There is also some controversy as to whether this process is associated with hematologic neoplasms or other reactive plasmacytic processes, such as Castleman disease, although current knowledge suggests that these processes are independent.[5]

Owing to the rare nature of this condition, there are neither guidelines nor consensus on the treatment of affected patients. Of the several treatments tested, most have been only partially effective, including oral and topical corticosteroids, topical calcineurin inhibitors, melphalan, immunoglobulins, rituximab and thalidomine, and phototherapy (psoralen and ultraviolet [UV] A and narrowband UVB therapy).[1,6,7]

In conclusion, cutaneous plasmacytosis is uncommon in white individuals, and usually has a chronic and benign long-term course. Nonetheless, follow-up of these patients is recommended owing to the scarcity of information about this pathological process.[6]

Conflicts of interest

The authors declare that they have no conflicts of interest.

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Systemic steroids in the management of moderate-to-severe hidradenitis suppurativa☆

Uso de los corticoides sistémicos en el tratamiento de la hidradenitis supurativa moderada-grave

To the Editor:

Hidradenitis Suppurativa (HS) is a debilitating chronic inflammatory disease of the apocrine gland-bearing skin, for which effective medical treatment remains elusive. Several immunological derangements are now identified in the disease.[1] Adalimumab is so far the sole European Medicines Agency approved drug. It shows encouraging, albeit suboptimal results.[2] More immunosuppressants are in the pipeline. Despite this shift towards anti-inflammatory therapies in HS, evidence is scarce regarding the use of systemic steroids (SS), with only a limited number of case reports[3–5] and series[6,7] available. We aimed to evaluate SS as adjuncts to other medical therapies in HS.

A retrospective cohort study was conducted. The setting was an Adnexal Skin Diseases Clinic in a tertiary Dermatology department in Lisbon. Data was captured by searching the electronic and written medical records of the clinic. Patients were eligible if they had moderate or severe HS, as defined by the International HS Severity Score (IHS4), treated with SS at least in one occasion. Primary endpoint was a clinical response as defined by the HS Clinical Response Score (HiSCR).[8] Changes in patient-reported outcomes (Dermatology Life Quality index – DLQI and pain Numeric Rating Scale - NRS) were also evaluated. For statistical analysis, a Wilcoxon signed-rank test at a level of significance of 0.05 was used with STATA/IC 15.1 (STATA Corp., Texas, USA).

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Table 1 Patients general data and treatment outcomes with adjunct systemic steroids. All scores present a pretreatment, a posttreatment and a delta value which is represented, respectively, by the letters i (initial), f (final) and Δ (change with treatment).

Patient Number	Gender	Age	Years with HS	Reason for initiation	Concurrent treatments	Peak PDN mg/kg	PDN span days	HiSCR	$ISH4_i$	$ISH4_f$	ΔISH4	$DLQI_i$	$DLQI_f$	ΔDLQI	NRS_i	NRS_f	ΔNRS
I	F	59	4	Disease cooling	Doxy	0.47	24	no	43	36	-16%	15	14	-7%	8	7.5	-6%
II[§]	M	48	2														
First therapeutic cycle				Sympt. relief	ADA - Doxy	0.46	40	no	56	56	0%	26	20	-23%	1	0	-100%
Second therapeutic cycle				Sympt. relief	ADA	0.24	30	no	56	56	0%	20	20	-5%	1	4	300%
Third therapeutic cycle				Preop.cooling	Ertapenem	0.66	30	yes	56	56	0%	20	21	-0%	4	1	-75%
III	M	44	1	Acute flare	Isotretinoin	0.35	12	yes	6	4	-33%	10	0	-100%	8	0	-100%
IV	M	18	1														
First therapeutic cycle				Disease cooling	Clind-Rif	0.7	60	yes	12	4	-67%	11	8	-27%	7	1.5	-79%
Second therapeutic cycle				Acute flare	ADA	0.24	30	yes	4	4	0%	4	2	-50%	1	0	-100%
V	M	58	1	Acute flare	Clind-Rif	0.28	60	yes	4	4	0%	4	1	-75%	2	1.5	-25%
VI	F	24	0	Disease cooling	ADA - Doxy	0.57	60	no	17	17	0%	11	12	-9%	6.5	6.5	0%
VII	M	58	5	Disease cooling	Clind-Rif	0.33	54	yes	16	10	-38%	16	12	-25%	6.5	4.5	-31%
VIII	F	44	19	Disease cooling	Acitretin	0.29	10	yes	20	8	-60%	8	5	-38%	0	0	0%
IX	F	46	31	Acute flare	ADA	0.5	29	no	4	2	-50%	8	8	0%	25	23	-8%
X	M	58	30	Disease cooling	Doxy	0.5	30	yes	31	24	-23%	23	17	-26%	5	2	-60%
XI	F	49	3	Disease cooling	ADA	0.47	90	yes	46	35	-34%	8	1	-88%	4	2	-50%
XII	M	67	2	Acute flare	ADA	0.42	15	yes	8	8	0%	9	11	22%	7	0	-100%
XIII	F	21	1	Disease cooling	IFX - Doxy	1	60	yes	6	2	-37%	3	3	0%	10	0	-100%
XIV	M	28	2	Disease cooling	ADA - Doxy	0.72	70	yes	14	12	-14%	16	10	-38%	4.5	1	-78%
XV	F	32	16	Disease cooling	Doxy	0.28	29	no	24	24	0%	19	19	0%	7	5	-29%
XVI[§]	F	54	2														
First therapeutic cycle				Disease cooling	Doxy	0.37	35	yes	4	4	0%	24	22	-8%	7	5	-29%
Second therapeutic cycle				Preop.cooling	Doxy	0.37	15	yes	4	0	100%	22	0	-100%	5	0	-100%
			Overall deviation*					**14/20**	**15**	**9**	**-40%**	**13**	**10.5**	**-19%**	**5.75**	**1.5**	**-74%**

*All scores (IHS4, DLQI and NRS) and their respective Δ are represented by median values. All changes were statistically significant (Wilcoxon signed-rank test, $p < 0.05$).

[§]Patient II and XV had only draining-fistulae. HiSCR was considered to be met despite absence of changes in IHS4, as treatment with PDN led to a clinically meaningful reduction in suppuration, inflammatory burden and overall disease severity.

HS – Hidradenitis Suppurativa; PDN – Prednisolone; Preop – Preoperative; Sympt – Symptomatic; ADA – Adalimumab; Doxy – Doxycycline; Clind-Rif – Clindamycin – Rifampicin; IFX – Infliximab.

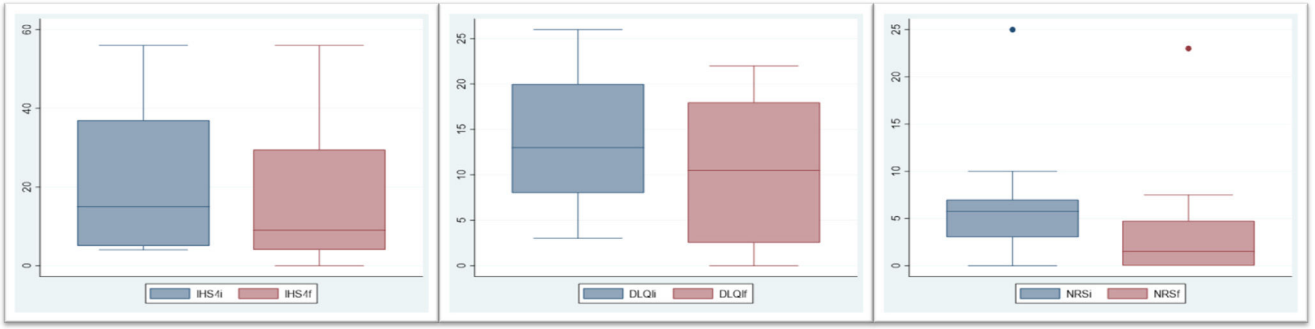

Figure 1 Pretreatment (i) and posttreatment (f) with adjunct systemic steroids. A statistically significant change was observed in both the IHS4 and the patient-reported outcomes (Wilcoxon signed-rank test, $p < 0.05$).

Among 121 HS patients followed at the clinic, 20 (16.5%) met eligibility criteria and 16 were analysed (Table 1). Four excluded due to poor compliance or lost to follow-up). Most patients were women (9/16), Caucasian (15/16), with a mean age of 45 years (18-67) and a mean duration of disease of 7.5 years (1-31). Most had severe disease (10/16; median IHS4 of 15).

Twenty cycles of adjunct systemic steroids were performed (three patients underwent >1 cycle). Therapy was initiated for disease cooling (11), acute flares (5), symptomatic relief (2) or preoperative cooling (2). The median maximum prednisolone dose was 0.44 mg/kg (0.28-1) and the median duration was 30 days (10-90). SS were mostly used in combination with doxycycline (9 cycles) followed by adalimumab (8 cycles). Most cycles (14/20, 70%) met the HiSCR. Those who did not, were performed in patients with higher disease activity (median IHS4 of 33.3 in non-responders vs 10 in responders). Median IHS4 reduced 40% ($p = 0.0012$) (Fig. 1). A significant improvement was observed in all patient-reported outcomes: median pain NRS and DLQIs reduced, respectively, 74% ($p = 0.0007$) and 19% ($p = 0.003$). Three patients had remarkable disease worsening shortly after steroid withdrawal. No treatment discontinuation or remarkable adverse events were noted.

There's a paucity of research regarding the use of systemic steroids in HS, albeit they are prescribed in more than 1% of patients' visits.[9] To the best of our knowledge, this is the largest series to date evaluating SS as adjunct therapy in HS. Our results suggest that a short-to-medium term taper can be beneficial to rapidly control the painful hyperinflammatory flares while conventional HS treatments achieve proper disease control. Previously, long-term low-dose SS have showed advantages,[7] but the risk of cumulative steroids' detrimental effects must be considered. Additionally, uncontrolled inflammation is thought to increase the risk of postsurgical complications.[10] In this series, 2 patients were treated preoperatively with favourable results and SS may thus be helpful to include in preoperative cooling strategies.

In conclusion, moderate-to-severe HS patients are likely to benefit from addition of SS to other medical therapies for both disease control and preoperative care. Due to limitations in the present study, regarding its retrospective design and sample size, a placebo-controlled trial is warranted to further clarify the role of SS in the anti-inflammatory strategies of HS.

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Actinic Cheilitis Treated With Daylight Photodynamic Therapy☆

Terapia fotodinámica con luz de día en el tratamiento de la queilitis actínica

To the Editor,

Actinic cheilitis is a chronic premalignant disease that generally affects the lower lip and that is considered equivalent to actinic keratoses of the skin.[1–3] Several treatments have been proposed for actinic cheilitis, but none of these are considered definitive. Daylight photodynamic therapy (PDT) is a widely used treatment for actinic keratoses and has shown efficacy similar to that of conventional PDT, with no or minimal pain.[4,5] The objective of our study was to describe the efficacy and safety of daylight PDT in the treatment of actinic cheilitis according to our experience. Between May and October 2018, 6 patients were treated. After gentle curettage, a roll of cotton wool was placed in the internal labial mucosa to expose the lower lip and sufficient quantity of methyl aminolevulinate cream was applied. Then exposure to ambient sunlight for 2 hours without occlusion was recommended. The rest of the skin was protected with factor 50+ sunscreen. After 2 hours of exposure, the treated area was washed and factor 50+ sunscreen was applied to the lip. Another session was performed 2 weeks later and the patients were examined after 2 months (Figs. 1 and 2). Patients were assessed with a clinical scale (affected area and complete/partial/no response) and a visual analogue scale (VAS) for pain. The characteristics of the study population and the outcomes of treatment are summarized in Table 1. Four of the patients (67%) showed complete response and 2 had a partial response with a mean reduction in the affected area of 58.3%. The mean score on the VAS was 0.5 out of 10.

Actinic cheilitis is a premalignant disease with a rate of transformation to squamous cell carcinoma (SCC) of 16.9% and a relative risk of 2.5 for developing this entity.[1,2] Furthermore, labial SCC has a 4-fold higher tendency for developing lymph node metastases compared with SCC on the skin.[6,7] Chronic exposure to ultraviolet radiation is the main risk factor implicated in the onset of actinic cheilitis, along with smoking and alcohol abuse.[3] Several different treatments have been used for actinic cheilitis, such as ablative methods (cryotherapy, CO_2 laser treatment), partial surgical resection, or vermilionectomy, and topical treatments such as imiquimod or 5-fluoracyl.[3,8] On the other hand, actinic keratoses are the skin equivalent of actinic cheilitis[3] and several different treatments similar to those listed above have been tried, although daylight PDT has also been used. This technique consists of a photosensitizing substance (MAL/5-ALA) that is activated by exposure to ambient sunlight (visible light) without the need for prior occlusion or exposure to red light from a lamp as is the case for conventional PDT.[4] Daylight PDT has been associated with efficacy rates similar to conventional PDT (clearance rate of 70% to 93% at 3 months after a single session[4,5]) with much better tolerance as there is no or minimal pain.[4] Conventional PDT has also been used in actinic cheilitis (15 previous studies)[3] with a mean clinical response of 60.25% and histological clearance of 47.4%.[3] Intense pain during treatment was the main side effect, requiring the administration of oral analgesics or local anesthetic.[3] Daylight PDT for actinic cheilitis has been used less frequently, and is only mentioned in 2 studies in the literature,[8,9] and in a description of 2 cases.[10] Fai et al.[8] treated 10 patients with actinic cheilitis with daylight PDT using aminolevulinate cream, obtaining total remission of the affected area in 70% of patients after 3 months; this was maintained in 50% of patients at 6 to 12 months of final follow-up. All patients showed partial remission that was maintained until the end of follow-up. In another study, 11 patients were treated, obtaining a cure rate of 91%,[9] and 2 isolated case reports with complete clearance after treatment.[10] No adverse effects were reported in any cases. Our results are very similar to those described previously in the literature. All patients experienced a reduction in the affected area after treatment, with complete resolution in 67% of patients. These results exceeded those obtained with other medical treatments such as 5-FU (clinical clearance in 30% to 40% of patients)[3] and were similar to imiquimod 5% cream[3] (40% to 100%). No side effects were detected other than mild discomfort, with the results obtained with the VAS for pain of 0 or 1 (range 0-10). Despite the limited number of patients, given the results obtained and those presented in previous studies, we consider that daylight PDT is a good alternative for treatment of actinic cheilitis. This technique obtains response rates similar to those of conventional PDT, without the associated pain. There is also no need for a PDT lamp, so it can be performed in any center.

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