Quality of Life in Patients with Cutaneous T-Cell Lymphoma☆

Calidad de vida en pacientes con linfomas cutáneos de células T

To the editor:

As is well known, primary cutaneous t-cell lymphomas has a chronic course with frequent relapses and patients may go through the stages of patches, plaques, tumors, or erythroderma and also multiple treatments are often needed.[1,2] The presence of these lesions, often extensive and visible, frequently has a detrimental effect on physical appearance, negatively affecting personal and work relationships.[3,4] On the other hand, the intense and sometimes irrepressible pruritus[3] that may sometimes be associated with these entities also clearly results in the deterioration of quality of life in patients with CTCL.[3]

Quality of life impact in CTCL has been evaluated in few occasions.[4] Thus, we initiated this study with the goal of determining the degree to which the quality of life is affected in patients with this diagnosis. We developed a descriptive, cross sectional, and analytical study in which we asked patients with CTCL who availed themselves of our hospital's Cutaneous Lymphoma Unit during the period between December of 2015 and July of 2016 to complete the Dermatology Quality of Life Index questionnaire (DLQI) in its Spanish version (upon authorization from its authors). This test has a minimum total result of 0 points (null impact on quality of life) and a maximum of 30 (extreme impact), and it analyzes the effect of the disease on quality of life in the last 7 days.[5] The patients and their results are classified according to lymphoma subtype, stage, sex, age, and other parameters. The statistical analysis was conducted using the SPSS-16 program in its Spanish version, and statistical significance was set at $P<0.05$ (Mann-Whitney test).

Table 1 Mean DLQI of each suctype LCCT.

CTCL SUBTYPE	Patients	Mean DLQI
MF IIA ≤	37 (82,9%)	1,2
MF IIIB ≥	10 (21,2%)	7,6
Lymphomatoid papulosis	5 (9,43%)	2,2
Anaplastic Large cell lymphoma	1 (1,8%)	8

We included 53 patients with CTCL, whose average age was 57.4 years, 71.6% (38) being men as compared to 28.3% (15) women. The most frequent subtype with 42 cases (79.2%) was classic mycosis fungoides (Figure 1). The average DLQI Test result of all patients was 3.87. Patients with MF in a stage equal to or less than IIA obtained an average result of 1.2, while those presenting an advanced stage showed an average result of 7.6 (moderate impact), these differences being statistically significant ($P<0.05$). Patients with lymphomatoid papulosis (all T1aN0M0B0) had an average result of 2.2 and the patient with CD30+ anaplastic large cell lymphoma an 8 (Table 1). Regarding the test analysis per question, the average values are summarized in Table 2.

CTCLs progresses insidiously and chronically and the visibility of lesions could mar the physical appearance of patients. Moreover, the pruritus these patients suffer can also interfere with the overall function and performance of the individual. Nevertheless, in our study we have obtained a mean DLQI test value of 3.87 for patients with CTCL, which implies that the impact on quality of life by this disease is low, being null (DLQI mean 1.2) in patients with stage IIA MF or less and moderate (DLQI mean 7.6) in patients with advanced stage MF (IIB or greater). On the other hand, the mean value for the first DLQI questions, for a result of 0 to 3, was 2.11, much greater than the rest of the values for the other test questions, which allows us to confirm that pruritus is the factor that most determines decline in the quality of life for these patients (Table 2).

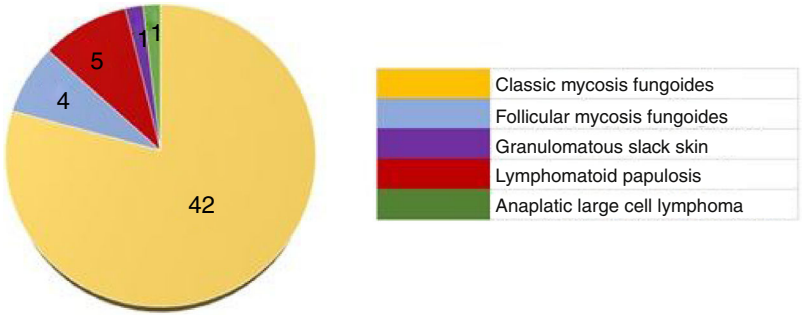

Figure 1 Distribution of each CTCL subtype.

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Table 2 Mean DLQI value of each CTCL subtype.

Question	Mean result (all patients)	MF stage IIA or less	MF stage IIB or greater
Over the last week, how itchy, sore, painful or stinging has your skin been?	2,11	0,71	2,8
Over the last week, how embarrased or self conscious have you been because of your skin?	1,17	0,18	2,2
Over the last week, how much has your skin interfered with you going shopping or looking after your home or garden?	0,09	0,04	0,31
Over the last week, how much has your skin influenced the clothes you wear?	0,09	0,03	0,44
Over the last week, how much has your skin affected any social or leisure activities?	0,07	0,04	0,42
Over the last week, how much has your made it difficult for you to do any sport?	0,05	0,04	0,34
Over the last week, has your skin prevented you from working or studying?	0,08	0,04	0,22
Over the last week, how much has your skin created problems with your partner or any of your close friends or relatives?	0,09	0,03	0,32
Over the last week, how much has your skin caused any sexual difficulties?	0,05	0,04	0,42
Over the last week, how much of a problem has the treatment for your skin been, for example by making your home messy, or by taking up time?	0,07	0,05	0,13
MEAN DLQI VALUE	3.87	1.2	7.6

There is a similar study[4] where a self-administrated questionnaire was answered by patients with CTCL. This study had only cualitative or dicotomic options to evaluate the psychosocial impact of the disease in different areas. They obtained high rates of patients affected in most areas studied (Social, laboral, sexual. . .), but the real intensity of each patient affectation not could be clarified.

As limitations of our study, we would like to highlight the cross-sectional design and the small sample of patients with lymphomatoid papulosis and CD30+ anaplastic large cell lymphoma. Even this study is only a ''snapshot'' in time, the minimal (cuantitative measured) decline in the quality of life for patients with CTCL indicated in our study is noteworthy. This probably may be due to characteristics of the disease itself, since in initial stages (those in which the patient remains for a prolonged period), the lesions are hardly visible and the symptomatology is tolerable. We also consider that, despite having found pruritus to be the factor that most determines the impact on quality of life in these patients, it was underrepresented in the DLQI (it only appears explicitly in one question), which would lead to a lower result regarding the true effect for the patient. Thus, specific and validated tests on the quality of life in patients with CTCL are needed to definitively know the degree to which the individuals suffering from it are affected.

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Clinical and Ultrasound Image of a Cutaneous Fibrolipomatous Hamartoma☆

Imagen clínica y ecográfica de un hamartoma fibrolipomatoso cutáneo

To the Editor:

Cutaneous fibrolipomatous hamartoma (CFH) is characterized by the presence of lobules of mature adipose tissue surrounded by fibrous septa in the mid-to-deep dermis. In the literature, this entity has several different names, although the most widely used is precalcaneal congenital fibrolipomatous hamartoma.[1]

Clinically, the condition presents as a soft, flesh-colored, asymptomatic, solitary mass. It is generally bilateral and symmetrical and located on the inner part of the plantar aspect of the heel, or precalcaneal region. Cases of unilateral involvement have been reported,[2,3] and, although this is a congenital lesion, it is sometimes first noticed years after birth.[3] Most cases are sporadic, although the possibility of autosomal dominant or X-linked inheritance has been postulated.[1,3,4]

The diagnosis is mainly clinical. However, in doubtful cases, such as unilateral involvement or uncharacteristic sites, biopsy is usually the approach taken. The differential diagnosis is with infantile hemangioma, congenital hemangioma, vascular malformations, lipoma, and neurofibroma. Skin imaging with high-frequency ultrasound can facilitate the differential diagnosis and confirm the clinical suspicion, thus obviating the need for invasive diagnostic tests.

Case Description

We present a clinical and ultrasound image of CFH in a 5-month-old girl with no personal history of interest. Clinically, the image shows an asymptomatic flesh-colored mass that was soft in consistency. The lesion had been present since birth and had grown in proportion with the patient. No similar change was observed on the contralateral foot (Fig. 1).

The patient was an only child, and the parents did not have similar abnormalities.

The ultrasound image confirmed the clinical suspicion and made it possible to rule out differential diagnoses (Fig. 2). We did not observe any masses or tumors or clusters of anechoic channels with or without internal flow. The lacunae did not disappear on compression of the skin by the transducer. Doppler mode did not reveal a signal in the interior or periphery of the lesion.

The parents were informed that the lesion was benign and did not require treatment. The lesion was to be reviewed only in the case of changes or onset of symptoms.

Discussion

Skin imaging with high-frequency ultrasound is increasingly used in childhood dermatologic diseases. The differential diagnosis of tumors is one of its main applications. Unlike tumors, hamartomas appear as thickening of the dermis and/or subcutaneous cellular tissue and not as masses. The echogenicity of a hamartoma depends on the most abundant element. In the case of CFH, the reticular dermis thickens owing to the presence of islands of fatty tissue between collagen fibers that can extend to the hypodermis. Ultrasound reveals hypoechoic islands surrounded by hyperechoic bundles in the deep dermis and in the subcutaneous cellular tissue. The literature contains few references to ultrasound assessment of CFH. Cambiaghi et al.[5] present a series of 3 cases and report on the usefulness of ultrasound in the differential diagnosis, although they do not describe or present their imaging findings. Grilo et al.[6] describe CFH as a poorly

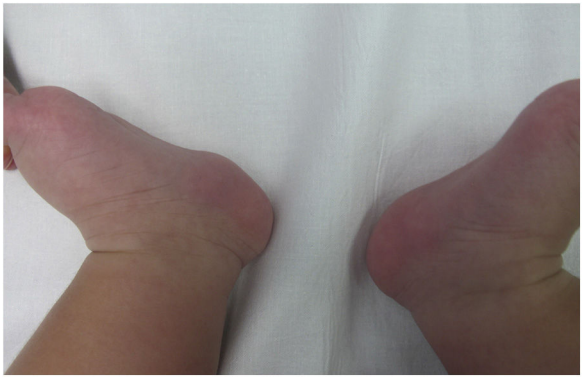

Figure 1 Clinical image of a fibrolipomatous hamartoma in a 5-month-old girl. The image shows a flesh-colored tumor with a soft consistency on the precalcaneal region of the left heel.

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