JPRAS Open 15 (2018) 81–85

Contents lists available at ScienceDirect

JPRAS Open

journal homepage: http://www.journals.elsevier.com/jpras-open

Case Report

A rare case of adult scalp pyoderma gangrenosum with cranial osteolysis

L. Aljohmani *, K. Abdul-Jalil, C. deBlacam, G.M. Murphy, J.B. O'Sullivan

Departments of Plastic and Reconstructive Surgery, Beaumont Hospital, Beaumont road, Dublin 9, Ireland

ARTICLE INFO

Article history:
Received 10 November 2017
Accepted 22 November 2017
Available online 5 January 2018

Keywords:
Pyoderma gangrenosum
Cranial osteolysis
Adult
Treatment
Scalp

ABSTRACT

Pyoderma gangrenosum (PG) is a rare and painful idiopathic skin condition that has one or more areas of chronic ulceration with well demarcated and undermined borders. Bone osteolysis (the pathological destruction of bone tissue) secondary to PG is a rare phenomenon with limited cases reported in children only.

This is the first case report of scalp PG with cranial osteolysis in an 80-year-old adult, with an initial presentation mimicking skin carcinoma. This case highlights the importance of a multidisciplinary team (MDT) meeting discussion, diagnosis of PG by exclusion and the successful treatment of this patient's PG eroding to the bone.

Case report

An 80-year-old Caucasian male presented to our services with painful scalp ulcerative lesions. The patient was a non-smoker with type-2 diabetes mellitus and hypertension. The patient sustained a traumatic, left parietal scalp wound 2 months prior to presentation. The traumatic scalp wound was managed with regular dressing by the public health nurse but healing was slow and the wound began to deteriorate (Figure 1). At presentation, the patient had also developed a new non-traumatic left frontal scalp ulcer. In the weeks prior to presentation both ulcers had become painful. The scalp lesions

* Corresponding author. Departments of Plastic and Reconstructive Surgery, Beaumont Hospital, Beaumont road, Dublin 9, Ireland.
E-mail address: aljohmal@tcd.ie (L. Aljohmani).

https://doi.org/10.1016/j.jpra.2017.11.006

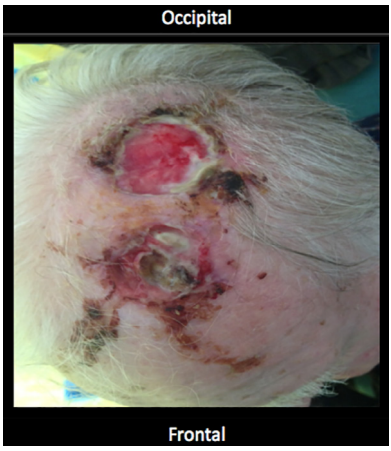

Figure 1. Photograph of scalp PG lesion over time, September 2015.

were eroding in nature with central ulceration and bleeding and according to the patient had gradually increased in size with associated bleeding of the ulcer. As a neoplastic lesion was suspected, the patient was admitted for oncology workup including radiological staging and punch biopsies for histological diagnosis.

Following multiple quadrant 4 mm punch biopsies (total of 8 were taken, 4 at each ulcer), results revealed inflammation and chronic ulceration of the tissue with no evidence of malignancy. A computed tomography (CT) of the brain revealed erosion of the outer table cortex only (Figure 2), at both left parietal and left frontal scalp lesions with normal brain parenchyma. After discussion of the case at the skin cancer MDT meeting it was decided to perform investigations for non-cancerous causes of scalp ulceration with input from dermatology and microbiology. Subsequent investigations (Table 1) were all negative and after careful consideration a clinical diagnosis of osteolytic PG of the scalp was made.

The patient commenced oral steroids; at post discharge he was followed up closely and his scalp ulcers slowly improved (Figure 3). His steroid regime involved oral prednisolone 40 mg daily for the

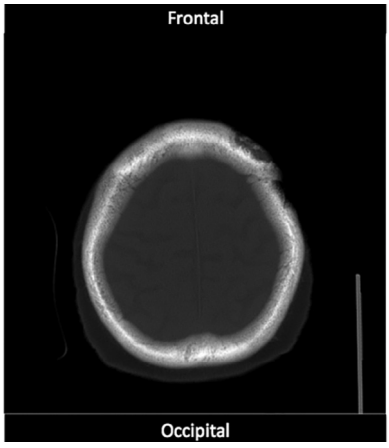

Figure 2. CT brain scan of time, correlating with Figure 1 photographic image of PG scalp lesion in September 2015, October 2015 and January 2016.

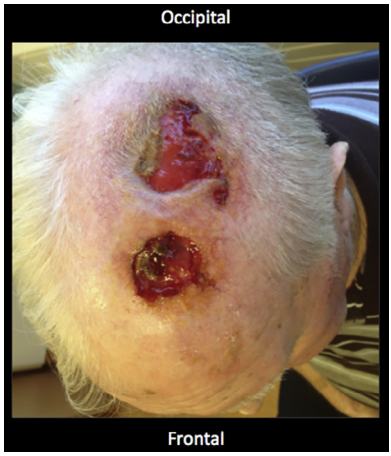

Figure 3. Photograph of scalp PG lesion over time, October 2015.

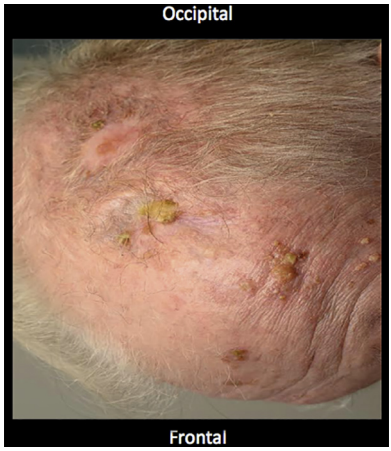

Figure 4. Photograph of scalp PG lesion over time, January 2016.

duration of 3 weeks followed by gradual tapering of dose by 5 mg per week, with the completion of the course at week 12. At one month after the commencement of steroids, his left frontal scalp ulcer had healed and healing of the left parietal scalp ulcer was progressing (Figure 1). The patient reported that pain from both ulcer sites had subsided after the ulcers had healed at 3 months (Figure 4). He had no complication from steroid therapy.

Discussion

Osteolytic PG of the scalp is an idiopathic cutaneous disorder. It is characterized by progressive painful ulceration, which causes local destruction of underlying structures. It is a benign lesion, which typically develops post trauma to the area. Histological examination of tissue biopsy shows non-specific inflammation with no malignancy features.[1]

PG is a condition diagnosed by exclusion of other underlying systemic disease or inflammatory process that may contribute to the development of skin ulceration or necrosis.[2–5] Approximately 50% of patients

Table 1
Investigations.

Test grouping	Test	Level	Reference range	Interpretation
Hematological	U&E	(Within normal limits)	(Within normal limits)	
	Albumin/creatinine ratio	<0.7	0.0–2.5 mg/Mmol	No diagnosis of micro/ macroalbuminuria diagnosis reached
	Lactate dehydrogenase (LDH)	311	208–378 IU/L	
	ESR	11	1–12 mm/h	
	FBC	(Within normal limits)	(Within normal limits)	
	LFT's	(Within normal limits)	(Within normal limits)	
	Iron study	(Within normal limits)	(Within normal limits)	
Serology	Rheumatoid arthritis factor (RF)	<12	0–20	Unlikely rheumatoid arthritis diagnosis to be reached 90% of rheumatoid is RF positive.
	Connective tissue disorder (CTD) screen – includes antibodies for DNA, RNP, SM, Ro, (52 & 60), La and other nuclear antigens	Negative	Within normal ranges	Connective tissue disorder unlikely to be diagnosed
	Anti nuclear factor (ANF)	Negative	Within normal range	Connective tissue disorder unlikely to be diagnosed
	Myeloperoxidase antibody (MPO)	0.1	0.0–3.5 IU/ml	Unlikely vacuities diagnosis to be reached
	Proteinase 3 (PR 3)	1	0–2 IU/ml	Unlikely vascular damage detected
	Serum protein electrophoresis (SPEP)	Borderline low	Within normal range	No diagnosis reached
	Immunoglobulin			No diagnosis reached – no
	IgG	5.8	6–15 g/l	contribution was made to any
	IgA	1.61	0.7–4 g/l	diagnosis to allergy- focused
	IgM	0.33	0.5–2.3 g/l	evaluation.
	Anti neutrophil cytoplasm AB (ANCA)	Negative	Within normal range	ANCA is positive in 90% of patients with generalized Wegener's granulomatosis or microscopic polyarteritis.
	Anti cardiolipin (IgG)	2	0–10 GPLU/ml	Anti-phospholipid syndrome
	Anti cardiolipin (IgM)	1	0–10 MPLU/ ml	unlikely,
	Beta 2 glycoprotein IgG	3	0–6 U/ml	
	Beta 2 glycoprotein	1	0–6 U/ml	
Microbiological	Blood culture		No growth	Unlikely infective cause
	Culture swab		No growth	detected
Endoscopy	OGD		Nil findings on scope	Unlikely intra esophageal or intra-colonic cause detected
	Colonoscopy		Nil findings on scope	

diagnosed with PG with skin ulceration or necrosis have an underlying systemic disease.[6] The condition was first described over 85 years ago and an infectious etiology was suspected. Subsequent microbiological investigations failed to elicit any microbial growth to support this concept.[7,8]

Several treatments have been described in the literature for cases with known PG including the use of immune-suppressive treatment for a period of 3–6 months depending on response and regular follow-up.[5,9–12] Surgical excision with split thickness skin-graft reconstruction has also been performed with variable results. PG is a condition where surgical resection may be detrimental due to increase in size of the affected area after resection and poor healing post-operatively. There is also a limited uptake of grafts over the affected area. Surgery has the added disadvantage of potentially initiating

a second surgically-triggered PG at the donor site[13,14] or at other sites (for example, the site where the split skin graft is harvested).

In the current case study, the patient made a significant recovery 1 month after initiation of steroid therapy with a total regression of the ulcers at 3 months; therefore, further treatment was not required. In this age group, it is prudent to consider preoperative steroid treatment if undergoing surgical treatment[9,15,16] and discuss the potential of recurrence with the patient.

There are two unique features in this case. First, the appearance of the second non-traumatic ulcer (left frontal scalp ulcer) completely separate to the initial ulcer. In addition, it is the first case report of adult scalp osteolytic PG which has been successfully treated with oral steroids.[17,18] The presentation and nature of the lesions created a high-level of suspicion of a malignant origin but effective MDT discussion and sufficient investigations allowed the correct diagnosis to be reached and a more appropriate management plan to be implemented. It is important to take a cautious approach to cancer-mimicking lesions such as those described and to consider that surgical intervention may not lead to the best outcome possible.

This case highlighted the importance of MDT meeting discussions, exclusion of malignancy and consideration of other causes of ulcerating scalp lesion such as PG.

In summary scalp PG with cranial osteolysis is a rare condition.[19] This case demonstrates that, with an accurate diagnosis, unnecessary surgical procedures can be avoided and a course of oral steroids may provide a safe and effective alternative treatment.

Conflict of interest

None.

Funding

None.

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