CASE REPORT

Forehead necrosis, one of the many facades of giant cell arteritis

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SUMMARY

Giant cell arteritis (GCA) is known to be a potentially blinding condition. Swift diagnosis can aid in preventing permanent visual loss and, more importantly, protect the contralateral eye. Classical symptoms include jaw claudication, myalgia and new-onset headache. We present two cases of GCA with scalp necrosis, a rare feature associated with this condition. In the first case, forehead necrosis preceded the visual symptoms by 2 days. In the second case it was noted a few weeks after the patient presented with profound unilateral loss of vision. Scalp necrosis is an important sign that should prompt those approached by these patients to consider GCA.

BACKGROUND

Giant cell arteritis (GCA) is a well-documented cause of visual loss. Prompt diagnosis is essential to reduce the prevalence of permanent, sometimes bilateral, loss of vision. It is not uncommon for scalp necrosis to precede visual loss in GCA. Rarer signs may present first to different specialties, therefore awareness of such signs is important.

CASE PRESENTATION**Case 1**

An 82-year-old man presented with bilateral visual loss of 3 days duration. Visual acuity was no perception of light in the right eye and perception of light in the left eye. Optic discs were pale and swollen bilaterally. The patient was noted to have right-sided forehead necrosis (figure 1), which had been present for 2 days prior to the visual loss.

Case 2

A 76-year-old woman with early dementia presented with a history of loss of vision in her left eye for approximately 2 weeks. This was associated with a right-sided temporal headache, and loss of appetite and weight, though no history of jaw claudication. Visual acuity was perception of light in the left eye and 0.6 logMAR in the right. The patient had an ocular history of right acute angle-closure glaucoma, with patent bilateral laser peripheral iridotomies and bilateral quiet pseudophakia. On examination, there was a left relative afferent pupillary defect and posterior capsular opacification of both intraocular lenses. Asteroid hyalosis was present in the left vitreous. The right optic cup-disc ratio was 0.9 with inferior nerve fibre loss and the left disc was pale. The patient was also noted to have scalp necrosis (figures 2 and 3) over both temporal areas, right more than the left.

Figure 1 Male patient—scalp necrosis, right side.

INVESTIGATIONS

In case 1, erythrocyte sedimentation rate (ESR) was 98 mm/h and C reactive protein (CRP) was 112 mg/L. A temporal artery biopsy was performed on the left side confirming GCA, with histology showing multinucleated giant cells, and inflammatory infiltrate primarily composed of CD4+ T cells and activated macrophages.

Figure 2 Female patient—scalp necrosis, right side.

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Figure 3 Female patient—scalp necrosis, left side.

Case 2 had a CRP of 31 mg/L and an ESR of 95 mm/h. A subsequent temporal artery biopsy was positive for GCA with presence of giant cells.

DIFFERENTIAL DIAGNOSIS

In these two cases GCA should have been immediately suspected.

Other possibilities, although unlikely, for this presentation prior to visual loss are:

1. Herpes zoster
2. Irritant contact dermatitis
3. Ulcerated skin tumours
4. Postirradiation ulcers
5. Microbial infections
6. Pyoderma gangrenosum
7. Factitial dermatitis

TREATMENT

In both cases, treatment was started immediately. In case 1, the patient was started on intravenous methylprednisolone 500 mg for 3 days, then oral prednisolone at 1 mg/kg once daily, which was slowly tapered down, guided by ESR and CRP response. Case 2 was started on oral prednisolone at 1 mg/kg once daily and this has been slowly tapered down, again guided by ESR and CRP.

OUTCOME AND FOLLOW-UP

Steroids were tapered down slowly in both cases, guided by ESR and CRP. The vision in the contralateral unaffected eye in case 2 remains stable. Cognitive impairment in case 2 has worsened significantly, her carers feel this has greatly accelerated in the

short-time preceding her visual loss. Steroid use is likely to be for a duration of greater than 1 year, with the patients' general practitioner or rheumatologists. The scalp necrosis resolved in both patients with no further local sequelae.

DISCUSSION

GCA is an inflammatory systemic vasculitis of unknown aetiology affecting medium-sized and large-sized arteries. It is the commonest vasculitis in adults and is commoner in the elderly. The superficial temporal artery is the commonest affected artery. It should be apparent that the ophthalmic, occipital, vertebral, posterior ciliary, proximal vertebral, aorta, carotids, subclavian and iliac arteries can be affected. Cognitive changes are also possible in GCA, due to cerebral hypoperfusion. Amid the classical symptoms of jaw claudication, myalgia, scalp tenderness and new-onset headache, include less common symptoms and signs such as scalp necrosis.

A major review of GCA reports scalp necrosis as a very rare presentation, with one review discovering 24 cases reported from 1946 to 1997 and another found 78 cases published from 1946 to 2007.^{1–3} One paper reports a case of tongue necrosis at presentation, secondary to GCA.⁴

A literature review indicated those with GCA and associated scalp necrosis have a higher incidence of visual loss (32% compared with 20% rate of visual disturbance in those without scalp necrosis).³ This paper also suggests that scalp necrosis after the start of steroid therapy may indicate inadequate steroid dosage. Complete resolution or improvement of scalp necrosis is observed in 75% of cases.²

Learning points

- ▶ Scalp necrosis can present before ocular symptoms in giant cell arteritis (GCA).
- ▶ It remains essential to maintain a high index of suspicion of GCA to help prevent permanent bilateral blindness.
- ▶ Prompt treatment with high-dose steroids is recommended to prevent further devastating visual loss and other neurological symptoms such as stroke.
- ▶ Scalp necrosis is thought to represent a more aggressive disease process and is associated with worse prognosis and higher mortality.

Competing interests None declared.

Patient consent Obtained.

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