

Bilateral scalp necrosis as a rare but devastating complication of giant cell arteritis

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Abstract Giant cell arteritis (GCA) is a medium to large vessel vasculitis of unknown aetiology. Commonly, it affects the temporal arteries and is known as temporal arteritis. It has an association with polymyalgia rheumatica and can result in severe complications such as loss of vision and rarely scalp necrosis. There are approximately 100 cases of scalp necrosis in patients with GCA published in the literature to date. We report a case of a man who presented with a 4-week history of bilateral scalp necrosis associated with headache, jaw claudication, temporal artery tenderness, and raised inflammatory markers. He did not have any visual loss. A diagnosis of GCA was made and he was started on high-dose steroids immediately. The scalp lesions did improve and his symptoms resolved without any visual loss but, sadly he died due to severe sepsis. This case report is important as it describes a rare but severe complication of a common large vessel vasculitis seen by both primary care physicians and rheumatologists. Prompt recognition and early treatment by the physician are crucial to the patient to prevent visual loss or a fatal stroke. It also highlights complications associated with steroids which are the mainstay of treatment for this condition.

Keywords Giant cell arteritis < rheumatic diseases · Vasculitis < rheumatic diseases

Case description

A 77-year-old Caucasian male with a background history of hypertension and atrial fibrillation presented with a 4-week history of a generalised headache, jaw claudication and a large

necrotic area over his scalp (Figs 1, 2, and 3). He denied any visual loss or systemic symptoms.

He had initially presented to his GP with a black painful nodule near the superior temporal line which was thought to be a ruptured superficial blood vessel. This had developed into a large, painful and bilateral scalp necrotic ulcer on admission to hospital.

Examination also revealed scalp tenderness and impalpable temporal arteries.

Blood tests showed an elevated C reactive protein (CRP) of 66 mg/L and erythrocyte sedimentation rate (ESR) of 74 mm/h.

Temporal biopsy was not deemed necessary for several reasons. Firstly, the diagnosis seemed obvious from a clinical point of view. Also, given the difficulty in palpating a temporal artery due to such advanced disease this seemed like a futile exercise.

Other differentials considered were skin necrosis due to warfarin therapy. However, given the fact that he had been on warfarin for the last 5 years this seemed unlikely. His INR was also within range.

Treatment with high dose steroids in the form of oral Prednisolone 60 mg once daily was initiated immediately with proton pump inhibitor (PPI) cover and bone protection. He was also referred to the plastic surgeons who wanted the lesions to heal before any potential skin grafting.

Once initiated on treatment, he was allowed home with regular outpatient follow-up and monitoring of ESR.

From a GCA point of view, he was doing well with an improvement in both his clinical symptoms and inflammatory markers. Steroids were being reduced slowly. He did not report any visual problems and his scalp ulceration was healing.

Sadly, he continued to have several admissions to hospital with urinary sepsis and eventually died due to its complications.

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Fig. 1 Image of bilateral scalp necrosis

Discussion

Cooke et al. described the first ever case of scalp necrosis due to GCA in 1947. Since then, a further 100 or so cases have been reported in the literature [1].

GCA is a chronic granulomatous vasculitis that affects any vessel but has a predilection for the extra-cranial branches of the aortic arch especially the temporal arteries, hence its other name temporal arteritis [1, 2]. The aetiology of giant cell arteritis is not completely understood but its prevalence increases with age.

Commonly, it presents with headache, scalp tenderness, jaw claudication and visual loss. In 50 % of all cases, it co-exists with polymyalgia rheumatica [3].

It can also cause tongue necrosis, stroke and myocardial infarction [3, 4].



Fig. 2 Image of bilateral scalp necrosis



Fig. 3 Image of bilateral scalp necrosis

Tenderness over a prominently dilated tortuous, thickened superficial artery and absent pulsation are the usual characteristic hallmarks of this condition [4–6].

Scalp necrosis is a very rare complication of giant cell arteritis and is usually co-existent with visual loss [1] and indicates advanced disease. Early diagnosis and treatment of this cutaneous complication are key to preventing other devastating complications [6].

Temporal artery biopsy is considered to be the gold standard for diagnosis. In around 5–10 % of cases, the temporal artery biopsy gives false negative results usually if the diseased section is not biopsied or the length of biopsy is inadequate [2–5].

Several other causes of scalp necrosis in older patients need to be considered in the differential, such as herpes zoster, irritant contact dermatitis, ulcerated skin tumours, post-irradiation ulcers, microbial infections and pyoderma gangrenosum [2].

GCA is very sensitive to steroid therapy which is the usual mainstay of treatment.

Our case has shown that prompt diagnosis and treatment prevented progression of the scalp necrosis and other catastrophic complications such as blindness and stroke. Sadly, he died from sepsis probably as a result of steroid-related immunosuppression.

Key learning points and lessons learnt

1. Scalp necrosis is a rare but serious complication of GCA which usually precedes or occurs alongside visual loss, and should be a warning skin to the clinician.
2. Early diagnosis and treatment in the form of steroids is crucial to the patient to prevent further severe complication.
3. High-dose steroids can cause severe complications.

4. Multi Disciplinary Team (MDT) approach is important including plastic surgeons and GPs.

Conflict of interest None.

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