

New Entities in Dermatology

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Abstract

Dermatology is a rapidly developing medical field, in which too many new advances in terms of diagnosis and therapy have occurred within the last several years. Technological progression of chemical science and imaging methods as well as new tools have made it possible to expose the mysterious aspects of diseases. Therefore, novel dermatologic entities have been described day by day. In this article we decided to search new entities reported electronically in Pubmed and EBSCO indexes in the last decade after year 2002. Twenty-six new entities were found out after our investigation. In this review, the reported entities have been classified according to the sections of dermatologic disorders. We think that this article would help to update the knowledge of dermatologists about new concepts.

INTRODUCTION

Dermatology is a rapidly developing medical field, in which too many new advances in terms of diagnosis and therapy have occurred within the last several years. As dermatology involves a wide range of diseases affecting all age groups, ranging from inherited diseases to acquired disorders such as cancers and aging these advances have been parallel to that of general medicine. Technologic progression of chemical science and imaging methods as well as new tools have made it possible to expose the mysterious aspects of diseases and thus many unknown disorders have been found out, whilst causes of known diseases have been clarified. Therefore, novel dermatologic entities have been described, although some of them have been remained limited to only one or a few cases, and added to the indexes.

In this context, we decided to search new entities reported electronically in Pubmed, EBSCO indexes in the last decade after year 2002. Twenty-six new entities were found out after our investigation. In this review, the reported entities have been classified according to the sections of dermatologic disorders. We think that this would help to update the knowledge of dermatologists.

PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

PEDIATRIC BLASCHKITIS

Controversy exists regarding the pathogenesis and classification of blaschkitis and lichen striatus within the spectrum of blaschkolinear dermatoses.

Keegan et al. reported two young children who developed relapsing, pruritic, papulovesicular eruptions in multiple bands along Blaschko lines on the neck, trunk, and extremities. Skin specimens in both revealed spongiotic dermatitis.(1) Denk reported a 2½-year-old girl presented with multiple grouped papules along the lines of Blaschko on her trunk. They differentiate this eruption from lichen striatus and propose the term blaschkitis in children (2).However Müller et al. reported that blaschkitis and lichen striatus had similar histopathological findings with respect to epidermal changes and composition of the inflammatory infiltrates. They propose these dermatoses are located in the same spectrum of a disease rather than being an entity.(3)

CIRCUMSCRIBED PALMAR OR PLANTAR HYPOKERATOSIS

Circumscribed palmar or plantar hypokeratosis is a rare benign epidermal malformation of the skin. Perez et al., have described firstly this entity as an asymptomatic, well-circumscribed, and depressed erythema persisting for many years on the palms or soles. Its main histopathologic feature shows a characteristic epidermal depression with an abrupt decrement in the thickness of the stratum corneum, with a sharp stair between normal and involved skin. (4). Urbina et al., using transmission electron microscopy, showed a reduction in keratin bundles and keratohyaline granules, and increased lipid in the horny layer, suggesting a primary disorder of keratinization.(5). Only in one recently reported case malignant transformation has been described. (6)

PRETIBIAL ANGIOPLASIA

Mistry et al presented 8 patients who had lesions that clinically resembled necrobiosis lipoidica but showed histopathological features of venous insufficiency on skin biopsy. The patients' ages ranged from 39 to 73 years. Only one patient was female. All patients had lesions on the legs, most on the anterior aspect. The clinical diagnosis was generally necrobiosis lipoidica; other clinical impressions included lichen planus, morphea, and Kaposi sarcoma. All patients had features on pathology resembling venous insufficiency and no features of other dermatoses. (7)

SAURIAN PAPULOSIS

Molina-Ruiz AM et al described a new epidermal keratinization disorder in two patients with a long-standing widespread cutaneous eruption. The eruption was characterized by symmetric erythematous, flat, discrete papules with a polygonal shape and fine scaling. The papules covered most of the skin surface and, in some areas of the trunk, they were arranged along the lines of cleavage, parallel to the ribs. There was no facial, mucosal, nail, or palmoplantar involvement; the teeth and hair were normal. The first patient had a sister with an identical eruption, and a brother of the second patient was said to have similar skin lesions. Histopathology revealed well-demarcated areas of compact eosinophilic orthokeratotic hyperkeratosis overlying a slightly acanthotic epidermis. Lesional skin showed weaker immunoreexpression for connexin 43 compared with normal skin. They propose the name "saurian papulosis" to describe this newly described clinicopathologic entity (8).

VESICULOBULLOUS DISEASES

ANTI-P200 PEMPHIGOID/ ANTI-LAMININ GAMMA1 PEMPHIGOID

Anti-p200 pemphigoid is a recently defined autoimmune subepidermal blistering disease characterized by circulating and tissue-bound autoantibodies to a 200-kDa protein (p200) of the dermal-epidermal junction (DEJ). While the exact identity of p200 remains unknown, it has been demonstrated to be immunologically and biochemically distinct from all major autoantigens of the DEJ, including bullous pemphigoid antigens 180 and 230, laminin 1, 5 and 6, alpha6beta4 integrin, and type VII collagen. Clinically, most reported cases present with tense blisters as well as urticarial papules and plaques, closely resembling bullous pemphigoid. Histopathological examination of lesional skin biopsies shows subepidermal split formation and superficial

inflammatory infiltrate typically dominated by neutrophils. Immunopathologically, linear deposits of immunoglobulin (Ig)G and C3 are detected along the DEJ by direct immunofluorescence microscopy of perilesional skin. Indirect immunofluorescence microscopy of patients' sera on NaCl-split human skin demonstrates circulating IgG autoantibodies labeling the dermal side of the split. (9)

Dainichi et al. reported 70 cases with this newly defined autoimmune subepidermal bullous disease and renamed this disease from the previously used anti-p200 pemphigoid to anti-laminin gamma1 pemphigoid, a new entity of an autoimmune bullous disease. (10)

ADNEXAL DISEASE

CHILDHOOD FLEXURAL COMEDONES

A new form of comedones, known as childhood flexural comedones, has recently been reported.(11) The authors described the development of comedones in the large skin folds during childhood, in which the comedones had 2 orifices connected via a thin layer of epidermis. The appearance of comedones is usually related to acne, hidradenitis suppurativa, chronic sun damage, or other types of cutaneous damage. It can also occur after molluscum contagiosum infection.(12)

WIDESPREAD POROKERATOTIC ADNEXAL OSTIAL NEVUS

Porokeratotic eccrine ostial and dermal duct nevus and , porokeratotic eccrine and hair follicle nevus, are rare disorders of keratinization with eccrine and hair follicle involvement.

Goddard et al. reported 5 cases with widespread skin involvement following the lines of Blaschko. Three patients had histologic involvement of both acrosyringia and acrotrichia. Based on the observation of overlapping histologic features, they propose the name "porokeratotic adnexal ostial nevus" to incorporate the previously described entities porokeratotic eccrine ostial and dermal duct nevus and porokeratotic eccrine and hair follicle nevus. (13)

NEVUS TRICHOLEMMOCYCTICUS

The names "trichilemmal cyst nevus" or "nevus trichilemmocysticus" were firstly proposed by Tancheva-Poor I et al. (14) They reported a 31 year-old woman with an organoid nevus characterized by multiple trichilemmal cysts arranged in a bandlike pattern. They realized that some histopathologic features of this complex nevus were

reminiscent of those of well-established organoid nevi such as nevus comedonicus, porokeratotic eccrine nevus, or hair follicle nevus, but the presence of multiple large trichilemmal cysts was a conspicuously distinctive abnormality. Afterwards Laralde et al. reported a nevus tricholemmocysticus in a 10 year-old girl. (15)

Metabolic and systemic diseases

NEPHROGENIC SYSTEMIC FIBROSIS

Nephrogenic systemic fibrosis (NSF) was first described in 2000 as a scleromyxedema-like illness in patients on chronic hemodialysis. This condition was formerly known as nephrogenic fibrosing dermopathy as it was initially observed in, and thought to solely affect, the skin. However, it is now known that several organs such as liver, lungs, muscles and heart may be involved. (16) The relationship between chronic kidney disease (CKD) and gadolinium contrast during MRI was postulated in 2006, and subsequently, virtually all published cases of NSF have had documented prior exposure to gadolinium-containing contrast agents.(17,18). Skin lesions may evolve into poorly demarcated thickened plaques that range from erythematous to hyperpigmented. With time, the skin becomes markedly indurated and tethered to the underlying fascia. The diagnosis of NSF is based on the presence of characteristic clinical features in the setting of chronic kidney disease, and substantiated by skin histology.(19)

Genodermatoses

PYODERMA GANGRENOSUM, ACNE, AND SUPPURATIVE HIDRADENITIS (PASH)

PAPA syndrome is a recently identified hereditary autoinflammatory syndrome clinically characterized by pyogenic arthritis, severe acne, and pyoderma gangrenosum. It is caused by mutations in the PSTPIP1 gene. Braun Falco et al. reported two patients with pyoderma gangrenosum and acute or remittent acne conglobata, but, in contrast to PAPA syndrome, lacked any episodes of pyogenic arthritis. Instead, they had suppurative hidradenitis. Mutations in PSTPIP1 exons 1 to 15 were excluded. In the promoter region, an increased repetition of the CCTG microsatellite motif was present on one allele in both patients. Alterations of the most commonly affected exons of the MEFV, NLRP3, and TNFRSF1A genes also were not detectable.(20) They propose the acronym “PASH” syndrome for this new autoinflammatory disease.

THE H SYNDROME

Molho-Pessach et al. reported 10 patients with cutaneous hyperpigmented, hypertrichotic, and indurated patches associated with hearing loss, short stature, cardiac anomalies, hepatosplenomegaly, scrotal masses, and hypogonadism. Laboratory evaluation of the patients revealed growth hormone deficiency and hypergonadotropic hypogonadism with azoospermia. Cutaneous histopathologic examination showed hyperpigmentation of the basal layer with seborrheic-keratosis-like acanthosis, histiocytic infiltration, and a perivascular mononuclear infiltrate with plasma cells and mast cells throughout the dermis and subcutaneous fat. They suggested this is a novel multisystemic autosomal recessive inherited disorder and call the name “H syndrome”.(21)

Hereditary, focal, transgressive palmoplantar keratoderma with associated clinical findings:

PIGMENTARY DISORDERS

ACQUIRED BRACHIAL CUTANEOUS DYSCHROMATOSIS

Acquired brachial cutaneous dyschromatosis (ABCD) was first described by Rongioletti and Rebra as a pigmentary disorder primarily of the dorsal aspects of the forearms, which occurred in a post-menopausal, woman, with a long-standing history of hypertension and ACEI usage. (23) It was characterized by asymptomatic, irregular, gray-brown patches with geographic borders, which were occasionally interspersed with hypopigmented, slightly atrophic macules. The histopathological examination of this entity reveals epidermal atrophy, basal-layer hyperpigmentation, solar elastosis, and superficial telangiectases.

Hu et al. described a case of ABCD and emphasize the effect of cumulative UV damage in the etiology of this entity. (24)

CONFETTI-LIKE MACULAR ATROPHY: A E?

Aksoy et al, reported two female patients with hypopigmented, atrophic, shiny macules on the upper limbs and upper trunk. Histopathological examinations revealed an atrophic epidermis with disorganized, hyalinized and coarse collagen bundles in the middle and lower dermis. Elastic fiber loss and fragmentation were detected in the upper dermis. They explained that their cases did not show depressed or herniated atrophic macules clinically but the macules were at the same level with the surrounding healthy skin and the histopathological findings showed the histopathological features of both atrophodermis and

anetoderma. They propose that their cases may represent a clinicopathological entity which has not been described before. (25)

HAIR, NAILS AND MUCOUS MEMBRANES

ALOPECIC AND ASEPTIC NODULES OF THE SCALP (PSEUDOCYST OF THE SCALP)

Alopecic and aseptic nodules of the scalp (AANS) is reported first in Japan as 'pseudocyst of the scalp'.(26) Abdennader studied on 15 cases with AANS and 7 cases with dissecant folliculitis. They found that in contrast to dissecting cellulitis of the scalp, the lesions in AANS are not fluctuating and the surrounding scalp is normal, there is no association with acne conglobata or hidradenitis suppurativa. In all the cases of AANS in which the puncture was positive, the material was sterile with negative bacteriological and mycological cultures. The associated alopecia is nonscarring and the histopathological examination is a nonspecific inflammation. Most of the cases with AANS responded well to doxycycline treatment. (27)

CONNECTIVE SKIN DISEASES

FACIAL BLASCHKITIS

Linear configurations of cutaneous lupus erythematosus or discoid lupus erythematosus have rarely been described. Treatment with systemic corticosteroids and anti-malarial agents resulted in remission.(28) Imhof et al reported a 15-year-old female with an erythematous cutaneous eruption on the right cheek and perioral area which is in a linear distribution following the lines of Blaschko. Histopathological findings and direct immunofluorescence were compatible with chronic cutaneous lupus erythematosus but treatment with topical steroids and systemic antimalarial agents over 2 months showed hardly any improvement contrary to similar cases reported in the literature in the past. Therefore, they recommended to call this new entity LE-like facial Blaschkitis of the adult.(29)

TOXIC EPIDERMAL NECROLYSIS-LIKE ACUTE CUTANEOUS LUPUS ERYTHEMATOSUS AND THE SPECTRUM OF THE ACUTE SYNDROME OF APOPTOTIC PAN-EPIDERMOLYSIS (ASAP)

Toxic epidermal necrolysis-like lesions have been described in the setting of lupus erythematosus, and have been considered as a specific hyperacute variant of cutaneous lupus erythematosus, with features different from classical drug-related toxic epidermal necrolysis (TEN).(30) The term "acute syndrome of apoptotic pan-epidermolysis" (ASAP) was proposed by Ting et al., to include all the clinical

situations of massive and acute epidermal cleavage resulting from apoptotic injury. ASAP comprises drug-induced TEN, TEN-like lupus erythematosus, and other conditions as acute graft versus host disease and TEN-like pseudoporphyria. (31)

Subcutaneous fat tissue diseases

LIPOATROPHIC LESIONS PRECEDED BY PAIN AND ERYTHEMA

Imamura S et al., reported an adult female patient well-defined, deeply depressed lesions on the abdominal walls, which were preceded by slight pain and erythema of a few days duration, without any history of trauma or injection. The depressed lesion showed a marked decrease of the subcutaneous fatty tissue with minimal cell infiltration. They found three more cases with this similar clinical and histopathological examination from the literature. They pointed that the most important differential diagnosis may be lipodystrophia centrifugalis abdominalis infantilis (LCAI), which is characterized by depressed lesions in the abdominal wall of infants or children. They propose this type of lipodystrophia is distinct from LCAI because their cases were adult and the lesions developed with pain or paresthesia with erythema before the development of the depression. (32)

NEOPLASMS OF THE SKIN

LINEAR SYRINGOMATOUS HAMARTOMA

White et al reported a 4-year-old girl presented with a linear, indurated area of dusky erythema and hyperpigmentation down the left leg, present since birth. The histopathological examination suggested syringomata. They suggested that the clinical course and appearances would be a novel entity and they coined the term 'linear syringomatous hamartoma. (33)

MULTIPLE IDIOPATHIC MUCOCUTANEOUS NEUROMAS

Multiple mucosal neuromas are uncommon and represent a key feature of the multiple endocrine neoplasia syndrome (type 2b), in which pheochromocytoma, medullary carcinoma of the thyroid, and gastrointestinal ganglioneuromas also occur; therefore, its early recognition can be life saving. Multiple mucocutaneous neuromas with a predilection for the face and distal extremities can also represent a manifestation of the PTEN hamartoma-tumor syndrome, which includes Cowden and Bannayan-Riley-Ruvalcaba syndromes.(34) Truncot et al. reported a patient with a history of 13 year multiple mucocutaneous neuromas.

They excluded the multiple endocrine neoplasia type 2b syndrome with the biological, morphological and genetic investigations.(35) They proposed the term multiple idiopathic mucocutaneous neuromas as a new entity.

CUTANEOUS ADULT T-CELL LEUKEMIA/LYMPHOMA

Adult T cell leukemia/lymphoma (ATLL) is a type of T-cell neoplasm etiologically associated with HTLV-1. (ATLL) has been divided into four subtypes: (i) acute; (ii) lymphoma; (iii) chronic; and (iv) smoldering. Smoldering variants often present with patches, plaques and papular skin lesions and the cases showing less than 5% abnormal T-lymphocytes in peripheral blood without involvement of other organs. Amano et al. studied the proviral DNA load of HTLV in smoldering ATLL group, in asymptomatic carrier and cutaneous ATLL group and define the cutaneous ATLL, which has less than 5% abnormal T lymphocyte in peripheral blood, a normal lymphocyte count (i.e. <4 x 10⁹/L), no hypercalcemia and lactate dehydrogenase values of up to 1.5 times the normal upper limit and at least one of the histologically proven skin lesions accompanying monoclonal integration of human T-cell lymphotropic virus type 1 (HTLV-1) proviral DNA in the skin lesion. (36)

EPITHELIAL SHEATH NEUROMA

Requena et al., described four examples of a peculiar cutaneous lesion characterized histopathologically by a proliferation of enlarged nerve fibers ensheathed by squamous epithelium involving the superficial dermis. The perineural epithelial sheaths were composed of uniform squamous epithelium with evidence of cornification in the form of dyskeratotic cells or resulting in orthokeratotic basket-weave corneocytes. Immunohistochemical studies confirmed the epithelial and neural nature of the two components of the lesions, with the nerve fibers expressing immunoreactivity for S-100 protein, neurofilaments, CD57, and nerve growth factor receptor, whereas the perineural epithelial sheaths showed immunoreactivity for cytokeratins. The authors propose the term “epithelial sheath neuroma” for this lesion. (37)

SUBCUTANEOUS PANNICULITIS-LIKE T-CELL LYMPHOMA

Subcutaneous Panniculitic - Like T-Cell Lymphoma (SPTCL) was described in 1991 by Gonzalez and colleagues as a group of lymphomas localized primarily in the subcutaneous tissue. It was included as a distinct entity since 2001 WHO/EORTC classification of hematolymphoid

classification; in 2008 WHO classification it continues to be recognized as a separate entity.(38) It accounts for less than 1% of all non-Hodgkin lymphomas. Clinically, patients present with fever and multiple subcutaneous nodules most commonly on the extremities and trunk. Laboratory abnormalities may not be severe even though more than 50% may show cytopenias. (39)

Infections of the skin

CHILDHOOD VULVAR TUBERCULID

Localized tuberculid has been infrequently reported in the literature. Most of these reports are of papulonecrotic tuberculids localized to the penis, an entity designated as “penis tuberculid.” In females, such a localized genital tuberculid has been documented by firstly by Pandi et al in 2007. They reported an 11-year-old girl with lichen scrofulosorum confined to the vulva, associated with cervical and inguinal tubercular lymphadenitis. (40)

PSYCOCUTANEOUS DISEASES

MORGELLONS DISEASE

The term “Morgellons disease” first publicly appeared on the Internet in 2002. The “index” case was the first modern case to which that label was appended: a sick child whose physical signs and symptoms were collectively unrecognized as an entity at local and regional medical facilities. As the child's illness persisted without recognition or resolution, the unaffected parent sought similar illness descriptions from historic medical references, eventually settling on “The Morgellons”, a label given to childhood cases described in France in 1674 by Sir Thomas Browne (41). The disease is characterized by fiber-like strands extruding from the skin in conjunction with various dermatologic and neuropsychiatric symptoms. 15, 625 family have registered to ‘The Morgellons disease research foundation’ website till January 2012. There are controversies about this disease whether it is delusional parasitosis or another disease with unknown etiology.

CONCLUSION

New entities continue to be described. It is important to be aware of these new disorders in terms of diagnosis and treatment. Additionally even an unusual case report may play a role in elucidating some undiscovered aspects of a known disease and also shed light on therapy, because there are still so many skin disorders with unexplained etiopathogenesis and uncertain therapy. We think that new entities should be shared by all dermatologists in order to

upgrade knowledge of skin disorders.

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