Case report

Scalp ulceration: a rare manifestation of giant cell arteritis

Louise McDonald,¹ Gavin Baker,² Olga Kerr¹¹Department of Dermatology, Belfast Health and Social Care Trust, Belfast, UK²Department of Histopathology, Belfast Health and Social Care Trust, Belfast, UK

Correspondence to

Dr Louise McDonald;
louisemcdonald@doctors.org.uk

Accepted 10 October 2019

SUMMARY

An 81-year-old woman presented with an enlarging, tender ulcer on her scalp over an 8-week period, attributing it to a prior graze with garden shears. C-reactive protein and erythrocyte sedimentation rate were elevated at 87.7 mg/L and 112 mm/hour, respectively. Incisional biopsies demonstrated ulceration and full thickness necrosis with no evidence of malignancy. Vasculitis was suggested as a likely cause of such extensive necrosis and subsequent temporal artery biopsy findings were consistent with giant cell arteritis. The patient was initially treated with high-dose oral prednisolone and achieved complete healing of the scalp necrosis within 12 months, with a gradual down-titration of steroid therapy thereafter. Scalp necrosis is a rare, potentially life-threatening complication of giant cell arteritis. This case highlights the importance of considering scalp necrosis as a manifestation of giant cell arteritis when assessing scalp ulceration. Prompt diagnosis and treatment can prevent significant morbidity and potential mortality.

BACKGROUND

This case highlights a very rare but important clinical manifestation of giant cell arteritis. Scalp ulceration and necrosis may be an isolated presenting feature and it is crucial to consider underlying giant cell arteritis in the differential diagnosis to avoid significant morbidity and potentially life-threatening complications.

CASE PRESENTATION

An 81-year-old woman presented with an 8-week history of a rapidly enlarging, exquisitely tender ulcer on her scalp. She had no prior history of skin malignancy and had herself attributed the ulcer to a possible slight graze with garden shears 1 month prior. On systemic questioning, she reported an episode of ipsilateral jaw claudication 2 months prior to the ulcer but denied any visual loss, headache, joint pains or malaise. Physical examination revealed an extensive, ulcerated and eroded area on her scalp extending from the anterior hair margin to the parieto-occipital junction. The area was not undermined and there was no associated clinical lymphadenopathy (figure 1). The remainder of her skin examination did not reveal any abnormality.

INVESTIGATIONS

Blood tests on initial presentation revealed a normal white cell count but both C-reactive protein (CRP)

and erythrocyte sedimentation rate (ESR) were elevated at 87.7 mg/L and 112 mm/hour, respectively. Swabs of the ulcerated area were negative on microbiology and virology testing.

An incisional skin biopsy was taken from both the anterior and superior pole of the lesion. Both biopsies demonstrated an inflammatory cell infiltrate with ulceration, vascularisation and full thickness necrosis extending to subcutaneous fat with no evidence of malignant change. Although vasculitis was not seen it was suggested as a likely cause of the extensive necrosis. A subsequent temporal artery biopsy demonstrated transmural inflammation, disruption of the elastic lamina and multinucleated giant cells consistent with giant cell arteritis. A cross-sectional image of the artery is shown in figure 2, showing giant cells, lymphocytic inflammation and complete obliteration of the lumen and the giant cells are seen at higher magnification in figure 3.

DIFFERENTIAL DIAGNOSIS

The differential for scalp ulceration presenting in an elderly person is broad. Initially a cutaneous malignancy such as a squamous cell carcinoma or premalignant lesion was felt to be most likely given the rapid evolution, friability and erosion. The more rare cutaneous angiosarcoma was also considered given its predilection for skin and superficial soft tissue areas but histopathology demonstrated no evidence of malignancy. Infective aetiologies such as herpes zoster, bacterial and fungal infection were excluded following negative microbiology and virology testing. The phenomenon of erosive pustular dermatosis describes exudate overlying eroded plaques or nodules on the scalp and can present as 'scalp ulceration'. Its cause is unknown and it may be preceded by trauma, inflammation, infection or malignancy but was not felt to fit clinically in this case. Pyoderma gangrenosum should always be considered in the context of rapidly progressive ulceration but the absence of an undermined violaceous edge did not fit with clinical appearances. Allergic contact dermatitis can cause scalp inflammation but history was not suggestive in this case. The history of jaw claudication and elevated ESR prompted concern regarding giant cell arteritis and the full thickness necrosis on skin biopsy and temporal artery biopsy findings confirmed the diagnosis.

TREATMENT

The patient was treated initially with high dose oral prednisolone commencing at a dose of 60 mg/

© BMJ Publishing Group Limited 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: McDonald L, Baker G, Kerr O. *BMJ Case Rep* 2019;**12**:e230795. doi:10.1136/bcr-2019-230795

Figure 1 Ulcerated lesion on left side of scalp with irregular border.

day (1 mg/kg/day) in line with British Society of Rheumatology (BSR) and British Health Professionals in Rheumatology (BHPR) guidelines for management of giant cell arteritis,¹ with twice weekly wound debridement and dressing applications. She also required regular analgesia in the initial months post-presentation as the ulcerated area was very tender.

OUTCOME AND FOLLOW-UP

The patient was followed up by both Dermatology and Rheumatology, with the rate of down-titration of oral prednisolone guided by scalp healing, clinical symptoms review and monitoring of inflammatory markers. The scalp ulceration re-epithelialised over a period of 6 months (figure 4) with complete healing achieved within 12 months. Throughout this period, the dose of oral prednisolone was continually reduced in keeping with BSR guidelines. Following normalisation of the ESR and CRP within the first month and progressive ulcer healing, oral prednisolone was successfully down-titrated within 9 months to a dose of 15 mg/day. Subsequent gradual tapering and maintenance of remission allowed successful cessation of steroid therapy after a total of 24 months. The scalp area at 24-month review remained healed (figure 5) and she has not had any disease recurrence.

Cognizant of the patient's age, gender and keen to mitigate against the impact on bone density of prolonged glucocorticoid therapy, the Rheumatology team had prescribed prophylactic

Figure 2 Image of cross section of artery showing giant cells (GC), lymphocytic inflammation (L) and complete obliteration of lumen (LU).

Figure 3 Image demonstrating giant cells at higher magnification.

bisphosphonate, calcium and vitamin D at the time of steroid initiation. A dual-energy X-ray absorptiometry (DEXA) scan undertaken as part of steroid monitoring in her second year of therapy, however, demonstrated osteoporosis. Despite steroid-weaning and cessation, she subsequently sustained a fractured neck of femur after an accidental fall shortly after her 2-year follow-up. Although a traumatic mechanism of injury may have explained the fracture, it is important to consider the possible added burden of her prolonged steroid therapy as a contributing factor in developing osteoporosis and fracture risk. Her recovery post hemi-arthroplasty was complicated by postoperative delirium and, despite prolonged in-patient rehabilitation, investigation and physiotherapy, she had residual issues with safety awareness and loss of confidence. The impact of this episode on her quality of life is evident in her personal reflections as part of the 'Patient's perspective' included later in the report and illustrates the need to consider the entirety of outcome including the potential impact of treatment as well as of the disease itself.

DISCUSSION

Giant cell arteritis is a chronic granulomatous vasculitis affecting medium-large cell vessels and as yet its precise aetiology and underlying pathogenesis are not fully understood. Onset may be abrupt or insidious and classical symptoms of headache, jaw claudication, joint pains or malaise may not always be present. Scalp necrosis is a rare cutaneous complication of giant cell arteritis developing as a result of multi-vessel occlusion of all four main arteries supplying the temporal scalp.²

Scalp necrosis was first described by Cooke *et al* in 1946 and the most recent meta-analysis of cases published between 1946

Figure 4 Previously ulcerated area 6 months post-commencement of corticosteroid therapy.

Figure 5 Previously ulcerated area 24 months postpresentation with complete healing evident.

and 2007 suggested that the presence of scalp necrosis conferred a higher incidence of both visual loss and other visual defects than in giant cell arteritis without scalp necrosis, possibly reflective of more extensive vascular involvement.³ The differential diagnoses for scalp ulceration are numerous and, in the absence of other suggestive features, the potential exists for delayed diagnosis and treatment of the underlying giant cell arteritis. In previously reported cases where scalp necrosis was the only presenting feature the diagnosis of giant cell arteritis was made approximately 1 month later than in those cases presenting without scalp necrosis.³ In this case, a high index of suspicion for giant cell arteritis ensured a focused history eliciting preceding jaw claudication and recognition that, while the minor trauma reported was a possible precipitating factor in ulceration, the degree of ensuing necrosis suggested a grossly compromised scalp arterial supply.

The need for early recognition, diagnosis and treatment of giant cell arteritis is enshrined in BSR and BHPH guidelines¹ and further emphasised in the updated EULAR recommendations for management of large vessel vasculitis.⁴ While glucocorticoid therapy remains the mainstay of treatment in giant cell arteritis, the comorbidities associated with chronic glucocorticoid therapy, especially in elderly patients, are well recognised.^{5,6} Determining treatment duration and rate of steroid tapering for individual patients remains a therapeutic challenge given a variable disease course, risk of relapse, the absence of valid biomarkers to evaluate response and the need to balance the risk of both disease and treatment-related adverse events.⁶ With a lack of consensus relating to the efficacy and safety of steroid-sparing options, it is hoped that further research will help determine efficacious and safer alternative therapies in this condition.^{6,7}

This case highlights the importance of considering scalp necrosis as a manifestation of giant cell arteritis when assessing scalp ulceration, especially in the context of a raised ESR. Prompt diagnosis and treatment with careful monitoring for adverse events is necessary to prevent significant morbidity and potential mortality.

Contributors LMD has authored this article in full. OK was the supervising Consultant Dermatologist during the case and has provided guidance and approved the content prior to submission for publication. GB is a Histopathology Specialist

Patient's perspective

After all of my treatment and recovery I found that my energy was lower and I had some balance issues. I had physiotherapy which helped a little but unfortunately I have never felt the same. I used to enjoy walking my dogs but I rarely go outside now. Carers come to my house four times a day now to help me with certain tasks and overall this had a big impact on my quality of life. The scalp ulcer was very painful and took a long time to heal. I am grateful that the cause was found because the pain improved after I started my treatment and as it healed.

Learning points

- ▶ The differential diagnosis for scalp ulceration in the elderly is broad and careful history and investigation to exclude infective, malignant and inflammatory causes is vital.
- ▶ A high index of suspicion for giant cell arteritis should be maintained in a patient presenting with scalp ulceration without obvious cause as it may be the only clinically evident sign.
- ▶ The presence of scalp necrosis in giant cell arteritis confers an increased risk of mortality. Prompt diagnosis and treatment may be life saving.
- ▶ It is important to be cognizant of the risks of long-term oral glucocorticoid therapy and appropriate monitoring of side effects and comorbidities is necessary to prevent further morbidity.

Registrar and was responsible for oversight of reporting of the temporal artery biopsy, providing commentary on findings and obtaining the most appropriate cross-sectional imaging for inclusion as well as reviewing content of case report prior to submission.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Dasgupta B, Borg FA, Hassan N, *et al.* BSR and BHPH guidelines for the management of giant cell arteritis. *Rheumatology* 2010;49:1594–7.
- 2 Dummer W, Zillikens D, Schulz A, *et al.* Scalp necrosis in temporal (giant cell) arteritis: implications for the dermatologic surgeon. *Clin Exp Dermatol* 1996;21:154–8.
- 3 Tsiannakas 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.
- 4 Hellmich B, Agueda A, Monti S, *et al.* 2018 update of the EULAR recommendations for the management of large vessel vasculitis. *Ann Rheum Dis* 2019;0:annrheumdis-2019-215672–2012.
- 5 Gale S, Wilson JC, Chia J, *et al.* Risk associated with cumulative oral glucocorticoid use in patients with giant cell arteritis in real-world databases from the USA and UK. *Rheumatol Ther* 2018;5:327–40.
- 6 Ponte C, Rodrigues AF, O'Neill L, *et al.* Giant cell arteritis: current treatment and management. *World J Clin Cases* 2015;3:484–94.
- 7 Roberts J, Clifford A. Update on the management of giant cell arteritis. *Ther Adv Chronic Dis* 2017;8:69–79.

Copyright 2019 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

Customer Service

If you have any further queries about your subscription, please contact our customer services team on +44 (0) 207111 1105 or via email at support@bmj.com.

Visit casereports.bmj.com for more articles like this and to become a Fellow