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Case Report

A Case of "Inflammatory Linear Verrucous Epidermal Nevus" (ILVEN) Treated with CO_2 Laser Ablation

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Abstract

The "inflammatory linear verrucous epidermal nevus" is a rare disease, consisting of hyperplasia of the normal components of the epidermis. Its clinical features include erythematous and hyperkeratosic, warty, sometimes psoriasiform or lichenoid patches with a typical linear arrangement. At present, there are no effective medical therapies available. Currently, the best therapeutic results are obtained with surgical excision or the latest laser therapy. The Authors present a 9-years old girl with an inflammatory linear verrucous epidermal nevus on her neck, successfully treated with CO_2 Laser ablation.

Introduction

The "inflammatory linear verrucous epidermal nevus" (ILVEN) is a rare hamartomatous lesion consisting of hyperplasia of the normal components of the epidermis. Children are more commonly affected, and it predominates in females [1].

Clinically, it is characterised by erythematous and hyperkeratosic, warty lesions, in a linear arrangement which follows the lines of Blaschko.

Typically, the lesions are pruritic and unilateral. They usually occur on a limb following the lines of Blaschko, more rarely on the trunk, in a curvilinear pattern, or in different areas [2, 3].

Case report

An otherwise healthy 9-years old girl showed up to our clinic for a curvilinear, erythematous, warty-like eruption on her neck. The lesion was pruritic and rough to the touch. It was present by less than one year and showed a rapid growth. The past medical and familial histories were insignificant. In the patient's family, no one had a similar lesion or other skin diseases.

The patient did not refer previous treatments of the lesion, except for 10-days of corticosteroids topical application without any beneficial effects.

During the clinical evaluation, no other lesions were observed in any other part of the body.

Routine blood tests showed no involvement of other body regions, a sign of systemic infection or inflammation.

Due to the rapid growth of the lesions, the proven ineffectiveness and invasiveness of medical treatment and other physical therapies, as well as the aesthetic and functional complications of surgery (Table1), accordingly with the patient and her parents, we decided to remove the lesion with a CO_2 laser.

Table 1: CO_2 laser vs. surgical excision

	CO_2 laser	Surgical excision
Controindications	-	+
Pre-operative treatment	-	+
Anaesthesia	-/+	+
Procedure time	Quick	Slow
Complications/side effects	Poor	Possible
Post operative treatment	-	+
Time of wound healing	Days	Weeks

Before starting the treatment, we made an incisional biopsy which confirmed the diagnosis of ILVEN. The histology showed acanthosis, papillomatosis, parakeratotic hyperkeratosis and an inflammatory infiltrate in the upper dermis.

Figure 1: A 9 years-old girl, with an ILVEN on her neck

We used the 10,600-nm CO2 pulsed laser at a frequency of 10 Hz and a level of 1.0-1.5 (Fig. 2).

By the hamartoma's size, we decided to remove the lesion in a singular session. The operation has been performed without anaesthesia and did not require any patient-preparation.

The laser application was rapid and well tolerated by the young patient (Fig. 3).

At the end of the laser treatment, no specific medication was necessary except for the local application of antibiotic ointment for about a week.

No side effects or complications (e.g. scar, pigment modification) were observed in the follow-up the treatment (after one week, one month and three months).

A long-term follow-up (24 months) did not show any signs of recurrence.

Discussion

First described by Unna in 1896 [4], the inflammatory linear verrucous epidermal nevus (ILVEN) is as congenital malformations or hamartomas derived from embryonic ectoderm. It is a rare disease, more common in female [5]. Even if familial cases have been reported, ILVEN is usually sporadic. It usually appears at birth or within the first five years of life, although an adult onset has been described too [6].

Figure 2: The patient during CO_2 laser ablation

Clinically, it presents with pruritic, erythematous and verrucous papules, in a linear distribution following Blaschko's lines. Usually, they are unilateral and localised to the left side of the body [7]. Extremities, especially the limb, are the more common localisation. The length of the nevus is highly variable, and in some cases it can involve the entire limb, causing nails alterations, such as subungual hyperkeratosis and local inflammation. More rarely, ILVEN has been described on the trunk with typical curvilinear transverse bands, which follow the Blaschko lines, sometimes stopping at the midline [8]. Different localisations, like genital or mucosal, are extremely rare [9-11].

Occasionally, as in the "Child Syndrome" ("Congenital naevus Hemidysplasia with inflammatory and Limb Defects"), the epidermal inflammatory hamartoma may be associated with skeletal-articular defects and visceral hypoplasias, which usually occur ipsilateral [12].

Figure 3: The patient immediately after the laser treatment

More rarely, the "epidermal nevus syndrome" has been described. It is characterised by complex developmental abnormalities of skin, eyes, nervous system, skeletal, urogenital and cardiovascular systems [13, 14].

The clinical course and prognosis are varied, depending on the individual characteristic and the possible association with more important organs alterations.

The inflammatory epidermal hamartoma is usually a chronic and progressive disease characterised by periodic inflammatory breakthroughs associated with increased pruritic symptoms, and rarely with microbial superinfection, eczema, or even necrosis. In the long term, the disease can stabilise and may even show spontaneous regression. On the other hand, even if extremely rare, ILVEN has seen to be associated with malignant transformation, such as basal or squamous cell carcinoma and keratoacanthoma [15-17].

The diagnosis of ILVEN is mainly clinical, supported by the medical history and histological examination. The histology is characterised by epidermal hyperplasia of normal components, with acanthosis, papillomatosis, hyperkeratosis and parakeratosis. Diffuse or perivascular inflammatory reactions have been reported in the papillary dermis [18].

If a more complex syndrome is suspected, it is recommended to exclude the involvement of other body parts with specific diagnostic exams (e.g. fundus examination, skeletal X-rays, ultrasounds, abdominal CT).

The treatment of NEVIL is very complex and often frustrating.

At present, there are no effective medical therapies available. Topical corticosteroids, dithranol and retinoids are beneficial in a small percentage of patients [9, 19, 20]. Topical vitamin D analogues, 5-FU and calcineurin inhibitors may be considered as therapeutic options [21, 22].

Currently, the best therapeutic results are obtained with the physical modalities, like surgical excision, cryotherapy, photodynamic and laser therapy. Among these, the last one seems to be the better therapeutic option, because of the low risk of complication and recurrences, and for the excellent aesthetical results [23-27].

In conclusion, due to the ineffectiveness of conventional drug therapies based on steroids and retinoids, and pending the outcome of case-control studies on the effects of new drugs such as derivatives of vitamin D, 5-FU or calcineurin inhibitors, we can state that the most effective therapeutic approach is represented by surgical exeresis and CO_2 laser therapy.

In particular, based on our clinical experience, laser therapy seems to be the best treatment. In fact, compared to surgery, laser therapy has no contraindications and requires no special patient-preparation, such as anaesthesia is not always necessary. The laser action is faster, less invasive and less destructive than surgery. Side effects are practically absent and, even more importantly, there is no contact therapy or any related complications. The laser lesions showed rapid re-epithelisation and the aesthetic and functional results were excellent. Our patient and her parents were very satisfied not only with the effectiveness of the operation but also with the speed of the treatment, the tolerability and absence of any special pre- or post-operative treatments or precautions except for short-term local antibiotic therapy.

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