Giant Cell Arteritis with Facial Edema Presenting with Delayed Jugular Venous Flow

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Abstract

The detection of abnormalities of the cranial arteries on magnetic resonance imaging (MRI) is useful for the diagnosis of giant cell arteritis (GCA). However, reports on the veins of GCA patients are rare. We report the case of an elderly woman with GCA who presented with facial edema. She presented with a one month history of headache and facial edema. After MRI and enhanced computed tomography revealed delayed blood flow in the left jugular vein, a temporal artery biopsy was performed. She was diagnosed with GCA based on the biopsy findings. Following corticosteroid therapy, her symptoms and venous flow improved. The present case indicates that delayed jugular venous flow can occur in GCA patients with facial edema.

Key words: giant cell arteritis, facial edema, jugular vein, prednisolone

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Introduction

Giant cell arteritis (GCA) is a systemic granulomatous vasculitis that occurs in older individuals (1-3). Early diagnosis and immediate treatment with corticosteroids are important to prevent the severe vascular complications of GCA, which include blindness and cerebrovascular accidents. The usual systemic features of the disease include headache, scalp tenderness, jaw claudication, malaise, and arthralgia in the proximal joints. Facial edema is sometimes observed in patients with GCA, and must be appreciated as another presentation of GCA, especially when it is accompanied by other symptoms (4, 5). GCA affects the large- and medium-sized arteries, predominantly the cranial arteries. The investigation of cranial arteries by magnetic resonance imaging (MRI) has been reported to be useful for the diagnosis in patients suspected of having GCA. However, few reports have focused on the imaging of the veins of GCA patients. We herein report a case of GCA in an elderly woman who presented with facial edema, in whom delayed blood flow in the jugular vein was revealed by MRI and enhanced computed tomography (CT). The case demonstrates that MRI might show delayed venous flow in patients with early-stage GCA who present with facial edema.

Case Report

A 73-year-old woman was referred to our hospital due to an approximately 1-month history of temporal headache, neck pain, facial edema, low-grade fever, and jaw claudication. On physical examination, the patient was conscious and alert. Her blood pressure, pulse rate, respiratory rate and temperature were 120/70 mmHg, 62 beats/min, 16 breaths/min, and 36.9°C, respectively. Her face was swollen (Fig. 1a), and her left jugular vein was distended. She had trismus and pain upon opening her mouth. She had no ophthalmological problems. Her breath and heart sounds were clear. The laboratory findings revealed a white blood cell count of 10,300/µL, a hemoglobin level of 10.7 g/dL, a C-reactive protein level of 10.7 g/dL, a C-reactive protein level of 9.68 mg/dL, and an erythrocyte sedimentation rate of 118 mm/h. Her prothrombin time (PT)-INR was 1.06, her activated partial thromboplastin time (aPTT) was 40.8 s, and her D-dimer level was 0.98 µg/mL.

The patient had previously undergone head MRI at another hospital, 1 week after the onset of her symptoms. T2-
Facial edema is sometimes recognized in GCA patients and may herald the presence of the disease (4, 5). Facial edema might be caused by the inflammatory involvement of the branches of the external carotid artery. Few reports have focused on the veins of GCA patients with or without facial edema. In the present case, MRI and enhanced CT demonstrated delayed jugular venous flow at the time at which the patient had facial edema. After the administration of PSL, which was used to treat the patient’s GCA, the patient’s facial edema improved and the delayed venous flow rapidly regressed. A few cases of venous thrombosis in GCA patients have been reported (6). There is an almost 2.5-fold increase in the risk of venous thromboembolism in patients with GCA in comparison to the general population (7). This suggests the possibility of phlebostasis in GCA patients. In the present case, MRI and enhanced CT did not show venous thrombus, and her PT-INR, aPTT, and D-dimer levels were normal. As a result, we did not suspect the presence of venous thrombus and did not investigate coagulopathy any further. The facial edema might have been caused by the widespread inflammatory involvement of branches of the external carotid artery (e.g., facial, internal maxillary, and ascending pharyngeal arteries), although such situations have only rarely been documented (8, 9). Alternatively, the abnormal release of vascular endothelial growth factor, a protein that both induces hyperpermeability and stimulates endothelial cell growth (10), could play a role in the development of local or distant swelling in some GCA patients (11, 12). The altered fluid distribution in the face might influence the local hemodynamics. The interstitial pressure might be elevated due to the increase of interstitial fluid volume. This elevation could place some stress on the veins, leading to the change of the jugular venous flow that was observed in the present case. Although the rate at which asymmetric blood flow in the jugular vein occurs in the general population is unknown, it is sometimes observed in the MRI scans of healthy individuals. The left jugular vein is considered to be easily affected - probably for anatomical reasons. This patient showed delayed venous flow in the left jugular vein,

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Discussion

The course of this patient suggested two important clinical issues: 1) GCA patients with facial edema can present with delayed jugular venous flow; and 2) MRI might show the delayed flow at an early stage of GCA with facial edema.

Figure 1. (a) Facial edema was observed on admission. (b) It diminished after the initiation of treatment with prednisolone (PSL).

Figure 2. Magnetic resonance T2-weighted imaging shows the high intensity of the left internal jugular vein (arrow).
which promptly improved after corticosteroid therapy. In this case, it seems that the patient’s delayed venous flow was influenced by GCA.

MRI might show delayed jugular venous flow at an early stage of GCA in patients presenting with facial edema. In this case, phlebostasis was observed on MRI just 1 week after the onset of symptoms, whereas no abnormal arterial findings were indicated. Klink et al. (13) reported that MRI of the cranial arteries is accurate and reproducible during the initial diagnosis in patients with suspected GCA, with a sensitivity of 88.7% and a specificity of 75.0% in comparison to the reference standard. In addition to the arterial findings, abnormal jugular venous flow might be detected by MRI at an early stage of GCA in patients presenting with facial edema.

In conclusion, delayed jugular venous flow can be detected in GCA patients with facial edema by MRI and enhanced CT. MRI might show delayed venous flow at an early stage of GCA in patients with facial edema. Further cases should be accumulated to determine how frequently delayed venous flow is observed in GCA patients with facial edema and to elucidate its meaning and pathogenesis.

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

References


