

CASE REPORT

A curable pseudo-dementia related to an atypical presentation of giant cell arteritis

CLÉMENT LAHAYE¹, MANUEL SANCHEZ², AUDREY ROUET³, ARIANE GROSS², NATHALIE FAUCHER², AGATHE RAYNAUD-SIMON², MATTHIEU LILAMAND²

¹Unité mobile de Gériatrie, CHU Clermont-Ferrand, 63000 Clermont-Ferrand, France

²Département de Gériatrie, APHP, Hôpital Bichat, 75018 Paris, France

³Département de Gériatrie, APHP, Hôpital de la Pitié-Salpêtrière, 75013 Paris, France

Address correspondence to: Clément Lahaye. Tel: +33 4 73 75 45 94; Fax: +33 4 73 75 45 99.
Email: clahaye@chu-clermontferrand.fr

Abstract

A 70-year-old patient was admitted with rapidly progressive cognitive decline associated with limitations in activities of daily living, weight loss and cerebellar ataxia. The diagnosis of giant cell arteritis (GCA) with vascular involvement was made, based on the presence of a metabolically active vasculitis of the brachiocephalic trunk on ¹⁸FDG-PET imaging. Temporal artery biopsy also revealed pan-arteritis. A progressive regression of cognitive disorders occurred under corticosteroid treatment and immunosuppressive therapy. Previously published case reports concerning this atypical presentation of GCA are scarce. They suggest that numerous cognitive symptoms, such as impairment of short-term memory, disorientation, delirium, impaired attention or visual hallucinations might be related to GCA. Thus, this diagnosis should be considered as a curable cause of unexplained cognitive impairment associated with weight loss and systemic inflammation.

Keywords: cerebral vasculitis, dementia, giant cell arteritis, older people, reversible cognitive impairment

Key points

- Rapid cognitive decline associated with systemic inflammation should suggest the diagnosis of an atypical giant cell arteritis (GCA).
- Positron emission tomography affirms the presence of active large vessel vasculitis.
- Glucocorticoid therapy may cure giant cell arteritis but also restore cognitive function.

Case report

Giant cell arteritis (GCA) is the most common form of vasculitis in older adults [1]. The typical clinical presentation associates headache, scalp tenderness and jaw claudication inflammatory arthralgia and constitutional symptoms. High-dose corticosteroid therapy generally allows a rapid recovery. However, atypical forms of GCA also exist, and a delayed diagnosis may lead to potentially serious consequences, especially in older patients.

A 70-year-old patient was admitted for rapidly progressive cognitive decline. He had a history of alcohol abuse, type

2 diabetes since 2015 and hypertension. Clinical examination revealed psychomotor slowing, apathy, asthenia, weight loss of 15 kg in 6 months, axial extrapyramidal syndrome without resting tremor, cerebellar ataxia and sarcopenia. There were neither disturbed alertness, nor fasciculation, abnormal movements, orthostatic hypotension, headache, abnormality of cranial nerves, sensitive or motor deficit. His Mini Mental State Examination (MMSE) score was 20/30, Frontal Assessment Battery (FAB) score was 13/18. He also presented with limitations in activities of daily living, both for basic and instrumental activities (4/6 on the Katz scale and 1/4 on the Lawton scale, respectively). Symbolic gestures

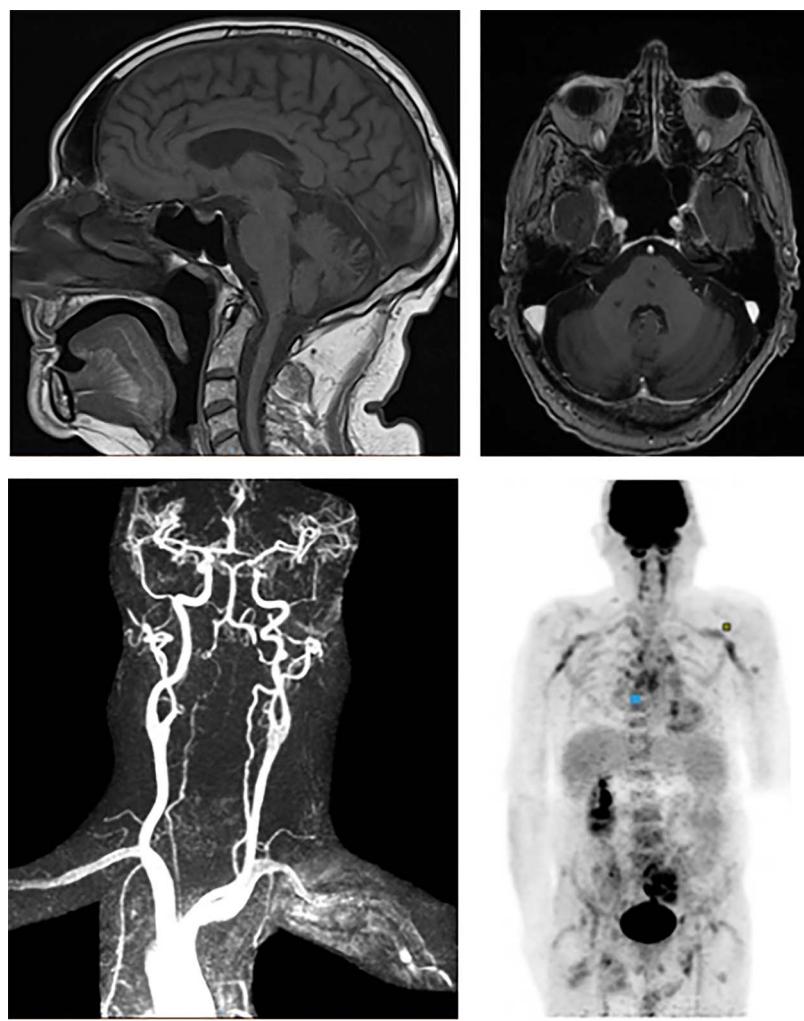


Figure 1. Brain and vascular imaging of the patient. Top left: cerebellar atrophy (MRI, T1 turbo spin echo, sagittal). Top right: microinfarcts of right middle cerebellar peduncle and pons (MRI, multiplanar reconstruction, axial). Bottom left: extensive right vertebral stenosis (magnetic resonance angiography). Bottom right: bilateral hypermetabolism of the arterial walls of the brachiocephalic arterial trunk, subclavian arteries, vertebral, maxillary and temporal arteries (Fluorine-18-Fluorodeoxyglucose positron emission tomography).

and visuo-constructive praxis were abnormal. There was no obvious depressive syndrome.

Blood analysis showed elevated C-reactive protein (27 mg/L). Thyroid-stimulating hormone, vitamins B1 (134 nmol/L), B6, B9 and B12 and liver enzymes were normal. Nuclear antibodies, anti-neutrophil cytoplasmic antibodies, serologies for HIV, TPHA/VDRL, *Bartonella*, rickettsiosis, *Coxiella* and brucellosis were negative. Three sputum tests for *Mycobacterium tuberculosis* as well as interferon- γ release assays excluded the diagnosis of tuberculosis. Cerebrospinal fluid analysis (CSF) showed 0.44 g/l of proteins, 3.3 mmol/l of glucose, 6 cells/mm³, negative culture. The polymerase chain reaction tests for John Cunningham virus, *Herpes simplex* virus 1 and 2 and varicella-zoster virus were negative, as well as antineuronal antibody, acetylcholine anti-receptor antibody and *Tropheryma Whipplei* polymerase chain reaction analysis.

The CSF levels of protein A β 42 (1109 pg/mL) and P-tau were normal (46 pg/mL), whereas total tau was elevated at 2019 pg/mL ($N < 350$ pg/mL).

Electroencephalogram and electromyogram of the four limbs were normal. Medullary MRI excluded spinal cord compression or myelopathy. Thoracoabdominopelvic computed tomography showed no specific abnormality. Brain MRI found cerebellar atrophy with multiple microinfarcts; besides, vascular MRI revealed a left vertebral stenosis with a V2 segment occlusion as well as an extensive right vertebral stenosis (Figure 1). The hypothesis of vascular dementia remained possible regarding the cerebellar lesions, although the timeline of cognitive decline seemed atypical. Furthermore, the presence of rapid weight loss argued against this diagnosis. Wernicke's encephalopathy was excluded, given the absence of delirium, ocular abnormality or vitamin B1 deficiency. However, the diagnosis of

paraneoplastic encephalitis was hypothesised. ¹⁸FDG-PET examination highlighted a metabolically active vasculitis of the brachiocephalic trunk, involving subclavian, vertebral and temporal arteries. Temporal artery biopsy showed an inflammatory pan-arteritis characterised by a destruction of the inner elastic lamina, an infiltration of the muscularis media and giant cells. Therefore, the final diagnosis was GCA with vertebral artery involvement, associated with multiple cerebellar strokes. The patient was prescribed three boluses of methylprednisolone, followed by 0.7 mg/kg/day of oral prednisone, associated with six courses of cyclophosphamide IV followed by azathioprine 100 mg/j. Inflammatory syndrome and anorexia disappeared within 1 week; cognitive performance and autonomy gradually improved (at 4 months MMSE 29/30, FAB 14/18, Katz scale 6/6 and Lawton scale 3/4).

GCA has exceptionally been described as a potentially curable dementia. Clinical manifestations from case reports suggest impairment of short-term memory, disorientation, delirium, impaired attention or visual hallucinations [2–4]. These neuropsychiatric symptoms are usually preceded by the classical manifestations of GCA. Several hypotheses have been evoked to explain these cognitive disorders: local inflammation due to vasculitis of the intracranial or meningeal vessels, systemic inflammation or subacute ischemia of the temporal and occipital lobes, derived from extracranial involvement of the vertebral arterial system [5, 6]. Although often incomplete, a gradual recovery of neuropsychiatric symptoms is expected under corticosteroid therapy.

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Declaration of Consent: Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

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