Case Report

A Variety of Atypical Manifestations in Giant Cell Arteritis

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Giant cell arteritis (GCA) is a chronic granulomatous vasculitis of large and medium size vessels in the elderly. A new-onset headache is the most frequent symptom. An anterior ischemic optic neuropathy (AION) is one of the most common causes of permanent visual loss. There are four cases with unusual presentation of giant cell arteritis, scalp abscess, prolonged transient monocular visual loss (TMVL), bilateral central retinal artery occlusion (CRAO), and chronic ear pain. All patients had pathologically proven giant cell arteritis, and three of them progressed to blindness in the end. Scalp abscess is a rare sign in GCA. Delay in diagnosis because confusion of the abscess after scalp ischemia with other cutaneous lesions may result in death. TMVL is the forewarning symptom of AION or CRAO in GCA. Early recognition of TMVL is important to make early diagnosis of GCA to prevent blindness. Spontaneous ear pain is extremely rare, and reports have documented delay in diagnosis of GCA resulting in irreversible blindness.

Keywords: *Bilateral central retinal artery occlusion, Chronic ear pain, Giant cell arteritis, Scalp abscess, Temporal arteritis*

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Giant cell arteritis (GCA) is a vasculitis of medium to large size of arteries. The condition is most prevalent in the elderly population. Cranial branches are usually involved and cause the consequent symptoms along the affected vessels. The common related symptoms are temple headache, jaw claudication, fever, weight loss, and malaise. The common presenting signs are decreased temporal artery pulsations, artery tenderness, and sudden visual loss from anterior ischemic optic neuropathy (AION) or central retinal artery occlusion (CRAO).

Case Report

Case 1: Progressive bilateral visual loss with scalp abscess

A 76-year-old Thai male presented to Prince of Songkla Ophthalmology Clinic following bilateral loss of vision, progressing over the previous two weeks. The patient reported his blindness had been preceded by a biparietal headache of three months duration. The vision loss progressed over one month beginning initially in the left eye, followed by the right eye, five days later. Three weeks prior to admission the patient developed a painful skin lesion on the right side of his scalp, and had noted some jaw claudication and voice changes. He had no other medical history aside from hypertension.

His eye examination showed no light perception in both eyes. His pupils were 5 mm in diameter without response to light. His right eye had Descemet's folds and occasional cells in the anterior chamber. The optic nerve heads were bilaterally pale. There was no tenderness on either temporal artery, but he had severe pain around his right scalp abscess (Fig. 1).

His erythrocyte sedimentation rate (ESR) was 126 mm/hr and C-reactive protein (CRP) was 4.8 mg/L. Magnetic Resonance Imaging (MRI) of his orbits and brain revealed enlargement and enhancement of both optic nerves. Magnetic Resonance Angiography (MRA) of the brain revealed arteriosclerotic changes in the intracranial part of the internal carotid artery.

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A bilateral temporal artery biopsy was performed and confirmed giant cell arteritis in both vessels. Pus from his abscess was cultured positive for *Staphylococcus aureus*.

He was treated with oral cloxacillin and oral prednisolone with resolution of his scalp pain and abscess over 1 week. The Descemet's folds and anterior chamber cells disappeared after one week of treatment. The visual acuity has not improved at two years follow-up.

Case 2: Transient monocular visual loss (TMVL)

A 70-year-old Thai female experienced intermittent blurred vision in her right eye for one month. It occurred 2-3 times a week and lasted 20 minutes to three hours. The blurriness was grayish and had never gone to dark. Her change in vision was associated with persistent bilateral parietal pain, especially when combing her hair. She was not able to chew solid food due to jaw claudication, and over the last month lost two kgs due to loss of appetite. She had no other vascular disease.

Physical examination revealed visual acuity of 20/40 in the right and 20/50 in the left eye. No relative afferent papillary defect was detected. Color test and intraocular pressure were normal in both eyes. Mild nuclear sclerosis was noted in both eyes. Both optic nerves and retina were normal. The temporal arteries were engorged, pulseless, and tender bilaterally.

ESR was 103 mm/hr, CRP was 4.8 mg/L. Temporal artery biopsy on the right side was positive for giant cell arteritis (Fig. 2). The patient was placed on oral prednisolone 1 mg/kg/day, which was subsequently tapered. The transient monocular visual loss, parietal pain, and jaw claudication resolved after first week of treatment. At 18 months follow-up she had no recurrence of symptoms.

Case 3: Bilateral central retinal artery occlusion (CRAO)

A 79-year-old Chinese male developed sequentially bilateral blindness. His initial symptom was transient monocular vision loss in the right eye, which lasted 20 minutes.

This occurred 2-3 times/day and progressed to blindness within one week. Three days prior to presentation, his left eye was also affected. He did not complain of headache or fever, but he felt dizzy and malaise. He reported anorexia of three months duration, and had lost five kgs. He had to eat the rice congee for two weeks due to jaw claudication.

His eye examination showed no light perception in both eyes. Fundoscopy revealed pallor retina with cherry red spots in both eyes (Fig. 3).

His ESR was 103 mm/hr and CRP was 4.8 mg/L. The temporal arteries were engorged, but not tender. Right temporal artery biopsy was positive for giant cell arteritis. He was treated with methylprednisolone intravenously for three days of admission, followed by

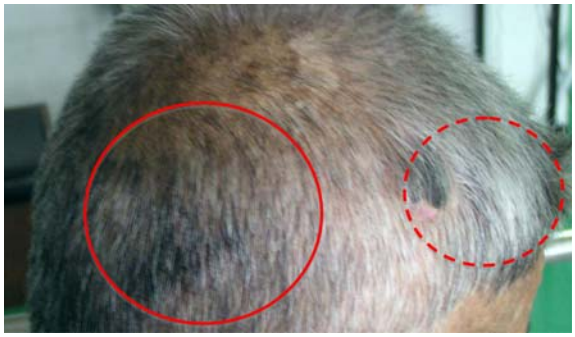

Fig. 1 Right scalp ischemia ( ) with abscess ( ) in case 1

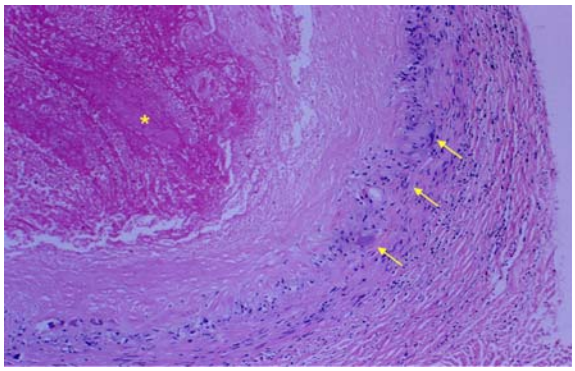

Fig. 2 Granulomatous inflammation including giant cell involved entire the circumferential and thickness of the arterial wall (→) with thrombosis (*) (H&E stain x40)

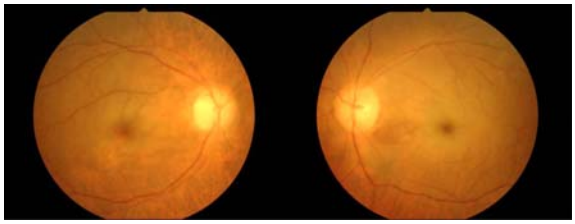

Fig. 3 Cherry red spots in bilateral central retinal arterial occlusion in case 3

oral prednisolone. At three months follow-up, though jaw claudication and anorexia had resolved, the vision had not improved.

Case 4: Persistent ear pain followed by sudden visual loss

An 83-year-old Thai male presented with a three-month history of left ear pain. The pain was aggravated by moving the left pinna, but was not worsened by chewing. He had also experienced bilateral frontal headache periodically for approximately three months. The patient reported that during the headache he was able to palpate vessels "like cords" on his scalp. He was seen by an otolaryngologsit, who treated him for otitis externa and noted no abnormalities on follow-up, by a dentist who treated his dental caries, and by a neurologist, and he was eventually admitted to the hospital for work up. During the hospital stay, he developed acute onset blindness in the left eye. His visual acuity was 20/30 in the right and no light perception in the left eye. The left optic nerve showed marked swelling. Arteritic ischemic optic neuropathy (AION) was diagnosed. He also had a low-grade fever, anorexia, jaw claudication, and had lost five kgs in two months.

ESR was 88 mm/hr and CRP was 19.2 mg/L. The right temporal artery biopsy was performed with a positive result. He was treated with methyl-prednisolone intravenously 20 mg/kg/day for three days followed by prednisolone orally. Though, most of his symptoms had resolved, the vision in the left eye remains no light perception (NLP) at six months follow-up.

Discussion

Giant cell arteritis is rarely diagnosed in Asia, as the incidence is 1.7 per 100,000 population[1]. These are the first four elderly patients diagnosed as GCA in three years since the start of neuro-ophthalmology clinic. No GCA has been diagnosed before since our hospital started 36 years ago.

In case 1, sequentially progressive bilateral visual loss over one month until blindness is unusual[2]. Typically, the affected patients experience sudden onset of visual loss or altitudinal field defect. Moreover, the signs of anterior segment ischemia, Descemet's fold and anterior chamber cells, which are the signs of anterior segment ischemia caused by ophthalmic artery and anterior ciliary involvement, are rarely reported[3-5]. Scalp necrosis leading to superficial scalp abscess is rare and is associated with a high risk of developing irreversible visual loss. The subepicranial abscess following extensive scalp necrosis was reported by Smitz S and Damme H in 2004[6], but this is the first report of superficial abscess with scalp necrosis.

In case 2, TMVL is one of the common forewarning symptoms of permanent visual loss in GCA. Compromised optic nerve circulation is more common cause of TMVL than affected retinal arterial flow in GCA because the incidence of AION is much higher than CRAO[3,7]. TMVL was found approximately 16-46% in GCA with ocular involvement, and more than half of patients develop permanent visual loss. The duration of TMVL usually lasts 2-10 minutes, but rarely persists for hours, as it did in the presented patient. The prolonged TMVL may be associated with chronic ocular hypoperfusion as the result of vascular lumen narrowing.

Case 3, a Chinese man who presented with sequentially bilateral CRAO is also rare in GCA. Bilateral CRAO was reported as the first case in a Chinese patient by Kwok AK et al in 1998 and three cases were found in the series of Hayreh SS et al in the same year[3,8]. Unilateral branch or central retinal artery occlusion can be found in about 10% of GCA patients. The simultaneous or sequentially bilateral CRAO has been reported in other conditions such as leukemia, post head injury, thrombotic thrombocytopenic purpura, and Wegener's granulomatosis. If this patient had presented to the hospital during his episodes of transient visual loss, the bilateral blindness might have been prevented.

In case 4, spontaneous chronic ear pain was the presenting symptom of GCA. A similar presenting complaint was reported in only one case by Coppetto JR in 1984, but that case had severe pain with lightheadedness, which lasted over one week. The ear pain was not related to jaw claudication as in the presented series. He postulated that the spontaneous ear pain might be an incomplete form of the lateral medullary syndrome, in which the arteritis affected the central nervous system[9].

In summary, the delay in diagnosis of GCA can cause several serious consequences, including irreversible visual loss. In addition, many rare ocular manifestations were seen in the case of delayed diagnosis. When an elderly present with forewarning symptoms such as TMVL or spontaneous ear pain, GCA should be considered even in Asian ethnics. The early diagnosis of giant cell arteritis and the prompt treatment are very important to prevent permanent blindness.

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References

1. Kobayashi S, Yano T, Matsumoto Y, Numano F, Nakajima N, Yasuda K, et al. Clinical and epidemiologic analysis of giant cell (temporal) arteritis from a nationwide survey in 1998 in Japan: the first government-supported nationwide survey. Arthritis Rheum 2003; 49: 594-8.
2. Galor A, Lee MS. Slowly progressive vision loss in giant cell arteritis. Arch Ophthalmol 2006; 124: 416-8.
3. Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of giant cell arteritis. Am J Ophthalmol 1998; 125: 509-20.
4. McKillop E, Tejwani D, Weir C, Jay J. Anterior segment ischaemia with giant cell arteritis. Can J Ophthalmol 2006; 41: 201-3.
5. Hamed LM, Guy JR, Moster ML, Bosley T. Giant cell arteritis in the ocular ischemic syndrome. Am J Ophthalmol 1992; 113: 702-5.
6. Smitz S, Van Damme H. Extensive scalp necrosis and subepicranial abscess in a patient with giant cell arteritis. J Am Geriatr Soc 2004; 52: 165-6.
7. Biousse V, Trobe JD. Transient monocular visual loss. Am J Ophthalmol 2005; 140: 717-21.
8. Kwok AK, Lam DS, Liew CT. Bilateral arteritic central retinal artery occlusion in a Chinese patient. Aust N Z J Ophthalmol 1998; 26: 175-6.
9. Coppeto JR. Spontaneous ear pain as the initial presenting manifestation of giant cell arteritis. J Neurol Neurosurg Psychiatry 1984; 47: 1059.

ความหลากหลายของลักษณะทางคลินิกที่พบได้ไม่บ่อยในผู้ป่วย giant cell arteritis

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Giant cell arteritis (GCA) เป็นการอักเสบเรื้อรังของหลอดเลือดขนาดกลางถึงขนาดใหญ่ มักพบในผู้ป่วยสูงอายุอาการที่พบบ่อยที่สุดเป็นอาการปวดขมับที่เพิ่งเกิดขึ้นใหม่ และเส้นประสาทตาส่วนหน้าขาดเลือดเป็นอาการแสดงที่ทำให้สูญเสียการมองเห็นที่พบได้บ่อย มีผู้ป่วย GCA ที่มีลักษณะทางคลินิกที่พบได้ไม่บ่อยจำนวนสี่รายได้แก่ ฝีที่หนังศีรษะ ตามัวข้างเดียวชั่วขณะที่มีระยะนานกว่าปกติ หลอดเลือดแดง ในจอตาอุดตันทั้งสองข้าง และรายสุดท้ายมีอาการปวดหูเรื้อรัง การตรวจทางพยาธิวิทยายืนยันว่า หลอดเลือดที่ขมับของผู้ป่วยทั้งสี่ราย เป็นการอักเสบแบบ GCA และมีผู้ป่วยสามรายตาบอดในที่สุด ฝีที่หนังศีรษะเป็นอาการแสดงที่พบน้อยมาก การที่คิดว่าเป็นจากสาเหตุอื่นและวินิจฉัยว่าเป็นจาก GCA ช้าอาจทำให้ผู้ป่วยเสียชีวิตได้ ส่วนผู้ป่วย ที่มีอาการตามัวชั่วขณะ อาจเป็นอาการนำในโรคเส้นประสาทตาส่วนหน้าขาดเลือด หรือ หลอดเลือดแดงในจอตาอุดตันที่จะพบใน GCA และสุดท้ายอาการปวดหูที่ตรวจไม่พบความผิดปกติในช่องหูเป็นอาการที่พบได้น้อยมาก มีรายงานที่ผ่านมาพบว่าอาการนี้ทำให้การวินิจฉัยช้าไปและส่งผลให้ผู้ป่วยตาบอดในที่สุด