CASE REPORT
SCALP NECROSIS IN GIANT CELL ARTERITIS AND REVIEW OF THE LITERATURE

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SUMMARY
A patient with giant cell arteritis (GCA) who developed scalp necrosis (SN) is described and 23 other cases in the English language literature are reviewed. SN is rare and occurs in older patients of mean age 77 yr. Thirteen patients presented to dermatologists. Nineteen (79%) had other serious complications of GCA: visual loss in 16, gangrene of the tongue in four and nasal septum necrosis in one. The mean interval between the onset of symptoms of GCA and SN was 3.0 months in the 19 cases which antedated corticosteroid therapy. SN resulted from active arteritis and no case was definitely linked to temporal artery biopsy. Scalp healing was complete or progressing satisfactorily in 18 cases (75%). SN is a potentially reversible complication of GCA and adequate corticosteroid therapy is mandatory. In the current case, SN related to inadequate dosage of prednisolone.

KEY WORDS: Scalp necrosis, Giant cell arteritis.

Fifty years ago, Cooke et al. [1] reported the first two cases of scalp necrosis (SN) in giant cell arteritis (GCA) and since then 21 other cases have been reported in the English language literature [2–19]. SN in GCA has been reported by rheumatologists on only one previous occasion [9] and only one case has been reported in a rheumatology journal [17]. The current patient is unusual in that she was already being treated for polymyalgia rheumatica (PMR).

CASE REPORT
A 78-yr-old woman presented to her general practitioner in August 1994 complaining of aching in her legs and shoulders, and stiffness in her neck and back. Her ESR was 127 mm/h and haemoglobin 11 G. A diagnosis of PMR was made and she was treated with 60 mg prednisolone daily with dramatic symptomatic relief. The dose of prednisolone was gradually reduced to 5 mg daily. Three months later, she had no symptoms or signs of PMR or GCA and her ESR was 7 mm/h.

The patient remained well on 5 mg prednisolone until 2 June 1995 when she developed a headache and loss of vision in her right eye. On admission to hospital on 5 June, the right temporal artery was tender with reduced pulsation and her ESR was 115 mm/h. A diagnosis of central retinal artery occlusion was made and prednisolone was increased to 60 mg daily. A right temporal artery biopsy was performed 14 days later, by which time the dose of prednisolone was 40 mg daily. At operation, the artery was noted to be thickened and non-pulsatile. Histology (Fig. 1) showed typical changes of GCA with gross narrowing of the lumen and extensive inflammatory cell infiltration of all the layers with collections of giant cells and fragmentation of the internal elastic lamina.

Ten weeks later (16 August), she complained of right periorbital, ear and scalp pain, and problems with washing her hair. Examination revealed an 8 × 11 cm crusting area of scalp in the right temporoparietal region with a clear line of separation which was then debrided to reveal a large area of SN (Fig. 2). At this stage, she was taking 15 mg prednisolone, but had reduced to 5 mg a month earlier: the dose was immediately increased to 30 mg daily.

The SN had almost healed by 11 October 1995 when her ESR was 20 mm/h and treatment was 20 mg prednisolone. However, on reduction to 17.5 mg, she became unwell, the SN was more extensive and the ESR rose to 87 mm/h. Since then, she has required between 20 and 30 mg prednisolone daily to control the SN. Azathioprine was added in April as a steroid-sparing drug. In June 1996, she was well on 20 mg prednisolone daily, her ESR was 10 mm/h and the SN was almost healed. She died later that month: the clinical diagnosis was pulmonary embolism, but a post-mortem was not obtained.

LITERATURE ANALYSIS
Analysis of the 24 reported cases of SN (including the above case) showed that 17 were women and seven men (ratio 2.4:1). Their mean age was 77 yr (range 66–86). Temporal artery biopsies were performed in 16 and were positive for GCA in 14. In another two patients, GCA was demonstrated on biopsy of the scalp lesion.

The most common complaint preceding SN was headache in 19 cases (79%). Eight had musculoskeletal symptoms, but only the above patient had a formal diagnosis of PMR. Sixteen (67%) had visual loss (bilateral in 11). One patient with total loss of vision also developed necrosis of the nasal septum and another had gangrene of the tongue. Two other patients also had tongue gangrene and another had incipient gangrene with tongue blanching. Thus, 19 (79%) had a major complication of GCA in addition to SN.

Nineteen patients developed SN before corticosteroid treatment, with a mean interval between the onset of GCA symptoms and SN of 3.0 months (range 0.5–11 months in 18 patients). Two developed SN within a month of steroid withdrawal due to side-effects. The three patients whose SN occurred whilst being treated with prednisolone include two in
whom SN developed within 6 weeks of commencing prednisolone [15, 17] and the current case. One patient [17] was on 35 mg prednisolone when SN developed overnight.

A common description of the scalp lesions was of crusting in the hair (10 cases) and in three of these the initial diagnosis was herpes zoster. The temporoparietal areas of the scalp were most commonly involved. Thirteen patients had bilateral lesions, including one patient with 80% scalp involvement. Healing was complete within a year in 15 cases and occurred within 6 months in at least six. Three others were healing well at 2, 6 and 10 months. Only one of these patients was not treated with prednisolone: 17 received initial doses of 30–100 mg daily (mean 56 mg in the 14 patients in whom the dose was given). Another patient was treated with 15 mg prednisolone plus azathioprine 100 mg daily.

Six patients had only partial or no healing of SN. Three were not treated with corticosteroids: one still had SN 3 yr after onset [1] and another for at least a year [10]. This patient [10] was the only one in whom skin grafting was performed. The third died of systemic GCA [1]. Three other patients died, one of causes unrelated to GCA after 9 weeks treatment [5]. The other two patients both had evidence of GCA at post-mortem: one had been treated with 15 mg prednisolone for a year [7] and the other with 40 mg to 15 mg for 3 months [11].

**DISCUSSION**

GCA has a peak onset between the ages of 60 and 75 yr. Healey and Wilske [20] found that 2/50 patients with biopsy-proven GCA presented with visual loss (4%) and two others subsequently lost vision, but none had SN. These patients with SN are older, have a high incidence of visual loss and tongue gangrene, and constitute a subgroup of severe GCA.

Thirteen of the 24 patients presented to dermatologists who recognize SN as a rare complication of GCA. Other skin manifestations of GCA included urticaria, oedema, erythema, vesicles, bullae, crusting, purpura and alopecia.

It is often questioned whether temporal artery biopsy may precipitate scalp ischaemia. An editorial review by

FIG. 1.—Histology of temporal artery × 45. Courtesy of Dr R. G. M. Letcher.

FIG. 2.—Scalp necrosis after debridement in August 1995.
Hall and Hunder [21], which described 652 biopsies of up to 6.5 mm in length, concluded that biopsy was a prudent and safe procedure. There is no doubt that SN was a result of active GCA in at least 21 of the 24 cases, as the skin lesions antedated biopsy. In two cases, GCA was demonstrated on biopsy of the scalp lesion [18]. In the current case, SN only became apparent 2 months after the biopsy when the ESR was high, indicating active arteritis. Since then, the SN increased in size when the patient was unwell with a high ESR and healed when the ESR was normal. One patient developed gangrene of the tongue beginning within hours of temporal artery biopsy and before steroid therapy; SN also occurred by 6 weeks, but there is insufficient detail to relate the SN to the biopsy [15].

There is no consensus on corticosteroid dosage and duration of treatment in GCA. Myles et al. [22] concluded that an initial dose of 20 mg prednisolone was sufficient in a series of 96 patients. Kyle and Hazleman [23] found that 40 mg was usually adequate, but higher doses were required occasionally in unusual clinical situations. They advise that alteration of steroid dose should be based predominantly on clinical symptoms and signs. Wilke and Hoffman [24] suggest that methotrexate should be added if the dose of prednisolone cannot be reduced to 15 mg daily after 2 months of treatment.

In conclusion, SN is a potentially reversible complication of GCA, but occurs against a background of severe arteritis; adequate corticosteroid therapy is mandatory.

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REFERENCES