no antimicrobianos como los antiinflamatorios (inhibición de la quimiotaxis de leucocitos, y activación y regulación de citoquinas inflamatorias en queratinocitos) y su actividad anticolagenasa, vía inhibición de las metaloproteinasas de la matriz dérmica. La metaloproteinasa 9 y su inhibidor, han sido involucrados en la EHH y también en la enfermedad de Darier[7,8].

La reciente publicación de 6 casos de EHH con dramática respuesta a doxiciclina[7], la escasez de efectos adversos, su accesibilidad, fácil manejo y bajo coste, nos llevaron a usarlo con excelente respuesta, nunca antes experimentada por nuestro paciente ni espontáneamente ni con otros tratamientos tópicos utilizados (dermocorticoides y ácido fusídico).

Conflicto de intereses

Los autores declaran no tener ningún conflicto de intereses

Bibliografía

1. Hu Z, Bonifas JM, Beech J, Bench G, Shigihara T, Ogawa H, et al. Mutations in ATP2C1, encoding a calcium pump, cause Hailey-Hailey disease. Nat Genet. 2000;24:61–5.
2. Dhitavat J, Fairclough RJ, Hovnanian A, Burge SM. Calcium pumps and keratinocytes: lessons from Darier's disease and Hailey-Hailey disease. Br J Dermatol. 2004;150:821–8.
3. Leinonen PT, Hägg PM, Peltonen S. Reevaluation of the normal epidermal calcium gradient, and analysis of calcium levels and ATP receptors in Hailey-Hailey and Darier epidermis. J Invest Dermatol. 2009;129:1379–87.
4. Burge SM. Hailey-Hailey disease: The clinical features, response to treatment and prognosis. Br J Dermatol. 1992;126:275–82.
5. Gisondi P, Sampogna F, Annessi G, Girolomoni G, Abeni D. Severe impairment of quality of life in Hailey-Hailey disease. Acta Derm Venereol. 2005;85:132–5.
6. Chiaravalloti A, Payette M. Hailey-Hailey disease and review of management. J Drugs Dermatol. 2014;13:1254–7.
7. Le Saché-de Peufeilhoux L, Raynaud E, Bouchardeau A, Fraitag S, Bodemer C. Familial benign chronic pemphigus and doxycycline: A review of 6 cases. J Eur Acad Dermatol Venereol. 2014;28:370–3.
8. Kobayashi T, Sakuraoka K, Hattori S, Hori H, Nagai Y, Tajima S, et al. Immunolocalization of human gelatinase (type IV collagenase, MMP-9) and tissue inhibitor of metalloproteinase 1 in Hailey-Hailey and Darier's diseases. Dermatology. 1996;193:110–4.

M.Á. Flores-Terry*, M.P. Cortina-de la Calle, M. López-Nieto y R. Cruz-Conde de Boom

Servicio de Dermatología, Hospital General Universitario de Ciudad Real, Ciudad Real, España

*Autor para correspondencia.
Correo electrónico: miguelterry85@hotmail.com (M.Á. Flores-Terry).

http://dx.doi.org/10.1016/j.ad.2015.10.015

Importance of educational sessions on cardiometabolic comorbidities. Awareness among psoriasis patients

Importancia de las sesiones educativas sobre comorbilidades cardiometabólicas. Conciencia entre los pacientes con psoriasis

Dear Editor:

There is strong evidence that psoriasis is associated with several cardiometabolic comorbidities, and that patients with psoriasis are at a higher risk of cardiovascular morbidity and mortality.[1–3] Understanding this is of crucial importance, not only for physicians but also for patients, as it can impact prognosis and patient quality of life.[4]

It has been shown that few patients with moderate to severe psoriasis are aware of their increased risk of atherothrombotic disease and metabolic syndrome.[5] Educational sessions are a recognized tool for informing and helping patients to understand the nature and course of their disease and the different treatments available, and can also help them to develop coping strategies.[6,7]

We performed an observational study to evaluate the impact of an educational session designed to promote knowledge among patients with psoriasis about their disease, lifestyle changes, and management of cardiometabolic comorbidities.

The educational session was held in the psoriasis unit of a Portuguese tertiary hospital. Briefly, it consisted of several oral presentations (30 min each) explaining the nature of psoriasis, introducing the various treatment options, exploring the association between psoriasis and cardiometabolic comorbidities/cardiovascular disease, and underlining the importance of monitoring and treating these. A questionnaire was created for the patients to complete before, immediately after, and 6 months after the session. The questionnaire included demographic information, questions regarding the association between psoriasis and cardiometabolic comorbidities/cardiovascular disease, and assessment of lifestyle and comorbidity management.

Seventy patients participated in the session and 53 completed all 3 questionnaires correctly. The demographic data, characteristics of disease, and treatments received are presented in Table 1. Regarding cardiometabolic comorbidities, 35.8%, 13.2%, and 35.8% of patients had a respective diagnosis of hypertension, diabetes mellitus, and dyslipidemia; 20.8% were obese (body mass index > 30); and 18.9% were active smokers (Table 1).

The McNemar test was used to assess significant improvements in knowledge between the different time points. A *P* value of less than or equal to .05 was considered statistically significant. A significant increase was observed in

Table 1 Characteristics of 53 patients who completed full questionnaires before and after a psoriasis education session.

Characteristics	
Age, mean (SD), y	51.3 (11.2)
Sex, No. (%) of patients	
Male	50.9 (27)
Level of education, No. (%) of patients	
No studies	1.9 (1)
Basic education (4th grade)	37.7 (20)
Basic education (9th grade)	17 (9)
Full secondary education	22.6 (12)
Third-level education	20.8 (11)
Body mass index, mean (SD)	26.7 (4.4)
Comorbidities/history	
Hypertension	35.8 (19)
Dyslipidemia	35.8 (19)
Diabetes mellitus	13.2 (7)
Obesity	20.8 (11)
Smoking	18.9 (10)
Family history of cardiovascular disease	15.1 (8)
Family history of psoriasis, No. (%) of patients	49.1 (26)
Disease duration, mean (SD), y	24.6 (13.2)
Psoriatic arthritis, No. (%) of patients	60.4 (32)
Current treatment, No. (%) of patients	
None	1.9 (1)
Topical	18.9 (10)
Phototherapy	7.5 (4)
Systemic	26.4 (14)
Biologic	39.6 (21)
Biologic + systemic	5.7 (3)

the percentage of correctly answered questions about the association between psoriasis and cardiometabolic comorbidities/cardiovascular disease on comparing the answers before and immediately after the session and before the session and 6 months later (Table 2). When logistic regression models adjusting for age, sex, and level of education were applied, the results were not altered.

Six months after the session, 60.4% of respondents stated that they had started to exercise or had increased their level of physical activity; 49.1% stated that they had lost weight (mean loss, 2.3 kg; maximum loss, 7 kg; minimum loss, 1 kg); and 50% stated that they had cut down on smoking, although none had quit.

After the session, over 90% of the participants stated that they were willing to consult their general practitioner to screen for cardiometabolic comorbidities or monitor those already diagnosed. This attitude led to newly diagnosed comorbidities (7 cases of dyslipidemia, 2 cases of hypertension, and 2 cases of diabetes mellitus) and therapeutic adjustments (in 4 cases of hypertension, 4 cases of dyslipidemia, and 1 case of diabetes mellitus).

Our results show that an educational session held with patients with psoriasis had a real impact on their health, leading to lifestyle changes (physical activity, weight loss, reduction of smoking), increased awareness of their disease and associated comorbidities, and new diagnoses of cardiovascular risk factors or therapeutic adjustments. Repeated educational sessions with the same group of patients might further improve these positive results.

Some limitations of this study must, however, be mentioned, mainly the limited number of participants and a potential response bias, either intentional or due to comprehension factors, associated with self-reported surveys.

In brief, this educational intervention proved to be a useful tool that had significant repercussions on patients' health, life, and ability to cope with the disease, and it should therefore be repeated with a larger number of patients.

Table 2 Improvements in awareness of association between psoriasis and cardiometabolic comorbidities/cardiovascular disease.[a]

	Before session	Immediately after session	*P*[b]	6 Months after session	*P*[c]
Hypertension	50.9	98.1	<.001	84.9	<.001
Dyslipidemia	47.2	98.1	<.001	81.1	<.001
Diabetes mellitus	39.6	94.3	<.001	75.5	<.001
Obesity	39.6	92.5	<.001	66	.004
Acute myocardial infarction	32.1	92.5	<.001	79.2	<.001
Stroke	30.2	88.7	<.001	73.6	<.001

[a] Results shown as the percentage of questionnaire respondents who were aware of the association between psoriasis and the comorbidities shown.
[b] Improvement immediately after the session.
[c] Maintenance of improvement 6 months after the session (McNemar test).

Conflict of interest

The authors declare no conflict of interest.

Bibliografía

1. Miller IM, Ellervik C, Yazdanyar S, Jemec GB. Meta-analysis of psoriasis, cardiovascular disease, and associated risk factors. J Am Acad Dermatol. 2013;69:1014–24.
2. Hugh J, Van Voorhees AS, Nijhawan RI, Bagel J, Lebwohl M, Blauvelt A, et al. From the Medical Board of the National Psoriasis Foundation: the risk of cardiovascular disease in individuals with psoriasis and the potential impact of current therapies. J Am Acad Dermatol. 2014;70:168–77.
3. González-Gay MA, González-Vela C, González-Juanatey C. Psoriasis: a skin disease associated with increased cardiovascular risk. Actas Dermosifiliogr. 2012;103:595–8.
4. Gisondi P, Girolomoni G. Cardiometabolic comorbidities and the approach to patients with psoriasis. Actas Dermosifiliogr. 2009;100 Suppl. 2:14–21.
5. Skiveren J, Philipsen P, Therming G. Patients with psoriasis have insufficient knowledge of their risk of atherothrombotic disease and metabolic syndrome. Clin Exp Dermatol. 2015;40:600–4.
6. Gallefoss F, Bakke PS. Impact of patient education and self-management on morbidity in asthmatics and patients with chronic obstructive pulmonary disease. Respir Med. 2000;94:279–87.
7. Bryan TJ, Estrada CA, Castiglioni A, Snyder ED. Impact of an educational intervention on provider knowledge, attitudes, and comfort level regarding counseling women ages 40–49 about breast cancer screening. J Multidiscip Healthc. 2015;8:209–16.

F. Mota[a,*], M. Selores[a,b,c], T. Torres[a,b,c]

[a] *Department of Dermatology, Centro Hospitalar do Porto, Portugal*
[b] *Dermatology Research Unit, Centro Hospitalar do Porto, Portugal*
[c] *Unit for Multidisciplinary Research in Biomedicine, Instituto de Ciências Biomédicas Abel Salazar, University of Porto, Portugal*

* Corresponding author.
E-mail address: fernandojrmota@gmail.com (F. Mota).

http://dx.doi.org/10.1016/j.ad.2015.12.011

A case of Birt-Hogg-Dubé syndrome presenting with a single pedunculated fibrofolliculoma and a novel FLCN gene mutation

Un caso de síndrome de Birt-Hogg-Dubé presentando con un único fibrofoliculoma pediculado y una novedosa mutación en el gen FLCN

To the Editor:

In 1997, Birt, Hogg, and Dubé described multiple firm papules on the scalp, forehead, face, and neck in 15 persons belonging to a family of 70 individuals spanning 3 generations.[1] The skin lesions were classified as 3 benign variants of hair follicle hamartomas: fibrofolliculomas, trichodiscomas, and acrochordons. This triad was later designated as the Birt-Hogg-Dubé syndrome (BHDS) (Online Mendelian Inheritance in Man catalog number, #135150).

A number of cases of BHDS have been identified since the original description was published, and significant associations have been reported, particularly with multiple lung cysts[2,3] and renal cancer.[2–4] A study in 2002 found that patients with BHDS have a 9.3-fold risk of developing renal tumors and a 32.3-fold risk of spontaneous pneumothorax.[2]

Case description

A 28-year-old man presented to our department with a 1-year history of skin lesions on the nose. The patient's past medical history was unremarkable, except for 2 spontaneous pneumothoraces at the age of 19 and 23 years. The patient also mentioned that his mother underwent right nephrectomy for a renal tumor at the age of 38 years.

Physical examination revealed 2 wine-colored, pedunculated papules, 1 measuring 3 mm on the right side of the columella, the other measuring 2 mm on the left alar rim (Fig. 1A and B). No other significant alterations were observed on examination.

Both lesions were excised. Histological examination of the papule from the left alar rim was compatible with a vascular hamartoma. The lesion from the columella showed features compatible with a fibrofolliculoma (Fig. 2A and B).

Sequencing of the coding exons (exons 4–14) and the intron-exon boundaries of the *FLCN* gene revealed a heterozygous *FLCN*:c.50G>C missense variant (p.Arg17Pro) in exon 4, both in the patient and in his mother. Numerous small cysts were observed in the basal regions of both lungs on computed tomography (CT) of the chest (Fig. 1C).

Applying the diagnostic criteria of the European BHD Consortium (Table 1),[5] we made a diagnosis of BHDS based on 1 major criterion (an *FLCN* germline mutation) and 2 minor criteria (multiple lung cysts and a first-degree relative with BHDS).

Currently, the patient remains under regular follow-up because of the increased risk of developing renal cancer. Additionally, the patient's first-degree relatives have been referred for gene analysis.

BHDS is an inherited autosomal dominant disorder caused by germline mutations of the folliculin (*FLCN*) gene located on chromosome 17(17p11.2).[5] BHDS-associated renal tumors display inactivation of the wild-type *FLCN* allele (for example, loss of heterozygosity, mutation, methylation), confirming that *FLCN* is a tumor suppressor gene that fits the classic 2-hit model.[6,7] This gene encodes folliculin, a 579 amino acid protein expressed in a variety of tissues