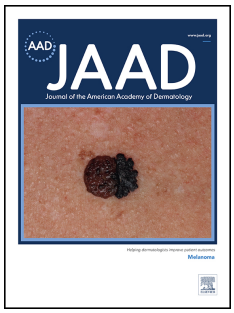
Since January 2020 Elsevier has created a COVID-19 resource centre with free information in English and Mandarin on the novel coronavirus COVID-19. The COVID-19 resource centre is hosted on Elsevier Connect, the company's public news and information website.

Elsevier hereby grants permission to make all its COVID-19-related research that is available on the COVID-19 resource centre - including this research content - immediately available in PubMed Central and other publicly funded repositories, such as the WHO COVID database with rights for unrestricted research re-use and analyses in any form or by any means with acknowledgement of the original source. These permissions are granted for free by Elsevier for as long as the COVID-19 resource centre remains active.

Journal Pre-proof



Broadspectrum Abnormal Localised Photosensitivity Syndrome

Sanaa Butt, MRCP, Amina Khalid, MRCP Derm, Angela Alani, MRCP Derm, Adam Fityan, FRCP, Hiva Fassihi, MD, Robert Dawe, MD, Sally Ibbotson, MD

PII: S0190-9622(20)32580-9

DOI: https://doi.org/10.1016/j.jaad.2020.08.119

Reference: YMJD 15185

To appear in: *Journal of the American Academy of Dermatology*

Received Date: 2 July 2020

Revised Date: 19 August 2020

Accepted Date: 31 August 2020

Please cite this article as: Butt S, Khalid A, Alani A, Fityan A, Fassihi H, Dawe R, Ibbotson S, Broadspectrum Abnormal Localised Photosensitivity Syndrome, *Journal of the American Academy of Dermatology* (2020), doi: https://doi.org/10.1016/j.jaad.2020.08.119.

This is a PDF file of an article that has undergone enhancements after acceptance, such as the addition of a cover page and metadata, and formatting for readability, but it is not yet the definitive version of record. This version will undergo additional copyediting, typesetting and review before it is published in its final form, but we are providing this version to give early visibility of the article. Please note that, during the production process, errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

© 2020 Published by Elsevier on behalf of the American Academy of Dermatology, Inc.

Journal Pre-proof

Broadspectrum Abnormal Localised Photosensitivity Syndrome

Sanaa Butt(MRCP)[1], Amina Khalid(MRCP Derm)[1], Angela Alani(MRCP Derm)[1], Adam Fityan(FRCP)[2], Hiva Fassihi(MD)[3], Robert Dawe(MD)[1], Sally Ibbotson(MD)[1]

[1]Photobiology Unit, Dermatology Department, University of Dundee, Ninewells Hospital & Medical School, Dundee, UK

[2] Department of Dermatology, University Hospital Southampton NHS Foundation Trust, Southampton, Hampshire, UK

[3] Department of Photodermatology, St John's Institute of Dermatology, Guy's and St Thomas' NHS Foundation Trust, London, UK

Corresponding author: Sanaa.butt@gmail.com

Word count: 500

Tables: 1
Figures: 1
Supplementary figures: 2 (https://data.mendeley.com/datasets/4vzpx255mt/1)
References: 5

Funding sources: none declared

Conflicts of interest: none declared

Journal Pre-proof

Journal Pre-proof

Dear Editor,

In recent years we have been increasingly aware of patients presenting with severe abnormal photosensitivity recurrently affecting fixed and limited areas, which can be provoked at these sites through phototesting. Monochromator phototesting has shown severely abnormal photosensitivity across a broadspectrum of wavebands but only at affected sites. We have thus coined the term Broadspectrum Abnormal Localised Photosensitivity Syndrome (BALPS) as a more accurate name. We have since realized this condition has been described in the past and termed 'fixed sunlight sensitivity'(FSS). We hope our suggested new diagnostic term better describes the clinical and photobiological features of this condition and should lead to increased recognition. We retrospectively studied 10 cases of BALPS seen through three specialist photodiagnostic units over a seven-year period in order to investigate their clinical characteristics, photodiagnostic investigations and histopathological findings to enable phenotyping of this patient cohort**(extended methods, see supplement).**

Eight of 10 patients were female. Mean age of onset was 37 years with mean time to presentation eight years following symptom onset. Limbs were a commonly affected site. Clinical features included erythema, oedema, blistering, intense burning sensation and pruritus**(see Table 1 and supplement for further clinical features)**. Phototesting**(detailed phototesting, Table 1)** showed broad-spectrum sensitivity at the affected sites with either normal or markedly less sensitivity noted at sites adjacent to these areas or at unaffected sites at 24 hours post irradiation**(figure 1)**. Common histopathologic findings were of epidermal spongiosis with lymphocyte exocytosis, mild dermal oedema and perivascular chronic lymphocytic infiltrates**(extended histopathology, see supplement).**

Journal Pre-proof

'Fixed sunlight sensitivity'(FSS) was first reported in 1975 by Emmet, describing a case of itchy, erythematous papular rash localised to sun-exposed sites on the face, reproduced by photoprovocation with longwave ultraviolet light[1]. Since then a handful of reports detail a similar clinical pattern, where triggers including food and drugs excluded and only sunlight remaining[2-4]. Emmett noted the difficulty in classifying this condition as it shares various characteristics with well recognised photodermatoses. One suggestion is to consider this as a localised form of CAD, given some similarities in phototesting and histopathology. However, typical photodistributed sites were not involved in our patients and only reproducible bizarrely localised sites were affected. In one of our patients UVA alone appeared to trigger the eruption, suggesting similarities to polymorphic light eruption, however, the clinical features did not correlate.

BALPS is also akin to a fixed drug eruption (FDE) given the localised recurrent nature, hence the term FSS used in the literature. In contrast to FDE we have not noted hyperpigmentation, nor could we incriminate any culprit drugs. Immunological memory is thought to be the pathogenesis implicated in FDE, with CD8+ T cells residing along the basement membrane primed and reactivated when reintroduced to the offending medication[5].

There may be a shared underlying pathogenesis in the form of a currently unidentified chromophore depositing in the skin of these patients, which absorbs the relevant wavelengths required to trigger a localised reaction. Due to the similar yet varied presentation of these cases, we wished to group them within this diagnostic entity of BALPS. We report on this diagnostic entity in order to raise awareness and facilitate identification of this fascinating patient cohort.

Journal Pre-proof

Figure 1

BALPS. Localised and fixed photosensitivity, only affecting right lower leg, with other sites unaffected. Monochromator phototesting on affected sites showed abnormal delayed erythema in UVB and UVA wavebands, 24 h after irradiation. UVA provocation testing on the affected site was markedly abnormal (grade 3 erythema) at 5J/cm^2. (Case 4, see supplement for further photos, Fig S1)

Journal Pre-proof

Journal Pre-proof

Patient	Age/ Gender/Skin Type^	Site	Time to onset/ clearance	Clinical features	Monochromator Non-affected site	Monochromator Affected site	UVA provocation Non-affected	UVA provocation affected	Management	Follow up
1	64/M/III	Right knee and right 5th finger, left forearm & lateral wrist	Hours/3-4 weeks	Erythema, oedema, tense yellow fluid filled blisters, desquamation and hypopigmentation	Back & left knee: Normal[&]	Right knee: UVA/visible sensitivity (365-460nm)	20 Jcm^{-2} negative	Left forearm 20 Jcm^{-2} grade 3 erythema	Photoprotection* Prophylactic narrowband UVB, "Psoracomb" device	Subjective: Improvement Objective: Improved, repeat testing showed only sensitivity at 365nm and lesser degree.
2	14/F/II	Thighs	2 days/ 2 weeks	Erythema, pruritus, oedema	Back: Normal[&]	Thigh: Normal[&]	Forearm: 20 Jcm^{-2} brown pigment (Normal)	Posterior thigh 20 Jcm^{-2} grade 3 erythema	Photoprotection* Prophylactic narrowband UVB	Subjective: Improvement Objective: Worsened, 5 Jcm^{-2} UVA grade 3 erythema + papules
3	43/F/II	Axilla, groins & abdomen	Few hours/24 hours	Erythema, oedema, blistering, 'burning' sensation	Back: Normal[&]	Inner thigh: UVB/UVA sensitivity (305-365nm)	Right front thigh: 10 Jcm^{-2} Negative	Right inner thigh: 5 Jcm^{-2} grade 3 erythema	Photoprotection* Prophylactic UVA1	Subjective: Improvement Objective: No change, repeat phototesting remained the same.
4	52/M/II	Right shin	Hours/2 days	Erythema, pruritus, blistering	Left shin: Normal[&]	Right shin: UVB/UVA sensitivity (305-365 nm)	Left shin: 5 Jcm^{-2} Negative	Right shin: 5 Jcm^{-2} grade 3 erythema	Photoprotection* Clobetasol propionate 0.05%	Subjective: Improvement Objective: No change, repeat phototesting remained the same.
5	50/F/II	Anterior & medial thighs	1-2 days/6 weeks	Erythema, pruritus, unilocular blistering	Back: Borderline at 305-400 nm	Thighs: UVB/UVA/visible greater sensitivity (305-400 nm)	Back: 5 Jcm^{-2} grade 3 erythema with papules	Right inner thigh: 5 Jcm^{-2} grade 4 response	Photoprotection* Clobetasol propionate 0.05%	Subjective: Improvement with sunscreen

								Right outer thigh: 5 Jcm^{-2} grade 3 erythema		Objective: No change, repeat phototesting remained the same.
6	37/F/IV	Buttocks & thighs	1 day/3-4 days	Burning sensation, macular erythema/purple discolouration	Back: Borderline at 335 nm	Thighs: UVB/UVA/visible sensitivity (305-400nm)	Not done	Not done	Photoprotection*	Further Investigation and follow up suspended due to Coronavirus pandemic
7	50/F/II	Buttocks, thighs, flanks below axillae	30 mins/2 weeks	Pruritus, erythema, burning sensation, urticated papules, resolves with purpuric change	Back: Normal[&]	Buttock: UVB/UVA sensitivity (305, 365 nm)	Not done	Buttock: 10Jcm^{-2} grade 3 erythema	Photoprotection* Prophylactic narrowband UVB, "Psoracomb" device – not tolerated Tacrolimus 0.1% Methotrexate 10mg weekly	Follow up suspended due to Coronavirus pandemic
8	56/F/II	Legs, back of thighs, knees, abdomen	unknown/2-3 weeks	Erythema, papules, blistering,	Back: Normal[&]	Thigh: UVB/UVA/visible sensitivity (305-400nm)	Forearm: 10Jcm^{-2} grade 1 erythema Back: 10Jcm^{-2} grade 2 erythema and papules	Thigh: 10Jcm^{-2} grade 3 erythema	Methotrexate 25mg once weekly + IM glucocorticoid injection for alternative condition Photoprotection*	Did not attend
9	31/F/II	Right buttock, inner thighs	2-3 days/4 weeks	Pruritus, erythema, burning sensation, oedema	Back: Normal[&]	Buttock: UVB/UVA sensitivity (305-340 nm)	Not done	Not done	Photoprotection* Clobetasol propionate 0.05%	Subjective: Improvement
10	52/F/II	Left buttock	18 hours/7 days	Erythema, desquamation	Back: minor UVB/UVA sensitivity (300-320 nm)	Left buttock: UVB/UVA/visible sensitivity (300-400 nm)	Not done	Not done	Photoprotection* Clobetasol propionate 0.05%	Subjective: Improvement

Table 1. Clinical features and phototesting results of patients with BALPS. [&]Within population reference range. *Photoprotection advice includes behavioural modification, environmental, clothing & topical sunscreen. ^Fitzpatrick skin phototype.

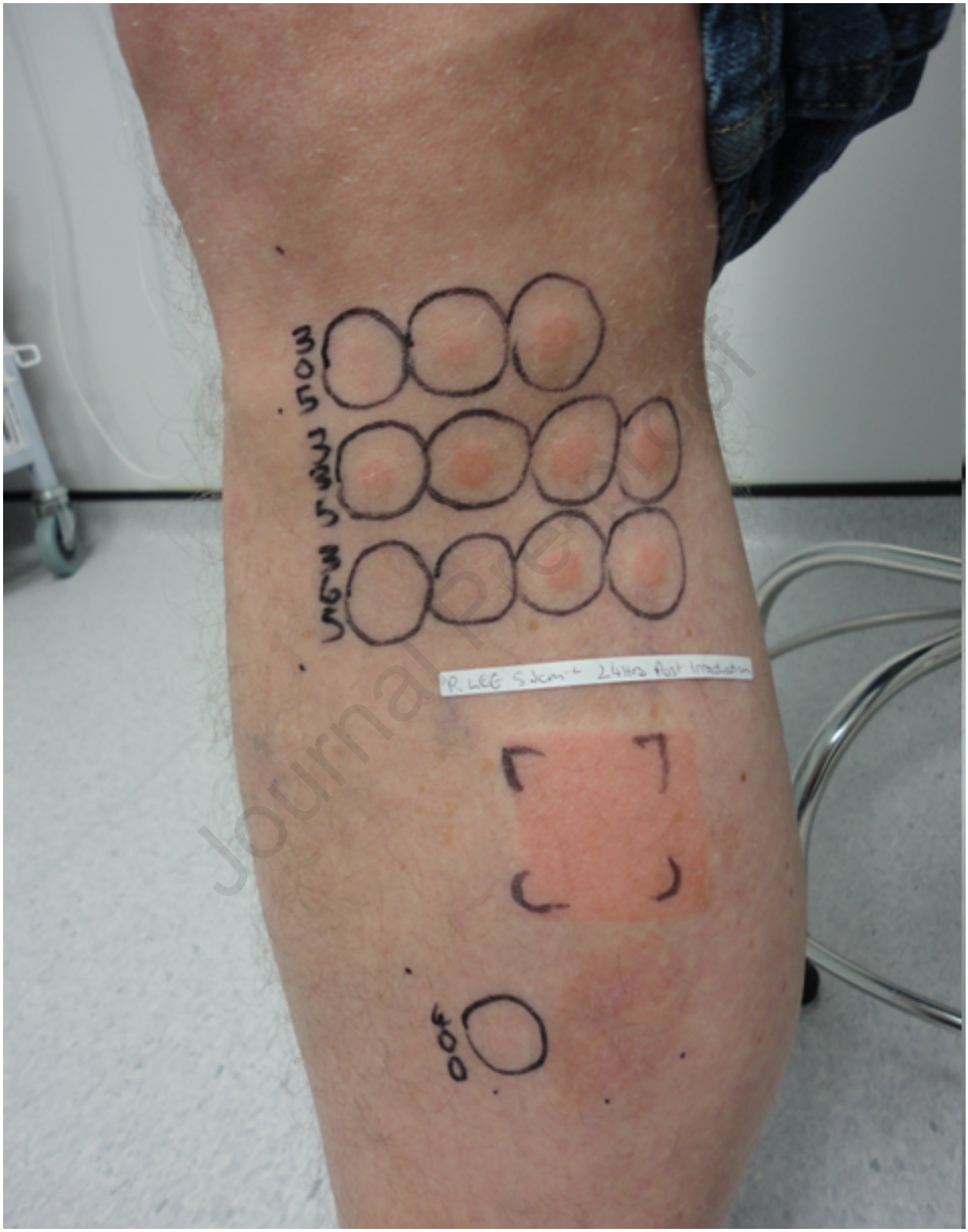
Journal Pre-proof

Journal Pre-proof

References

1. Emmett EA. Fixed long ultraviolet eruption. *Arch Dermatol*. 1975; Feb;**111**(2):212-4.
2. Valdivieso R, Cañarte C. It is not a fixed drug eruption, it is a fixed "sunlight" eruption. *Int J Dermatol*. 2010; Dec;**49**(12):1421-3
3. Gamé D, Bassas J, Grau C et al. Fixed sunlight eruption: a new idiopathic photodermatosis rather than a variant of fixed drug eruption. *Photodermatol Photoimmunol Photomed.* 2017. Jul;**33**(4):222-224
4. Valdeolivas-Casillas N, Piteiro-Bermejo AB, Trasobares-Marugán L et al. Fixed sunlight eruption: A case report. *J Eur Acad Dermatol Venereol.* 2016 May;**30**(5):894-5
5. Shiohara T. Fixed drug eruption: Pathogenesis and diagnostic tests. *Curr Opin Allergy Clin Immunol.* 2009 Aug;**9**(4):316-21.

Journal Pre-proof



Journal Pre-proof