CASE REPORT

A Case of Giant Cell Arteritis that Presented with Buccal Skin Ulceration along the Facial Artery

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Abstract:
A 68-year-old man presented with right buccal ulceration along the facial artery, temporal pain, lagophthalmos, diplopia, and tongue deviation to the right. Contrast-enhanced computed tomography showed bilateral temporal artery and right maxillary artery wall thickening, and a diagnosis of giant cell arteritis (GCA) was made according to the American College of Rheumatology 1990 criteria. Treatment with corticosteroids ameliorated his symptoms. This is the first report of GCA with buccal skin ulceration along a facial artery. Because a delayed diagnosis can lead to irreversible damage, it is essential to notice rare symptoms, such as skin ulceration and multiple cranial neuropathy-like symptoms.

Key words: giant cell arteritis (GCA), facial artery, maxillary artery, buccal skin ulceration, trismus

Introduction
Giant cell arteritis (GCA) is granulomatous vasculitis of medium to large vessels that mostly affects the elderly. Its main vascular targets are extracranial branches of the carotid artery, aorta and its branches (1). Typical symptoms of GCA include a low-grade fever, headaches, scalp tenderness and jaw claudication. However, because of the numerous organs supplied by the aorta and carotid arteries, GCA can produce a broad range of symptoms. The lack of awareness of the atypical manifestations may result in a delay in both the diagnosis and treatment, which can lead to irreversible consequences, such as vision loss, limb anoxia, stroke or even death. In this regard, it is important to recognize the less-frequent complications of GCA.

We herein report a very rare case of GCA presenting with cheek ulceration along the facial artery and multiple cranial neuropathy-like symptoms.

Case Presentation
A 68-year-old man was admitted to our hospital because of skin ulcers on his cheek area and difficulty opening his mouth. He had no notable medical history, including hypertension, diabetes mellitus, or dyslipidaemia. He had been a regular smoker of 10 cigarettes daily for 50 years.

One month prior to admission, the patient presented with upper right cheek to temporal pain and lower eyelid rash. Seventeen days before admission, the pain from the right cheek to the temporal region increased, and swelling and redness of the right face and trismus appeared. He visited a dentist, where a mandibular fracture was suspected, but he had no history of trauma, and there was no fracture on computed tomography (CT). A blood test showed an elevated C-reactive protein (CRP) level of 6.73 mg/dL.

Under suspicion of right maxillary odontitis, the patient was admitted to the dental clinic and started on treatment with ampicillin 3 g/day and clindamycin 1,200 mg/day. Fifteen days before admission, the antibiotics was changed to meropenem 1 g/day, to which no response was observed. Twelve days prior to admission, the ulcer under the right eyelid expanded in a downward band, and crusting was observed. Herpes zoster was suspected, and acyclovir 750 mg/day was started, but the symptoms continued to worsen.
Seven days prior to admission, the patient was referred to the Department of Dermatology at our hospital. Vasculitis was suspected due to the presence of an ulcer along the course of the facial artery on right cheek. A biopsy of the skin ulcer revealed findings suggestive of vasculitis. Since bilateral temporal pain was also observed, GCA was suspected, and the patient was referred to our department and admitted.

On admission, his chief complaint was right cheek pain, temporal pain, and trismus. A physical examination revealed an ulcer along the course of the facial artery on right cheek (Fig. 1A). He had difficulty opening the mouth (Fig. 2A), difficulty closing the right eye (Fig. 2B), double vision in the left and right gaze direction and tongue that was deviated to the right (Fig. 2C), which suggested that the patient had III, V, VII, and XII cranial nerve impairment. No obvious abnormalities were noted on other cranial nerve examinations or in any cerebellar functions. The bilateral temporal
Figure 3. Contrast-computed tomography scan of the head. A: Wall thickening was observed in the right temporal artery, and mild wall thickening was observed in the left temporal artery. The lower panel shows a magnified view of the right temporal artery (arrow). B: Wall thickening and narrowing of the lumen were observed in the starting portion of the right maxillary artery. The lower panel shows a magnified view of the right maxillary artery (arrow).

The laboratory study results were as follows: white blood cell count, 13,590/μL; erythrocyte sedimentation rate, 79 mm/h; creatinine (Cre), 0.8 mg/dL; serum IgG, 1,116 mg/dL. There were no abnormalities on the coagulation test. Antinuclear antibody, antiphospholipid antibodies, cryoglobulin, and anti-neutrophil cytoplasmic antibody testing were all negative. The urinalysis was normal. Contrast-enhanced CT of the cervicothoracic and abdominal pelvic regions showed emphysema, but there was no evidence of large vessel vasculitis. Some calcified lesions were observed in the descending thoracic aorta and abdominal aorta, but no obvious calcification, thrombus or aneurysm was observed in the other arteries. Contrast-enhanced CT of the head showed bilateral temporal artery wall thickening (Fig. 3A) and right maxillary artery wall thickening (Fig. 3B). Head MRI showed no significant findings in the cranial vessels and no evidence of cerebral infarction or cerebral haemorrhaging. A spinal fluid examination showed no obvious evidence of infection.

According to the American College of Rheumatology 1,990 criteria (2), he was diagnosed with GCA, based on an onset over 50 years old, new onset of temporal pain and erythrocyte sedimentation rate >50 mm/h. There was no evidence for a diagnosis of any other disease, including infections and malignancies. Diseases that needed to be differentiated included ANCA-negative ANCA-associated vasculitis (AAV), polyarteritis nodosa (PAN), varicella zoster virus (VZV) infection, and local bacterial infection. The diagnosis of AAV was ruled out because the lesions in our case were mainly observed in the medium to large vessels, with no findings of small artery damage in the ears, nose, upper respiratory tract, pulmonary, or kidney. Although it was difficult to definitively rule out the diagnosis of PAN, the absence of findings of visceral infarction or small intra-abdominal aneurysm, which are more likely to be seen in PAN, and the presence of temporal artery findings, which are less likely to be seen in PAN, made the diagnosis of GCA more appropriate than PN. VZV and bacterial infections were excluded because skin biopsies showed no evidence of infection, and broad-spectrum antimicrobials and antivirals were ineffective.

At 3 days after admission, treatment was started with methylprednisolone 1 g for 3 days, followed by oral high-dose prednisolone (PSL) (50 mg/day, 1 mg/kg/day). His cheek pain and temporal pain resolved promptly after the initiation of treatment. A right temporal artery biopsy was performed 10 days after the start of treatment (Fig. 4) and did not reveal findings characteristic of GCA, such as the infiltration of multinucleated giant cells or fragmentation of the
Discussion

We encountered a rare case of GCA presented with skin ulceration along the facial artery and various cranial neuropathy-like symptoms, such as double vision, difficulty in jaw opening, difficulty in eye closure and tongue deviation.

Because of the high incidence of temporal artery involvement, GCA used to be called temporal arteritis. With the increased use of imaging, including ultrasonography or positron emission tomography in combination with CT, it has become clear that the other branches of external carotid arteries are frequently involved, although some cases are asymptomatic. In our case, CT images and skin ulcer along the facial artery indicated the involvement of the temporal, maxillary, and facial arteries. All of these arteries are branches of the external carotid artery. In a report using colour Doppler ultrasonography, facial artery involvement was found in 40.9% of patients with GCA (5), and a histologically proven case of facial artery involvement in GCA has also been reported (6). In addition, 29% of patients with GCA reportedly had maxillary artery involvement on positron emission tomography (7). Although there are scattered reports of skin ulceration of the scalp in GCA (8-10), extensive skin ulceration along the course of the facial artery, as in our case, has not been reported. Skin ulcers may be seen in cases of coexisting atherosclerotic lesions or arterial thrombi, but in the present case, the atherosclerotic findings were mild, and no obvious arterial thrombi were observed.
Although we were unable to clearly identify what caused the tendency to develop buccal ulcers in this patient, we suspect that it was due to the high degree of inflammation of vasculitis.

Cranial nerve palsies can occur as complications of GCA. Most of these reports concern oculomotor nerve palsy (11-14), with only limited reports describing multiple cranial neuropathies due to GCA. Fytili et al. (15) described a 68-year-old man with abducens, laryngeal and acoustic nerve involvement, Ross et al. (16) described an 80-year-old man with oculomotor, trochlear and abducens nerve involvement and a 73-year-old man with oculomotor, trigeminal and abducens nerve involvement.

Our case presented with diplopia, jaw trismus, lagophthalmos and tongue deviation. Diplopia is reported in 1%-19% of patients with GCA. Diplopia can result from ischemia of the ocular motor nerves or extraocular muscles or brainstem (17). Diplopia usually appears during the active phase of arteritis and typically precedes vision loss. Unlike visual loss, diplopia is reported to be transient and can be improved with treatment within 24 h up to a few months (18). In our case, diplopia improved within a week after the start of treatment.

Jaw trismus is less frequently reported than jaw claudication. However, in a retrospective study from Israel, 6.8% of patients complained of a reduction in jaw opening in GCA (19). The precise pathological mechanism underlying trismus is unknown, but this symptom may result from an ischemic maxillary artery supplying the masseter muscles or impairment of the trigeminal or facial nerve.

To our knowledge, lagophthalmos in GCA has not been reported. The orbicularis oculi muscle, which closes the eyelids, is innervated by the facial nerve and receives its blood from the angular artery, a terminal branch of the facial artery. In our case, eye closure may have been impaired by ischemia of the orbicularis oculi muscle or facial nerve. In addition, to our knowledge, deviation of the tongue associated with GCA has not been previously reported. Ischemia of the lingual artery, a branch of the external carotid artery, or hypoglossal nerve impairment is thought to be a possible cause.

Although our patient had a high degree of functional impairment at the time of the diagnosis, a prompt diagnosis and treatment led to a favourable prognosis without irreversible damage. Even in cases with atypical findings, such as the present case, it is necessary to consider GCA as a differential diagnosis.

The authors state that they have no Conflict of Interest (COI).

References

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