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Case Report

Unusual systemic conditions in a patient with giant cell arteritis

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ABSTRACT

Giant cell arteritis is a systemic vasculitis that causes inflammation in medium- and large-sized blood vessels. The condition can lead to irreversible blindness if not recognised and treated promptly with high-dose steroids. Clinical manifestations typically include headache, jaw pain, fever, and fatigue. However, unusual manifestations of the disease have been reported, including pulmonary nodules, uveitis, pericarditis, and stroke. We report a case of biopsy-confirmed giant cell arteritis in a patient found to have renal cell carcinoma, exhibiting these unusual manifestations simultaneously. This case report demonstrates the atypical presentation that giant cell arteritis may have and the importance of having a high clinical suspicion for the condition.

KEYWORDS: Giant cell arteritis; renal cell carcinoma; pulmonary nodules; uveitis; pericarditis

Introduction

Giant cell arteritis (GCA) is a systemic vasculitis that causes inflammation in medium- and large-sized blood vessels [1]. Common clinical manifestations of GCA include headaches, fatigue, fevers, and weight loss associated with elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) [2]. While the diagnosis of GCA can be made based on the above constellation of symptoms, many disease associations and possible unusual manifestations of GCA have been reported including renal cell carcinoma (RCC), uveitis, pericarditis, and stroke [1–4]. We report a unique case of biopsy-confirmed GCA in a patient exhibiting these systemic findings simultaneously.

Case presentation

A 69-year-old Caucasian male with a history of RCC status after left nephrectomy over 25 years ago, began experiencing cough, weight loss, night sweats, and fatigue. His primary care physician prescribed azithromycin, but the symptoms persisted. The patient then presented to an emergency room (ER) the following week where he was treated with oral steroids and antibiotics and was referred to a pulmonologist. The pulmonologist prescribed a 4-week steroid taper, intranasal steroids, and antibiotics that resulted in minimal improvement of his symptoms.

Four weeks later, the patient experienced fever of 101.4°F and went back to the ER where a computed tomography (CT) chest was performed that revealed bilateral pulmonary nodules, the largest of which was a lingular lesion measuring 6 mm × 15 mm. A CT of the abdomen and pelvis showed a new and indeterminate lesion measuring 2.6 cm at the lateral upper pole of the right kidney. At this time, the patient noted intermittent diplopia and described an episode of complete bilateral vision loss lasting under 1 min. An autoimmune workup was ordered which was negative for antinuclear antibody, cyclic citrullinated peptide, rheumatoid factor, and C3 with normal angiotensin converting enzyme, negative antismooth muscle, antismith, anti-SSA, and SSB. Furthermore, anti-neutrophil cytoplasmic antibodies (ANCA) testing was negative on laboratory evaluation and myeloperoxidase (MPO)-ANCA and proteinase 3 were within normal limits. Abnormal findings included elevated ESR at 97 mm/h (normal: 0.0–10.0) and an elevated CRP at 18.31 mg/l (normal: <0.50). Ophthalmology was consulted for significant conjunctival injection, which led to a diagnosis of bilateral uveitis and optic neuritis. During the admission, he was treated with piperacillin-tazobactam and azithromycin and his fever resolved while he was in-patient. He was also started on prednisolone eye drops and 1 g solumedrol daily and was discharged.

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Three days later, the patient noted further worsening of vision in the left eye and presented to our ER. The patient denied history of mucocutaneous, genital ulceration, and skin nodules. Visual acuity was 20/80 in the right eye and 20/250 in the left eye. Pupils were bilaterally sluggishly reactive to light with a left afferent pupillary defect. Visual fields were normal by confrontation in the right eye and constricted in the left eye. Slit lamp examination was notable for trace pigmented cell bilaterally in the anterior chamber. Fundus examination revealed swelling of the optic nerve in the inferior aspect of the right eye and chalky white optic nerve swelling in the left eye with inferotemporal splinter haemorrhage (Figure 1). Magnetic resonance imaging (MRI) of the brain showed a subacute left pontine infarct without any dural enhancement and moderate tortuosity of the vertebral basilar system with evidence of right vertebral artery wall thickening, suggestive of atherosclerosis.

Lumbar puncture revealed opening pressure of 25 cm H_2O, glucose of 137 mg/dl, protein of 34 mg/dl but was negative for white blood cells, gram stain, and culture. Bronchoscopic biopsy of the pulmonary lesions was attempted but could not be completed due to their location. Bronchoalveolar lavage revealed a possible infectious pattern, including 59% neutrophils, 21% lymphocytes, and 20% monocytes. Cytology was negative for malignant cells. Gram stain revealed Gram-positive cocci, Gram-positive bacilli, and few Gram-negative bacilli.

Lastly, a transthoracic echocardiogram revealed a small circumferential pericardial effusion. GCA was suspected due to the patient's history of left eye amaurosis, fevers, weight loss, and elevated ESR and CRP. The patient was treated with intravenous methylprednisone 1 g daily for 2 days and then was transitioned to oral prednisone 80 mg daily. Left temporal artery biopsy revealed an inflammatory infiltrate

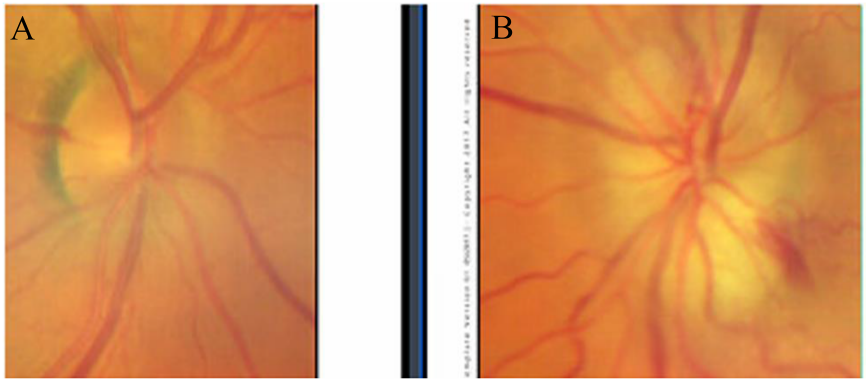

Figure 1. Fundoscopic images of (a) the right eye with normal disc, drusen, and nevus and (b) the left eye with pallid chalky white disc swelling, inferotemporal splinter haemorrhage, and drusen.

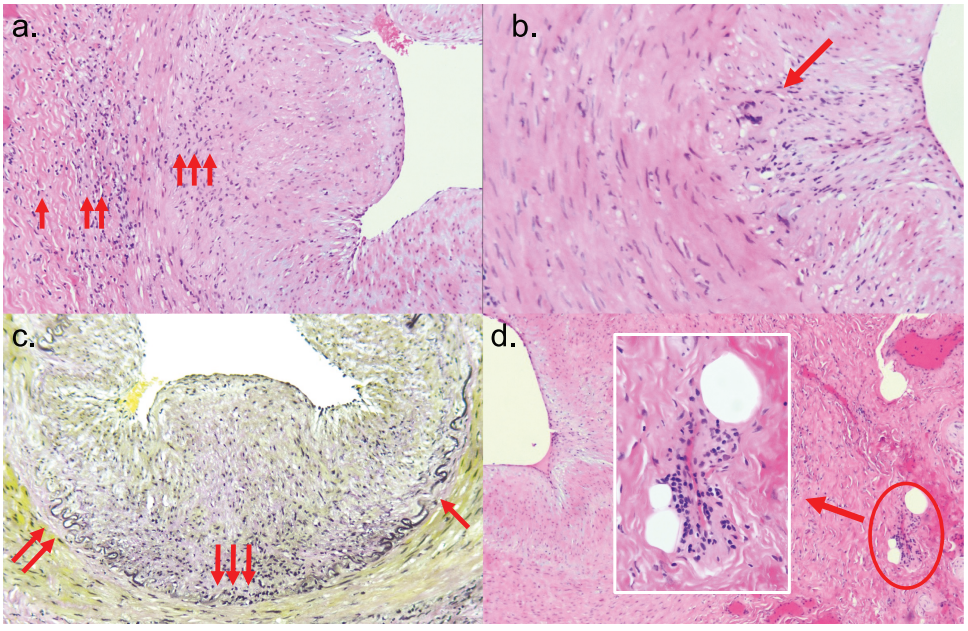

Figure 2. Image of the temporal artery biopsy with H&E stain at 100× magnification demonstrating (a) an inflammatory infiltrate within the adventitia (single arrow), media (double arrow), and intima (triple arrow), (b) scarring of the intima with the presence of a multinucleated giant cell, (b), (c) focally maintained (single arrow), focally fragmented (double arrow), and absence (triple arrow) of elastic lamina interna, and (d) inflammatory infiltrate surrounding a small vessel within the vasa vasorum, suspicious for involvement by GCA.

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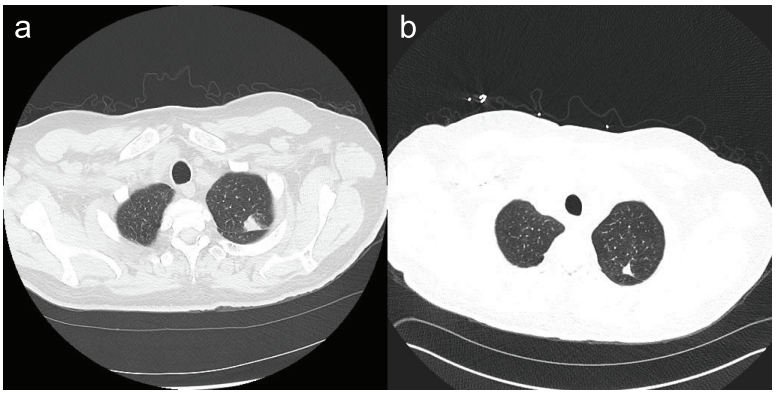

Figure 3. (a) CT of chest from outside the hospital showing the focal nodular area of consolidation within the left upper lobe posteriorly, measuring 1.6 × 1.1 cm and (b) CT of chest after both antibiotic and steroid treatment; left upper lobe posterior consolidative opacity adjacent to the fissure has decreased.

within the adventitia, media, and intima, the focal presence of multinucleated giant cells, areas of fragmented or absent elastic lamina, and inflammatory infiltrate surrounding vessels in the vasa vasorum, consistent with GCA (Figure 2). A CT of chest while on steroid therapy revealed a significant decrease in pulmonary nodule size (Figure 3) and continued resolution of fever and cough. Ultimately, an acid-fast culture grew nontuberculous mycobacterium avium complex (MAC). However, by the time the culture resulted, the patient was already treated with both short-course antibiotics and steroids and his fever and pulmonary symptoms resolved. His follow-up positron emission tomography/computed tomography (PET/CT) scan and chest x-rays have continued to show resolution of pulmonary findings, despite not being treated with long-term antibiotic therapy for MAC.

Of note, urinalysis revealed haematuria with 20–50 red blood cells and moderate blood. Given his history of left RCC and recent CT revealing a new right renal lesion, the patient underwent an MRI of abdomen which confirmed the diagnosis of RCC in the right kidney. He underwent a right partial nephrectomy. Pathology was consistent with clear cell RCC.

The patient experienced improvement in his vision and was placed on tocilizumab, 162 mg subcutaneously weekly, with a prednisone taper. At his last follow-up, his vision was 20/25 in the right eye and 20/60 in the left eye. Fundus examination revealed a normal appearing right optic nerve and pallor of the left optic nerve.

Discussion

Our patient presented with multiple concurrent systemic findings in the setting of high ESR and CRP and was diagnosed with GCA. While elevated ESR and CRP can be seen in other inflammatory states or in malignancy, the presence of visual loss and chalky white disc swelling greatly increased the likelihood of GCA in this patient. By the 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology criteria, the patient met criteria for classification of GCA via sudden vision loss, CRP greater than 10 mg/l, and positive temporal artery biopsy [5].

The underlying pathophysiological mechanisms that account for his many other concurrent systemic findings including pulmonary nodules, cerebrovascular accident, pericarditis, uveitis, and RCC remain uncertain. Uveitis as a presenting symptom of GCA is rare and has been previously reported in four prior reports [4]. Three patients were diagnosed with posterior uveitis, and one had anterior uveitis. All had associated headaches and elevated ESR along with visual loss that increased the suspicion for GCA. The mechanism relating the two conditions is unclear; however, these reports demonstrate that uveitis can potentially delay the diagnosis and appropriate treatment of GCA, leading to permanent visual deficits [4]. Posterior circulation strokes have been reported in GCA due to ischaemia involving the vertebral or posterior cerebral arteries; a large case series reported that patients with biopsy-proven GCA and traditional cardiovascular risk factors or permanent visual loss were at increased risk of suffering vertebrobasilar strokes [3]. Our patient's vertebral arteries did show evidence of atherosclerosis on MRI and may have put him at higher risk of stroke in association with GCA.

Pericarditis has been described as another rare manifestation of GCA [2]. In a cross-sectional analysis involving 25,940 patients (4329 with GCA and 21,611 age-matched controls), Tiosano *et al.* report a 1.22% incidence of pericarditis in patients with GCA compared to only 0.33% of controls [2]. A potential explanation for this may be an underlying autoinflammatory process triggering a T-helper 17 cell response, resulting in excessive production of interleukin-1 [2].

Paraneoplastic vasculitis can occur in association with systemic malignancy, including RCC [6]. The occurrence of GCA in the context of relapse of the patient's RCC suggests the possibility of GCA-related vasculitis caused by pre-existing cancer. While the exact relationship between RCC and paraneoplastic vasculitides is poorly understood, RCC is known to secrete a number of pro-inflammatory cytokines such as interleukin-6 and tumor necrosis factor-α that have been implicated in the pathogenesis of GCA [6, 7]. Notably, the patient was placed on an interleukin-6 inhibitor, tocilizumab, and has been clinically improving.

Finally, whether the pulmonary nodules and GCA are directly related remains controversial. In a 2018 case report, Pinho dos Santos *et al.* described a similar patient with a diagnosis of GCA who presented with pulmonary nodules [1]. After thorough workup including biopsy, which revealed the

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presence of a chronic inflammatory infiltrate without giant cells, it was determined that the nodules were most likely related to GCA given clinical response to steroid therapy [1]. Our patient's bronchoalveolar lavage and acid-fast culture suggest an infectious aetiology; however, the patient's pulmonary findings resolved despite not being on long-term antibiotic therapy for MAC. It remains unclear whether the pulmonary findings were solely infectious or if there was a component of inflammatory manifestation given the improvement on steroids and tocilizumab.

This is the first known report of a patient presenting with multiple simultaneous pathologies that have a rare association with GCA including pulmonary nodules, uveitis, stroke, pericarditis, and RCC. This case illustrates that GCA can manifest alongside several other diseases, emphasising the need for physicians to maintain a heightened awareness to diagnose the condition and prevent irreversible vision loss. Further research is needed to determine whether GCA is directly linked to these other pathologies, especially RCC, and to elucidate the underlying mechanisms driving their presentation.

Conflict of interest

None declared.

Funding

None declared.

Patient consent

Signed informed consent was obtained from the patient regarding the use of patient information for the purposes of writing a case report publication.

Ethical approval

Not applicable.

References

[1] Dos Santos DP, Fonseca JP, Romão V *et al.*. Pulmonary nodules in a patient with giant cell arteritis. *Pulmonology* 2019;**25**:109–13.

[2] Tiosano S, Adler Y, Azrielant S *et al.*. Pericarditis among giant cell arteritis patients: from myth to reality. *Clin Cardiol* 2018;**41**:623–7.

[3] Gonzalez-Gay MA, Vazquez-Rodriguez TR, Gomez-Acebo I *et al.*. Strokes at time of disease diagnosis in a series of 287 patients with biopsy-proven giant cell arteritis. *Medicine* 2009;**88**:227–35.

[4] Slemp SN, Martin SE, Burgett RA *et al.*. Giant cell arteritis presenting with uveitis. *Ocul Immunol Inflamm* 2013;**22**:391–3.

[5] Ponte C, Grayson PC, Robson JC. For the DCVAS Study Group *et al.* 2022 American College of Rheumatology/EULAR classification criteria for giant cell arteritis. *Ann Rheumatic Dis* 2022;**81**:1647–53.

[6] Moldovan T, Boynton D, Kuperus J *et al.*. Incidence and clinical relevance of paraneoplastic syndromes in patients with renal cell carcinoma. *J Urol Oncol* 2023;**41**:392.e11.

[7] Gogusev J, Augusti M, Chrétien Y *et al.*. Interleukin-6 and TNFα production in human renal cell carcinoma. *Kidney Int* 1993;**44**:585–92.

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