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CASE REPORT
Other

Case report of a peculiar aneurysm of the ascending aorta: when there is much more beyond an incidental finding

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Background Aneurysms of the thoracic aorta are common in male patients around the VI–VII decade of life and most have a degenerative aetiology; otherwise, the occurrence of this disease at a younger age should prompt the search of rarer causes. We report a singular case of ascending aortic aneurysm (AAA) in a young man.

Case summary A large AAA accompanied by multivessel dilatation and renal failure of unknown onset was incidentally found in a 23-year-old male during the diagnostic work-up after a car accident. A systemic disease was therefore suspected, and a full clinical investigation revealed the uncommon diagnosis of sarcoidosis accompanied by large vessel vasculitis.

Discussion Only a few reports in the literature describe the concurrence of sarcoidosis and large vessel vasculitis (Takayasu arteritis), which may share non-specific immunoinflammatory abnormalities. This case underlines the importance of a multisystem diagnostic approach even in front of an incidental finding that is inconsistent with patient's age.

Keywords Aortic diseases • Vasculitis • Multivessel diseases • Case report

Learning points

- The concurrence of sarcoidosis and large vessel vasculitis (Takayasu arteritis) has been rarely described: aortitis may be one of the possible manifestations of sarcoidosis in the context of multisystem involvement.
- In the cases described in the literature, the diagnosis of sarcoidosis usually precedes that of large vessel vasculitis, with a delay of several years. The time-point of clinical presentation in our patient is singular: both sarcoidosis and large vessel vasculitis appeared in a subacute/chronic phase.
- Two possible hypotheses may explain the association of sarcoidosis and large vessel vasculitis: they could be two related diseases, sharing common non-specific immunoinflammatory abnormalities (in particular, the same pattern of chronic granulomatous inflammation) or granulomatous vasculitis may be a complication of sarcoidosis.

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Introduction

Degenerative thoracic aortic aneurysms are most common in the VI–VII life decade, mainly in males with other risk factors for atherosclerosis.[1] Conversely, an aortic disease in a young patient should prompt further evaluation to investigate rarer aetiologies. Aortitis is a rare manifestation of sarcoidosis which, as a multisystem disease, can affect the heart (with various manifestations including heart block and arrhythmias, heart failure, valvular dysfunction, simulated infarction, and pericardial disease[2]) kidneys, and vessels, beyond the most common pulmonary localization. A few reports[3–8] describe the concurrence of sarcoidosis and large vessel vasculitis, namely Takayasu arteritis (TA). The latter is an uncommon chronic vasculitis of unknown aetiology, which primarily affects the aorta and its main branches; it is more common in female and Asian descent. The onset of symptoms in TA tends to be subacute, which often leads to a delay in the diagnosis from months to years, during which time the vascular disease may progress, leading to narrowing, occlusion, or dilation of the arteries. The clinical presentation accounts symptoms of reduced blood flow, arterial pain, and tenderness, or non-specific constitutional symptoms. In most cases, the diagnosis is based upon suggestive clinical features and specific imaging findings of the aorta and/or its branches. Additionally, TA or other forms of large-vessel vasculitis must be considered either in patients incidentally found to have findings suspicious for vasculitis on imaging obtained for other clinical indications or when vasculitis is found on histologic examination of surgically removed segments of arteries.[9] Even if rarely described in conjunction, sarcoidosis and TA may be related as they are characterized by similar non-specific immunoinflammatory abnormalities, expressed in the form of chronic granulomatous inflammation.

Case presentation

A 23-year-old male presented to the Emergency Department of our hospital after a car accident with a low-impact collision. He was of African descent (born in Cameroon) and his past medical history was unremarkable; he did not use any medication, was in good clinical status and conducted an active life, playing in a football team.

At the first medical evaluation, the patient was alert, oriented, and collaborative (Glasgow Coma Scale—GCS 15), eupnoeic, and haemodynamically stable (arterial blood pressure 110/60 mmHg). The physical examination revealed tenderness at palpation of the thorax, cervical processes, left pulmonary base, where the breathing sounds were reduced, and left hypochondrium; he also complained pain on the left knee. The neurologic examination was unremarkable.

The first electrocardiogram showed sinus rhythm and normal QRS, S-T segment, and T waves (*Figure1*).

The initial lab tests revealed moderate arterial hypoxaemia (PaO_2 72 mmHg) and mild anaemia (haemoglobin 12 g/dL); troponin values were in the normal range (TnI < 0.02 μg/L).

The patient underwent an immediate thoraco-abdominal computed tomography (CT), which unexpectedly revealed a large ascending aortic aneurysm (AAA) with a maximum diameter of 61 mm in the suprabulbar portion, accompanied by aneurysmatic deformations of the coeliac trunk (maximum diameter 18 mm) and superior mesenteric artery (19 mm) (*Figure 2A and* B). Signs of an acute aortic syndrome were excluded.[10] As collateral findings, numerous confluent lymph nodes were observed in the prevascular space, along the internal and external left mammary chains, in paratracheal, infra- and subcarinal regions and at the pulmonary hilum (*Figure 2C*). A small traumatic pulmonary contusion was also noticed at the left infero-posterior lobe, with limited associated pleural effusion.

Timeline

Timeline	Description
Day 0: presentation	Admission to the Emergency Department after a car accident with a low-impact collision
Day 0-1 h after admission	Computed tomographic evaluation: incidental diagnosis of multiple aneurysms involving ascending aorta, coeliac trunk, and mesenteric artery; multiple lymphadenopathies
	Blood tests: previously undiagnosed renal failure
Day 1-18 h after admission	Admission to a medical ward for further evaluation
Days 2–19	Clinical and haemodynamic stability
	Diagnostic work-up, including: • Echocardiography • Serologic and autoimmunity markers evaluation • Magnetic resonance angiography • Coronary computed tomography angiography • Positron emission tomography • Fibrobronchoscopy with fine needle biopsy
Day 20	Cardiac surgery: replacement of the ascending aorta with a vascular prosthesis and remodelling of the sino-tubular junction
Day 21	Postoperative monitoring in intensive care unit
Day 25	Subxiphoid drainage of pericardial effusion
Day 34	Hospital discharge to physical rehabilitation centre

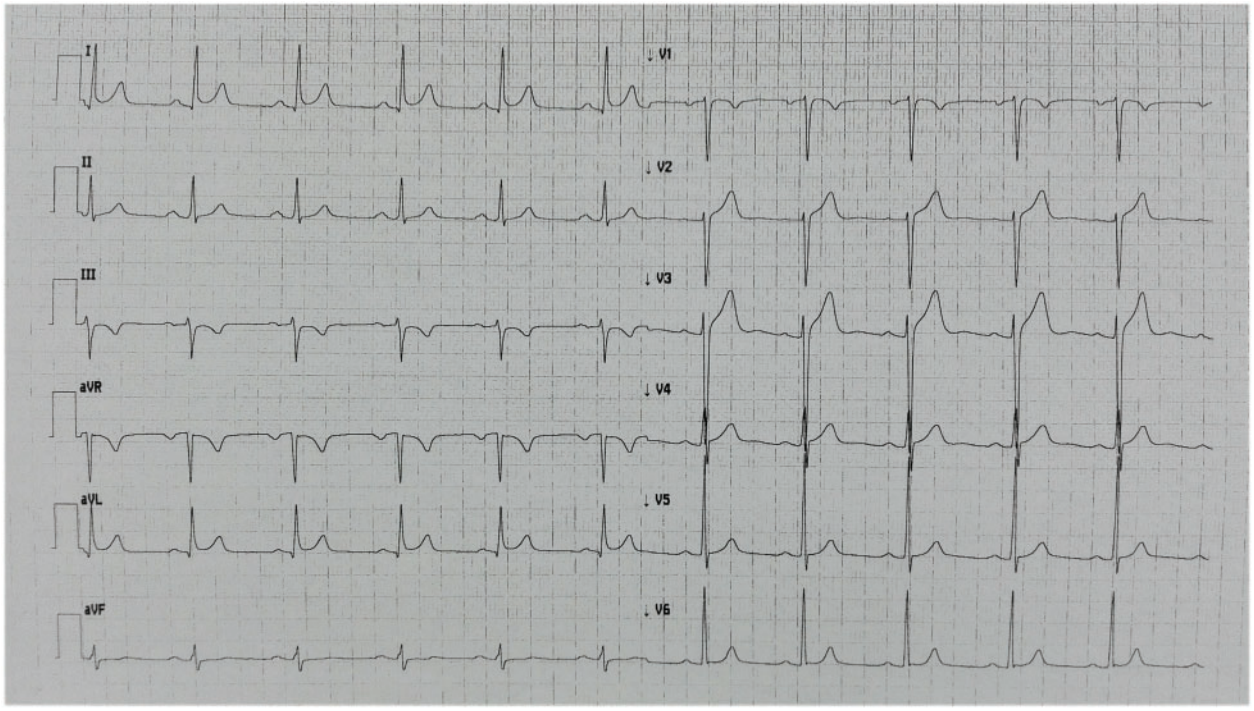

Figure 1 First 12-lead electrocardiogram showing normal sinus rhythm and normal QRS, S-T segment, and T waves.

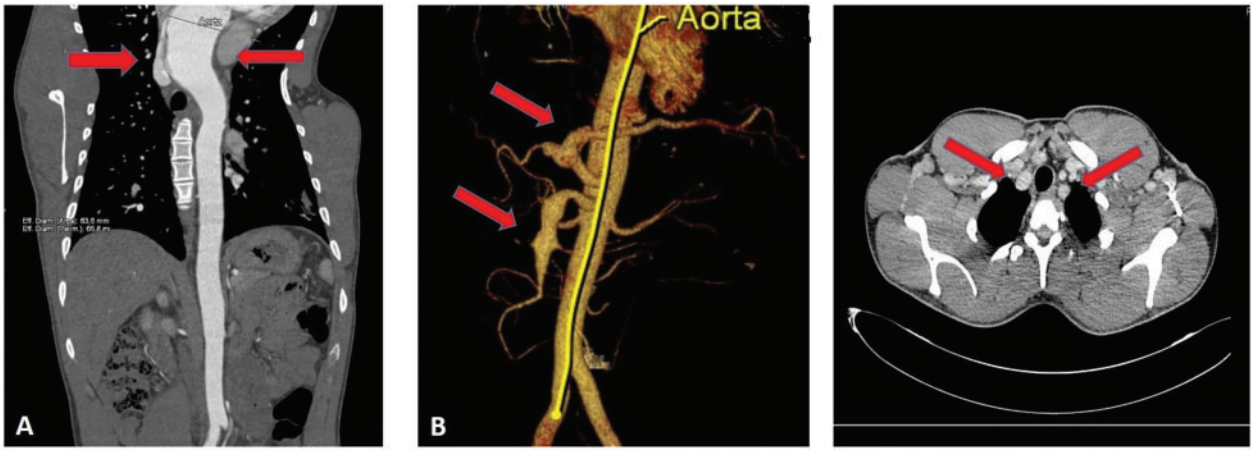

Figure 2 Thoraco-abdominal computed tomography. (*A*) Ascending aortic aneurysm with a 61-mm maximum diameter in the suprabulbar portion (red arrows); (*B*) aneurysmatic dilatation of the coeliac trunk (18 mm, superior red arrow) and of superior mesenteric artery (19 mm, inferior red arrow); (*C*) multiple lymph nodes in paratracheal position can be noted (red arrows).

Complete blood tests revealed previously unknown renal failure (serum creatinine 1.89 mg/dL), and elevation of creatine phosphokinase (674 U/L) and myoglobin (200 ng/mL) levels, with persistently normal TnI values (0.01 μg/L). Complete blood count was normal, with stable Hb values at repeated evaluations, and inflammation markers were unremarkable (procalcitonin 0.06 ng/mL; C-reactive protein < 9 mg/L).

Transthoracic echocardiography confirmed the presence of aortic root dilatation (40 mm at the sinuses' of Valsalva level), AAA (63 mm in the proximal ascending aorta), and mild-to-moderate aortic regurgitation through a tricuspid aortic valve (*Figure 3*). Dimensions and function of the right and left ventricles were normal.

Given the stability of the clinical presentation, the patient was admitted to a medical ward for further evaluation.

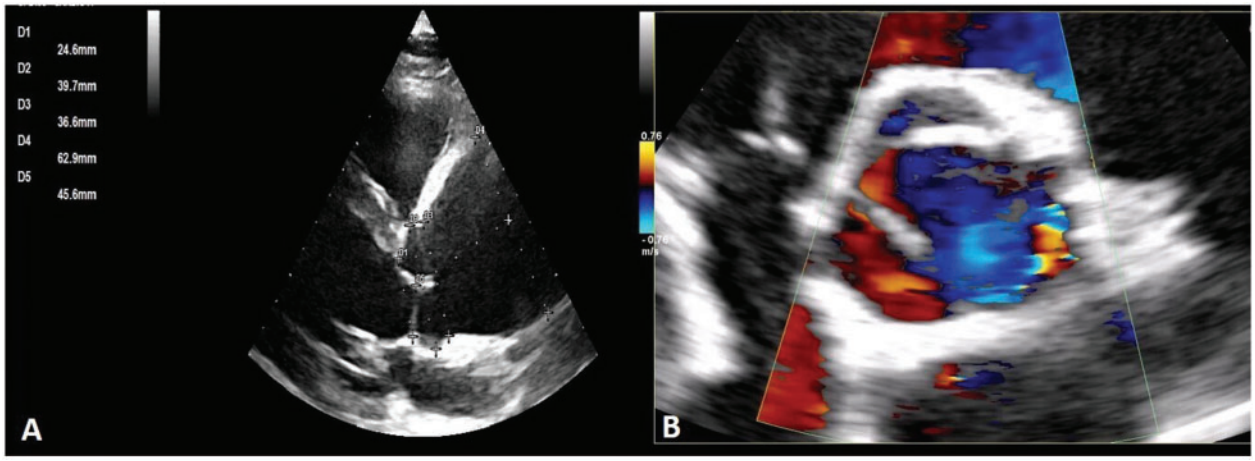

Figure 3 Transthoracic echocardiography. (*A*) Parasternal long-axis view showing aortic root dilatation (39.7 mm) and proximal ascending aortic aneurysm (36 mm). (*B*) Tricuspid aortic valve in parasternal short-axis view with colour Doppler.

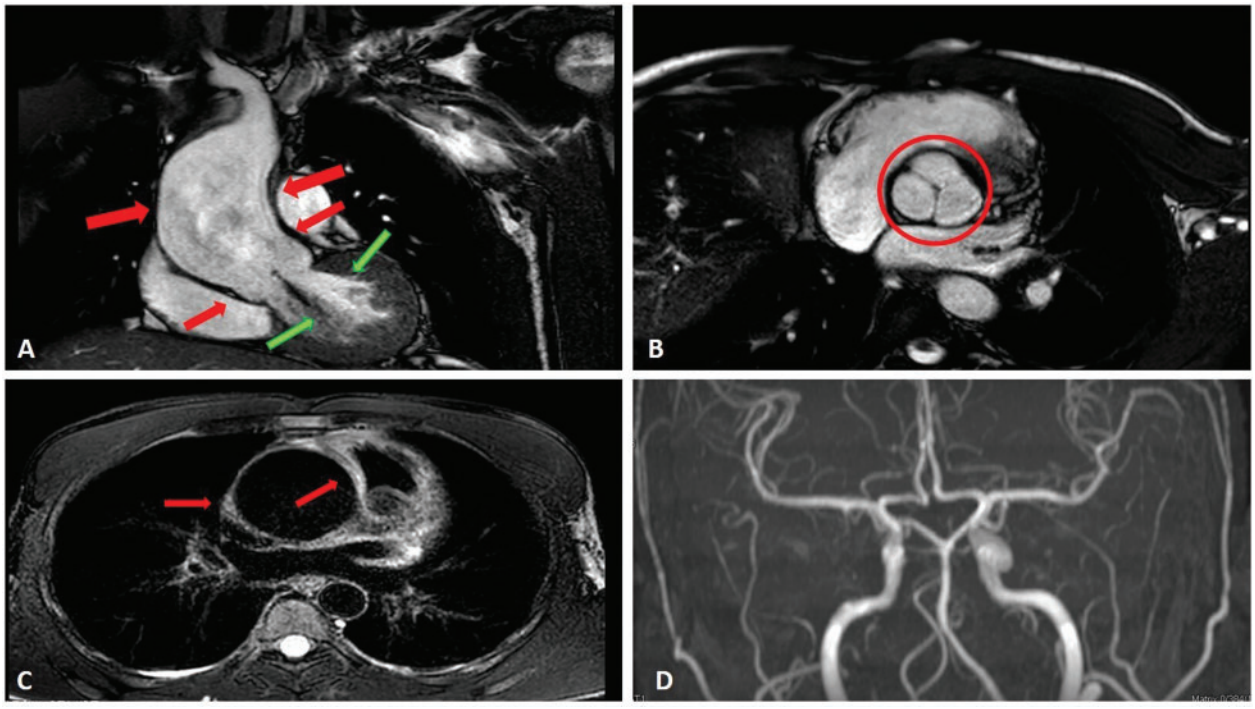

Figure 4 Magnetic resonance imaging. (*A*) Bulbar ectasia (44 mm, inferior red arrows), suprabulbar ascending aortic aneurysm (64 mm, superior red arrows), and mild aortic regurgitation (regurgitant fraction 29%, green arrows); (*B*) normal tricuspid aortic valve (red circle); (*C*) thickening of the ascending aortic wall (~5 mm, red arrows), in the absence of oedema or signs of dissection; (*D*) normal intracranial arterial vessels.

In the suspicion of aortitis, a complete evaluation of the aorta using magnetic resonance angiography (MRA) was performed, including the cerebral district. At the thoracic level, MRA revealed a homogeneous circumferential thickening of the ascending aortic wall (~5 mm), without oedema nor signs of dissection; bulbar ectasia, suprabulbar AAA, and mild aortic regurgitation in a tricuspid aortic valve were confirmed. The intracranial vascular system was normal (*Figure 4*).

A coronary CT angiography showed normal epicardial vessels (*Figure 5*).

The patient also underwent a functional evaluation through positron emission tomography with 18-fluorodeoxyglucose, which

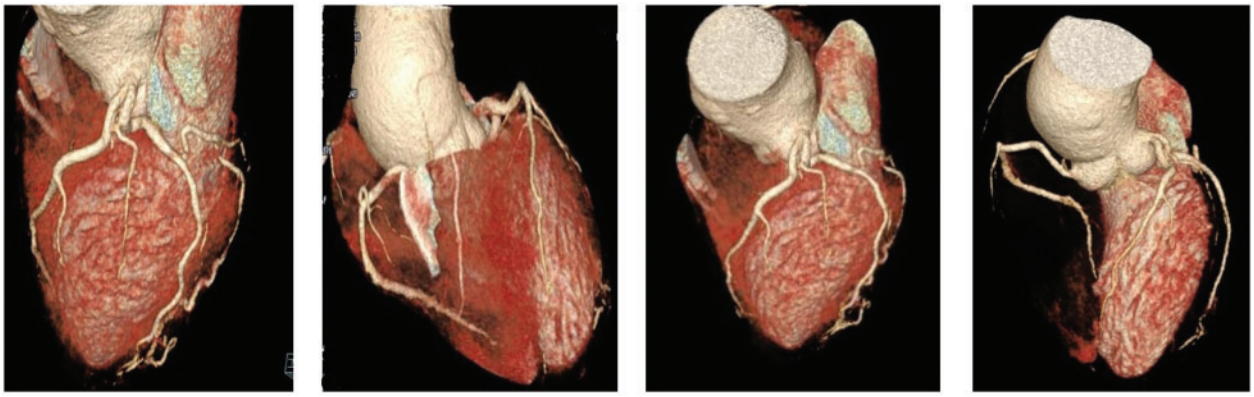

Figure 5 Coronary computed tomography angiography showing normal epicardial coronary arteries.

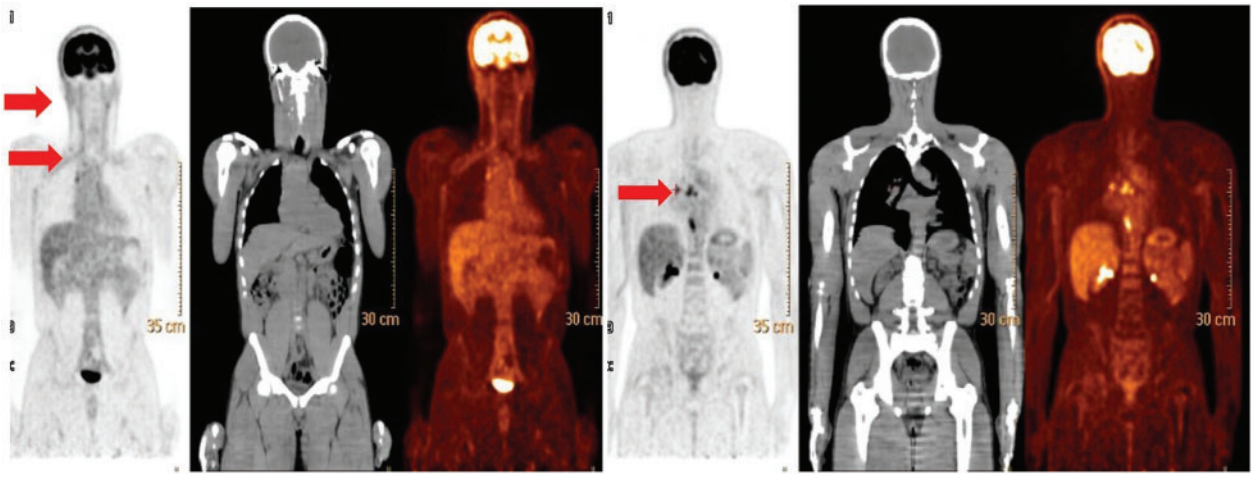

Figure 6 Positron emission tomography with 18-fluorodeoxyglucose showing (*A*) moderate hypermetabolism in the ascending aorta (lower red arrow) and mild hypermetabolism in the carotid (upper red arrow) as well as subclavian, iliac, and femoral arteries, and (*B*) intense hypermetabolism corresponding to lymph nodes, particularly at right pulmonary hilum.

exhibited moderate hypermetabolism in the ascending aortic wall and mild hypermetabolism in carotid, subclavian, iliac, and femoral arteries, compatible with an inflammatory process involving vessels, though no longer intensively active. On the other hand, intensely hypermetabolic areas were highlighted corresponding to lymph nodes, particularly at the right pulmonary hilum (*Figure 6*).

Main autoimmunity markers (Anti-Neutrophil Cytoplasmic, Antinuclear and Extractable Nuclear Antigen antibodies) were negative and also extensive serological research of possible infectious agents (including Hepatitis-B and -C viruses, Human Immunodeficiency Virus 1–2, *Treponema pallidum*, *Chlamydia trachomatis*, *Borrelia burgdorferi*, *Rickettsia conorii*, and *Rickettsia mooseri*) did not show any positivity, with the exception for Quantiferon test (deposing for a previous exposition to Koch Bacillus, without signs of active disease). Angiotensin-converting enzyme (ACE) levels were in the normal range (ACE 55.6 U/L, with reference range 42–138 U/L).

The investigations on renal failure (creatinine levels stabilized in a range of 1.8–1.9 mg/dL) suggested a pattern of tubulointerstitial injury, with high β2-microglobulin levels (3.1 mg/L); morphologically, the kidneys appeared structurally subverted with poor corticomedullary differentiation and diffuse cortical hyperechogenicity at an abdominal ultrasound scan, suggesting a non-recent onset of renal failure. A mild polyclonal hypergammaglobulinemia (22.8%) was also highlighted, with increased levels of IgG (17–1 g/L), without monoclonal components at serum immunofixation.

Eventually, the patient underwent a fibrobronchoscopy with bronchoalveolar lavage and transbronchial needle aspiration of the

enlarged lymph nodes: the cytologic examination revealed lymphocytes and granulomatous epithelioid cells, organized in non-caseous granulomas. The diagnosis of sarcoidosis with nodal, renal, and vascular involvement was therefore established.

The patient electively underwent valve-sparing aortic root replacement with a vascular prosthesis (straight vascular prosthesis no. 26) and aortic valvuloplasty with remodelling of the sino-tubular junction. Systemic steroid therapy was started just after surgery. The timing of this therapy was a challenging decision: on one side, the unfavourable disease characteristics would recommend a prompt start of steroid therapy,[11] even before surgery; on the other hand, there was no evidence of current active inflammation. Thus, the risk of surgical wound complications under steroid therapy was considered more dangerous and the treatment initiation postponed after surgery (the patient was given methylprednisolone 40 mg daily starting from the 7th postoperative day). The postoperative clinical course was complicated by pericardial effusion, which required subxiphoid drainage. Eventually, the patient was discharged in good clinical status and with a tapering program of steroid therapy on the 13th postoperative day.

Discussion

Considering the young age of the patient and the multivessel involvement, systemic vasculitis and genetic diseases affecting the aorta were the main differential diagnoses to be contemplated.

The patient had no history of familial sudden death nor collagenopathies, thus excluding the need for genetic testing. Marfan scoring according to revised Ghent criteria[9] was 6, thus inconclusive for the diagnosis of systemic involvement. No robust clinical element argued in favour of non-syndromic forms of thoracic aortic aneurysm and Loeys–Dietz syndrome, which were therefore excluded. The whole imaging tests performed confirmed the presence of a tricuspid aortic valve and normal aortic diameter beyond the isthmus, thus excluding also aneurysms associated with bicuspid aortic valve and coarctation of the aorta.

Therefore, this complex clinical picture ultimately suggested a systemic large-vessel vasculitis, without features of current activity, coexisting with sarcoidosis that involved hilar lymph nodes and kidneys with tubulointerstitial damage.

The coexistence of sarcoidosis and TA has been occasionally reported.[2–7] Even if a direct interrelation has not yet been demonstrated, TA-like granulomatous vasculitis may be, in fact, a complication of sarcoidosis. Indeed, both diseases are characterized by nonspecific immunoinflammatory abnormalities, sharing the same pattern of chronic granulomatous inflammation. In the previously described cases, the diagnosis of sarcoidosis usually preceded that of large vessel vasculitis, with a time lag of several years. Consistently, the onset of sarcoidosis in our patient dated presumably a long time before, as suggested by the poor renal corticomedullary differentiation. The revision of the literature concludes that two hypotheses may explain this association of pathologies: sarcoidosis and TA could be two related diseases, or the granulomatous vasculitis may be a complication of sarcoidosis.

Patient perspective

This case underlines the importance of a complete and multi-organ clinical evaluation, even in front of an occasional clinical finding when an AAA is encountered in a young patient such ours. The identification of the rare aetiology of this AAA implied controversial therapeutic decisions (like the timing of steroid therapy initiation).

Lead author biography

Nicole Ceschia is a resident in Cardiology at the Department of Experimental and Clinical Medicine, University of Florence.

Supplementary material

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

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: Collected from the patient at the time of in-hospital stay, subsequently lost to follow-up. The authors confirm that witnessed verbal consent for submission and publication of this case report including images and associated text has been obtained from the patients detailed in this case report. This has been discussed with the editors.

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