



XXXV

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Auditorio y Centro de Congresos Víctor Villegas, Murcia



DERMATOSIS PARANEOPLÁSICAS

UNA OPORTUNIDAD DE COLABORACIÓN ENTRE MEDICINA INTERNA Y MEDICINA EXTERNA

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Murcia



DERMATOSIS PARANEOPLÁSICAS

- Es aquel conjunto de manifestaciones cutáneas que se presentan antes, durante o después del inicio de una patología tumoral con la que no guardan una relación de dependencia metastásica
- Son las segundas en frecuencia, tras las de origen endocrinológico

DERMATOSIS PARANEOPLÁSICAS

CONTINUING MEDICAL EDUCATION

Clinical and pathologic findings of paraneoplastic dermatoses

Vinh Q. Chung, MD, MPharm Sci,^a Samuel L. Moschella, MD,^b
Artur Zembowicz, MD, PhD,^a and Vincent Liu, MD^c
Boston and Burlington, Massachusetts and Iowa City, Iowa

Paraneoplastic dermatoses comprise a heterogeneous group of noninherited skin conditions that manifest internal malignancy. Familiarity with paraneoplastic dermatoses is important to both clinician and pathologist alike, as recognition of such a condition offers opportunity for early diagnosis and treatment of internal malignancy; monitoring for tumor recurrence; and insight into pathophysiology which may yield possible clues to treatment. Herein are reviewed 16 of the best established paraneoplastic dermatoses that display distinctive clinical and pathologic findings. (J Am Acad Dermatol 2006;54:745-62.)

Actas Dermosifiliogr. 2013;104(4):285-298



ACTAS
Dermo-Sifiliográficas

Full English text available at
www.elsevier.es/ad



REVISIÓN

Alertas cutáneas en malignidades sistémicas (parte I)

M. Yuste-Chaves* y P. Unamuno-Pérez

Actas Dermosifiliogr. 2013;104(7):543-553



ACTAS
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Full English text available at
www.actasdermo.org



REVISIÓN

Alertas cutáneas en malignidades sistémicas (parte 2)

M. Yuste Chaves* y P. Unamuno Pérez

1º síntoma en el 1% de los pacientes con neoplasias internas

POSTULADOS DE CURTH

- 1. Comienzo simultáneo**
- 2. Curso paralelo de las dos afecciones**
- 3. Neoplasia uniforme, en tipo celular o lugar de emplazamiento**
- 4. Asociación estadística**
- 5. Asociación genética**



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Dermatol Clin 26 (2008) 45–57



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Dermatol Clin 26 (2008) 59–68

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^cDepart



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Dermatol Clin 26 (2008) 69–87

Genodermatoses with Cutaneous Tumors and Internal Malignancies

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DERMATOLOGIC
CLINICS

Dermatol Clin 26 (2008) 1–15

DERMATOLOGIC
CLINICS

Manifestations of Internal Malignancies: An Overview

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DERMATOLOGIC
CLINICS

or neoplasms

Associated malignancy
plasma cell disorder
gastric adenocarcinoma

Hodgkin's disease
epithelial carcinoma
squamous cell carcinoma
of aerodigestive tract

lung, esophageal, breast
carcinoma
questionable association with
adenocarcinoma of stomach,
colon, and breast
cutaneous T-cell lymphoma

and breast

is leukemia,
gic malignancies
gastrointestinal
opharyngeal
osteosarcoma

lymphoma,
cytic leukemia,
case
sial, rarely
na
cancer

sial, multiple

sial

multiple

gynecancies

stic tumors

a of lung

Pruritus

No abnormality

Hodgkin's disease

Tipos de dermatosis paraneoplásicas*

- Relacionadas con síndromes neoplásicos hereditarios.
- Relacionados con síndromes de inmunodeficiencia heredados.
- Relacionados con tumores secretores de hormonas.
- Relacionados con otros tipos de tumores.

TABLE 2. Cutaneous Manifestations of Internal Malignancy

INHERITED SYNDROMES
1) Cowden syndrome
2) Gardner syndrome
3) Peutz-Jeghers syndrome
4) Muir-Torre syndrome
5) Howel-Evans syndrome
6) Birt-Hogg-Dubé syndrome
7) Hereditary leiomyomatosis/renal cell cancer syndrome
8) Melanoma/pancreatic cancer syndrome
9) Multiple mucosal neuromas syndrome
10) Neurofibromatosis type 1
11) Inherited immunodeficiency syndromes
HORMONE-SECRETING TUMORS
1) Ectopic ACTH syndrome
2) Carcinoid syndrome
3) Multiple endocrine neoplasia syndrome
4) Glucagonoma syndrome

PROLIFERATIVE AND INFLAMMATORY DERMATOSES
1) Acquired hypertrophicosis lanuginosa
2) Acanthosis nigricans
3) Sign of Leser-Trelat
4) Tripe palms
5) Bazex syndrome
6) Primary systemic amyloidosis
7) Scleromyxedema
8) Sweet syndrome
9) Pyoderma gangrenosum
10) Blistering disorders
11) Dermatomyositis
12) Clubbing and related disorders
13) Cutaneous leukocytoclastic vasculitis
14) Coagulopathies
15) Figurate erythemas
16) Extramammary Paget disease
17) Infectious disorders
18) Generalized pruritus, ichthyosis, and exfoliative dermatitis
19) Pigmentary disorders
20) Miscellaneous skin, hair, and nail disorders

* Modificado de *CA Cancer J Clin* 2009; 59; 73-98

Clinical and pathologic findings of paraneoplastic dermatoses

Vinh Q. Chung, MD, MPharm Sci,^a Samuel L. Moschella, MD,^b
Artur Zembowicz, MD, PhD,^a and Vincent Liu, MD^c
Boston and Burlington, Massachusetts and Iowa City, Iowa

Paraneoplastic dermatoses comprise a heterogeneous group of noninherited skin conditions that manifest internal malignancy. Familiarity with paraneoplastic dermatoses is important to both clinician and pathologist alike, as recognition of such a condition offers opportunity for early diagnosis and treatment of internal malignancy; monitoring for tumor recurrence; and insight into pathophysiology which may yield possible clues to treatment. Herein are reviewed 16 of the best established paraneoplastic dermatoses that display distinctive clinical and pathologic findings. (J Am Acad Dermatol 2006;54:745-62.)

Table 1. Paraneoplastic dermatoses

Disease	Clinical presentation	Histologic features	Internal malignancy	Strength of association	Epidemiology	Timing	Courses
Acanthosis nigricans	Hyperpigmented, velvety papillomatosis, often in axillae	Hyperkeratosis, papillomatosis, acanthosis with elongated dermal projections	Most are adenocarcinomas: intra-abdominal, 70%-90%; gastric, 50%-60%	Usually with insulin resistance	Age > 40 y	Before malignancy, 20%; during, 60%	Closely parallel
Acquired ichthyosis	Diffuse rhomboidal scales on trunk and extensor surfaces	Orthokeratosis, mild acanthosis, thin or absent granular layer	Hodgkin's lymphoma, 70%-80%	NWE	Both genders; older age	Usually concomitant or after, but also before malignancy	Closely parallel
Tripe palms	Rugose, velvety palms; may be associated with AN	Hyperkeratosis, acanthosis, papillomatosis	Lung CA most often; gastric CA second in frequency, especially with AN	>90%	Both genders; older age	60% before or during malignancy	NWE
Leser-Trélat sign	Increase in size and number of seborrheic keratoses; pruritus; AN in 20% of cases	Seborrheic keratoses; may resemble papilloma in AN	GI adenocarcinoma, 1 in 3 cases; lymphoproliferative, 1 in 5	NWE	Both genders; older age	Before, during, or after malignancy	NWE
Bazex syndrome	Scaly, erythematous plaques in acral areas	Hyperparakeratosis, acanthosis, mononuclear perivascular infiltrate	SCC of oropharynx, larynx, esophagus, lungs	Nearly every case	Mostly male; age 60-70 y	60% before, 20% during, 15% after	Closely parallel
Dermatomyositis	Periorbital heliotrope rash; flat-topped papules over knuckles	Interface dermatitis with variable mucin deposition	Increased ovarian CA in women	25%-30%	Gender unclear; age, 50s, 60s, older than in idiopathic dermatomyositis	Before or during; most within 1 y	Closely parallel
Paraneoplastic pemphigus	Pruritic, polymorphous lesions, bullae, papules; plaques on trunk, oral areas	Suprabasilar acantholysis; vacuolar degeneration; DIF shows IgG intercellularly and at the dermal-epidermal junction	3/4 lymphoproliferative: 42% non-Hodgkin, 29% CLL	Nearly every case	Mean age, 57 y	50% before	Parallel with benign tumors; not parallel with malignancy
Erythema gyratum repens	Rapidly progressing erythematous rings on trunk, proximal parts of extremities	Hyperkeratosis, superficial dermal perivascular lymphohistiocytic infiltrate	Bronchial CA most frequent (32%)	82%	2 M:1 F; mean age, 63 y; Caucasian	80% before	Closely parallel

Table 1. Cont'd

Disease	Clinical presentation	Histologic features	Internal malignancy	Strength of association	Epidemiology	Timing	Courses
Necrolytic migratory erythema	Migratory erythematous macules or papules in annular configuration on face, abdomen, thighs, perianal and oral areas	Parakeratosis, keratinocytes with pyknotic nuclei, epidermal pallor	Pancreatic alpha-cell tumor	Nearly every case	Both genders; age, 50s, 60s	Early or late	Not parallel
Sweet's syndrome	Erythematous tender plaques or nodules in upper extremities; fever, neutrophilia	Dense and extensive neutrophilic infiltrate with vasculopathy	85% AML and lymphoma	20%	Gender NWE median age, 52 y	2/3 before, 40% within 1 mo	Closely parallel
Pyoderma gangrenosum	Discrete ulcers with undermined borders; pathergy	Dense dermal neutrophilic infiltrate, epidermal necrosis	AML most frequent, multiple myeloma next; solid tumors rare	7%	F > M; mean age, 45-52 y	NWE	NWE
Multicentric reticulohistiocytosis	Pink or brown papules and nodules on hands, face; symmetric arthropathy	Nodular infiltrate of histiocytes and multicellular giant cells with eosinophilic ground glass cytoplasm	No predominant cancer type (hematologic, breast, ovarian, gastric, cervical)	20%	1 M:1.85 F; mean age, 50 y; 80% Caucasian	Unknown	Not parallel
Necrobiotic xanthogranuloma	Red-orange dermal nodules, plaques that ulcerate on periorbital area, trunk	Mid dermis to panniculus: palisading xanthogranuloma, Touton giant cells, necrobiosis	Hematologic and lymphoproliferative	80% with benign IgG monoclonal gammopathy	Both genders; mean age, 54-56 y	NWE	NWE
Scleromyxedema	2- to 4-mm-diameter waxy papules distributed symmetrically on arms, hands, face (leonine facies)	Fibroblast proliferation; mild perivascular lymphocytic infiltrate, with increased mucin	No clear specific malignancy; multiple myeloma, Hodgkin's, non-Hodgkin's	80% have monoclonal gammopathy IgG-λ	Both genders; age, 30-50 y	NWE	NWE
Cutaneous amyloidosis	Smooth, waxy, nonpruritic papules and plaques: "pinch purpura" on eyelids	Apple-green birefringence with Congo red when stained with purple and crystal violet	Multiple myeloma, thyroid or multiple endocrine neoplasia	13%-16% have multiple myeloma	Systemic: M > F, average age, 65 y; localized: NWE	NWE	NWE
Hypertrichosis lanuginosa acquisita	Soft, downy, nonpigmented lanugo hair on face, trunk, axillae	"Mantle hair" structures that extend parallel to epidermis	Men: lung first, colorectal second; women: colorectal first, lung or breast second	Nearly every case	3 F:1 M; age, 40-70 y	Late in course of malignancy but possible before	NWE

DERMATOSIS PARANEOPLASICAS ESPECÍFICAS

- Acantosis nigricans maligna
- Acroqueratosis paraneoplásica de Bazex
- Eritema gyratum repens
- Hipertrichosis lanuginosa adquirida
- Eritema necrolítico migratorio (síndrome de glucagonoma)
- Pénfigo paraneoplásico
- Síndrome carcinoide

DERMATOSIS PARANEOPLÁSICAS FACULTATIVAS

- Eritrodermias
- Herpes zoster
- Dermatitis ampollosas (penfigoide ampolloso, herpes gestationis, pénfigo, dermatitis herpetiforme)
- Paniculitis pancreática
- Amiloidosis sistémica
- Hirsutismo

DERMATOSIS PARANEOPLÁSICAS FACULTATIVAS

- Ictiosis adquirida
- Dermatomiositis del adulto
- Tromboflebitis migrans
- Paquidermoperiostosis adquirida
- Prurigo simple y nodular
- Eritema anular centrífugo
- Dermatitis neutrofílicas (síndrome de Sweet, pioderma gangrenoso)

DERMATOSIS PARANEOPLÁSICAS FACULTATIVAS

- Queratosis seborreicas eruptivas. S. de Lesser Trelat
- Reticulo histiocitosis multicéntrica
- Escleromixedema
- Xantogranuloma necrobiótico y otros xantomas
- Calcinosis cutánea tumoral
- Vasculitis paraneoplásicas
- Síndrome acral vascular paraneoplásico

DERMATOSIS Y CÁNCER

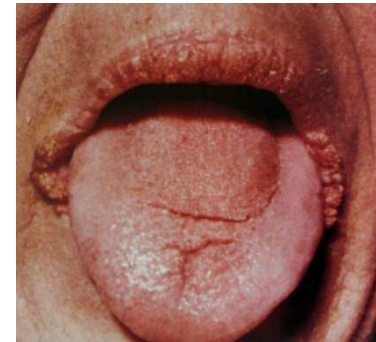
- DERMATOSIS PARANEOPLÁSICAS ESPECÍFICAS
- DERMATOSIS PARANEOPLÁSICAS FACULTATIVAS
- **GENODERMATOSIS CON PROPENSIÓN A MALIGNIDAD**
- **INDICADORES DE EXPOSICIÓN A CARCINÓGENOS**

DERMATOSIS PARANEOPLASICAS **ESPECÍFICAS**

- **Acantosis nigricans maligna**
- **Acroqueratosis paraneoplásica de Bazex**
- **Eritema gyratum repens**
- **Hipertrichosis lanuginosa adquirida**
- **Eritema necrolítico migratorio (síndrome del glucagonoma)**
- **Pénfigo paraneoplásico**
- **Síndrome carcinoide**

ACANTOSIS NIGRICANS MALIGNA

- **Placas simétricas de piel engrosada, aspera, pardo- grisácea**
- **Predilección por pliegues y flexuras**
- **1/3 afecta mucosas (oral, genital, conjuntival)**
- **Diagnóstico diferencial: Obesidad, resistencia insulina, fármacos (corticoides, estrógenos...)**



TIPOS D ACANTOSIS NIGRICANS

- **Benigna**
- **Pseudoacantosis nigricans**
- **Síndrómica**
- **Maligna**



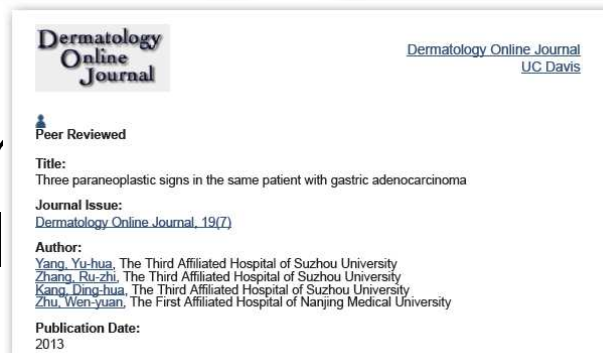
ACANTOSIS NIGRICANS MALIGNA



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- Asociación
queratoderm
Favorece el



ser-Trélat y
triple palma.
N Maligna

ACANTOSIS NIGRICANS MALIGNA

- Se asocia sobre todo con carcinomas del tracto gastrointestinal (90%), especialmente ADENOCA. GÁSTRICO.
- Niveles elevados de (TGF- α), que estimulan el factor de crecimiento epidérmico (EGF) receptor y la hormona melanocito estimulante
- Otros :Ca. Mama y pulmón
- Con frecuencia es el signo presentación del tumor (>50%)
- Curso paralelo al tumor



Malignant acanthosis nigricans in a patient with a gastrointestinal stromal tumor

Keon Woo Park, Do Hyoung Lim, and Soon Il Lee

KJIM

Acantosis nigricans como manifestación inicial paraneoplásica de adenocarcinoma gástrico

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Gastroenterol Hepatol. 2007;30(1):15-8

Triple Palma



- Aspecto peculiar de palmas, con un aspecto exagerado de los dermatoglifos. La piel se encuentra engrosada, hiperqueratósica y con superficie suave.
- Presenta múltiples sinonimias (acantosis nigricans palmaris, paquidermatoglifia, etc.) y puede manifestarse sin la patología tumoral. Pero es muy frecuente que aparezca con el inicio de una neoplasia, especialmente **gástrica o pulmonar**.

**Ca. Pulmón
Ca. Gástrico**

90%

**Asociado a A. Nigricans
y
Signo de Lesser-Trelat**



Signo-síndrome de Leser-Trélat

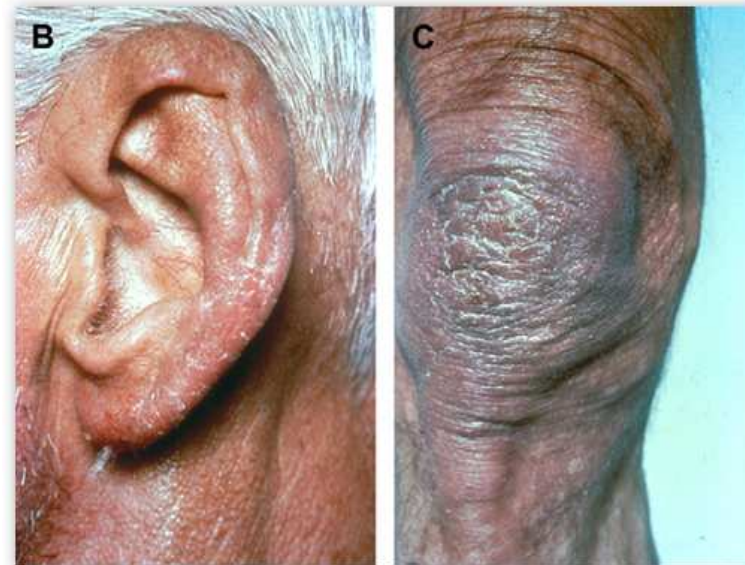
- Este cuadro está determinado por la existencia de QS múltiples, lesión tumoral benigna presente en un alto porcentaje de la población normal, pero de la que difiere por la aparición de gran cantidad de elementos, en forma cuasi explosiva, muy diseminadas y pruriginosas. Atributos estos de los que carece la primera forma mencionada. Sin embargo algunos trabajos mencionan la existencia de esa sintomatología sólo en un 40 - 50% de los enfermos.



**Adenocarcinoma gástrico
(y otros GI)**

ACROQUERATOSIS PARANEOPLÁSICA DE BAZEX

- Dermatitis psoriasiforme acral (periungueal, palmar, plantar, nariz, pabellones auriculares, mejillas). En ocasiones se extienden a codos, rodillas y regiones malares.
- Eritemato violáceas. Simétricas.
- Suele preceder al tumor. Curso paralelo.
- Más frecuente en varones



CASOS CLÍNICOS

Actas Dermosifiliogr 2006;97(3):196-9

Acroqueratosis paraneoplásica con lesiones ampollosas asociada a carcinoma epidermoide esofágico

Miguel Cabanillas, Lidia Pérez-Pérez, Dolores Sánchez-Aguilar, Virginia Fernández-Redondo y Jaime Toribio
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Clásicamente se han considerado tres fases en esta enfermedad⁹. La primera se caracteriza por la aparición de eritema y descamación psoriasiforme en los dedos de las manos y pies, que afecta también a los pabellones auriculares y al dorso nasal, con alteraciones distróficas ungueales frecuentes (hiperqueratosis subungueal y onicólisis). En este momento la neoplasia suele ser asintomática. En una segunda fase, la erupción se extiende a palmas y plantas, mostrando una queratodermia difusa de tonalidad violácea, y coincidiendo frecuentemente con los primeros signos y síntomas de la enfermedad neoplásica. Finalmente, en un tercer estadio, la afectación cutánea se extiende a áreas proximales como rodillas, muslos y brazos, donde se muestran placas eritematosas mal definidas. En esta fase es frecuente constatar ya la existencia de metástasis ganglionares o viscerales.



ACROQUERATOSIS PARANEOPLÁSICA DE BAZEX



- Se asocia casi siempre con Tumores de origen epitelial: CARCINOMA EPIDERMÓIDE

Netherlands
The Journal of Medicine

NOVEMBER 2013, VOL. 71, NO 9
481

PHOTO QUIZ

Skin lesions in a patient with head and neck cancer

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p.

le
y

- En el 63% de los casos precede a la neoplasia

Síndrome de Bazex (acroqueratosis paraneoplásica)

J.M. Fernández-Recio^a, I.M. Rodríguez-Navarro^b, A.J. Chaves Álvarez^b, D. de Argila Fernández-Durán^b, P. López Vallejo^c y F. Monje Gil^d

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SEMERGEN. 2007;33(6):305-7



Erythema Gydatum Repens: A Rare Paraneoplastic Rash

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Submission history: Submitted September 7, 2012
Reprints available: [link]
DOI: 10.5811/wes

Erythema
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Imagen de la se

Diagnóstico
adenocarci

Paraneoplast

Antoni Bennà
José Manuel M

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J EADV
Journal of the European Academy of Dermatology and Venereology

DOI: 10.1111/j.1468-3083.2012.04663.x

J EADV

SHORT REPORT

Erythema gyratum repens is not an obligate paraneoplastic disease: a systematic review of the literature and personal experience

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Abstract

Background Erythema gyratum repens (EGR) is a rare clinical entity that is considered to be an obligatory paraneoplastic disease. According to the literature, an underlying neoplasm can be detected in 82% of the cases.

Objectives The aim of this systematic review was to evaluate the association of EGR with malignancies or other non-neoplastic conditions.

Methods The medical records of patients seen at the Section of Dermatology, University of Genoa between 1990 and 2010, in whom a diagnosis of EGR had been made, were reviewed for evidence of systemic associations. A systematic search of the Cochrane library, EMBASE, Pubmed and MEDLINE databases was also conducted. Key search term used in the review was 'erythema gyratum repens'.

Results Four patients with a diagnosis of EGR have been retrieved from our medical records. One case was idiopathic, one was associated with a bronchial carcinoma and two were associated with drug-intake. One hundred and twelve original cases of EGR were selected from the literature for detailed review. Among these, 58 cases (70%) were associated with an underlying neoplasm, 25 cases (30%) were non-paraneoplastic and 29 cases have been considered as different dermatoses mimicking EGR in their clinical presentation ('EGR-like' eruption).

Conclusion EGR should no longer be considered as an obligate paraneoplastic syndrome as the cases that are not associated with neoplasm are more than expected. In addition to searching an underlying neoplasm, dermatologists should be aware about the possibility of other associations including also drug-intake.

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J EADV 2014, 28, 112-115

Rev Clin Esp. 2014 Oct;214(7):425-427. doi: 10.1016/j.rce.2014.05.009. Epub 2014 Jun 20.

Erythema gyratum repens: Not always a paraneoplastic disease.

[Article in English, Spanish]

Galán-Gutiérrez M¹, Martínez-Peinado CM¹, Ruiz-Villaverde R².

European Journal of Dermatology

ACCUEIL | NUMÉRO EN COURS | S'ABONNER | ARCHIVES | ACHETER UN NUMÉRO

Lupus erythematosus gyratus repens

Volume 17, numéro 1, January-February 2007

J Drugs Dermatol. 2003 Jun;2(3):315-7.

Erythema gyratum repens in a case of resolving psoriasis.

Bryan ME¹, Lienhart K, Smoller BR, Johnson SM.

Med Klin (Munich). 2002 Dec 15;97(12):759.

[Erythema gyratum repens. Drug hypersensitivity after azathioprine in a patient with type 1 autoimmune hepatitis].

[Article in German]

von Rainer Günther ZB, Nasser S, Hinrichsen H, Fölsch UR.



CED

Clinical and
Experimental Dermatology



Clinical dermatology • Concise report

CED
Clinical and Experimental Dermatology

Erythema gyratum repens associated with pityriasis rubra pilaris

N. Almaani, A. Robson, R. Sarkany and W. A. D. Griffiths

St John's Institute of Dermatology, Guy's and St Thomas' NHS Foundation Trust, London, UK

doi:10.1111/j.1365-2230.2010.03861.x

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- **Aparición en pocas semanas de vello tipo lanugo fetal (fino y canoso), que puede alcanzar hasta 10-15 cm.**
- **Cara, pabellones auriculares. Tronco. Extremidades.**
- **Predilección por sexo femenino (70%).**
- **T. Colo-rectales, pulmón y mama. En varones T. colon**
- **Procesos benignos: Cirrosis biliar primaria**
- **Suele preceder al tumor en mucho tiempo. Habitualmente no hay curso paralelo.**
- **Gran valor en diagnóstico precoz. Supervivencia inferior a 3 años**

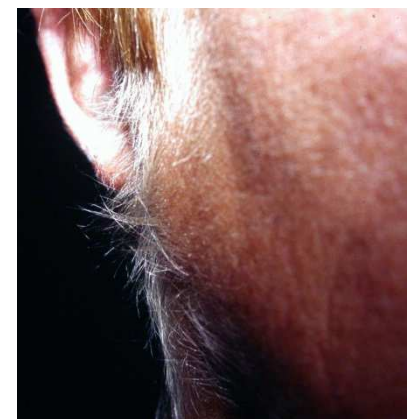
CASOS BREVES

Hipertrichosis lanuginosa adquirida paraneoplásica

Juan Sánchez-Estella, Manuela Yuste, Juan C. Santos y M.ª Teresa Alonso

Unidad de Dermatología. Hospital Virgen de la Concha. Zamora. España.

Actas Dermosifiliogr 2005;96(7):459-61



Eritema necrolítico migratorio como marcador de síndrome del glucagonoma

Necrolytic migratory erythema as clue to the diagnosis of glucagonoma syndrome

N. Ormaechea-Pérez, M^aA Arregui-Murua, A. López-Pestaña, M^a López-Núñez, A. Jaka-Moreno, A. Tuneu-Valls
Servicio de Dermatología. Hospital Donostia. San Sebastián. Guipúzcoa. España.

- 90% con Glucagonomas (crecimiento lento) _ca. de céls α del páncreas (o tumor ectópico).
- Suele ser la 1^a manifestación del síndrome
- TAC, RNM o ECO. Gammagrafía con análogos de somatoestatina
- 50% metástasis en el momento del diagnóstico
- Hay otras causas: S. pseudoglucagonoma
- Ambas clínicamente iguales

- | | |
|--------------------------------|--------------------------------------|
| - Enfermedad celiaca | - Enfermedad inflamatoria intestinal |
| - Malabsorción | - Abuso de heroína |
| - Hepatitis | - Abscesos odontogénicos |
| - Pancreatitis crónica | - Iatrogénicos |
| - Procesos malignos digestivos | - Déficit de zinc |



**Eritema necrolítico migratorio como
marcador de síndrome del glucagonoma**

*Necrolytic migratory erythema as clue to the diagnosis of glucagonoma
syndrome*

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- **Máculas eritematosas de crecimiento centrífugo, con infiltración progresiva y aparición de vesículas y ampollas**
- **Afecta sobre todo tronco y cintura pélvica, fundamentalmente periné y áreas intertriginosas**
- **Glositis y boqueras**
- **Pérdida peso, anemia, hiperglucemia (controlable con dieta)**





Paraneoplastic pemphigus with eosinophilic spongiosis and autoantibodies against desmocollins 2 and 3

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Australas J Dermatol. 2013 Nov;54(4):241-50. doi: 10.1111/j.1440-0960.2012.00921.x. Epub 2012 Jul 3.

Paraneoplastic pemphigus.

Yong AA¹, Tey HL.

Author information

Abstract

Paraneoplastic pemphigus (PNP) is a distinct autoimmune blistering disease that can affect multiple organs other than the skin. It occurs in association with certain neoplasms, among which lymphoproliferative diseases are most commonly associated. The clinical presentation of PNP consists typically of painful, severe oral erosions that may be accompanied by a generalised cutaneous eruption and systemic involvement. The eruption may be of different morphology, consisting of lesions that resemble pemphigus, pemphigoid, erythema multiforme or graft versus host disease, as well as lesions resembling lichen planus. Similarly, the histological findings also show considerable variability. PNP is characterised by the presence of autoantibodies against various antigens: desmoplakin I (250 kd), bullous pemphigoid antigen I (230 kd), desmoplakin II (210 kd), envoplakin (210 kd), periplakin (190 kd), plectin (500 kd) and a 170-kd protein. This 170-kd protein has recently been identified as alpha-2-macroglobulin-like-1, a broad range protease inhibitor expressed in stratified epithelia and other tissue damaged in PNP. The prognosis of PNP is poor and the disease is often fatal. Immunosuppressive agents are often required to decrease blistering, and treating the underlying malignancy with chemotherapy may control autoantibody production. The prognosis is better when PNP is associated with benign tumours and these should be surgically excised when possible.

IFI + con epitelio de esófago de mono y vejiga de rata



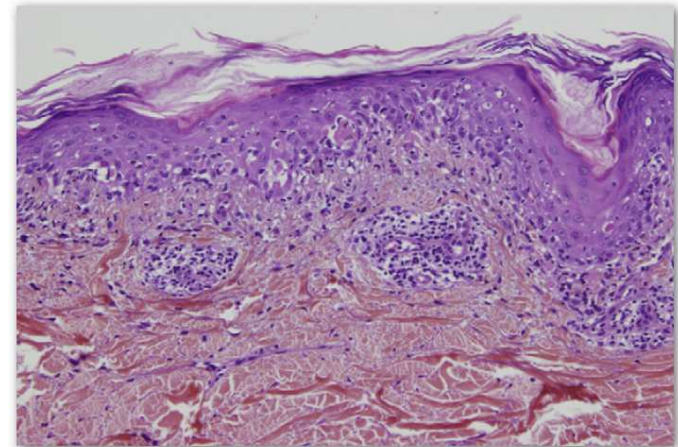
- **Afectación mucosa dolorosa, constante e intensa**
- **Estomatitis ± conjuntivitis refractaria a tratamiento**
- **Lesiones cutáneas variables: desde eritema, lesiones anulares de tipo EEM/STJ y ampollas de penfigo/Penfigoide**
- **Liquen Plano**



Síndrome multiorgánico autoinmune paraneoplásico asociado a linfoma folicular

Follicular Lymphoma With Paraneoplastic Autoimmune Multiorgan Syndrome

Actas Dermosifiliogr. 2012;103:244-6.



PIEL (BARC). 2014;29(9):552-555



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Caso clínico

**Pénfigo y bronquiolitis obliterante como
manifestaciones de síndrome paraneoplásico
autoinmune en un paciente con linfoma folicular**





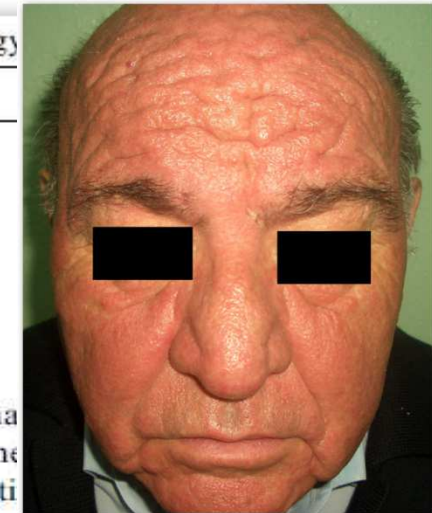
- **Etiopatogenia: ¿reacción cruzada con AC anti-Ags tumorales?**
- **Curso paralelo al tumor?. Alta mortalidad incluso tras tratamiento del tumor, incluyendo procesos benignos Timoma/enf. Castelman.**
- **Complicaciones respiratorias (Bronquiolitis obliterante) como causa de muerte.**
- **Neoplasias asociadas:**
 - **Linfoma no-Hodgkin, LLC, Enfermedad de Castleman**
 - **Sarcomas**
 - **Carcinoma escamoso broncogénico**

Cutaneous manifestations of the malignant carcinoid**SÍNDROME**

- Sin motivo aparente o por estrés físico o psíquico, alcohol o compresión del tumor

Classification of vascular abnormalities heralding an internal malignancy according to etiology

Etiology	Vascular abnormality	Associated malignancies
Vascular dilatation	Flushing	Carcinoid tumor Medullary thyroid carcinoma Systemic mastocytosis Pheochromocytoma Renal cell carcinoma Pancreatic tumors (VIPoma) Pancoast tumor (in HS) Superior mediastinal neurinoma Myeloma (in POEMS syndrome)
	Telangiectasia	Several malignancies (in the setting of liver metastases) Breast cancer Bronchogenic carcinoma Carcinoid tumor Adenocarcinoma of the hepatic duct MAE
Vascular inflammation	Vasculitis	Several malignancies, most commonly hematopoietic
Vascular occlusion	Trousseau	Most commonly pancreas, lung, prostate, stomach, and colon
	Mondor's disease	Mostly breast carcinoma
	Deep vein thrombosis	Several malignancies, especially advanced stages Mucin-secreting adenocarcinomas are the most common.
	Purpura	Several malignancies, mostly hematopoietic Lymphomas are most common in ITP. Gastric and breast carcinomas are most common in TTP.
	Cutaneous ischemia	Several malignancies, including carcinomas of pancreas, stomach, small bowel, ovary, kidney, and lymphoma and leukemia In the setting of cryoglobulinemia, lymphomas and multiple myeloma are the most common associated cancers.



DERMATOSIS PARANEOPLÁSICAS FACULTATIVAS

- ➡ • Ictiosis adquirida
- ➡ • Dermatomiositis del adulto
- Tromboflebitis migrans
- Paquidermoperiostosis adquirida
- Prurigo simple y nodular
- Eritema anular centrífugo
- ➡ • Dermatosis neutrofílicas (síndrome de Sweet, pioderma gangrenoso)

SD. TROUSSEAU: TROMBOFLEBITIS MIGRATORIA)



Brotos inflamatorios recidivantes en red venosa superficial de extremidades. Puede afectar sobre todo EESS y tronco

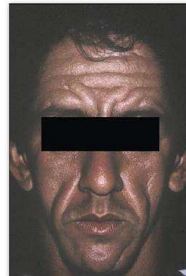


Asociaciones

- Páncreas
 - Pulmón
 - Próstata
 - Gástrico
 - Colon
- } 50%

PAQUIDERMOPERIOSTOSIS

- Eritema periungueal y engrosamiento progresivo de la porción distal de manos y pies (acropaquia). Uñas redondas y convexas.
- Artritis dedos, muñecas, tobillos. Periostitis.
- Asociación más frecuente: Neoplasias broncopulmonares



PRURITO

Prurigo simple y prurigo nodular de Hyde

- Asociación prurito persistente intenso y malignidad: 3% - 47%
- Descartar procesos linfoproliferativos (sobre todo linfoma de Hodgkin)
- Obstrucción maligna de vías biliares.
- Policitemia rubra vera: Prurito acuagénico
- Prurito nasal: tumor cerebral
- Dificultad: diferenciar prurito senil por xerosis de prurito paraneoplásico



Eritema anular centrífugo



Pulmón, mama y tracto GI alto

Five-year malignancy incidence in patients with chronic pruritus: A population-based cohort study aimed at limiting unnecessary screening practices

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Philadelphia, Pennsylvania

Background: The incidence of malignancy in patients with chronic pruritus and nondiseased skin is unknown.

Objective: We sought to assess the hazard ratio (HR) of incident overall malignancy and incident malignancy by subtype in patients with chronic pruritus during the 5 years after diagnosis.

Methods: A population-based cohort study was performed in the Health Improvement Network. In all, 8744 patients with chronic pruritus were matched with 31,580 patients without chronic pruritus based on sex, age, and practice. Primary outcomes were HR of incident malignancy and HR of malignancy subtypes.

Results: The fully adjusted HR for incident malignancy in patients with chronic pruritus was 1.14 (95% confidence interval 0.98-1.33). The fully adjusted HR for incident hematologic malignancy and incident bile duct malignancy in patients with chronic pruritus was 2.02 (95% confidence interval 1.48-2.75) and 3.73 (95% confidence interval 1.55-8.97), respectively. The incidence of hematologic malignancy and cholangiocarcinoma in patients with chronic pruritus was 0.0016 and 0.0003 per person-year, respectively.

Limitations: Potential for misclassification and detection biases is a limitation.

Conclusions: Chronic pruritus without concomitant skin changes is a risk factor for having undiagnosed hematologic and bile duct malignancies, but not other malignancies. The overall incidence of these malignancies in patients with chronic pruritus is very low. (J Am Acad Dermatol 2014;70:651-8.)

Acquired Ichthyosis Triggered by an Osseous Hemangiopericytoma: A Case Report and Review of the Literature

Aikaterini Patsatsi^a Aikaterini Kyriakou^a Vasilios Karavasilis^b
Fragkiski Tsatsou^a Georgios Lazaridis^b Dimitrios Kalabalikis^a
Dimitrios Sotiriadis^a

Ann Dermatol Venereol. 2012 Jan;139(1):9-14. doi: 10.1016/j.annder.2011.10.394. Epub 2011 Dec 3.

[Acquired ichthyosis and haematological malignancies: five cases].

[Article in French]

Berrady R¹, Baybay H, Khammar Z, Lahlou M, Lamchacht L, Galloui S, El Hatimi A, Mernissi FZ, Bono W.

ICTIOSIS ADQUIRIDA



- Las formas adquiridas son menos frecuentes que las congénitas por lo que un inicio brusco en adultos nos debe alertar
- No confundir con xerosis senil
- Más frecuente en varones
- Prurito
- Suele respetar palmas, plantas y flexuras



Ictiosis paraneoplásica

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^a Servicio de Dermatología. Instituto Valenciano de Oncología. Valencia. España.



- Con frecuencia precede al tumor. Curso paralelo.
- **Linfoma de Hodgkin.** Linfomas no Hodgkin, sarcoma de Kaposi, leiomiosarcoma y carcinomas de mama, pulmón, ovario y cérvix.
- Etiopatogenia desconocida: ¿Déficit vit A? ¿Déficit de AGs libres?
- También se ve en procesos no neoplásicos: lepra, sarcoidosis, trastornos tiroideos, hiperparatiroidismo, DM, fallo renal crónico, trasplante de médula ósea e infección por VIH, malnutrición, fármacos (alopurinol, cimetidina, ac. Nicotínico, cimetidina, hipocolesteromiantes, triparanol, butirofenonas o clofacimina y enf. Autoinmunes (LES y Dermatomiositis)