Scalp necrosis, one of the symptoms of giant cell arteritis.

Thirasak Saeko MD,
Pawinee Rerknimitr MD MsC,
Pravit Asawanonda MD PhD.

ABSTRACT:

SAEKO T, REKNIMITR P, ASAWANONDA P. SCALP NECROSIS, ONE OF THE SYMPTOMS OF GIANT CELL ARTERITIS. THAI J DERMATOL 2017; 33: 161-166.

DIVISION OF DERMATOLOGY, SKIN AND IMMUNOLOGY RESEARCH UNIT, DEPARTMENT OF MEDICINE, FACULTY OF MEDICINE, CHULALONGKORN UNIVERSITY, BANGKOK, THAILAND.

Giant cell arteritis (GCA) is an inflammatory systemic vasculitis that affected both medium and large-sized arteries. The etiology is unknown and it occurs mostly in elderly patients. The common presentations include headaches over the temporal and the occipital regions and tenderness around the areas. Fever, malaise, jaw claudication, polymyalgia rheumatica and partial or complete loss of vision are occasionally observed. Another drastic complication is an abrupt development of scalp necrosis.

We present a case of a 77- year-old male with severe headache and visual loss for 7 days and an abrupt appearance of necrotic patches on the scalp for 5 days.

Key words: scalp necrosis, giant cell arteritis, vasculitis

บทคัดย่อ:

ธีระศักดิ์ แซ่โก้ ภาวิณี ฤกษ์นิมิตร ประวิตร อัศวานนท์ หนังศีรษะตาย, หนึ่งในอาการแสดงของโรคหลอดเลือดเทมพอรัลอักเสบ วารสารโรคผิวหนัง 2560; 33: 161-166.

หน่วยวิจัยผิวหนังและภูมิคุ้มกัน สาขาตจวิทยา ภาควิชาอายุรศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย

From: Division of Dermatology, Department of Medicine, King Chulalongkorn Memorial Hospital, Bangkok, Thailand.
Corresponding author: Thirasak Saeko MD., email: ko_thirasak@hotmail.com

โรคหลอดเลือดเทมเพอรัลอักเสบ เป็นโรคที่เกิดจากการอักเสบทั้งระบบของเส้นเลือดแดงขนาดกลาง ถึงขนาดใหญ่ ซึ่งสาเหตุยังไม่ทราบแน่ชัด พบบ่อยในผู้ป่วยสูงวัย ผู้ป่วยส่วนมากจะมาด้วยอาการแสดงคือ ปวดศีรษะรุนแรงบริเวณขมับข้างใดข้างหนึ่ง หรือทั้งสองข้างและบริเวณท้ายทอย มีอาการเจ็บขมับตามแนวเส้นเลือดบริเวณขมับ อาการไข้ อ่อนเพลีย รู้สึกไม่สบายตัว ปวดบริเวณกราม ปวดเมื่อยตามตัว และอาจจะมองเห็นภาพไม่ชัดเจน หรือมองไม่เห็นซึ่งเป็นอาการแสดงของโรคนี้ อาการอื่นที่รุนแรงคือพบผื่นที่เกิดจากผิวหนังบริเวณหนังศีรษะตาย

รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยอายุ 77 ปี ที่มาด้วยอาการปวดศีรษะรุนแรงและตามองไม่เห็นมา 7 วัน และพบผื่นผิวหนังตายที่ศีรษะอย่างเฉียบพลันมา 5 วัน

คำสำคัญ: โรคหลอดเลือดเทมพอรัลอักเสบ, หนังศรีษะตาย, เส้นเลือดอักเสบ

Introduction

Giant cell arteritis (GCA) is an inflammatory systemic vasculitis occurs primarily in the elderly. It is known as a potentially blinding condition.[1] Early diagnosis can help preventing permanent visual loss. The patients usually present with headaches over the temporal and the occipital regions, fever, malaise, jaw claudication, myalgia and finally, loss of vision. Another drastic but rare complication is an abrupt development of scalp necrosis.

Case report

A 77-year-old male presented with progressive headache for 1 month. He had bi-temporal headache without history of weakness or visual loss. He developed more severe headache and right-eye visual loss for 7 days. Five days prior to the presentation, he noticed more brownish patches on the scalp. These skin lesions had abrupt onset. Neither family history of skin abnormalities nor concurrent medications were reported. His underlying disease was hypertension. The clinical examinations were multiple brownish reticulated necrotic patches on frontal and temporal scalp (Figure A). On ophthalmologic examination, right eye ptosis with complete ophthalmoplegia, no light perception, corneal edema with cell in anterior chamber and the right fundus could not be evaluated. Laboratory analysis revealed an elevated erythrocyte sedimentation rate, ESR: 90mm/hr (0-15mm/hr), and a raised C-reactive protein level, CRP: 31mg/L (<5mg/L). Other routine laboratory tests, including complete blood cell count, liver and renal function tests and serum electrolytes were normal. Magnetic resonance imaging of the brain showed the flattened posterior aspect of the right globe associated with small anterior segment. Ocular hypotension cannot be excluded. Doppler ultrasonography of the extracranial carotid arteries showed increased thickness of intima-media complex of both common carotid arteries. Skin biopsy from the necrotic right scalp

showed ulcer covered with crusted and debris, focal necrosis of the epidermis and necrosis of the sweat glands in dermis, prominent follicular epithelium necrosis with peri-follicular neutrophillic infiltration (Figure B). Temporal artery biopsy show the intimal thickening, disruption of internal elastic lumina, foci of vascular wall necrosis, infiltrates of lymphocytes and histiocytes and focal calcification are seen (Figure C-D). His final diagnosis was giant cell arteritis with scalp necrosis.

Treatment with intravenous methylprednisolone 1 g daily for four days, then oral prednisolone at 1 mg/kg once daily with slow taper was administered followed by low-dose aspirin.[2,3,4] The vision remains stable and the scalp necrosis resolved with no further local sequelae.

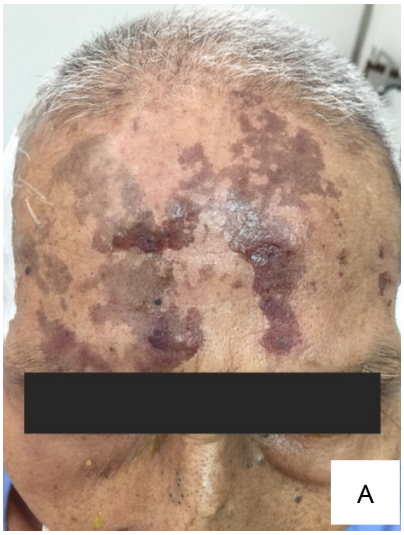


Figure A Multiple brownish reticulated necrotic patches on frontal and temporal scalp.

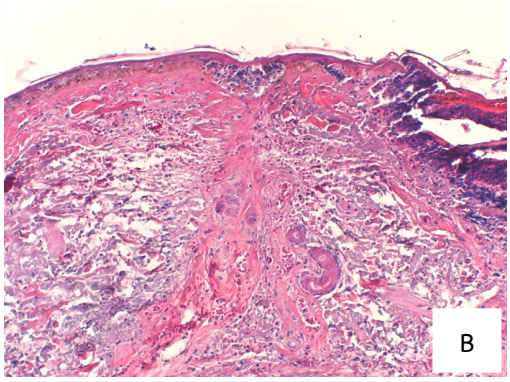
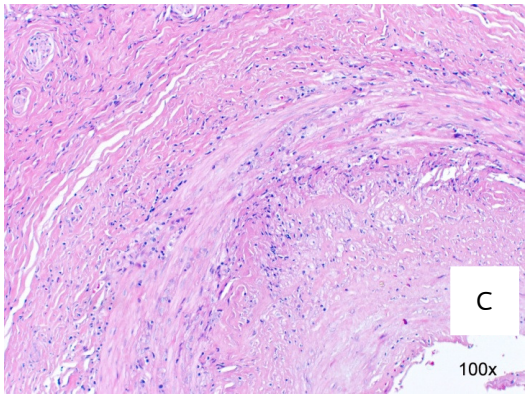


Figure B Skin biopsy from the necrotic right scalp: ulcer with crust and debris, focal epidermal and sweat glands necrosis with neutrophils infiltration (H&E 100x).



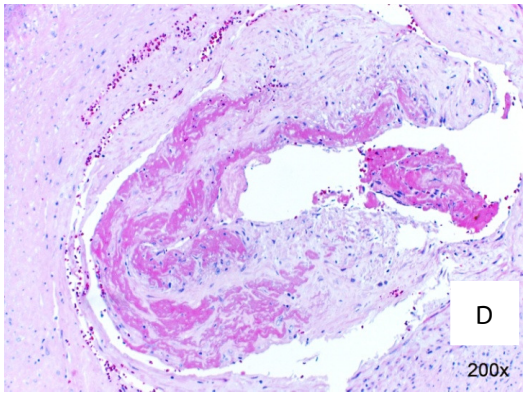


Figure C, D Temporal artery biopsy: inflammatory cells mainly lymphocytes and neutrophils infiltration in the vessel wall (H&E 100x, 200x).

โฆษณา 4 สี

Discussion

The clinical and laboratory findings of our patient meet at least 3 of the 5 American College of Rheumatology diagnostic criteria for GCA[5] i.e. age at disease onset of 50 years or older, a new onset of headache, an elevated ESR and a positive artery biopsy result. He was diagnosed with giant cell arteritis with scalp necrosis, a rare feature associated with this condition.

GCA is an inflammatory systemic vasculitis that affected both medium and large-sized arteries.[1] The etiology is unknown. It occurs mostly in the elderly patients and women are slightly more affected (age range: 54-90 years; female-to-male ratio: 1.62:1).[6] Most of the patients come with headaches over the temporal and the occipital regions and tenderness around the areas. Fever, malaise, jaw claudication, polymyalgia rheumatica and partial or complete loss of vision are commonly observed. Another drastic complication is an abrupt development of scalp necrosis. Patients with giant cell arteritis and associated scalp necrosis have a higher incidence of visual loss.[1,6,7] Early diagnosis and rapid start of treatment might prevent scalp necrosis and associated increase of mortality.

The histology of giant cell arteritis illustrated the infiltration of inflammatory cells including lymphocytes, macrophages and giant cells in an arterial wall. The inflammatory cells were normally found in group between the adventitia and the muscularis media. Moreover, an inflammation of the vascular wall was found as skip lesions. About 50% of the cases multinucleated giant cells are detected but this is not a constant feature.[8] The biopsy result could aid the clinical diagnosis, but the definite diagnosis could be made without a positive result as the biopsy was probably taken from a non-inflamed specimen.

Nowadays, there is a standard recommendation for treatment of giant cell arteritis. An early treatment with corticosteroid is a mainstay at an initial dosage of prednisolone 1 mg/kg body weight per day.[9] Physicians should prompt their treatment without waiting for an imaging and a biopsy result. The response of treatment was assessed by clinical symptoms and laboratory results (ESR and CRP). Dosage adjustment could be done if there were any worsening signs.[10] Even though, corticosteroid is a mainstay treatment, it could not reverse the visual impairment.[11] Additionally, physicians should beware of the side effects of long-term corticosteroid usage.

In conclusion, our patient presented with severe headache, visual loss and abrupt appearance of scalp necrosis. A biopsy from temporal artery showed an inflammation in the vascular wall. A laboratory resulted showed

elevated ESR and CRP. An early treatment with corticosteroid for 4 days was given and our patient's status was improved. Unfortunately, the visual impairment could not be reversed. Therefore, giant cell arteritis (GCA) is a condition that needs an early diagnosis and prompt treatment to prevent any other severe complications.

กิตติกรรมประกาศ

รายงานผู้ป่วยฉบับนี้ สำเร็จลงได้ด้วยดี เนื่องจากได้รับความกรุณาจาก ผู้ช่วยศาสตราจารย์แพทย์หญิงภาวิณี ฤกษ์นิมิตร และศาสตราจารย์นายแพทย์ ประวิตร อัศวานนท์ ที่กรุณาให้คำแนะนำ ปรึกษาแนวคิดตลอดจนปรับปรุงแก้ไขข้อบกพร่องต่างๆ ด้วยความเอาใจใส่อย่างดียิ่ง มาโดยตลอดจนเสร็จสมบูรณ์ ผู้เขียนขอกราบขอบพระคุณเป็นอย่างสูงไว้ ณ ที่นี้

อนึ่ง ผู้เขียนหวังว่า รายงานผู้ป่วยฉบับนี้จะเป็นประโยชน์ต่อผู้ที่สนใจและเป็นแนวทางในการหาความรู้เพิ่มเติมต่อไปในอนาคต หากมีข้อผิดพลาดประการใดต้องขออภัยมา ณ ที่นี้ด้วย

Reference

1. Palmer VE, Young-Zvandasara T, Vusirikala B. Forehead necrosis, one of the many facades of giant cell arteritis. BMJ Case Rep 2015; 172:65-6.
2. Bhatti MT, Tabandeh H. Giant cell arteritis: diagnosis and management. Curr Opin Ophthalmol 2001; 12:393-9.
3. Dasgupta B, Borg FA, Hassan N, et al. BSR and BHPR guidelines for the management of giant cell arteritis. Rheumatology (Oxford) 2010; 49:1594-7.
4. Nesher G, Berkun Y, Mates M, Baras M, Rubinow A, Sonnenblick M. Low-dose aspirin and prevention of cranial ischemic complications in giant cell arteritis. Arthritis Rheum 2004; 50:1332-7.
5. Hunder GG, Bloch DA, Michel BA, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 1990; 33:1122-8.
6. Tsianakas A, Ehrchen JM, Presser D, et al. Scalp necrosis in giant cell arteritis: case report and review of the relevance of this cutaneous sign of large-vessel vasculitis. J Am Acad Dermatol 2009; 61:701-6.
7. Currey J. Scalp necrosis in giant cell arteritis and review of the literature. Br J Rheumatol 1997; 36:814-6.
8. Poller DN, van Wyk Q, Jeffrey MJ. The importance of skip lesions in temporal arteritis. J Clin Pathol 2000; 53:137-9.
9. Mukhtyar C, Guillevin L, Cid MC, et al. EULAR recommendations for the management of large vessel vasculitis. Ann Rheum Dis 2009; 68:318-23.
10. Vaith P, Warnatz K. [Clinical and serological findings of giant-cell arteritis]. Z Rheumatol 2009; 68:124-31.
11. Proven A, Gabriel SE, Orces C, O'Fallon WM, Hunder GG. Glucocorticoid therapy in giant cell arteritis: duration and adverse outcomes. Arthritis Rheum 2003; 49:703-8.
12. Aui-Aree N, Tungsinmunkong K, Hirunpat S, Ratanasukon M, Wangsupadilok B. A variety of atypical manifestations in giant cell arteritis. J Med Assoc Thai 2010; 93:629-32.