski sign bilaterally. Sensory examination showed patchy impaired response to pinprick and vibration testing in the lower limbs; there was no neurological deficit of the upper limbs. A magnetic resonance imaging (MRI) whole spine with contrast and a computed tomography of the thorax, abdomen and pelvis (CT TAP) were performed urgently to exclude any malignancy. An MRI scan revealed extensive dural thickening within the upper thoracic spine with significant spinal cord compression *(Fig. 1)*. The CT TAP revealed an incidental finding of ongoing aortitis with enhancement of the ascending aorta and of the aortic arch.

The following day the patient underwent a thoracic laminectomy for urgent decompression of the spinal cord. Tissue samples were sent for histology, culture and sensitivities. Histology findings were consistent with a non-specific chronic inflammatory process, while cultures detected *Enterobacter cloacae* and *Pseudomonas aeruginosa*. The patient was started on intravenous (IV) dexamethasone and appropriate IV antibiotics. Specialist input from rheumatology, infectious diseases, neurosurgery and neurology was sought. A positron emission tomography/computed tomography (PET-CT) scan was carried out and confirmed the finding of ongoing large-vessel vasculitis with increased uptake *(Fig. 2)*.

Blood work including a vasculitis and autoimmune screen was taken *(Table 1)*. This revealed an elevated erythrocyte sedimentation rate (ESR) and a positive anti-neutrophil

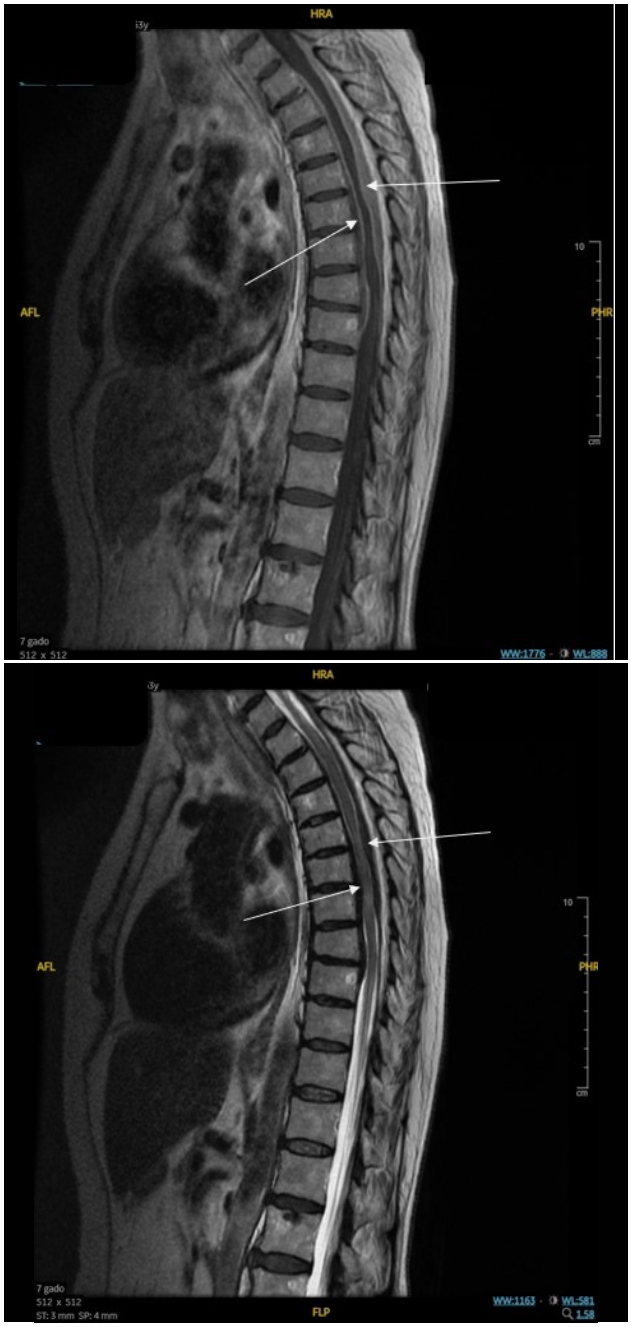


Figure 1. T1 (top) and T2 (bottom) weighted MRI spine showing marked dural thickening between the T2 to T9 levels.

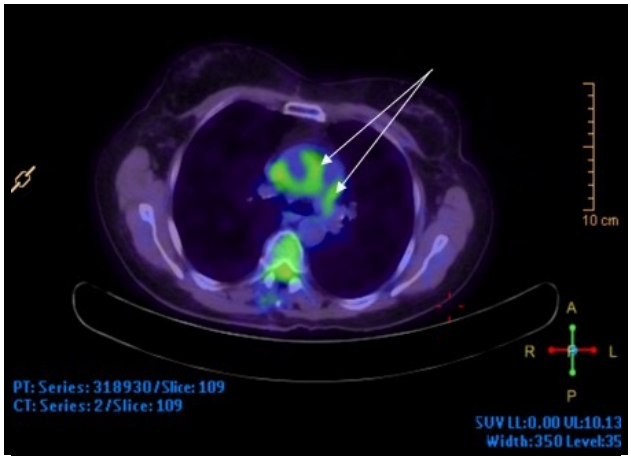


Figure 2. PET Scan showing increased uptake at arch of aorta indicative of aortitis.

Investigation	Result	Reference range
C-reactive protein (mg/l)	47.5	0–5
ESR (mm 1st hour)	75	18–22
Creatine kinase (U/l)	52	26–192
White blood cell count (×10⁹/l)	6.84	4.3–11.4
Eosinophil count (×10⁹/l)	0.08	0–0.6
Anti-nuclear antibody	Pos 1/320 titre	-
Complement 3 (mg/l)	1590	900–1800
Complement 4 (mg/l)	206	100–400
Anti-myeloperoxidase antibody (U/ml)	>200	0–20
Anti-proteinase 3 antibody (U/ml)	<2	0–20
Total extractable nuclear antigen (RU/ml)	1.	0–19.9
Rheumatoid factor immunoglobulin M (IgM) (IU/ml)	11.4	0–15.9
Anti-double stranded DNA antibody (DsDNA) (IU/ml)	11.8	0–100
Cryoglobulins	Negative	-
Immunoglobulin G4 (IgG4) (g/l)	0.62	0.04–0.86
Immunoglobulin G4 (IgG4) repeat test (g/l)	0.71	0.04–0.86
Urinalysis	No proteinuria	-

Table 1. *Laboratory findings during presenting admission.*

cytoplasmic antibody (ANCA); anti-myeloperoxidase. However, it was noted that the patient had a long-standing history of positive ANCA and was previously thoroughly investigated for small vessel vasculitis. She had no ear, nose or throat signs or symptoms, no lung involvement (normal high-resolution chest CT), no vasculitic rash and a normal eosinophil count. Also of note, IgG4 serum levels were within normal range on two separate tests.

The patient responded well to steroid therapy, with mobility improving gradually with the help of physical therapy. Neurological examination carried out one week after starting treatment showed improvement in lower limb power almost back to baseline. A repeat MRI of the whole spine was taken two weeks after the operation which showed that the previous dural thickening had resolved to normal appearance. The patient was switched from dexamethasone to prednisolone 10 mg daily and continued IV antibiotics for a total of three weeks. After four weeks of inpatient management the patient was discharged from hospital and was transferred to a rehabilitation centre to continue physiotherapy.

The patient was followed up regularly at rheumatology and neurology outpatient clinics. Following a bone mineral density scan which showed evidence of osteoporosis, prednisolone was decreased to 5 mg on alternating days, and the patient was started on zoledronic acid. For follow-up imaging, a PET-CT scan and MRI whole spine scan were carried out at three and four months respectively following discharge from hospital. The PET-CT showed moderate improvement but persistent metabolic uptake from the aortic arch, indicating ongoing vasculitis. The MRI of the spine showed an increase in dural thickening at T6/T7 when compared to the previous MRI. The results of this case were discussed at a Neuroscience team meeting, and despite worsening dural thickness, it was decided not to carry out any further operations and to manage with immunosuppression, provided that the patient did not deteriorate clinically. Following rheumatology follow-up, the patient was started on methotrexate 10 mg weekly for two weeks then it was increased to 15 mg weekly, and prednisolone was increased to 10 mg daily. To assess the response of increased immunosuppression, a repeat MRI of the spine was carried out that showed decreased dural thickness and degree of enhancement when compared to previous imaging *(Fig. 3)*. In view of this improvement, prednisolone was gradually tapered down until it was stopped, and methotrexate was reduced to 12.5 mg weekly. The patient remained clinically stable on this regimen of immunosuppression.

Two years following discharge from hospital, the patient

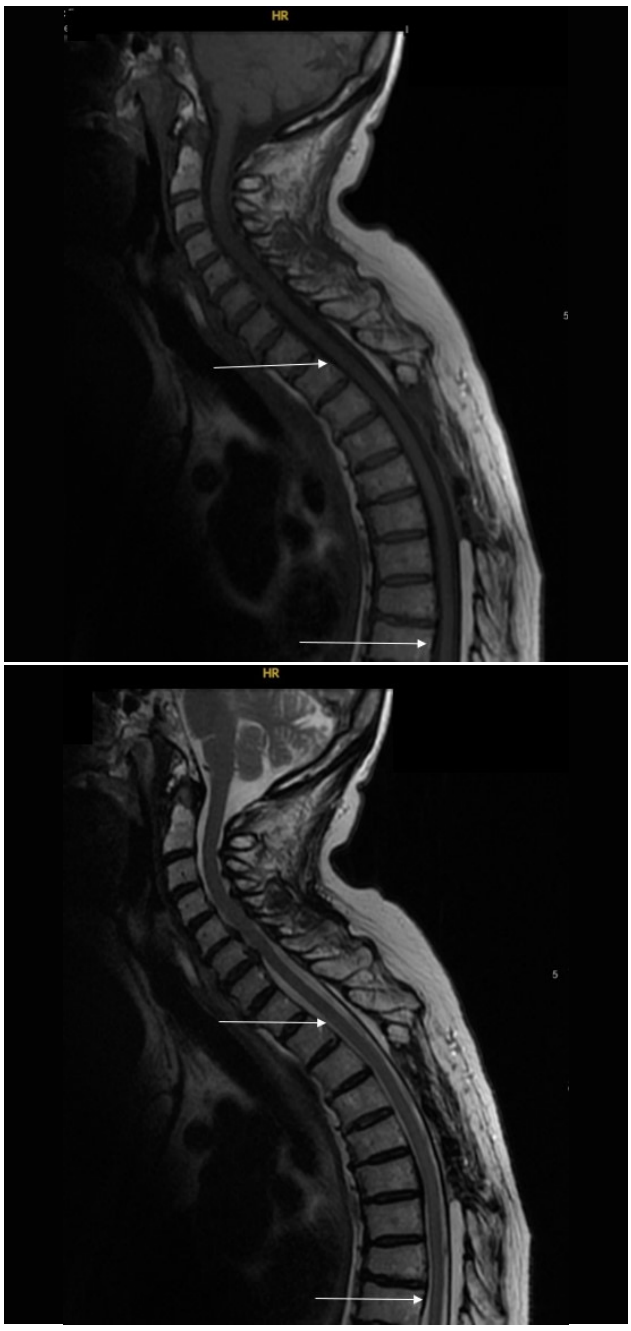


Figure 3. T1 (top) and T2 (bottom) imaging of the thoracic spine (T2-T9) three years post-laminectomy and ongoing immunosuppression.

was admitted under medical care in view of severe shortness of breath on exertion and bilateral pitting oedema. A CT of the thorax was carried out to rule out interstitial lung disease as a complication; however, this was normal. On examination she was noted to have an ejection systolic murmur radiating to the carotids, and bilateral chest crackles. An echocardiogram showed mild aortic valve stenosis and severe aortic regurgitation, which was thought to be a complication of the aortitis. An aortic valve replacement was attempted; however, it was abandoned in view of extensive fibrotic adhesions overlying the aorta. A pericardial biopsy taken at the time of the surgery showed fibroconnective tissue with inflammation. The patient was referred for a transcatheter aortic valve implantation (TAVI).

Three years after admission the patient still attends regular rheumatology outpatient's follow-up. She is currently stable in terms of disease progression; she is on regular methotrexate and has regained normal mobility. She has been discharged from neurological follow-up and is currently waiting for her TAVI to be carried out.

DISCUSSION

This case portrays the initial, uncommon presentation of TA and its long-term complications. TA typically presents either at the early systemic phase with non-specific symptoms of fever, lethargy, weight loss, night sweats and myalgias, or the late occlusive phase, with symptoms of ischaemia secondary to arterial occlusion[1]. Symptoms can vary depending on the arteries involved such as limb claudication, angina and hypertension. Neurological symptoms typically occur due to occlusion of related arteries, causing stroke, syncope and dizziness[1,2]. Thoracic myelopathy caused by compression is a documented but uncommon presentation[3,4]. This can occur either by spinal cord compression due to mass effect from aortic inflammation or by inflammatory extension to the dura as was seen in this case[5]. One of the main differential diagnoses early in the case was that of an IgG4-related disease, as this too can present with aortitis and pachymeningitis[6,7]. However, this was ruled out due to normal IgG4 serum levels on two separate tests and non-specific histological findings. As the samples cultured bacterial growth, this added to the diagnostic difficulty. The patient did not show signs and symptoms of systemic infection, and improved clinically with immunosuppression, supporting a primarily inflammatory process with possible secondary colonisation or contamination.

Although this patient does not fall under the European Alliance of Associations for Rheumatology (EULAR) research criteria for TA[8], given the presence of large-vessel vasculitis with high inflammatory markers and associated presence of pachymeningitis, a clinical diagnosis of TA was made. Regarding treatment, due to the acute presentation of spinal cord compression urgent neurosurgical decompression was needed. Following this, histology confirmed an ongoing inflammatory process which guided prompt initiation of immunosuppression with intravenous corticosteroids. Treatment was subsequently effectively tapered down with oral steroids and eventually a steroid-sparing agent; methotrexate[2]. Immunosuppression with such agents is the mainstay of treatment in TA[2]. Although evidence of management of spinal compression in TA is limited, a number of cases report that a similar approach of emergency decompressive laminectomy, followed by immunosuppression, has proved effective[4,5].

This case, in addition to highlighting the atypical clinical presentation of TA, clearly illustrates the progressive course of the disease and the subsequent development of complications over time. From the aortitis, the patient developed severe aortic regurgitation, presenting to

hospital with congestive heart failure, shortness of breath on exertion and bilateral limb oedema. The pathophysiology of fibrosis at the aortic root in TA is as follows: in the early phase, the active inflammation consists of mononuclear cell infiltration and oedema with fragmentation of the elastic fibres at the medio-adventitial junction where the tunica media and tunica adventitia, the two outer layers of a blood vessel wall, meet[9]. Laminar medial necrosis may occur. Later, there is reactive fibrosis in the intima and increased ground substance in the intimal medial junction[9]. The patient required surgical intervention for an aortic valve replacement; however, this was unsuccessful in view of inflammatory fibrotic changes of the aorta. She is now waiting for a TAVI to be carried out.

Another complication observed, secondary to the treatment given for TA, is that the patient developed glucocorticoid-induced osteoporosis following steroid use for immunosuppression. The pathophysiology involves direct effects on bone cells: glucocorticoids increase the receptor activator of nuclear factor-kappa B ligand (RANKL)-mediated osteoclastogenesis and prolong osteoclast lifespan leading to increased bone resorption, while simultaneously suppressing osteoblastogenesis and promoting osteoblast and osteocyte apoptosis, resulting in decreased bone formation[10]. Therefore, blocking osteoclast activity with RANKL inhibitors (e.g. denosumab) or bisphosphonates is effective in preventing glucocorticoid-induced osteoporosis[10]. In this case, the patient was treated with bisphosphonates and followed up regularly with bone density scans.

In conclusion, spastic paraplegia can be an uncommon and misleading presentation of TA. Initial investigations showing pachymeningitis can mislead physicians to a primary spine lesion; evidence of concurrent large-vessel inflammation guides the diagnosis towards TA. Appropriate investigation with imaging such as MRI and PET-CT scans, and blood work including inflammatory markers, a vasculitic and immunology screen is necessary to rule out potential differential diagnosis. Prompt management with thoracic laminectomy for spinal cord decompression followed by immunosuppression is essential for optimal neurological recovery. A swift and effective multidisciplinary approach to management will warrant ideal outcomes. Appropriate long-term follow-up is necessary to manage complications arising from both the aortitis itself and prolonged immunosuppressive therapy.

REFERENCES

1. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. *J Clin Pathol* 2002;**55**:481–486. doi: 10.1136/jcp.55.7.481
2. Somashekar A, Leung YT. Updates in the diagnosis and management of Takayasu's arteritis. *Postgrad Med* 2023;**135**:14–21. doi: 10.1080/00325481.2022.2159723
3. Chandrasekar N, Thandapani M, Govindasamy R, Rudrappa S, Vaithialingam B. Thoracic compressive myelopathy in a patient with Takayasu arteritis. *Cureus* 2023;**15**:e33496. doi: 10.7759/cureus.33496
4. Murugan C, Kavishwar RA, Ramachandran K, Shetty AP, B T P, Rajasekaran S. Spinal Hypertrophic Pachymeningitis Causing Thoracic Myelopathy in a Patient of Takayasu Arteritis: A Case Report. *JBJS Case Connect* 2021;**11**. doi: 10.2106/JBJS.CC.21.00520
5. Kim HA, Kim JH, Won JH, Suh CH. An unusual clinical manifestation of Takayasu's arteritis: spinal cord compression. *Joint Bone Spine* 2009;**76**:209–212. doi: 10.1016/j.jbspin.2008.09.007
6. Gautier F, Neumann L, Adle-Biassette H, Rubenstein E, Bernat AL, Chimon A, et al. Pachymeningitis associated with IgG4-related disease and ANCA positivity: case report and review of the literature. *Autoimmun Rev* 2023;**22**:103285. doi: 10.1016/j.autrev.2023.103285
7. Slade SJ, Bauer EM, Stone VV, Dave AJ. Spinal IgG4-related hypertrophic pachymeningitis with spinal cord compression: case report and literature review. *World Neurosurg* 2019;**130**:65–70. doi: 10.1016/j.wneu.2019.06.130
8. Grayson PC, Ponte C, Suppiah R, Robson JC, Gribbons KB, Judge A, et al. DCVAS Study Group. 2022 American College of Rheumatology/EULAR classification criteria for Takayasu arteritis. *Ann Rheum Dis* 2022;**81**:1654–1660. doi: 10.1136/ard-2022-223482
9. Joseph G, Goel R, Thomson VS, Joseph E, Danda D. Takayasu arteritis: JACC Focus Seminar 3/4. *J Am Coll Cardiol* 2023;**81**:172–186. doi:10.1016/j.jacc.2022.09.051
10. Hofbauer LC, Compston JE, Saag KG, Rauner M, Tsourdi E. Glucocorticoid-induced osteoporosis: novel concepts and clinical implications. *Lancet Diabetes Endocrinol* 2025;**13**:964–979. doi: 10.1016/S2213-8587(25)00251-7.