LETTERS TO THE EDITOR

Smooth Muscle Hamartoma Mimicking Multiple Eccrine Hidrocystomas

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To the Editor:

Eccrine hidrocystomas are rare cystic ducal lesions of the eccrine sweat gland. They were described by Robinson1 in 1983 as multiple lesions that appeared on the face of women who worked in hot and humid environments. In 1973, Smith and Chernosky2 described another group of patients in whom the lesion presented singly or in small numbers. Since then, these forms of presentation have become known as the classic or Robinson type and the solitary or Smith type, respectively. They present clinically as transparent or slightly blueish vesicular papular lesions measuring between 2 and 4 mm, on the face and torso. The lesions typically improve in winter and worsen in summer when they are exacerbated by sweat-producing stimuli, such as exercise and a warm humid environment; they may also be exacerbated by hormonal abnormalities such as hyperthyroidism.3

Smooth muscle hamartoma is a benign process characterized by a proliferation of bundles of smooth muscle in the dermis. Almost all cases are congenital but acquired cases, such as ours, also exist. These acquired forms are extremely rare and very few cases have been reported in the literature; to date, we have found only 1 published case of lesions on the face.4

We present the case of a 38-year-old man who visited our department due to the progressive appearance over 4 years of multiple translucent, skin-colored papular lesions grouped on the right cheek (Figure 1). These lesions increased in size in summer and on exercise (associated with heat and sweat) and improved in winter. A clinical diagnosis of multiple eccrine hidrocystomas was made and a biopsy was performed for confirmation. The biopsy revealed a smooth muscle hamartoma compressing the eccrine glands, thereby causing cystic ectasia of both the secretory coils and the eccrine ducts. Immunostaining with anti-actin antibodies and Masson-trichrome stain was positive in muscle fibers (Figure 2). Treatment was initiated with antiperspirant drugs (aluminum chlorohydrate, 20%) and produced a slight improvement; the patient’s symptoms have since remained stable with no new lesions or growth of the existing lesions.

Although the pathogenesis of eccrine hidrocystoma is not well understood, some authors consider that the lesion is due to retained sweat resulting from obstruction of the excretory part of the sweat gland; it would also explain why the lesions were unilateral in our patient—unlike previously published cases.

Treatment of solitary or Smith-type forms will essentially consist of surgical removal of the lesions. In multiple or Robinson-type forms, however, surgical treatment is not appropriate; several other therapeutic options have therefore been tried, with varying results. Patients should firstly be advised to avoid situations that trigger worsening of the lesions, such as exercise and very hot environments. Oral atropine has also been used, but the systemic side effects outweigh the benefits. However, topical

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Figure 1. Multiple flesh-colored papules grouped on the right cheek.

Figure 2. Masson-trichrome stain. ×10. Histology revealed a smooth muscle hamartoma compressing the eccrine glands and causing cystic ectasia of both the secretory coil and the eccrine ducts.
application of 1% atropine has produced a response in some patients, but not in others. Other treatments include the use of CO2 lasers or pulsed dye laser, destruction using electrosiccation with a fine needle electrode, and injections of botulinum toxin. In cases where there was an underlying disorder such as hyperthyroidism, resolution of the disorder led to remission of the lesions.

REFERENCES

Disseminated Lobular Capillary Hemangiomas

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To the Editor:

Lobular capillary hemangiomas are very common benign vascular lesions that frequently occur following injuries to the gums, lips, nasal mucosa, face, fingers, and toes. More rarely these hemangiomas present in multiple or multifocal form, in some cases forming part of a paraneoplastic syndrome, but in other cases with no apparent underlying disease. Rarer varieties have also been described, such as subcutaneous or intravenous lesions. Lobular capillary hemangiomas have also been reported following the use of certain drugs such as oral retinoids.1,2

We describe the case of a male patient aged 74 years with a history of long-standing malignant hypertension, chronic renal insufficiency due to nephroangiosclerosis, anemia of inflammation, and colonic polyposis with multiple polypectomies leading to histologic evidence of tubular or tubulovillous adenomas. The patient was being treated with carvedilol, torsemide, acetylsalicylic acid, nifedipine, enalapril, and erythropoietin.

The patient reported the appearance, over a period of 4 to 5 years, of sessile cutaneous lesions and subcutaneous nodular lesions, soft in consistency, reddish-violet in color, variable in size (measuring 0.5 to 2 cm in diameter), and located mainly on the trunk and neck (Figure 1). There was no involvement of the mucosa.

Blood tests were normal with the exception of a hemoglobin level of 9.4 mg/dL. Biochemical analysis revealed creatinine of 3.22 mg/dL, urea of 114 mg/dL, and uric acid of 8.24 mg/dL, with all other values within the normal range.

Several lesions were excised for histology studies, which revealed Figure 1. Multiple sessile, violaceous lesions on the trunk.

Figure 1. Multiple sessile, violaceous lesions on the trunk.
lobulated dermal lesions composed of thin-walled vascular channels lined with endothelial cells in nests or clusters, separated from each other by a fibrous stroma (Figure 2).

A computed tomography scan of the chest, abdomen, and pelvis showed no findings of interest. Colonoscopy revealed 6 new polyps, all of which were removed.

Single photon emission computed tomography (SPECT) with red blood cells labeled with technetium 99m (Tc-99m) revealed a number of subcutaneous nodules suggestive of hemangiomas in the dorsal, lumbar, and left maxillary regions and in the limbs. A similar nodule was also encountered in the posterior segment of the upper right pulmonary lobe (Figure 3).

We conclude that the appearance of multiple lobular capillary hemangiomas is rare. Cases have been described of lesions appearing de novo in patients with no underlying disease. Other cases have been reported in which the lesions were associated with previous injuries, due, for example, to the removal or electrocautery of a single lesion resulting either from a burn or from exfoliative dermatitis. Lesions have also been reported to appear following treatment with methotrexate, etretinate, or granulocyte-colony stimulating factor. Cases have been observed in which lesions were associated with malignant processes such as Hodgkin disease, chronic lymphatic leukemia, malignant melanoma and myeloma, immunodeficiencies (such as an interleukin-2 deficiency), and chronic inflammatory or infectious processes such as glomerulonephritis, hypertension, and cerebrovascular accidents.

Intestinal hemangiomas have been reported in patients with multiple cutaneous lobular capillary hemangiomas, although this association only occurs in less than 2% of cases. This possibility should be taken into account in patients presenting with associated symptoms such as melena or progressive anemia. SPECT imaging with Tc-99m-labeled red blood cells is very useful for detecting hemangiomas. In our patient, in addition to the subcutaneous lobular capillary hemangiomas, SPECT led to detection of the same kind of lesion in the lung.

The possible trigger for the angiomatous lesions could be the multiple chronic systemic disease presented by the patient.

Most lesions resolve spontaneously; if not, they can be treated using cryotherapy, electrocautery, surgical excision, or pulsed dye laser. In the case of our patient, lesions causing discomfort were excised and then a wait-and-see attitude was adopted.

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Letters to the Editor


Umbilical Pilonidal Sinus as a Possible Complication of Depilation

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To the Editor:

Pilonidal sinus is a chronic inflammatory disorder caused by a hair fragment penetrating the skin and producing a foreign body reaction leading to the formation of a sinus coated with granulation tissue. The disease commonly occurs in the sacrococcygeal region, but can also develop in other locations where an anatomical cleft facilitates the accumulation of hair; these locations include between the breasts, the axilla, the perineum, or in the spaces between the fingers (in the case of barbers in particular). The occurrence of the disease in the navel is rare.

A 28-year-old man came to our clinic with inflammation and suppuration in his navel that had commenced some 2 months previously. Meticulous examination revealed a sinus tract from which a number of hair fragments were extracted. The patient, who was hirsute and whose weight was appropriate for his height, had been shaving his body with a razor since about 4 months previously. The removal of the hairs from the cavity alleviated the symptoms, and no recurrence was evident 6 months later.

Most cases of umbilical pilonidal sinus present as recurrent omphalitis with pain, suppuration and bleeding, or even as an umbilical mass. Pilonidal sinus typically affects young, hirsute men, often with poor personal hygiene. Obesity and sweating are other factors that facilitate hair entry in the epidermis.¹ The literature does not refer to depilation as a risk factor for the development of umbilical pilonidal sinus, possibly because the interest in depilation among men is a fairly recent development. In our patient, fragments of hair cut from the chest and abdomen very likely settled within the navel, resulting in the formation of the pilonidal sinus.

Diagnosis is clinical and based on the detection of hairs nesting deep within the navel. Pathology reveals a foreign-body granuloma, with an epithelium-lined tract leading to an area of fibrosis and granulation tissue enveloping the hair fragments. This entity should be included in the differential diagnosis of umbilical lesions, such as, for example, epidermal cysts, umbilical hernias, pyogenic granulomas, endometriosis, omphalomesenteric duct remnants, urachal anomalies, and metastatic tumors.²

Most patients are cured by conservative treatment involving the extraction of the hair fragments and other debris from the cavity and, if necessary, the administration of oral antibiotics.³,⁴ Omphalectomy should only be resorted to for difficult-to-treat cases. In order to avoid the possibility of recurrence, navel reconstruction is not recommended, it being preferable to allow the surgical wound to heal by second intention.

References


Pigmented Eccrine Poroma

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To the Editor:

Eccrine poroma is a rare tumor that displays variable morphology.

Its clinical variability means it can adopt the appearance of other cutaneous tumors, whereby diagnosis is only rarely made on a clinical basis and must be confirmed by pathology.
Letters to the Editor

We present a case of pigmented eccrine poroma.

A 42-year-old woman was referred to us for the assessment of a pigmented lesion on the palmar surface of the third finger of the right hand. She had no relevant medical or surgical history and was not receiving any treatment.

The patient reported that the lesion had appeared on the palmar surface of the second phalanx of the third finger of the right hand 10 months previously. The lesion had grown slowly and had bled several times in the last month when knocked.

Examination revealed a pigmented, papular, oval-shaped lesion, 5 mm in diameter, that was uniform, clearly defined, and had an irregular verrucous surface. No ulceration was present (Figure 1).

Histopathology revealed thickening of the epidermis in broad anastomosed bands of small homogeneous, cuboidal cells with round basophilic nuclei. The edges of the tumor were clearly defined with no evidence of peripheral palisading, and intercellular accumulations of pigment were present (Figures 2 and 3).

These clinicopathologic findings led to diagnosis of pigmented eccrine poroma.

The lesion was completely excised and the patient has progressed favorably to date with no recurrence of the lesion.

Pigmented eccrine poroma is a rare skin adnexal tumor and accounts for less than 1% of primary cutaneous lesions. The term poroma refers to a group of benign skin adnexal tumors with poroid or distal ductal differentiation which can be of eccrine or apocrine lineage.

The various histopathologic variants of poroma (hidroacanthoma simplex, classic poroma, dermal duct tumor, and apocrine poroma) have clinically distinct morphologies, and there have been occasional descriptions of forms showing combinations of different neoplastic patterns.

Classic poroma is a tumor that generally occurs in patients aged over 40 years, with a slight predominance amongst women. These tumors have generally been reported to be found on the palms of the hands (10%) and soles of the feet (65%), although they can occur in many other locations.

Clinically, poroma can vary widely in form, consistency, and color. In most cases they occur as a solitary lesion in the form of an asymptomatic papule or nodule (with the possibility of bleeding and pain following trauma), in well-defined, sessile, or pediculate form, sometimes developing a papillomatous surface, and measuring between 1 and 5 cm in diameter. The color of the lesion can range from that of normal skin, or similar, to that of pyogenic granuloma or hemangioma. Exceptional cases of pigmented forms have been described, as is the case here.

Histologically, there is a discrete proliferation of cuboidal keratinocytes with rounded or oval monomorphic nuclei and scarce cytoplasm, while the uniform poroid cells form islands. These cells are similar to those of the peripheral cell layer in the most distal portion of the eccrine and apocrine duct. Clearly defined areas of necrosis in the center of the islands of tumor cells are characteristic of the condition. Cystic spaces can also present along with areas of keratinization, clear and pallid cells, dendritic melanocytes, and melanin, etc.

Poroma behaves biologically like a benign neoplasm, with the possibility of local persistence if incompletely excised.

It is important to note that the clinical characteristics of poroma are not specific. It can adopt the appearance of other cutaneous tumors, and as a result, differential diagnosis should include conditions such as pyogenic granuloma, hemangioma, seborrheic keratosis, fibroma, and even nodular and amelanotic melanomas. Clinical examination rarely proves sufficient for diagnosis and histologic confirmation must be obtained.

References

Low-Dose Isotretinoin For Treatment of Chronic Discoid Lupus in Women of Childbearing Age

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To the Editor:

We present the case of a 29-year-old woman diagnosed with systemic lupus erythematosus in 1988 on the basis of malar erythema, photosensitivity, arthralgia, thrombocytopenia, lymphopenia, and positive results for antinuclear and anti-DNA antibodies. She consulted in August 2004 with multiple erythematous plaques with atrophic centers on the upper region of the trunk and face, along with erythematous lesions with a hyperkeratotic appearance on both palms (Figure 1), with onset several months previously. Both types of lesion were compatible with lupus. At the time of consultation the patient was being treated with prednisone, mycophenolate, and chloroquine, without improvement in the cutaneous lesions. Treatment was prescribed with isotretinoin 40 mg/d, with agreement from the patient to use effective means of contraception. A rapid response was observed and the drug was well tolerated, so treatment was maintained for 6 months. The palmar lesions reappeared from 1 month after the drug was withdrawn. Topical treatment was administered for approximately a year, with no significant changes in triglycerides or hepatic enzymes. The patient used safe methods of contraception during the treatment period. There were no side effects apart from cheilitis and slight xerosis on the face.

Chronic discoid lesions are one of the most common forms of cutaneous lupus erythematosus. These are most commonly found on the face, scalp, and the ears, although palmar lesions are also possible.

Chronic discoid lupus erythematosus can be treated with various topical drugs, like potent corticosteroids, or imiquimod 5%, as well as systemic treatments like thalidomide, hydroxychloroquine, or acitretin. The last 2 are first-line drugs with similar levels of effectiveness, although acitretin has more associated side effects. Side effects include cutaneous xerosis, cheilitis, gastrointestinal disorders, increased serum levels of triglycerides, and high risk of teratogenic effects, which oblige patients to use contraceptive measures during treatment and for 2 years after the drug is discontinued.

Meanwhile, isotretinoin has been shown to be effective in the treatment of chronic discoid lupus erythematosus, and it can also be employed as a maintenance treatment at doses of 40 mg on alternate days. This treatment has similar side effects to acitretin but a lower risk of teratogenesis, which means that female patients prescribed the drug need only take contraceptive measures during treatment and for a month after this is suspended. In the case described here, the patient was planning a pregnancy in the medium term, whereby acitretin was rejected in favor of isotretinoin.

Figure 1. Hyperkeratotic erythematous lesions on both palms.

Figure 2. Improvement of lesions following 5 months of treatment with isotretinoin (20 mg/d).
Letters to the Editor

Given that most women with chronic discoid lupus are of childbearing age, it is encouraging to encounter a treatment that allows for patients to plan pregnancies more easily.

Similarly, we stress that maintenance treatment with lower doses of isotretinoin (20 mg/d) can be useful, allowing lesions to be controlled with reduced systemic side effects.

References

Purpura After Application of a Eutectic Mixture of Local Anesthetic

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To the Editor:

Eutectic mixture of local anesthetic (EMLA) is a topical cream containing lidocaine (25 mg/mL) and prilocaine (25 mg/mL) as active ingredients in an excipient of polyoxyethylene and carboxypolymethylene, which produces a pH of 9.4. EMLA is approved by the US Food and Drug Administration for patients aged from 3 years, to be applied on unbroken skin. This product is marketed in Spain as a cream (in 5 g and 30 g tubes) and as 4-cm self-adhesive patches, and it is frequently used as a topical anesthetic by dermatologists and pediatricians.

The most important side effects in terms of frequency include local blanching and erythema in the area of application, while less-common effects include contact urticaria, irritant dermatitis, allergic contact eczema, and purpura. Systemic complications are exceptionally rare, although the presence of methemoglobinemia—attributed to the metabolite of prilocaine—has been described in children, leading to headaches, blurred vision, unstable walking, cyanosis, and methemoglobin in the blood.

We report episodes of purpura as a result of EMLA use in 2 pediatric patients and review the literature, in which only a few cases of this effect have been described. The 2 patients, aged 9 years and 18 months, were both girls with a history of atopic dermatitis. They were prescribed EMLA with an occlusive dressing for 90 minutes prior to curettage of multiple molluscum contagiosum lesions. When the plastic film was lifted an asymptomatic petechial eruption was seen in the occluded zone (Figure) that resolved without treatment or sequelae within 2 weeks. Biopsy was not considered necessary given the characteristic clinical appearance of the lesion.

Purpura as a reaction to EMLA is a rare complication which we have found in only 25 published cases.1-4 The exact pathogenic mechanism is not known, but it appears to have a toxic rather than allergic basis.3 Although no patch tests were carried out on our patients, other authors report negative results from such tests both in immediate readings (30 minutes) and those taken at 48 and 72 hours.3

It is well known that EMLA cream can have a direct effect on blood vessels, causing blanching and erythema, and that it can even produce structural changes in the vessel walls that lead to extravasation of red blood cells and, consequently, purpura. Atopic dermatitis probably acts as a risk factor, given that abnormalities in the barrier function of the skin favor greater absorption of the drug.1 The fragility of the skin and blood vessels in children—as shown by purpuric eruptions resulting from efforts like vomiting, coughing, and crying—and the potential trauma from the plastic in the occlusive dressing can also influence the appearance of this uncommon side effect.1

Figure. Purpura in the area treated with eutectic mixture of local anesthetic.
Letters to the Editor

References

Unilateral Nevoid Hyperkeratosis of the Nipple and Areola Treated With Topical Calcitriol

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To the Editor:

Hyperkeratosis of the nipple and areola (HNA) is an uncommon condition characterized by verrucose thickening and café-au-lait pigmentation of the nipple and areola.1 It was first described by Tauber in 1923 and approximately 70 cases have been published to date in the medical literature. Several therapeutic approaches have been used to treat this condition, although no gold standard has yet been established.

We report the case of a 35-year-old man with a 5-year history of mildly pruriginous dermatosis on the nipple and areola of the left breast. The dermatosis started as an erythematous area and progressed to form a hyperpigmented plaque with thickening of the skin during the 18 months before the visit. The patient applied topical fluocinolone acetonide for 2 years, with some initial improvement; however, shortly after the patient started the treatment, the dermatosis spread.

Physical examination revealed a verrucose, hyperpigmented, hyperkeratotic, oval plaque measuring 3.8 cm in diameter and surrounded by a café-au-lait macule measuring 15 cm in diameter (Figure 1). The remainder of the examination was unremarkable.

The initial clinical diagnosis was seborrheic keratosis. A punch biopsy was performed, and staining with hematoxylin-eosin revealed the presence of hyperkeratosis, acanthosis, and papillomatosis (Figure 2). Based on the combination of histopathology and clinical findings, a diagnosis of unilateral nevoid hyperkeratosis of the nipple and areola (NHNA) was made.

Treatment was started with topical calcitriol, 0.3%. The ointment was applied twice per day for 6 months and the hyperkeratosis resolved completely, although it left residual hyperpigmentation of the skin. The patient showed no side effects after applying the medication.

HNA has been classified in 3 groups: HNA that presents as an extension of an epidermal nevus; HNA associated with other dermatoses such as acanthosis nigricans, cutaneous T-cell lymphoma, Darier disease, chronic eczema, ichthyosis, and ichthyosiform erythroderma; and idiopathic HNA, also known as NHNA.1-6

NHNA is more common in female patients (80%) aged 10 to 30.1,2,4,7 To date, only 11 cases have been reported in men.2,3,6,8,9 The lesions are generally bilateral and affect the nipple and areola in 72% of cases and the nipple only in 28%.5

The cause is unknown, but several factors, mainly endocrine factors,6,7 seem to affect the development of this condition. Clinically, it is characterized by verrucose, hyperkeratotic, and hyperpigmented lesions on the areola...
that extend to the nipples.\textsuperscript{3} The lesions are generally asymptomatic,\textsuperscript{5,6} although a case of NHNA reported by Pérez-Izquierdo et al\textsuperscript{6} prevented lactation.

The typical histopathologic findings are hyperkeratosis with the formation of keratotic plugs, acanthosis, and papillomatosis.\textsuperscript{7}

The differential clinical diagnoses to be considered are Paget disease, superficial basal cell carcinoma, dermatophytosis, and Bowen disease. The differential histopathologic diagnoses are verrucous epidermal nevus and acanthosis nigricans.\textsuperscript{1}

Some therapeutic options that have proven effective in the treatment of this condition are salicylic acid, 6%, lactic acid lotion, topical corticosteroids, topical tretinoin, oral vitamin A, and topical calcipotriol.\textsuperscript{8,10} Kubota et al\textsuperscript{9} reported favorable cosmetic results with cryotherapy using liquid nitrogen. Surgical resection is also effective.\textsuperscript{1}

This case is interesting, as NHNA is an uncommon condition, especially in men. Furthermore, although vitamin D analogues have traditionally been used in the past to treat this condition, the case presented here is the first in which topical calcitriol has been used. This agent inhibits cell proliferation and induces keratinocyte differentiation; therefore, it can be an option for therapy, although studies with larger numbers of patients are necessary.

References