原文題目(出處)	A 78-year-old woman with bilateral tongue necrosis. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;111:15-19
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<內文>

Introduction

Chief complaint

A 78-year-old woman presented to the emergency department complaining of a **sore tongue**.

Present illness

The patient reported pain of the right head, neck, face, and shoulder, especially while **eating** (ranked 8).

She also reported **fatigue**, and **visual blurring** that had developed 2 months before, **weight loss** over the preceding 8 weeks (from 67 to 60 kg; a **10.4%** reduction), and **tongue pain** of 4-weeks' duration (ranked 10).

10 days before presentation at the **emergency room**, she had undergone a complete blood count (**CBC**) and **c**omputed tomography (**CT**) of the head to rule out any underlying systemic conditions and space-occupying lesions.

CBC and **CT** results at that time were **normal**, except for a ***mild anemia***：hemoglobin hemoglobin, 10.7 g/dL（normal：12-16 g/dL for women）.

Her **platelet** count was **normal**：397 x 10^9/L（normal：140–400 x 10^9/L）

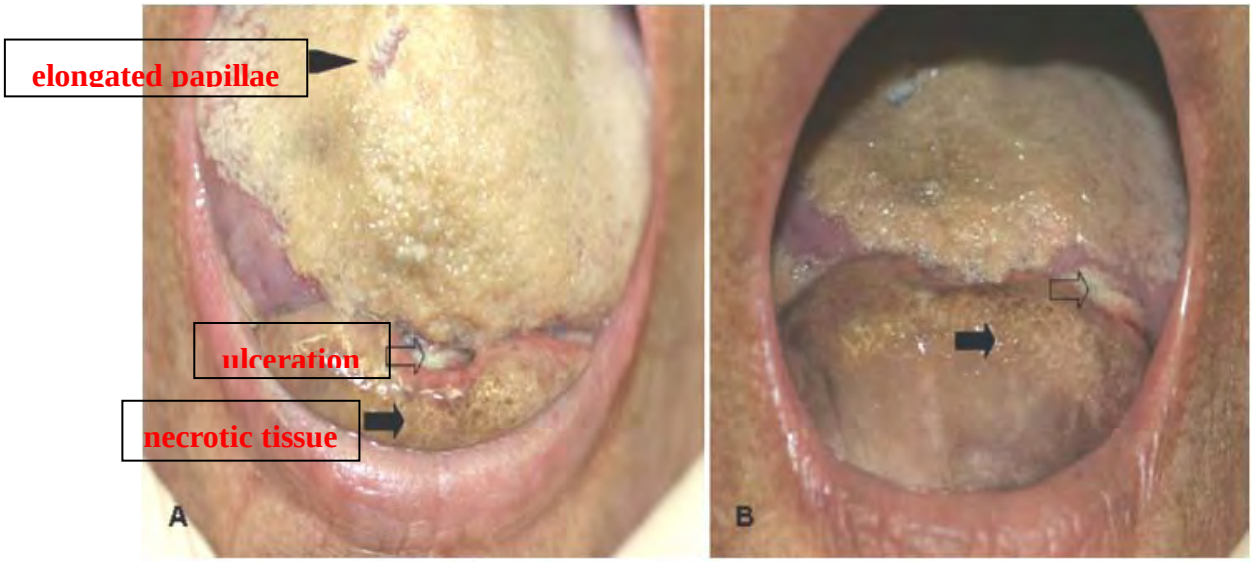
Medical history

A. **Hypertension**： **atenolol**, 100 mg/d, and **enalapril**, 5 mg/d
B. **Osteoporosis**： **calcium** and **vitamin D** (with no past bisphosphonate treatment)
C. **Gastroesophageal reflux disease**： **omeprazole**, 20 mg/d
D. **Admission**： 17 years ago
   she had suffered from **ovarian carcinoma** that was treated by **chemotherapy** and **oophorectomy** without therapeutic irradiation.
E. No known hypersensitivities or drug allergies

Clinical examination

The anterior third of the tongue was **enlarged bilaterally** and **painful to palpation**.

The tissue was **necrotic**, **without bleeding**. An **ulcerated fissure** was noted at its **proximal border**. The dorsal surface of the tongue was **yellowish-gray** with **elongated papillae** that **could not be wiped off**.



DIFFERENTIAL DIAGNOSIS

Trauma

Denied electrical, thermal, and chemical injury

Secondary to biting is rare

No oral parafunctional habits

Infectious diseases

Epstei Barr virus- and **cytomegalovirus (CMV)**-associated necrosis in patients with HIV infection

Neoplasms

Squamous cell carcinoma is the most common primary malignant neoplasm

Metastasis is accounting for 1% of oral malignancies, a quarter in the tongue

Vascular compromise related to vasculitic disorders

Giant cell arteritis (GCA) also known as temporal arteritis and cranial arteritis, is a chronic vasculitis of large and medium-sized arteries, with both localized and systemic inflammatory features.（10 to 70 / 100,000）

Older than 50 years (average：70 years), women more(1.4 to 3.0：1)

Serious ischemic complications, such as visual disturbances and sudden blindness owing to ischemic optic neuropathy, stroke, aneurysm of the aorta, infarction of the intestine, renal insufficiency, and myocardial infarction.

Clinical features：

a gradual worsening of diffuse unilateral headaches and scalp tenderness, facial pain or sore throat without headache, temporal artery abnormalities, chest pain, fever, anorexia, weight loss, and scalp necrosis.

Anemia and elevated platelet count are often noted

Cardiovascular and cerebrovascular disease

6% to 50% of patients with **antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis** (**Wegener's granulomatosis [WG]**), but only rarely do the presenting features include ulceration.

Vasopressin therapy for control of acute bleeding has been reported as a possible cause of tongue necrosis.

Radical neck dissection with ligation of the external carotid artery in patients with **subsequent radiotherapy** to the neck has been reported as a cause of unilateral tongue necrosis

Other causes of tongue necrosis include complications from **arterial embolization** and **anticancer chemotherapy or radiotherapy**.

Other systemic conditions

Patients with other systemic conditions associated with vascular involvement, such as **systemic lupus erythematosus**, **Raynaud's phenomenon**, and **anti-phospholipid syndrome**.

DIAGNOSIS AND MANAGEMENT

1. Elevated **erythrocyte sedimentation rate**（ESR）：69 mm/h(normal：1-20 mm/h)
2. Elevated **C-reactive protein** ：6.1 mg/L(normal：0-1 mg/L)
3. **Mild normocytic anemia** with hemoglobin level of 10.2 g/Dl
4. **Mean corpuscular volume** of 86.1μm^3 (normal：77-91 μm^3)
5. **Leukocyte**：15.6 x 10^9/L (normal ：4-10 x 10^9/L)
6. **Platelet**：572 x 10^9/L
7. **Creatinine** ：0.63 mg/dL (normal： 0.60 –1.06 mg/dL)
8. Cytoplasmic and perinuclear ANCA were negative
9. A **microbial culture** from the tongue revealed normal oral flora
10. Negative for herpes **simplex virus-1, herpes virus-2, and CMV** infection
11. **CMV antibodies** were not detected in the serum
12. **Plain occlusal mandibular** and **chest** radiographs were without any pathologic findings
13. Color-coded duplex sonography of the **temporal arteries** revealed occlusion of the left artery and normal flow of the right artery

According to **American College of Rheumatology (ACR) criteria**, at least 3 of the following 5 criteria are needed for a **diagnosis of GCA**：

(1) **Age older than 50 years at disease onset**（O）

(2) **New onset of localized headache**（O）

(3) **Temporal artery tenderness or a decreased temporal artery pulse**（O）

(4) **ESR higher than 50 mm/h**（O）

(5) A biopsy sample, including artery, showing **necrotizing arteritis**, characterized by a predominance of mononuclear cell infiltrates or a granulomatous process with multinucleated giant cells.

Our final diagnosis was **GCA**.

- Treatment
  1. **Prednisone**, 60 mg/d was initiated
  2. Topical **nystatin** and **chlorhexidine**
  3. **Opioid** analgesics

On the sixth day of hospitalization, the necrotic tissue underwent self-amputation

Histopathological examination of the amputated tissue

Revealed **necrosis** and **acute inflammation** with **no evidence of fungal**

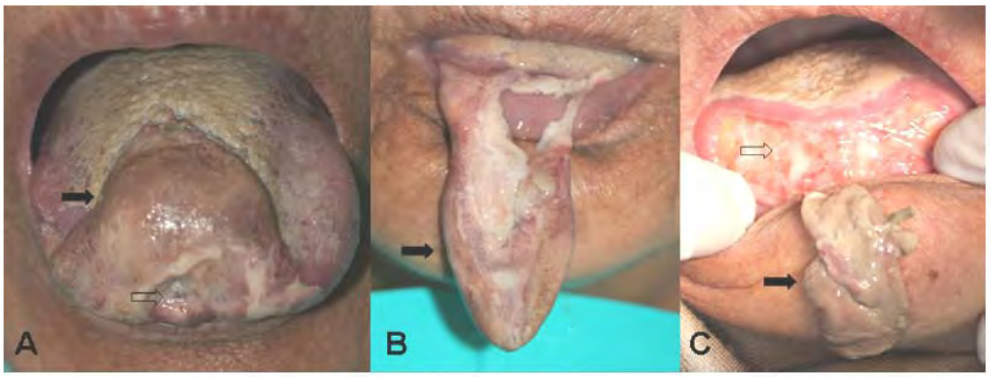
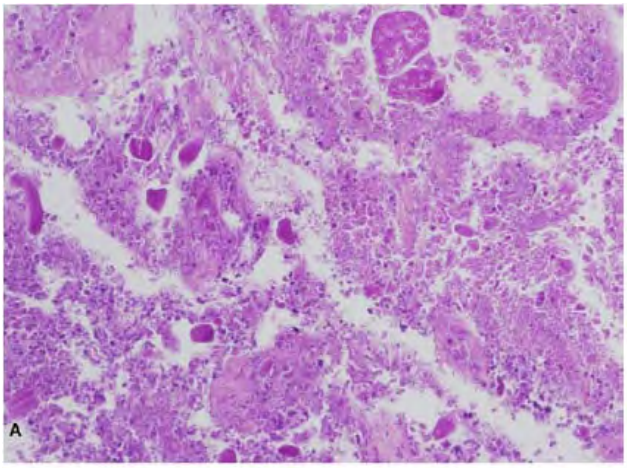
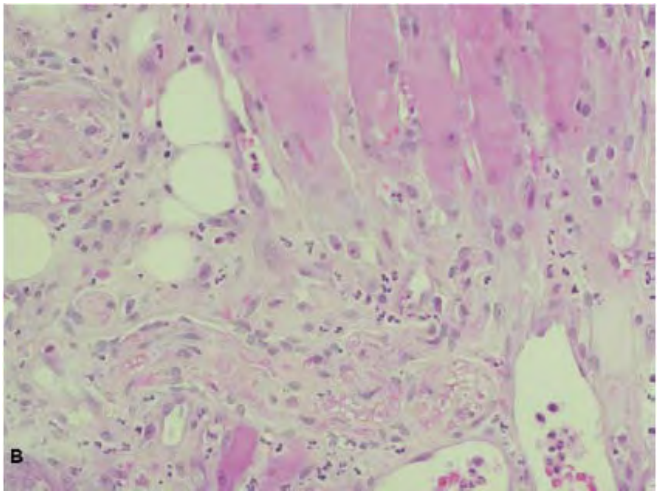


Fig. 2. The patient's tongue at **A**, third, **B**, fifth, and **C**, sixth day of hospitalization demonstrating auto-amputation of the necrotic tissue and exposure of underlying ulcerated tissue.



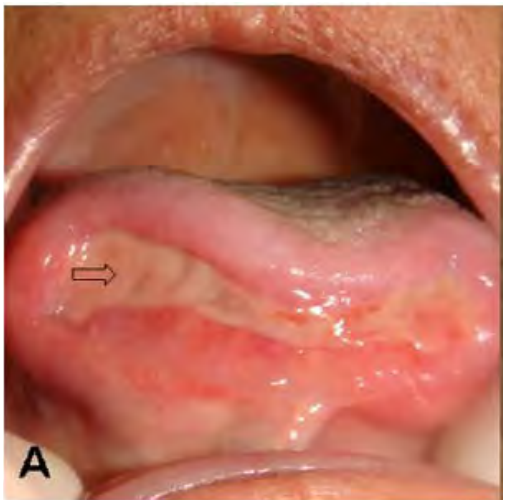
organisms

self-amputated necrotic material with **acute inflammation** and **bacterial colonization**



Viable tissue with a**cute inflammation within biopsied tongue** tissue proximal to necrosis

With progressive healing of the tongue after auto-amputation and with diminishing pain in the head, face, and neck, the patient was **discharged after 2 weeks** of hospitalization.



The patient was continued on **60 mg** of **prednisone** per day. Full epithelization was evident **a month** later

A **tapering** of the **steroid therapy** was instituted with continuation of **topical antifungal therapy**.

After **12 months**, the patient was in good health and showed a gain in weight (70 kg).

Her tongue morphology was **normal** with minimal limitations during function.

DISCUSSION

Orofacial manifestations of GCA include **temporal pain**, **jaw claudication and pain**, and **diplopia**.

Dental pain, dysphagia, glossitis, **necrosis of lip or tongue**, and facial swelling have been also reported.

Temporal pain may vary in character from throbbing, burning, boring.

Jaw claudication, which affects up to **65%** of the patients（the maxillary artery and masticatory muscle ischemia and pain）

Pain of the temporalis and masseter muscles on chewing is virtually pathognomonic of GCA.

In our patient, despite the absence of a **temporal artery biopsy**, a diagnosis of GCA was rendered with confidence based the presence of 4 of 5 ACR GCA diagnostic criteria.

The further course of GCA can be predicted according to the **initial systemic inflammatory response** of the patient, which is determined by the following 5 parameters：

1. **Sedimentation rate greater than 100 mm/h**
2. **Thrombocytosis：400 x 10^9/L**
3. **Hemoglobin below：11 g/dL**
4. **Leukocytosis greater than：11 x 10^9/L**
5. **Fever higher than 37.5°C**

Patients with a strong response (at least 4 positive parameters) may have a prolonged disease course with more flare-ups, and require higher steroid doses than moderate and weakly responding patients.

Our patient showed 3 positive inflammatory response parameters (**thrombocytosis, anemia, and leukocytosis**), and was therefore classified as a **moderate** inflammatory response with a **favorable** predicted course. Tapering of the steroid dose was instituted in the **sixth week** of therapy.

Because of the rich blood supply from the lingual, facial, pharyngeal, and palatine arteries, as well as collateral circulation, **ischemia (infarction) and necrosis of the tongue** is generally **rare**, which is most often **unilateral**.

GCA is by far **the most common** cause of **unilateral or bilateral tongue necrosis**.

In fact, in the **absence** of a history of physical or chemical trauma or head and neck irradiation therapy, **bilateral necrosis of the tongue** should be regarded as **pathognomonic of GCA**.

Rapid diagnosis of GCA is essential to **prevent** development of further **serious ocular or cardiovascular complications**.

題號	題目
1	Of all parameters to predict GCA according to the initial systemic inflammatory response of the patient, which one is false? **(A)** Leukocytosis **(B)** Hemoglobin **(C)** Thrombocytosis **(D)** Mean corpuscular volume
答案(D)	出處：
題號	題目
2	According to American College of Rheumatology (ACR) criteria for a diagnosis of GCA, Which one is false? **(A)** ESR higher than 70 mm/h **(B)** Age older than 50 years at disease onset **(C)** New onset of localized headache **(D)** Temporal artery tenderness or a decreased temporal artery pulse
答案(A)	出處：