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LETTER TO THE EDITOR Open Access

FDG-PET/CT as a useful tool for disease activity assessment in large vessel vasculitis in childhood

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The hybrid imaging modality, fluoro-D-glucose positron emission tomography/computed tomography (FDG-PET/CT), facilitates high-resolution anatomical imaging, while concurrently visualizing metabolically active processes.

FDG-PET/CT has become a widely established tool to assist in the detection of inflammatory processes in large vessels, such as Takayasu arteritis (TA), often presenting with a nonspecific clinical presentation [1, 2].

We retrospectively present four females, aged from 13 to 17 years, suffering from Takayasu arteritis (TA). Despite a multiregional referral area of our tertiary hospital, just these 4 patients were diagnosed and treated with between 2015 and 2022, emphasizing the rare incidence of this disease. One patient is of Asian descent, while the others are of Caucasian ancestry. Two of the girls have a history of chronic inflammatory bowel disease, and the Asian patient had a previous diagnosis of Kawasaki disease at the age of two.

All cases presented with fever, fatigue, and nonspecific symptoms, along with elevated inflammatory markers (Table 1). Prior to initiating treatment with prednisolone and weight-adapted maintenance therapy with tocilizumab, two patients underwent FDG-PET/CT in addition to conventional imaging techniques, including ultrasound and MRI (Table 2). Distinct patterns of extracranial large vessel vasculitis were observed across the different imaging modalities. A follow-up FDG-PET/CT was performed after a minimum of 12 months, showing reduced or, in some cases, no inflammation in the previously affected vessels, as illustrated in Figs. 1, 2, 3, 4 and 5.

Glucocorticoids should be used cautiously, followed by maintenance therapy with tocilizumab. While efficacy and safety of tocilizumab in treating Takayasu arteritis (TA) have been established, there is still insufficient data regarding the optimal timing for discontinuation of the medication [3]. FDG-PET/CT, an investigator-independent technique that clearly visualizes inflammation, supports appropriate therapy adjustments, in contrast to examiner-dependent modalities such as ultrasound. While MRI revealed possible chronic but no inflammatory alterations especially at the last follow-up, FDG-PET/CT was able to identify active large vessel vasculitis. Our results are

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Table 1 Patient characteristics

Parameter	Patients			
	Patient 1	**Patient 2**	**Patient 3**	**Patient 4**
Gender	female	female	female	female
Age at diagnosis of TA	17 y	13 y	13 y	13 y
Symptoms at initial manifestation	Fever	Fever	Fever	Fever
	Fatigue	Fatigue	Fatigue	Fatigue
	Headache, Art. Hypertension	Malaise, Erythema nodosum	Night sweats, Weight loss	Palpatory reduced pulse left upper extremity, Blood pressure difference lower to upper extremity
Laboratory finding				
CrP (N: < 5 mg/l)	27	63	89	149
ESR (mm/h)	83	88	76	> 100
SAA (N: < 40 mg/l)	105	> 120	> 120	> 120
ANA	negative	negative	negative	positive
Initial therapy	Prednisolon	Prednisolon	Prednisolon	Prednisolon
Maintenance therapy	Tocilizumab	Tocilizumab	Tocilizumab	Tocilizumab
Duration of tocilizumab	22 months	12 months	61 months	More than 96 months, still ongoing
Previous illnesses	Crohn`s disease	Uveitis intermedia		Kawasaki Syndrome
	Akne inversa	Juvenile idiopathic arthritis		
	Psoriasis inversa	Celiac disease		
Previous medication	Adalimumab	Adalimumab		
	Anakinra	Methotrexat		
	Infliximab	Prednisolon		
	Methotrexat			
	Ustekinumab			
	Vedolizumab			
Last medication before TA onset	Vedolizumab	Adalimumab		

Table 2 Performed imaging modalities

Parameter	Patients			
	Patient 1	**Patient 2**	**Patient 3**	**Patient 4**
FDG-PET/CT				
at the beginning of maintenance therapy	X	X		
under maintenance therapy	X	X	X	X
after stopping maintenance therapy		X		
Ultrasound				
at the beginning of maintenance therapy	X	X	X	X
under maintenance therapy	X	X	X	X
after stopping maintenance therapy				
MRI				
at the beginning of maintenance therapy	X		X	X
under maintenance therapy	X			X
after stopping maintenance therapy	X		X	X

consistent with studies on large vessel vasculitis in adults, suggesting FDG-PET/CT to be a valuable tool for therapy monitoring, in addition to other clinical and laboratory parameters [4]. However, FDG-PET/CT is not yet part of the standard of care diagnostics for TA in children, despite improvements in examination protocols and reduced radiation exposure [5]. This case series demonstrates that FDG-PET/CT provides valuable additional information in selected cases, offering deeper insight into inflammatory activity when MRI results are inconclusive. Nevertheless, further studies on the use of FDG-PET/CT in pediatric populations are needed.

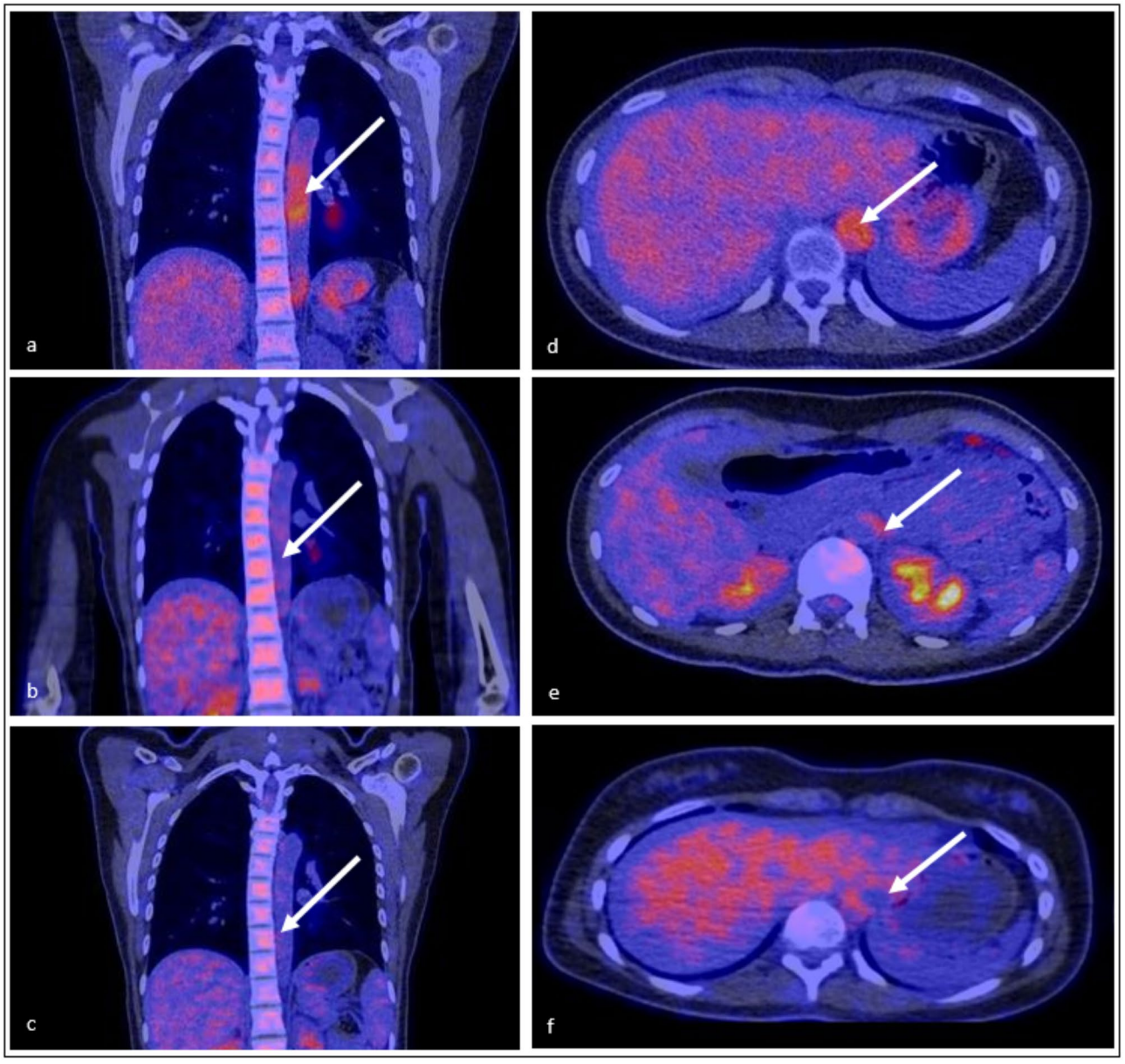

Fig. 1 TA with predominantly aortitis. Patient #1 presenting with FDG enrichment in the vessel wall of the descending thoracic aorta (**a** and **d**). 6 months after the start of therapy, unchanged evidence of moderately increased FDG accumulation in the proximal ascending aorta and in the thoracic descending aorta, but slightly decreasing accumulation in the proximal abdominal descending aorta (**b** and **e**). No further nuclide accumulation in vessel walls detectable 18 months after start of therapy, overall no florid inflammatory activity (**c** and **f**)

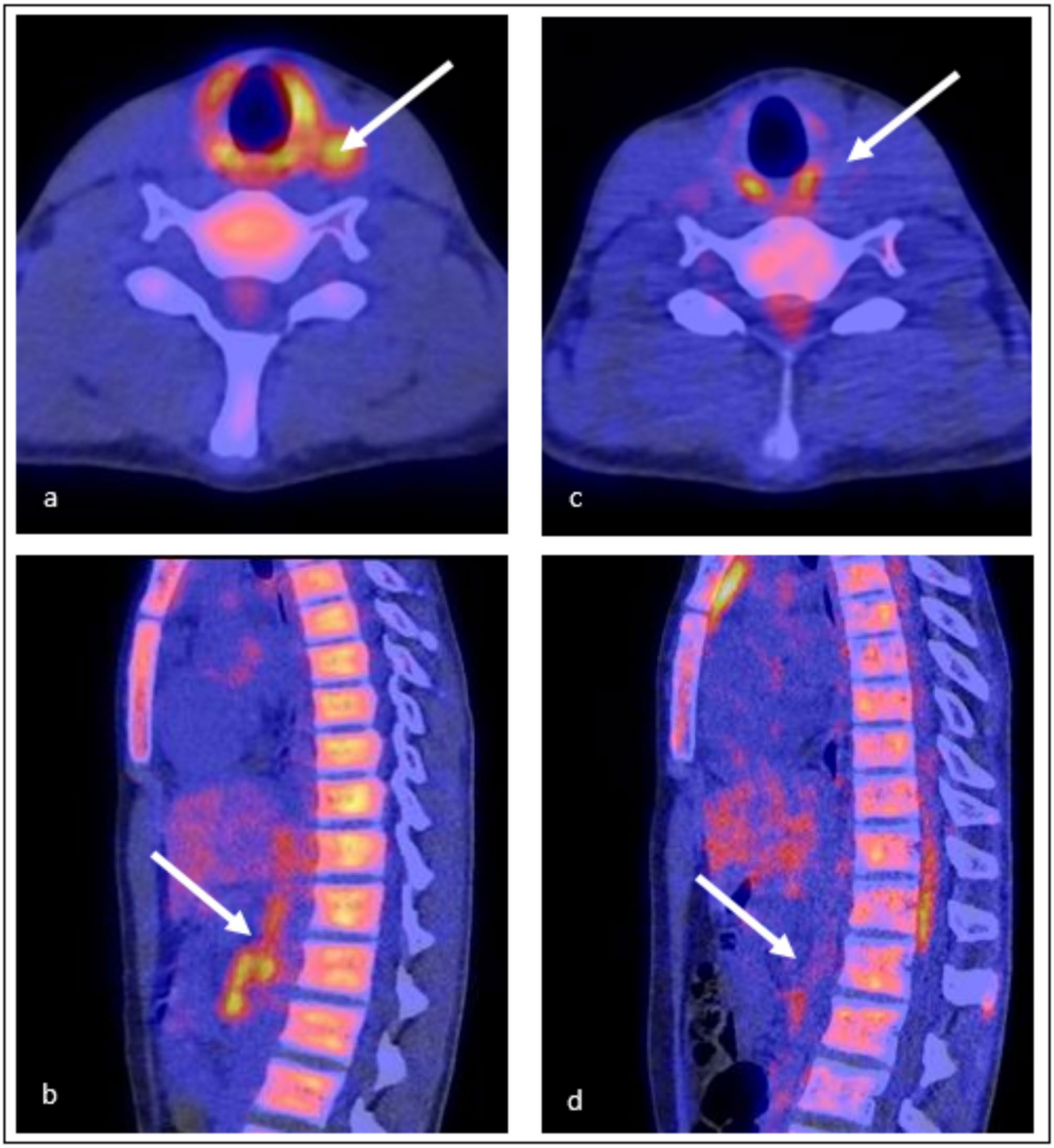

Fig. 2 TA of carotid artery. Patient #2 has shown ahort-distance circular nuclide accumulation in projection onto the left common carotid artery (**a**). Circularly accentuated FDG storage in the course of the abdominal aorta as well as long-distance storage in the course of the proximal superior mesenteric artery (**b**). FDG-PET/CT 12 months later has shown any significant nuclide enrichment (**c**, **d**)

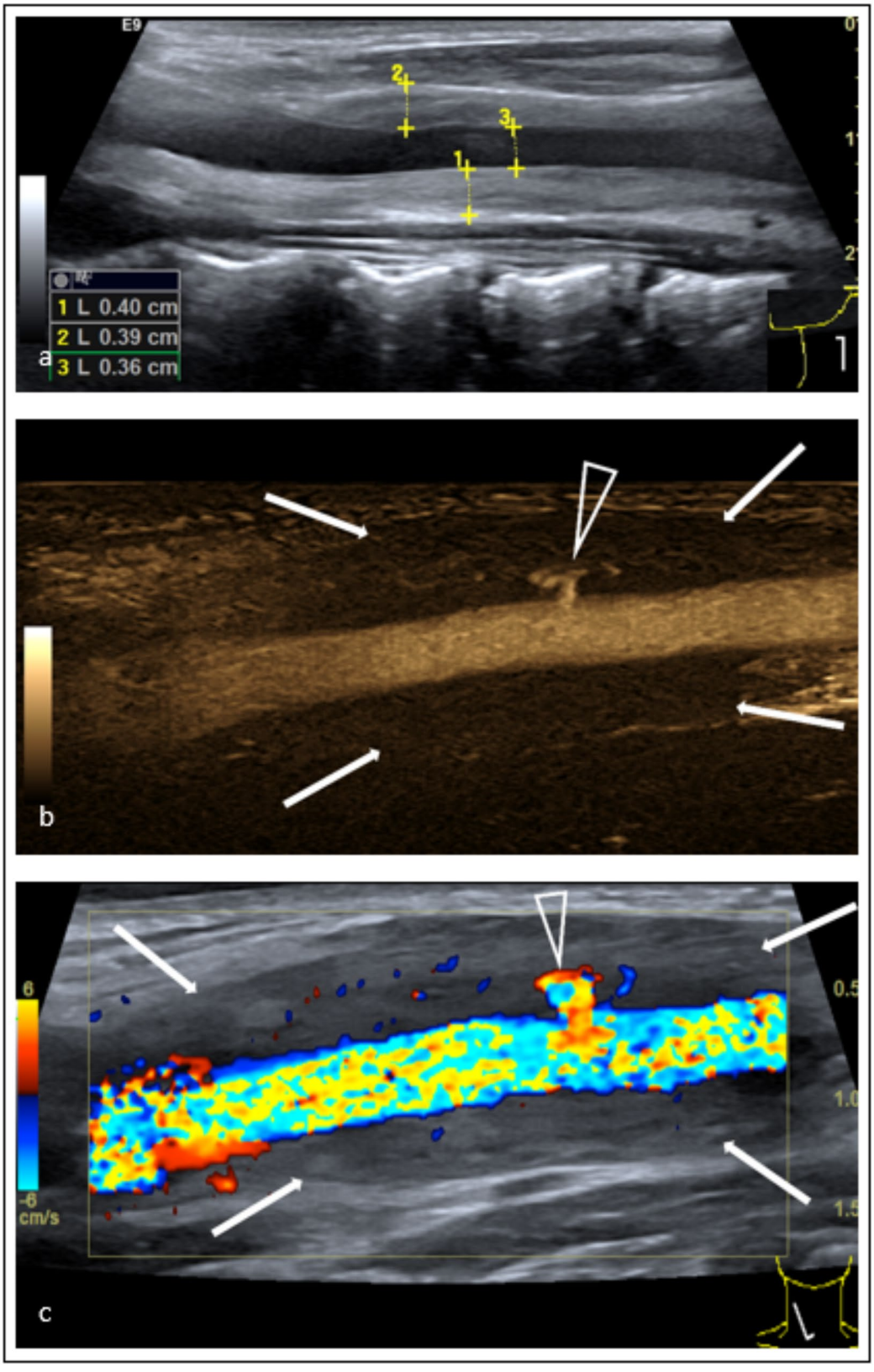

Fig. 3 Ultrasonographic evidence of vessel wall inflammation. In patient #3 initially underwent an ultrasound of several vessels, as shown in this figure of the left arteria carotis ext. Clearly echogenic thickened vessel wall, up to approx. max. 4 mm in diameter (white arrows in Fig. 5b and c) with a vessel lumen of approx. 3.6 mm (**a**). In B-flow and Doppler mode, a small arterial vessel (white arrowhead) can be found in the thickened wall around the right carotid artery (**b** and **c**)

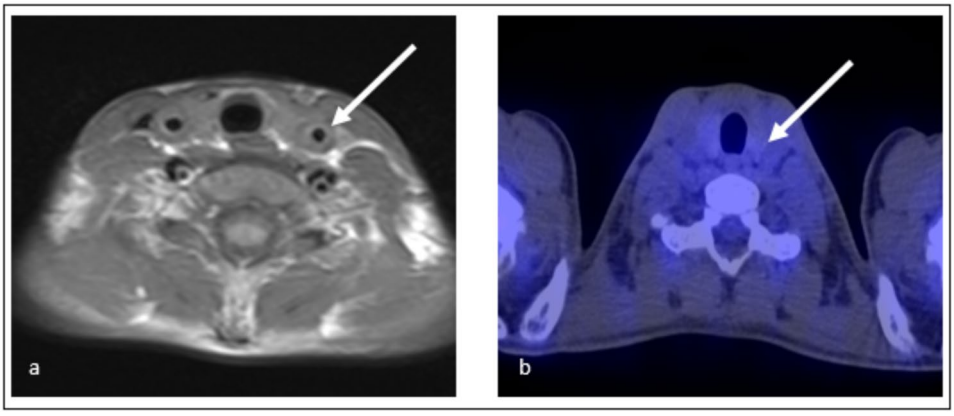

Fig. 4 TA of common and internal carotid artery. Same patient #3 as shown in Fig. 3. Here we demonstrate MRI of the neck. Axial T1-weighted unenhanced image demonstrates pronounced cuff around the common carotid artery, extending into the internal carotid artery on the left side (**a**). 21 months later, FDG-PET/CT shows no evidence of florid vasculitis (**b**)

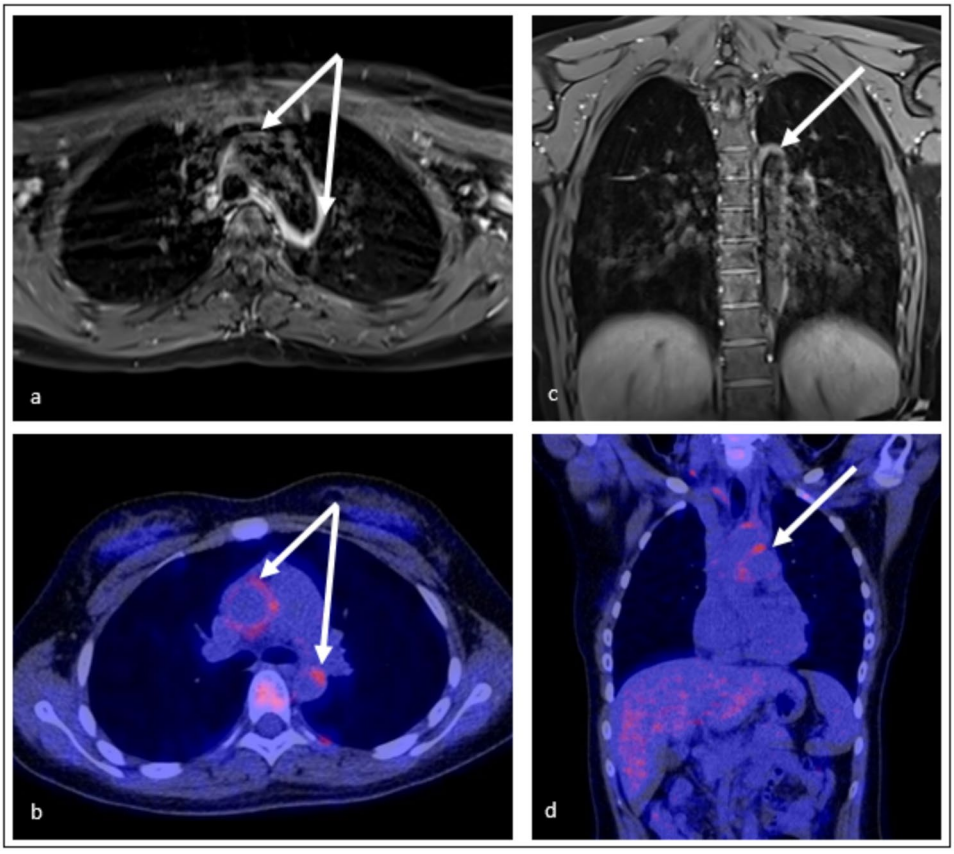

Fig. 5 Diffuse and prolonged TA. In patient #4 a MRI of thoracic vessels was performed. Axial and coronal T1-weighted postcontrast images with fat saturation show partly emphasised, partly thickened vessel walls of the ascending aorta, the aortic arch, the descending aorta and the supraaortic branches, in most areas mentioned before with pathological contrast agent uptake (**a** and **c**). Corresponding to this, FDG-PET/CT 3 years later showed a still indicated vessel-associated tracer storage in the wall of the ascending aorta and the aortic arch in the sense of a slight residual floridity (**b** and **d**)

Abbreviations

ANA	Anti-nuclear-antibodies
CrP	C-reactive protein
ESR	Estimated sedimentation rate
FDG-PET/CT	Fluorodeoxyglucose-positron emission tomography/computer tomography
MRI	Magnetic resonance imaging
SAA	Serum amyloid A
TA	Takayasu arteriitis

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Author contributions
TK analyzed and summarized all patient data. TK, AH and JR had substantial impact to the conception and design of the work; AA, JJ and OR performed examinations and interpretated data. All Authors have drafted the work or substantively revised it. All authors read and approved the final manuscript.

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