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ORAL AND INTESTINAL MANIFESTATIONS OF GIANT CELL ARTERITIS

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ABSTRACT

Background: Giant cell arteritis (GCA) is the most common primary vasculitis in individuals over 50 years of age. GCA typically affects large- and medium-sized arteries and is classically associated with cranial manifestations of ischaemia, such as headaches (notably in the temporal region), jaw claudication and visual symptoms that can lead to blindness. Extracranial symptoms are less frequently reported and are related to involvement of the thoracic and abdominal aorta and its main branches. Classic diagnostic tools (such as temporal artery ultrasonography and temporal artery biopsy) can be negative in extracranial GCA.

Case description: We report a difficult diagnosis of GCA in a 75-year-old woman who developed painful tongue ulcers that responded to prednisone treatment. However, a comprehensive diagnostic work-up did not lead to a certain diagnosis of GCA and corticosteroids were stopped after one month. A few months later, the patient suffered from mechanical ileus secondary to ischaemic stenosis of the small bowel. Pathological examination of the small bowel resection, and a second FDG-PET/CT, led to the diagnosis of extracranial GCA.

Conclusions: We present a case of CGA which sequentially affected cranial and extracranial arteries and illustrate pitfalls in the diagnosis of this polymorph condition.

KEYWORDS

Giant cell arteritis, lingual ulcerations, mesenteric ischaemia

LEARNING POINTS

- Giant cells arteritis (GCA) should be considered in older adults with symptoms and signs suggestive of tongue ischaemia, including tongue pain, inflammation, swelling and ulceration.
- GCA may present with extracranial manifestations due to involvement of the aorta and its main branches, including mesenteric ischaemia.
- Classic diagnostic tools for cranial GCA (temporal artery ultrasound and biopsy) may be negative in extracranial GCA, and FDG-PET CT should be obtained when this form of the disease is suspected.

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CASE DESCRIPTION

A 75-year-old woman was admitted to the hospital for severe tongue pain and a painful, burning mouth. The symptoms had been present for five days and prevented oral intake. She had been treated with oral fluconazole and topical nystatin without improvement. She had no fever. Additionally, she reported epigastric and lower abdominal pain lasting for a week. Past medical story was notable for mild chronic obstructive pulmonary disease and hypothyroidism. On admission, she was hypertensive (160/70 mmHg). The other vital signs were within the normal range. Oral examination revealed an inflamed, erythematous tongue with whitish plaques on the upper surface. There was no adenopathy, and the rest of the physical examination was unremarkable. Blood tests showed an elevated C-reactive protein (237 mg/l) and mild leucocytosis (13,100 cells/mm^3). Polymerase chain reaction for herpes simplex virus 1 and 2 was negative on a buccal swab. Despite intensive local care, the patient continued to complain of severe pain, impairing eating. An oesophagogastroduodenoscopy performed a few days after admission was normal. Biopsies from the duodenum, stomach and oesophagus showed mild chronic oesophagitis and mild inactive chronic pangastritis. There was no intestinal metaplasia or dysplasia, and *Helicobacter pylori* was absent. Tongue inflammation resolved progressively but two painful ulcers developed on the lateral border of the tongue *(Fig. 1)*. Further enquiries revealed occasional bitemporal headaches (worsening for 2-3 weeks), intermittent right monocular diplopia for 1-2 weeks and fatigue. Giant cell arteritis (GCA) leading to tongue ischaemia was suspected and prednisone 1 mg/kg/day was initiated, with rapid improvement of tongue pain and general symptoms. Ultrasonography of the temporal arteries was unremarkable. Autoimmune antibodies (ANCA, ANA, anti-SSA, anti-SSB) were absent. Fundoscopy was normal. A PET CT scan with injection of 18 F-fluorodeoxyglucose, performed two days after the start of high-dose glucocorticoids, did not show increased uptake of the aorta or main branches, nor any other hypermetabolic target. A temporal artery biopsy performed after seven days of prednisone showed only a small, localised arteritis focus, not specific for GCA. A pan-endoscopy with biopsies and MRI of the facial and cervical regions were normal. Biopsies of the tongue ulcers revealed acute inflammation; there was no dysplasia, but herpes simplex virus was detected by immunohistochemistry in the ulcer. Despite initial improvement with prednisone, it was felt that the diagnosis of GCA was not certain. Oral valacyclovir was administered for ten days, and prednisone was tapered over one month.

The evolution of the ulcers was favourable, but the patient still complained of intermittent, non-specific abdominal pain. Five months later, she was admitted at the hospital for severe bouts of abdominal pain and vomiting. Blood tests showed a mild leucocytosis (13,900 cells/mm^3) and C-reactive protein was moderately elevated (40 mg/l). Liver function tests were within the normal range. A CT-scan of the abdomen showed mechanical small bowel obstruction, and the patient

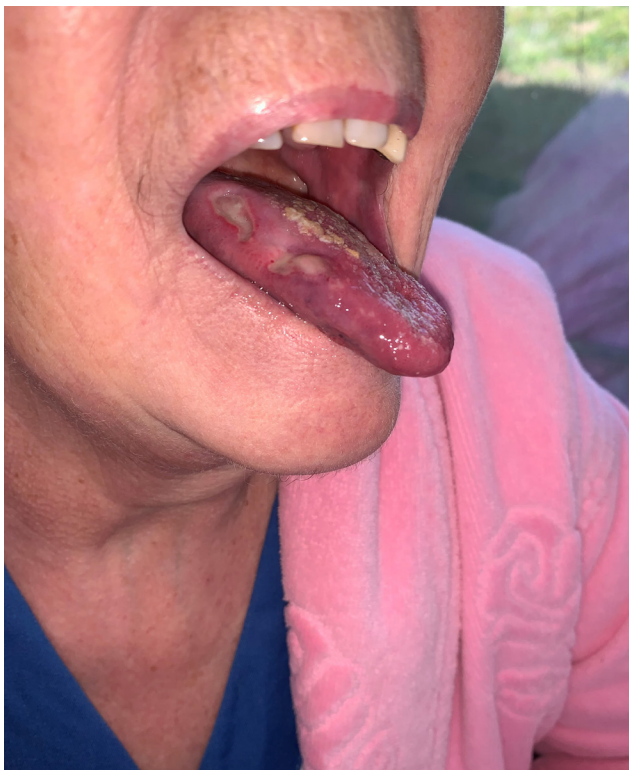

Figure 1. Presence of two ulcerative lesions on the right lateral border of the tongue.

underwent segmental small bowel resection. The pathology of the surgical specimen showed three stenotic areas, suggesting subacute ischaemia. Analysis of the mesenteric arteries revealed focal inflammation of the vessel wall with giant cells, consistent with GCA *(Fig. 2)*. A second PET CT scan performed seven months after the initial presentation showed homogeneous hypermetabolism involving the aortic arch, the thoracic aorta, carotid arteries, vertebral arteries, proximal subclavian and axillary arteries, the abdominal aorta, the iliac arteries and both femoral arteries. The total vasculitis score was 20/21. There were interspinous bursitis, enthesopathies and symphysitis suggestive of associated polymyalgia rheumatica.

Prednisone was restarted at 60 mg/day. Tocilizumab was added to the treatment two months later, allowing progressive tapering of prednisone. The clinical and biological evolution was favourable.

DISCUSSION

GCA is the most common primary systemic vasculitis in patients over 50 years old[1], with an incidence increasing with age. GCA primarily affects large and medium-sized arteries, typically involving cranial branches with a predilection for the temporal arteries, causing headaches, jaw claudication and visual disturbances. It later progresses to blindness in up to 20% of untreated cases, the most dreaded complication of the disease[1]. Tongue ischaemia is a classical, albeit rarer finding. The diagnosis of GCA is based on clinical classification criteria (2022 ACR/EULAR), laboratory tests (elevated sedimentation rate or C-reactive protein), imaging (temporal artery ultrasound 'halo sign', FDG-PET of the

aorta) and temporal artery biopsy[2]. The sensitivity of the new criteria is 87%, with a specificity of 94.8%[3].

Extracranial manifestations, secondary to inflammation of the aorta and its main branches – often bilaterally – is present in only 9% of cases, affecting the abdomen, thorax and pelvis[2]. Extracranial GCA can present without cranial involvement, with prominent systemic symptoms such as low-grade fever, fatigue and weight lost. In this variant, classical diagnostic investigations (temporal ultrasound and temporal artery biopsy) are often normal. The first description of mesenteric ischaemia associated with CGA was made in 1965 by Hamrin et al.[4]. The prevalence of mesenteric vasculitis and ischaemia is likely underestimated in CGA, as it may occur without overt abdominal symptoms, as shown in post-mortem studies[5]. In GCA, vascular involvement is proximal, unlike other vasculitides where inflammation is more distal (eosinophilic granulomatosis with polyangiitis, Behçet's disease). Due to significant collateral circulation in the mesenteric vascular beds, patients tend to remain asymptomatic, and multiple vessels must be affected before clinical symptoms manifest. The reference diagnosis standard is still angiography, but less invasive techniques may be useful due to the proximal nature of the arteritis, such as ultrasound, MRI and CT scans. The mortality risk for mesenteric ischaemia in CGA may be as high as 33%, possibly due to delayed diagnosis[5]. In a study of 28 cases of mesenteric ischaemia associated with GCA, only 46% of patients presented with an occlusive syndrome, and less than 15% had typical abdominal angina or gastrointestinal bleeding[5]. Grayson et al. showed that mesenteric artery angiographic lesions are present in 18% of CGA cases[6]. Scola et al. explained the low frequency of clinical manifestations of mesenteric involvement in GCA by the important collateral circulation[7]. The splanchnic vasculature consists of three main vessels (celiac artery, superior and inferior mesenteric arteries), interconnected by the artery of Drummond and the arc of Riolan, which help maintain intestinal perfusion in the event of occlusion of the

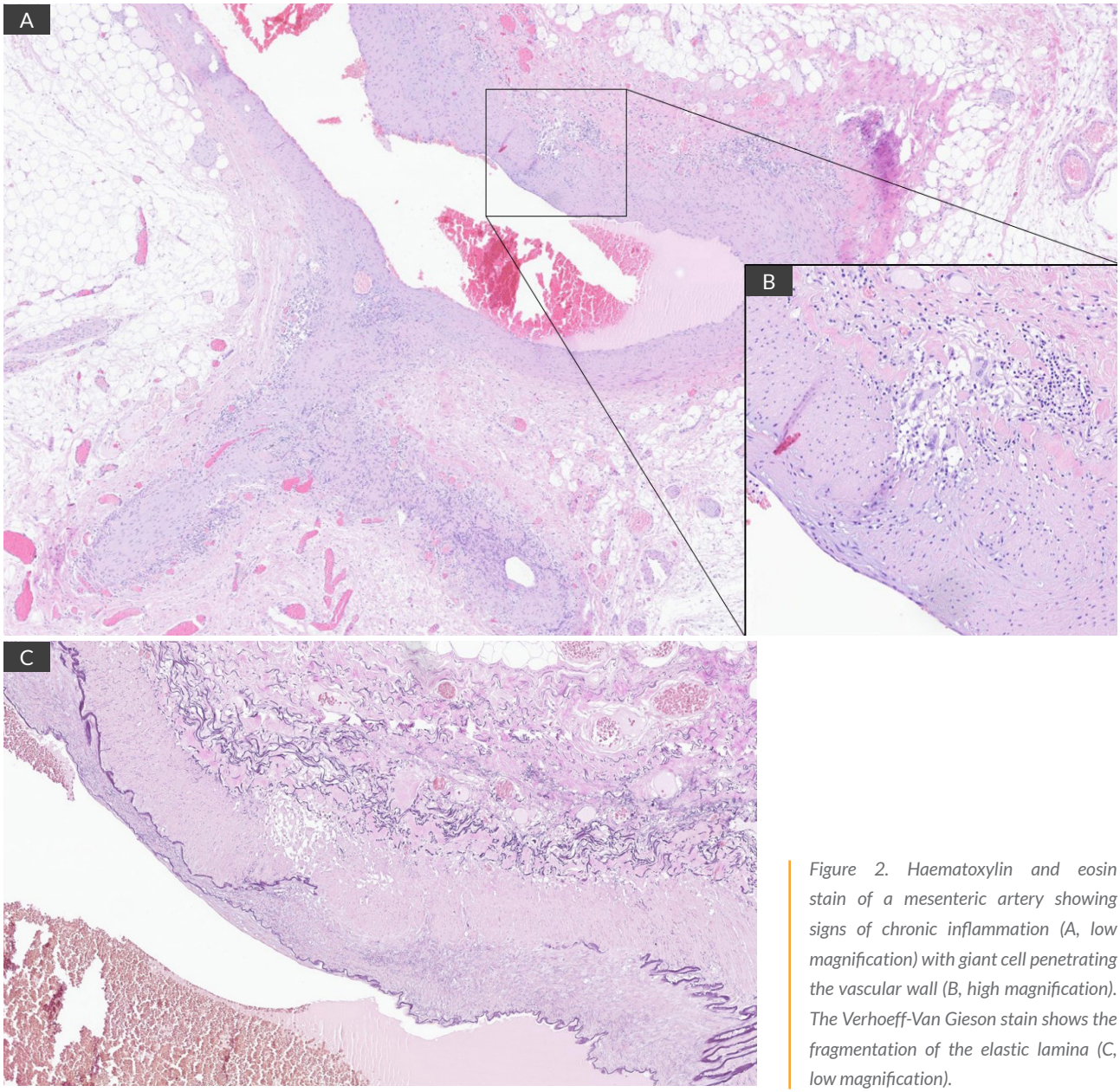

Figure 2. Haematoxylin and eosin stain of a mesenteric artery showing signs of chronic inflammation (A, low magnification) with giant cell penetrating the vascular wall (B, high magnification). The Verhoeff-Van Gieson stain shows the fragmentation of the elastic lamina (C, low magnification).

main vessels. Due to this anatomical configuration, multiple vessels in the splanchnic circulation must be significantly affected to produce clinical symptoms. Other vasculitides, such as IgA vasculitis and ANCA-associated vasculitis, which can present with acute digestive symptoms, involve smaller vessels beyond Drummond's artery and Riolan's arc[7].

Tongue ulcers are a classic but rare manifestation of GCA, with only a few dozen cases reported[8], due to the rich collateral lingual vascularisation[3]. The differential diagnosis of lingual ulcers includes malignancies, infections, drug toxicity (including chemotherapy), radiation therapy, embolisation and other vasculitides (polyarteritis nodosa, granulomatosis with polyangiitis)[8-10]. DeBord et al. recommended screening for GCA in any unexplained lingual necrosis in patients over 50 years old[10]. Due to the tongue's rich vascularisation, tongue ischaemia is related to extensive involvement of cranial arteries and is related to a poorer prognosis[8]. The involvement is usually located on the anterior two-thirds of the tongue. Rapid administration of corticosteroids generally results in complete healing of the tongue ulcers[9].

Our patient initially presented with the cranial form of GCA, though abdominal pain was already present and probably due to mesenteric ischaemia. Her story illustrates pitfalls in the diagnosis of GCA. The initial presentation, though some typical signs and symptoms were present, did not lead to a certain diagnosis of GCA despite an intensive diagnostic work-up including ultrasonography, PET CT and histology. The positive immunohistochemistry for herpes simplex virus in the tongue ulcer, which was probably a false positive or an accompanying finding without clinical signification, was an additional distractor. Non-specific abdominal complaints were overlooked until ischaemic stenosis finally led to small bowel obstruction.

CONCLUSION

GCA should be considered in the presence of severe tongue inflammation and ulcers in the older adult. Raised awareness for extracranial forms of CGA in the differential diagnosis of unexplained abdominal symptoms in older adults is also warranted to avoid delay in the introduction of appropriate, life-saving treatment. Finally, better knowledge of the interplay between cranial and extracranial forms of GCA, and refined diagnostic techniques and algorithms are needed to prevent complications of this treatable condition.

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