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Cutaneous Manifestations of Giant Cell Arteritis

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Abstract

A 68 year old woman with advanced giant cell arteritis presented with progressive tongue, lip, and scalp necrosis. She was initially misdiagnosed and treated for oral thrush by her primary care physician and emergency department personnel. She was ultimately correctly diagnosed with giant cell arteritis after she developed a left central retinal artery occlusion, ischemic optic neuropathy, and third cranial nerve palsy. She was treated with corticosteroids and the vasculitis was arrested. She was left with permanent defects in her tongue and lip as well as blindness in the left eye.

Introduction

Giant cell arteritis is a systemic vasculitis primarily affecting the extracranial branches of the carotid arteries. It occurs in patients over 50 years old and increases in incidence with age. It is an idiopathic autoimmune reaction directed toward the walls of medium and large arteries of the head and neck. Resulting inflammation leads to vascular occlusion with secondary signs and symptoms of headache, jaw claudication, scalp tenderness, and ischemic events to the eye resulting in blindness. Cutaneous infarction of the scalp and oral mucosa are rare complications seen in advanced cases. We present a case of a delayed diagnosis of giant cell arteritis presenting with oral mucosal and scalp necrosis and visual loss.

Case Report

A 68 year old woman presented to the emergency department with pain and swelling of her lips and tongue (Figure 1). Past medical history included only chronic obstructive pulmonary disease from smoking. One week prior she was given the diagnosis of "oral thrush" by her primary care physician thought to be secondary to several intramuscular corticosteroid injections she received for worsening arthritic symptoms of the hands and knees over the preceding few months. She was given oral diflucan and discharged home. The next day she returned to the emergency department with sudden loss of vision of the left eye and progressive left upper eyelid ptosis. She was referred to an ophthalmologist who noted a visual acuity of no light perception of the left eye, left upper eyelid ptosis, and a total pupil involving left CN III palsy (Figure 2). Funduscopic examination revealed a left central retinal artery occlusion and ischemic optic neuropathy. She was immediately started on oral prednisone 1.25mg/kg/day, and presented for a temporal artery biopsy four days later. Examination was unchanged with the exception of new bilateral eschars of the temples (Figures 3,4). A left temporal artery biopsy confirmed the diagnosis of giant cell arteritis (Figure 5). One year later, the patient remained blind in the left eye with partial loss of the tongue and lips, but was otherwise stable.



Figure 2. Total pupil involving left cranial nerve III palsy.

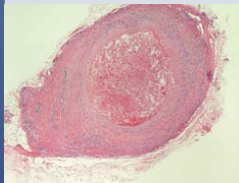


Figure 3. 68 year old woman with advanced giant cell arteritis: tongue and lip necrosis, left cranial nerve III palsy and blindness, scalp necrosis of temples.

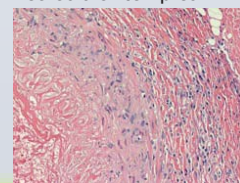


Figure 5. Temporal artery cross-section showing occlusion of vascular lumen (a. H&E; 40X) and diffuse granulomatous inflammation of arterial wall (b. H&E; 400X).



Figure 4. Bilateral scalp necrosis of temples.



Figure 1. 68 year old woman with tongue and lip necrosis.

Discussion

Scalp and oral mucosal infarction are rare complications of giant cell arteritis. Scalp necrosis involves diffuse arteriolar vasculitis of the reticular dermis. It is associated with a higher incidence of visual loss (64% vs. 36%), a higher mortality rate secondary to cerebral or coronary artery occlusion (41%), as well as tongue and lip necrosis.¹ While concomitant arteritis typically responds rapidly to corticosteroids, skin necrosis may continue for several months before clearing. The differential diagnosis of scalp ulceration in older patients includes herpes zoster, irritant contact dermatitis, ulcerated skin tumors, postirradiation ulcers, microbial infections, pyoderma gangrenosum, and giant cell arteritis.² Elderly patients presenting with dermatologic signs suspicious for giant cell arteritis should be started on systemic corticosteroids immediately to avert visual loss or other neurologic sequelae from the disease. Subsequent temporal artery biopsy should then be performed within several days of instituting steroid treatment to confirm the diagnosis.

References

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