XXXV Congreso Nacional de la Sociedad Española de Medicina Interna (SEMI)

IV Congreso Ibérico de Medicina Interna

II Congreso de la Sociedad de Medicina Interna de la Región de Murcia

19-21 de Noviembre de 2014

Auditorio y Centro de Congresos Víctor Villegas, Murcia

DERMATOSIS PARANEOPLÁSICAS

UNA OPORTUNIDAD DE COLABORACIÓN ENTRE MEDICINA INTERNA Y MEDICINA EXTERNA

José Frías Iniesta
Hospital Clínico Universitario Virgen de la Arrixaca
Murcia

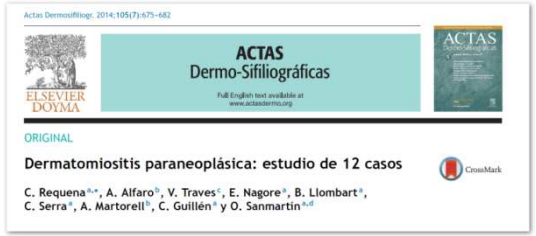
Dermatomiositis Paraneoplásica

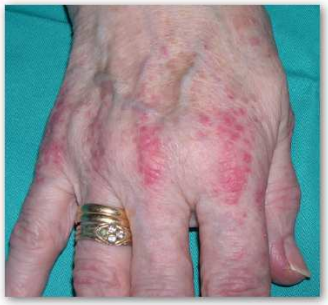
- 15-20% de los casos
- Más asociado a dermatomiositis (32%) que a miositis (14.9%). En más del 50% de los casos es la forma amiopática
- La forma juvenil no se asocia con neoplasias
- Suele manifestarse tras el debut del cáncer o como reflejo de su recaída. Curso paralelo. Media de 7 meses
- Si es previa al Ca.

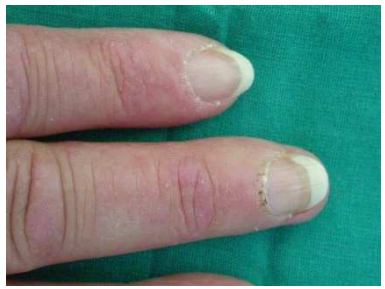
  este aparece el 1º año

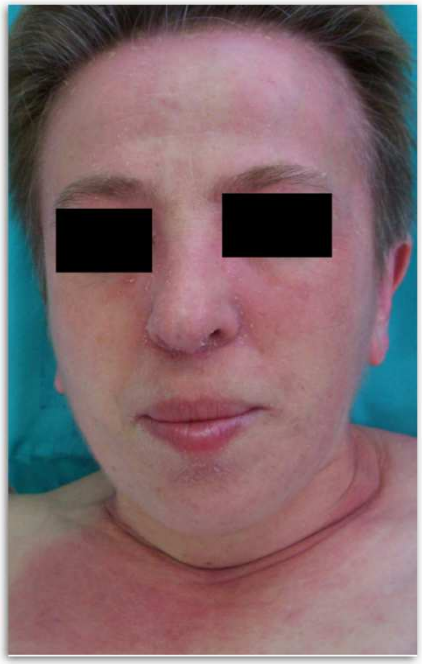
Dermatomiositis Paraneoplásica

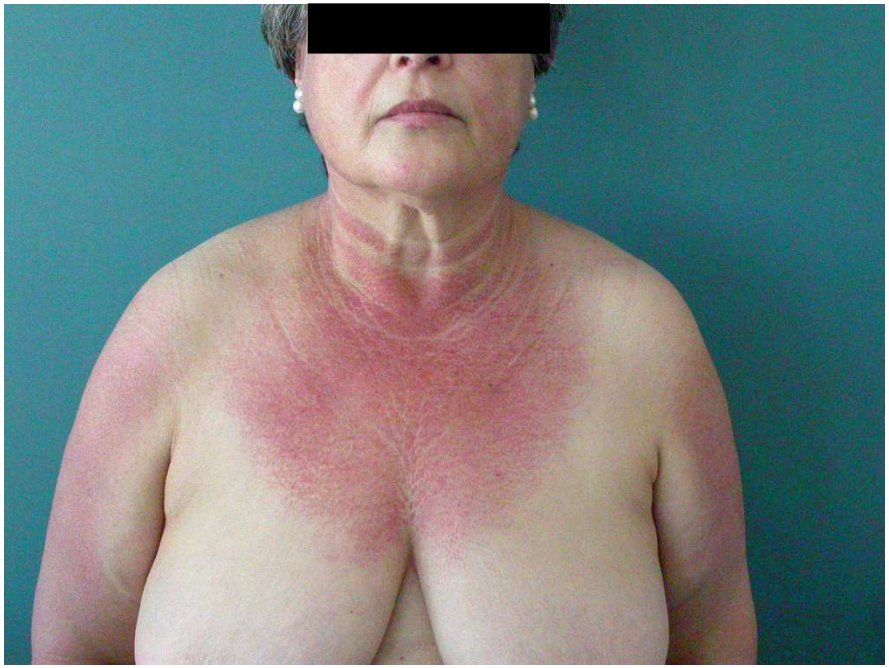
- No existe ningún rasgo clínico, histológico o analítico absolutamente específico de paraneoplasia en dermatomiositis
- Erupción eritemato violácea fotodistribuida, a veces poiquilodérmica
- Cara, V torácica y superficies de extensión de mmss
- Eritema y edema periocular
- Pápulas de Gottron y afectación cuticular

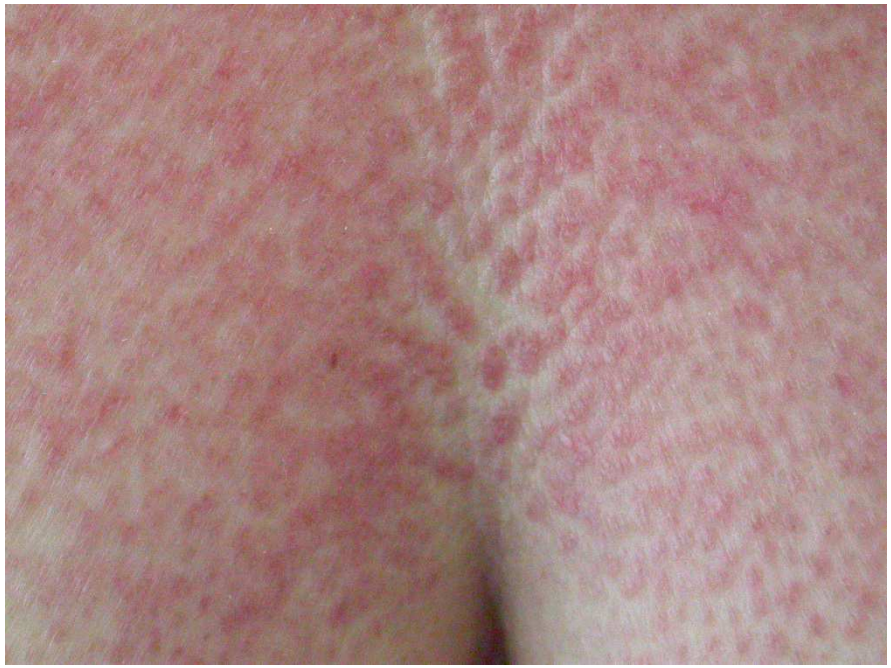


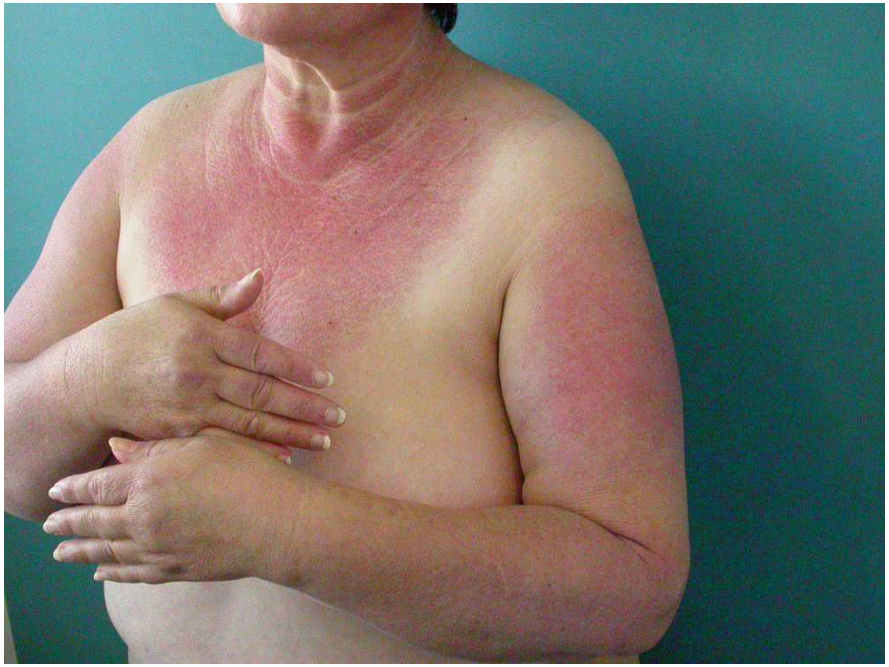


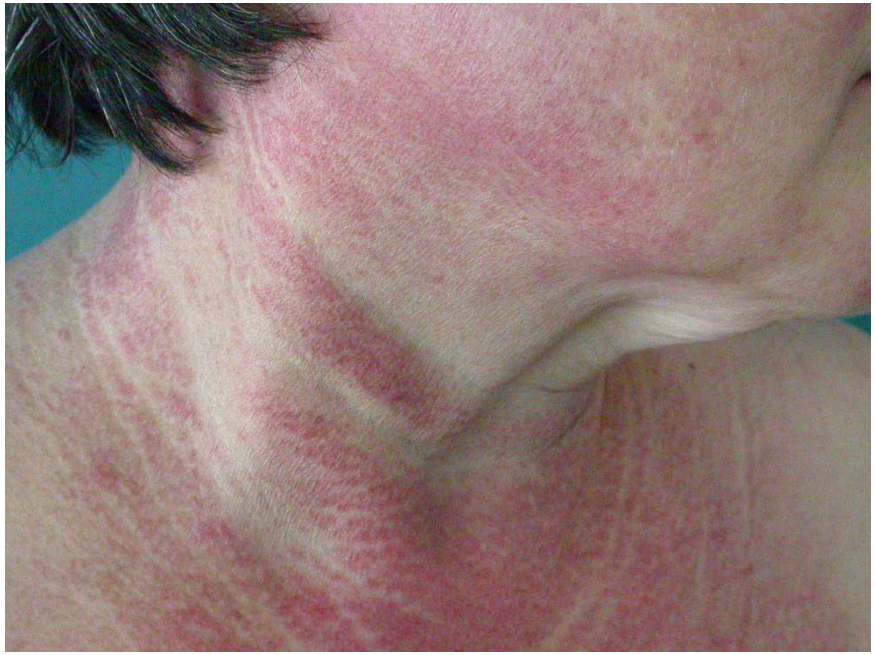


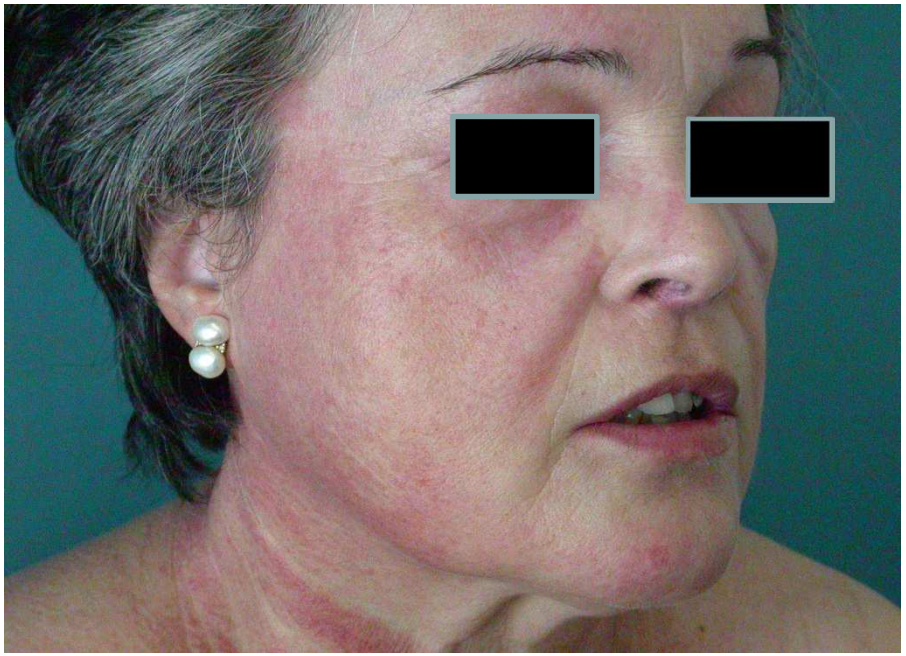


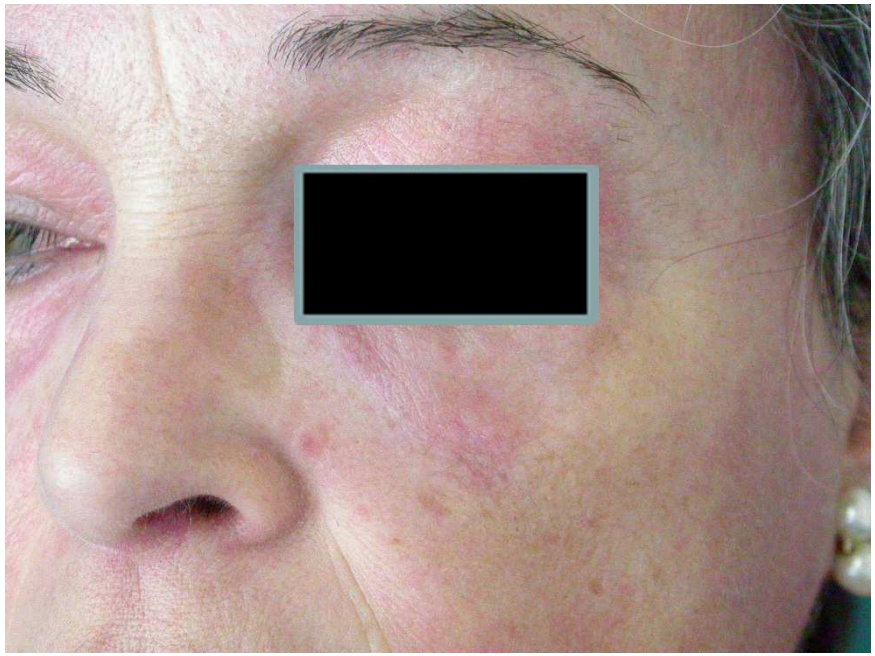


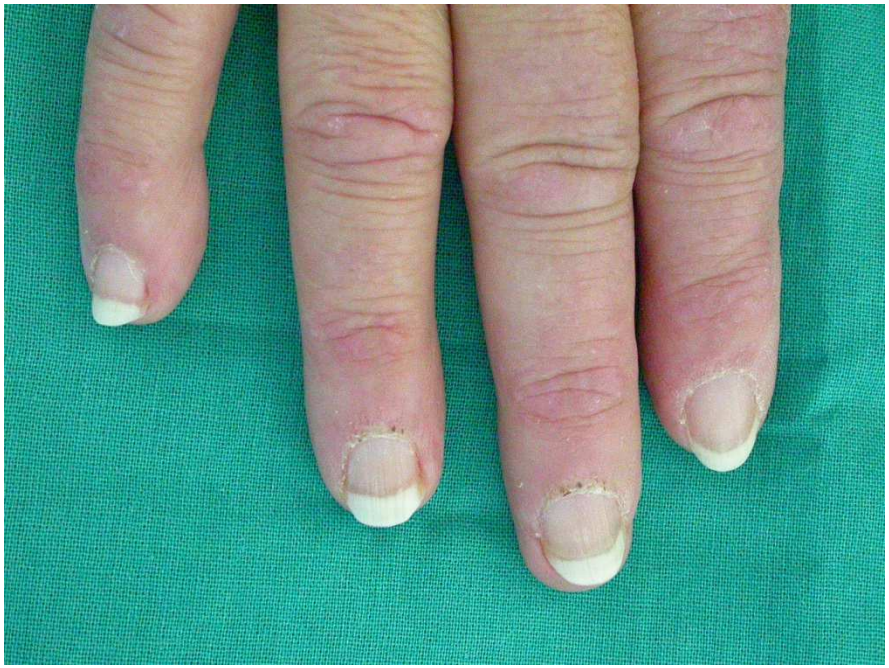


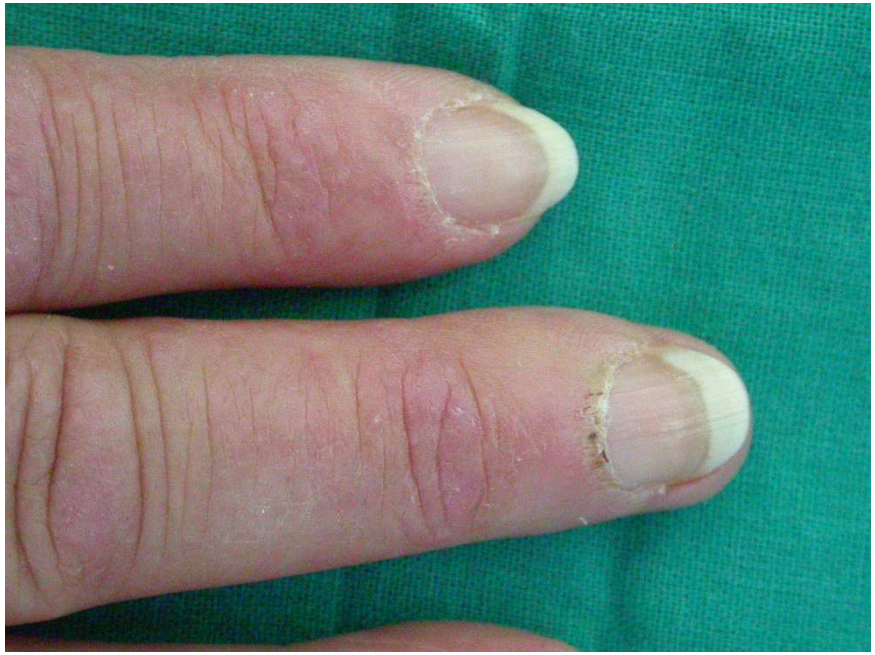












Derm plásica
Crit pecha

- **Sugie**
  - **Con**
  - **Var**
  - **Inte**
  - **Nec**
  - **Disf**
  - **Pres**
  - **Mal**

Incidence and predictive factors for malignancies with dermatomyositis: a cohort from southern China

D. Chen, S. Yuan, X. Wu, H. Li, Q. Qiu, Z. Zhan, Y. Ye, F. Lian, L. Liang, H. Xu, X. Yang

Department of Rheumatology and Clinical Immunology, the First Affiliated Hospital of Sun Yat-Sen University, Guangzhou, China.

Abstract

Objective

We aimed to explore the incidence of malignancy in dermatomyositis and assess the potential risk factors of occurrence of malignancy in DM from southern China.

Methods

A retrospective cohort study of patients admitted in the 1st affiliated university hospital between 2003 and 2012 was performed. Demographic information, clinical symptoms, laboratory findings, medications were documented. The endpoint of the study was defined as occurrence of malignancy or death.

Results

For this approximately 10-year retrospective study, 60 out of 246 dermatomyositis patients developed malignancies with the overall incidence of 24.4%. Nasopharyngeal carcinoma (NPC) and ovarian carcinoma were the most common malignant disease, accounting for 35% (21/60) and 15% (9/60) of malignancies, respectively. Lung and colon were followed as the third most common carcinoma (5 out of 60, 8.3%).

Among these 60 patients with malignancies, 39 (65.0%, 39/60) cases occurred within 1 year after DM diagnosis. Subsequently, malignancies were detected in 13 (21.7%, 13/60) patients during the second year and 8 (13.3%, 8/60) during the third year. One patient developed cancer at the 35th month after DM as the latest.

The logistic regression multivariate analysis indicated that male gender [odds ratio (OR) = 3.76, 95% confidence interval (CI) 1.86–7.61, p<0.01], dysphagia (OR= 2.21, 95%CI 1.10–4.48, p=0.03) and elevated erythrocyte sedimentation rate (ESR) (OR= 2.37, 95% CI 1.18–4.75, p=0.02) were risk factors for the occurrence of malignancies, while interstitial lung disease (ILD) acted as a protective factor (OR=0.13, 95%CI 0.06–0.28, p<0.01).

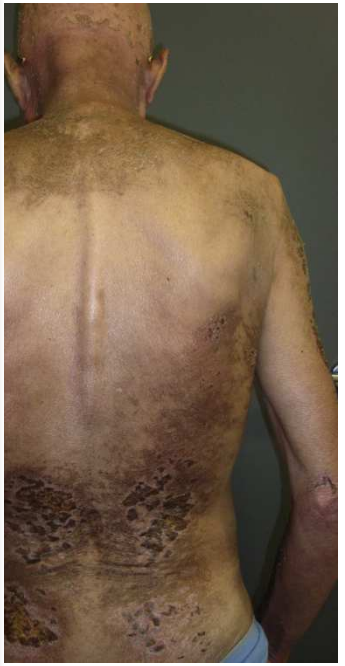
Conclusion

It was necessary to carry out routine malignancy screening for Chinese DM patients due to its high incidence. Nasopharyngeal carcinoma and ovarian cancer were the most common malignant disease. The risk of malignancy was highest in the first year after DM diagnosis and reduced thereafter. Extensive work-ups for malignancy screening should be carried out at the first year. Male gender, dysphagia and elevated ESR were risk factors for occurrence of malignancy. The presence of ILD could diminish the risk of coexisting of malignancy.

Key words

prediction analysis, dermatomyositis, malignancy

Clinical and Experimental Rheumatology 2014; **32**: 615-621.



CLINICAL AND LABORATORY INVESTI

Meta-analysis of the as polymyositis with cance

J. Wang,[1] G. Guo,[1] G. Chen,[2] B. Wu,[3] L. Lu[3]

[1]Shanghai Children's Hospital, Shanghai Jiao Tong University,
[2]Department of Internal Medicine III, University of Erlangen-N
[3]Shanghai Renji Hospital, Shanghai Jiao Tong University School
[4]Cincinnati Children's Hospital Medical Center and College of

British Journal of Dermatol

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Usefulness of Anti-p155 Autoantibody for Diagnosing Cancer-Associated Dermatomyositis

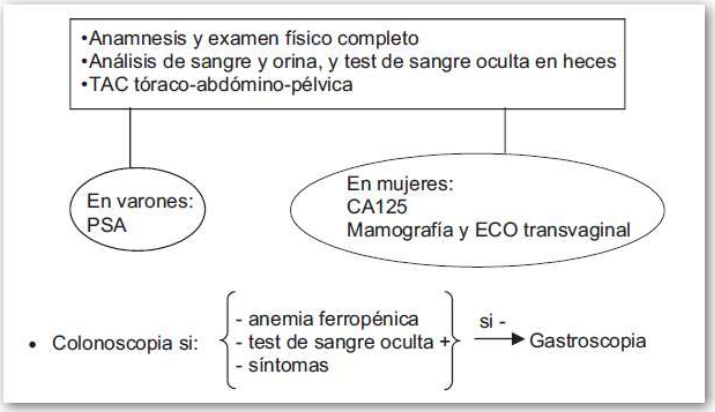
A Systematic Review and Meta-Analysis

Ernesto Trallero-Araguás,[1] Jose Ángel Rodrigo-Pendás,[2] Albert Selva-O'Callaghan,[2] Xavier Martínez-Gómez,[2] Xavier Bosch,[3] Moisés Labrador-Horrillo,[2] Josep Maria Grau-Junyent,[4] and Miquel Vilardell-Tarrés[2]

ARTHRITIS & RHEUMATISM
Vol. 64, No. 2, February 2012, pp 523–532

Dermatomiositis Paraneoplásica

- Si Ac p155 +: estudio de extensión
- Si estudio de extensión -: repetir cada 3 años
- Si Ac p155 -: controles según edad del paciente
- Estudio de extensión mediante PET con resultados similares a estudio convencional

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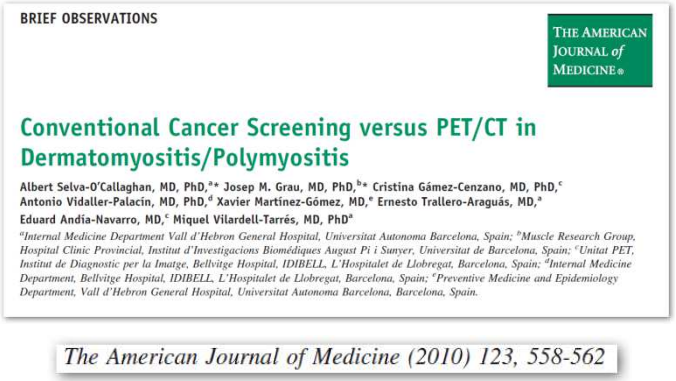

BRIEF OBSERVATIONS

THE AMERICAN JOURNAL of MEDICINE

Conventional Cancer Screening versus PET/CT in Dermatomyositis/Polymyositis

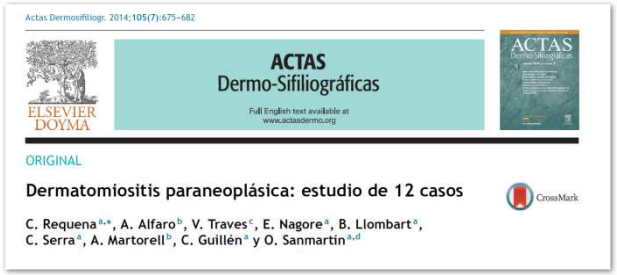
Albert Selva-O'Callaghan, MD, PhD,[a,*] Josep M. Grau, MD, PhD,[b,*] Cristina Gámez-Cenzano, MD, PhD,[c] Antonio Vidaller-Palacín, MD, PhD,[d] Xavier Martínez-Gómez, MD,[e] Ernesto Trallero-Araguás, MD,[a] Eduard Andia-Navarro, MD,[c] Miquel Vilardell-Tarrés, MD, PhD[a]

[a]Internal Medicine Department Vall d'Hebron General Hospital, Universitat Autonoma Barcelona, Spain; [b]Muscle Research Group, Hospital Clinic Provincial, Institut d'Investigacions Biomédiques August Pi i Sunyer, Universitat de Barcelona, Spain; [c]Unitat PET, Institut de Diagnostic per la Imatge, Bellvitge Hospital, IDIBELL, L'Hospitalet de Llobregat, Barcelona, Spain; [d]Internal Medicine Department, Bellvitge Hospital, IDIBELL, L'Hospitalet de Llobregat, Barcelona, Spain; [e]Preventive Medicine and Epidemiology Department, Vall d'Hebron General Hospital, Universitat Autonoma Barcelona, Barcelona, Spain.

The American Journal of Medicine (2010) 123, 558-562

Dermatomiositis Paraneoplásica

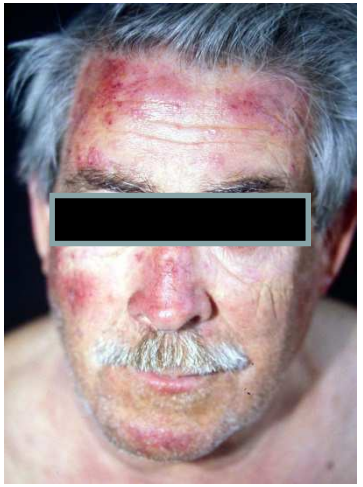
- En Europa los cánceres asociados a dermatomiositis del adulto son por orden de frecuencia :cáncer de ovario, pulmón, mama, colorrectal, estómagoy páncreas (otros: próstata, linfomas no Hodgkin, etc.)
- Por sexos en las mujeres los más frecuentes son el de mama y ovario y en varones el de pulmón y el colorrectal.
- Sin embargo, si nos encontramos con un paciente que proceda del Sudeste asiático, el perfil de riesgo cambia, ya que allí el cáncer más frecuentemente asociado a la dermatomiositis paraneoplásica es el cáncer de cavum

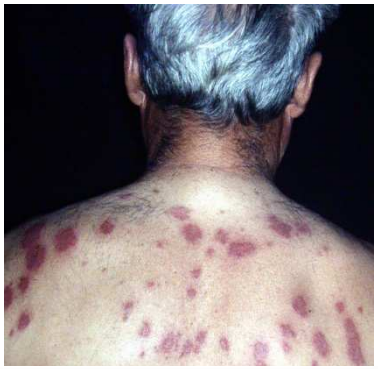


Sweet syndrome: Clinical presentation, associations, and response to treatment in 77 patients

Nicole M. Rochet, BS,[a] Rahul N. Chavan, MD, PhD,[a] Mark A. Cappel, MD,[a] David A. Wada, MD,[b] and Lawrence E. Gibson, MD[a]
Rochester, Minnesota, and Salt Lake City, Utah

- # DERMATOSIS AGUDA FEBRIL NEUTROFILICA





CRITERIOS MAYORES

1.-PLACAS O NODULOS ERITEMATOVIOLÁCEOS. DOLOROSOS DE INICIO BRUSCO.
2.-INFILTRADO NEUTROFILICO DÉRMICO, SIN VASCULITIS.

CRITERIOS MENORES

1.-CUADRO PRODROMICO DE FIEBRE O INFECCION > 38º.
2.- ASOCIACIÓN CON NEOPLASIA HEMATOLÓGICA O VISCERAL SUBYACENTE, ENF. INFLAMATORIA, EMBARAZO, INFECCIÓN GASTROINTESTINAL O DE VÍAS RESPIRATORIAS O VACUNACIÓN
3.-BUENA RESPUESTA A ESTEROIDES SISTEMICOS
4.- ANOMALÍAS ANALÍTICAS INICIALES (3 o 4):
- VSG > 20 MM/H
- ELEVACIÓN DE PROTEINA C REACTIVA
- LEUCOCITOSIS > $8x10^9/L$
- NEUTROFILIA > 70%

Conclusions: Sweet syndrome is a distinctive disorder with certain clinical and histologic characteristics, which usually has a complete response to systemic corticosteroids. It is important to evaluate Sweet syndrome patients who have laboratory evidence of anemia for an underlying malignancy. (J Am Acad Dermatol 2013;69:557-64.)

LOS 2 CRITERIOS MAYORES Y AL MENOS 2 MENORES

NB.:SINTOMAS SISTEMICOS POR INFILTRADOS NEUTROFILICOS

INTERNATIONAL JOURNAL OF ONCOLOGY 42: 1516-1522, 2013

Insight into Sweet's syndrome and associated-malignancy: A review of the current literature

SHAHZAD RAZA[1], ROBERT S. KIRKLAND[2], ANAND A. PATEL[2], JAMES R. SHORTRIDGE[1] and CARL FRETER[1]

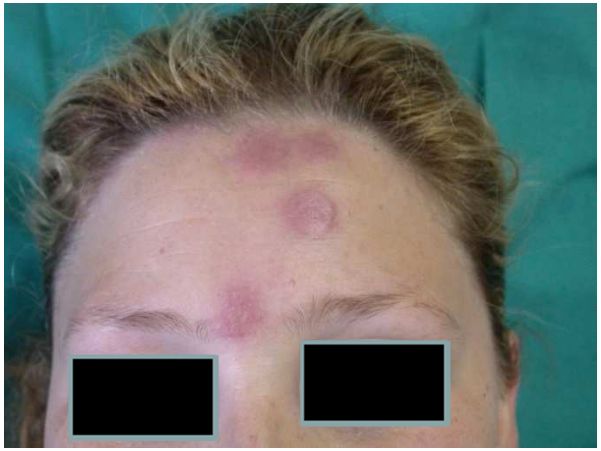
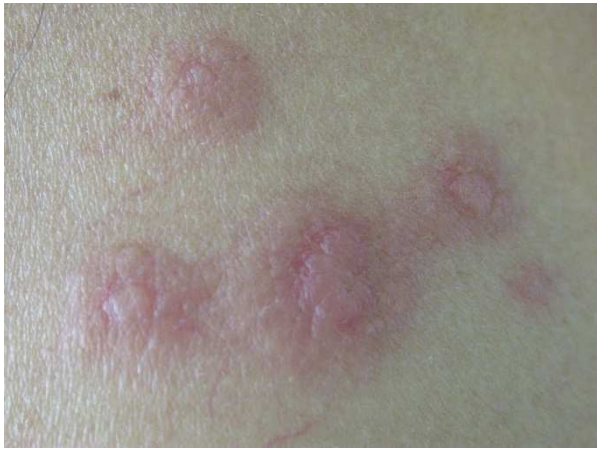
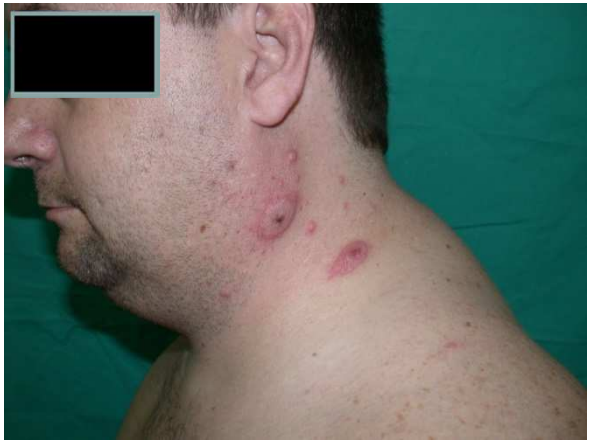
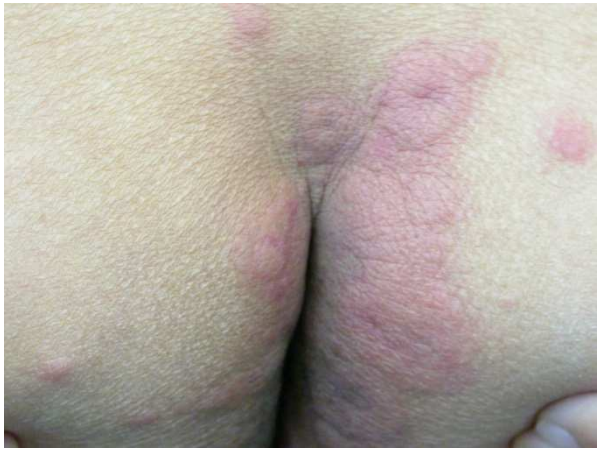
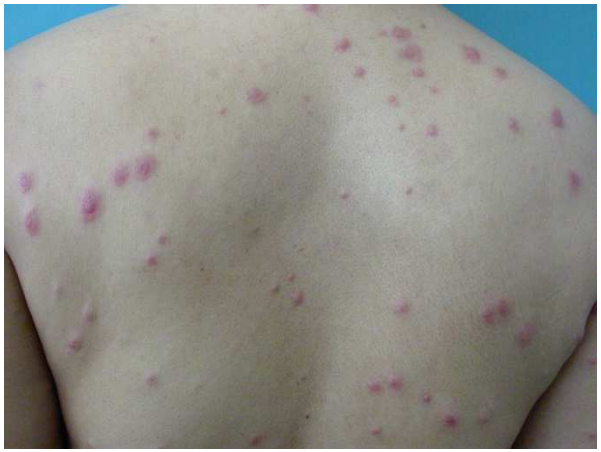
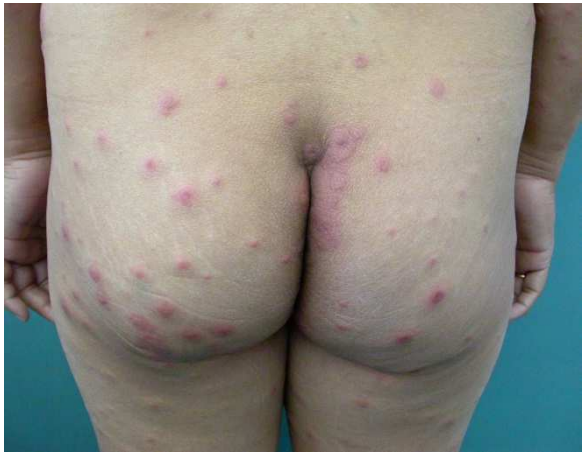
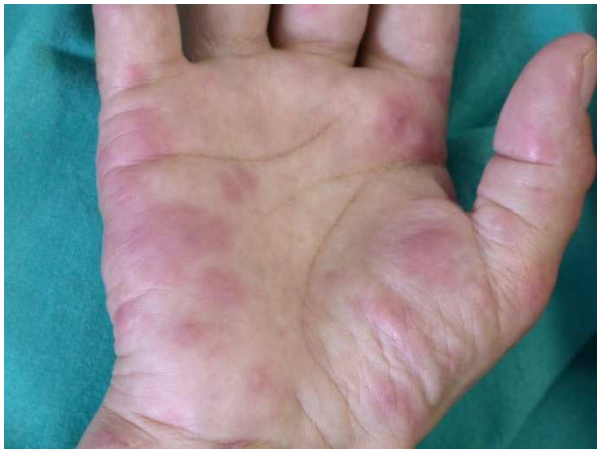
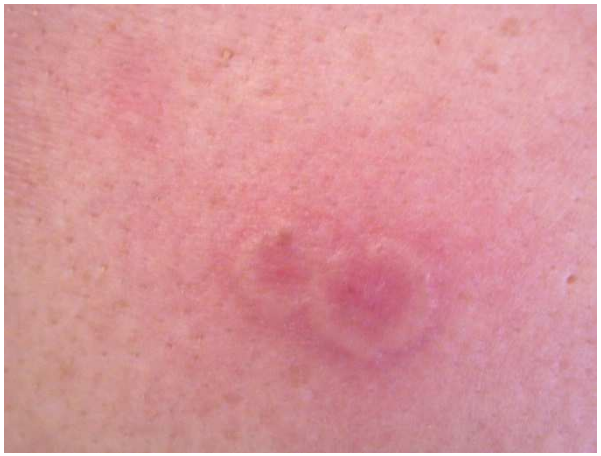
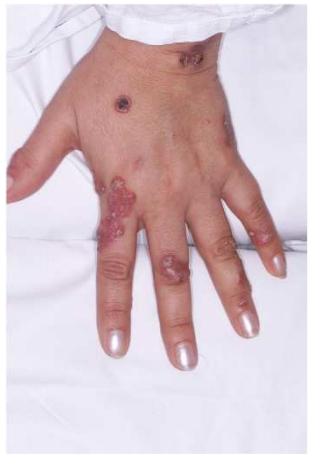
[1]Division of Hematology and Oncology, Ellis Fischel Cancer Center;
[2]University of Missouri School of Medicine, Columbia, MO 65203, USA

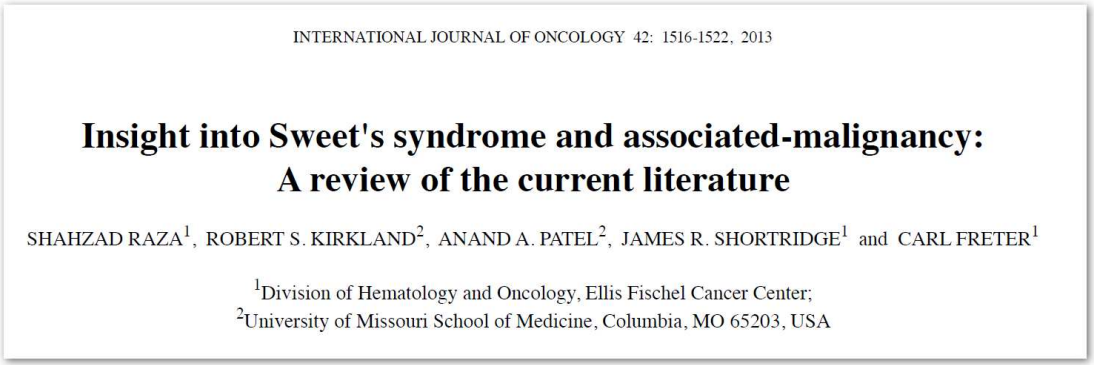
DERMATOSIS AGUDA FEBRIL NEUTROFILICA

- SS clásico o idiopático: habitualmente precedido por infección de vías respiratorias, puede estar asociado a enf. Inflamatoria intestinal o embarazo
- SS paraneoplásico: 21%. 15% Hematológicas y 6% sólidos
- SS causado por fármacos:

Diagnositc criteria for medications-induced Sweet syndrome	
1.	Acute onset of painful erythematous plaques or nodules
2.	Dermal neutrophilic infiltrate without evidence of vasculitis on histopathological examination
3.	Temperature >38˚C
4.	Temporal relationship between drug ingestion and clinical presentation or temporally-related recurrence after rechallenge
5.	Temporally related resolution of lesions after drug withdrawal or treatment with systemic corticosteroids

Anticancer drugs associated with Sweet's syndrome	
1.	Granulocyte colony stimulating factor (23,26,32,33)
2.	Bortezomib (34,35)
3.	Azacitidine (36,37)
4.	Decitibine (36,37)
5.	Imatinib mesylate (38-40)
6.	Lenalidomide (41-44)
7.	All-trans retinoic acid (21,45,46)



INTERNATIONAL JOURNAL OF ONCOLOGY 42: 1516-1522, 2013

Insight into Sweet's syndrome and associated-malignancy: A review of the current literature

SHAHZAD RAZA[1], ROBERT S. KIRKLAND[2], ANAND A. PATEL[2], JAMES R. SHORTRIDGE[1] and CARL FRETER[1]

[1]Division of Hematology and Oncology, Ellis Fischel Cancer Center;
[2]University of Missouri School of Medicine, Columbia, MO 65203, USA

- **79% % SON IDIOPATICOS/FÁRMACOS**
- **21% ASOCIADOS A CÁNCERES: 81% ENFERMEDADES HEMATOLOGICAS MALIGNAS, CON UN 42% DE LMA Y T. SÓLIDOS, 57% ADENOCA. DE TRACTO GENITOURINARIO, MAMA Y GASTROINTESTINAL**
- **SUELE SER EL PRIMER SIGNO DE UN CÁNCER NO DIAGNOSTICADO O DE SU RECURRENCIA**
- **EL S.SWEET ASOCIADO A HEMOPATIAS, PRESENTA NEUTROFILIA SOLO EN EL 42%**
- **SÍNTOMAS EXTRACUTANEOS EN EL 50% DE LOS CASOS ASOCIADOS A MALIGNIDAD**

INTERNATIONAL JOURNAL OF ONCOLOGY 42: 1516-1522, 2013

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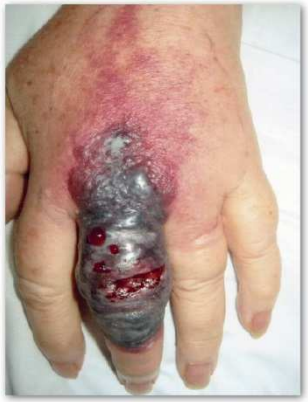
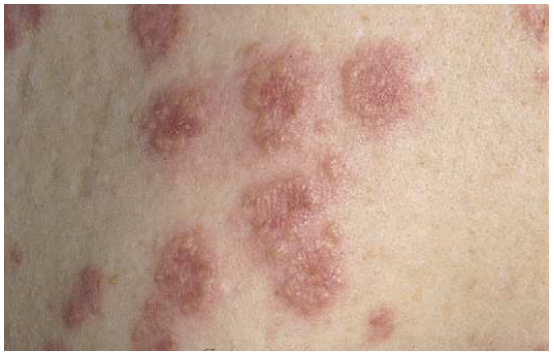
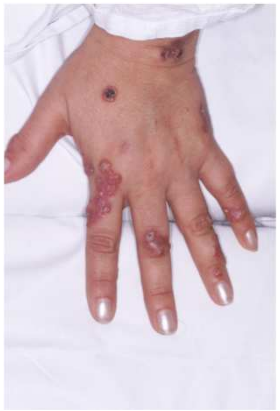
[1]Division of Hematology and Oncology, Ellis Fischel Cancer Center;
[2]University of Missouri School of Medicine, Columbia, MO 65203, USA

- **A favor de neoplasia asociada:**
  - **Ausencia de proceso respiratorio previo. Solo 20%.**
  - **Ausencia de fiebre y neutrofilia**
  - **Anemia. 85%. Anomalías recuento plaquetario (68%). Neutropenia.**
  - **Mayor gravedad e intensidad de las lesiones**
  - **Mayor tendencia a recurrencias**
  - **Mayor incidencia en manifestaciones extracutáneas**







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Actas Dermosifiliogr. 2007;98:102-

CASOS CLÍNICO

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Subcutaneous Sweet syndrome in the setting of myeloid disorders: A case series and review of the literature

May P. Chan, MD,[a,b] Lyn M. Duncan, MD,[c] and Rosalynn M. Nazarian, MD[c]
Ann Arbor, Michigan, and Boston, Massachusetts

Tunis Med. 2012 Aug-Sep;90(8-9):636-40.

[Vasculitis in Sweet's Syndrome].

[Article in French]

Chelly I[1], Zehani A, Mbazaa A, Nfoussi H, Azouz H, Haouet S, Kchir N, Zitouna M.

⊕ **Author information**

Abstract

BACKGROUND: Also called acute febrile neutrophilic dermatosis, Sweet's syndrome is non-infective dermatoses that exhibit a predominantly neutrophilic inflammatory infiltrate. Absence of vasculitis is a histologic criterion for diagnosis. However, recent reports suggest that vasculitis should not exclude the diagnosis.

AIM: To describe their clinical, pathological and therapeutic characteristics.

METHODS: We report a series of 47 cases of Sweet's syndrome, collected in our institution between 1997 and 2011.

RESULTS: The patient population consisted of 11 males and 36 females. The mean age was 47 years (28-74). An associated disease process was seen in 10 patients: inflammatory disease (3 cases), inflammatory bowel disease (2 cases), tuberculosis (3 cases) and diabetes (3 cases). One case of pregnancy was observed. In the prodromal phase, functional symptoms were reported in 38 cases (80.8%). Cutaneous lesions consisted of erythematous plaques or nodules. Lesions were located mainly on the upper or lower extremities. All biopsy specimens demonstrated a dermal infiltrate composed predominantly of neutrophils. Fibrinoid necrosis and intramural inflammation were observed in 8 cases.

CONCLUSION: The dermatosis can precede, follow, or appear concurrent with the diagnosis of the patient's diseases which requires careful surveillance.

Sweet syndrome," an acute necrotizing neutrophilic dermatosis. This subtype is also characterized by the rapid onset of progressive erythematous, warm, edematous cutaneous lesions with deep-tissue neutrophilic infiltration and soft-tissue necrosis, in the absence of infectious cause. Awareness of this entity and early dermatologic consultation is critical as debridement results in expansion of the process, resulting in additional and aggressive resection—a vicious cycle with significant possible morbidity. (J Am Acad Dermatol 2012;67:945-54.)

CLINICAL AND LABORATORY INVESTIGATIONS

BJD
British Journal of Dermatology

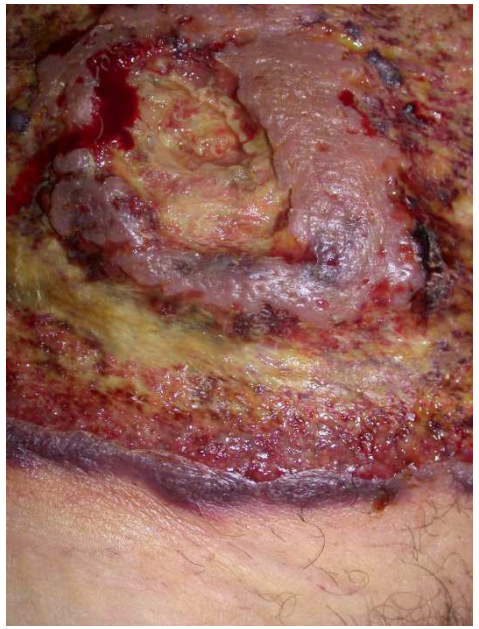
Pyoderma gangrenosum: a retrospective review of patient characteristics, comorbidities and therapy in 103 patients

A.M. Binus, A.A. Qureshi, V.W. Li* and L.S. Winterfield*

*Department of Dermatology and *Pyoderma Gangrenosum Clinic, Brigham and Women's Hospital and Harvard Medical School, Boston, MA 02115, U.S.A.*

BJD © 2011 British Association of Dermatologists 2011 **165,** pp1244–1250

- Poco especificidad diagnóstica, clínica o patológicamente
- Predominio femenino 76%
- El 78% de la úlceras se sitúan en mmii y el 67% tiene más de 2.
- 34% asociado a enfermedad inflamatoria intestinal
- 20% trastornos hematológicos- **10.7% malignos**
- 14% depresión
- 19% artritis seronegativa
- 11% psoriasis
- 9% hepatitis



Incidence, Mortality, and Disease Associations of Pyoderma Gangrenosum in the United Kingdom: A Retrospective Cohort Study

Sinéad M. Langan[1], Richard W. Groves[2], Tim R. Card[3] and Martin C. Gulliford[4]

Journal of Investigative Dermatology (2012) **132,** 2166–2170

- 3 veces mayor la tasa de mortalidad

J Dermatol. 2012 Dec;39(12):1010-5. doi: 10.1111/j.1346-8138.2012.01676.x. Epub 2012 Sep 14.

Bullous pyoderma gangrenosum: a case report and review of the published work.

Sakiyama M[1], Kobayashi T, Nagata Y, Fujimoto N, Satoh T, Tajima S.

⊕ Author information

Abstract

Pyoderma gangrenosum (PG) is an ulcerative skin disorder characterized by neutrophilic infiltrations. PG is generally classified into four types: (i) ulcerative; (ii) pustular; (iii) bullous; and (iv) vegetative. Among them, bullous PG is known as a rare type. Herein, we report a case of bullous PG together with a summary of the 12 PG cases treated in our department over the previous 15 years, and we review 38 well-documented bullous PG cases (65.8% female; aged 18-80 years [mean ± standard deviation, 51.6 ± 16.8]) in the published work, including the present case, from 1972-2011. Although the disease most frequently associated with PG is inflammatory bowel disease, bullous PG is most commonly associated with hematological disorders (25/38, 65.8%), which indicates the characteristic pathophysiology specific to bullous PG.

Pioderma gangrenoso: revisión de cinco casos

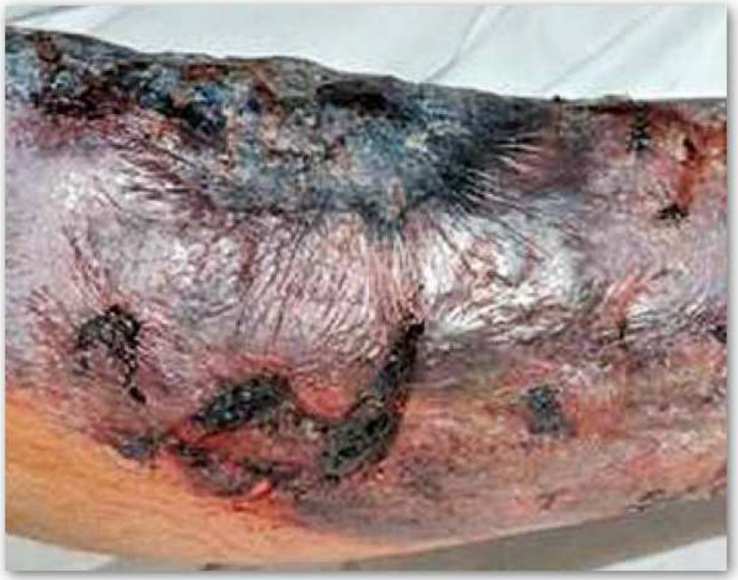
R. Haro[a], D.H. Todaro[b], A.L. Aguilar-Shea[b], J.M. Revelles[a] y L. Requena[a]

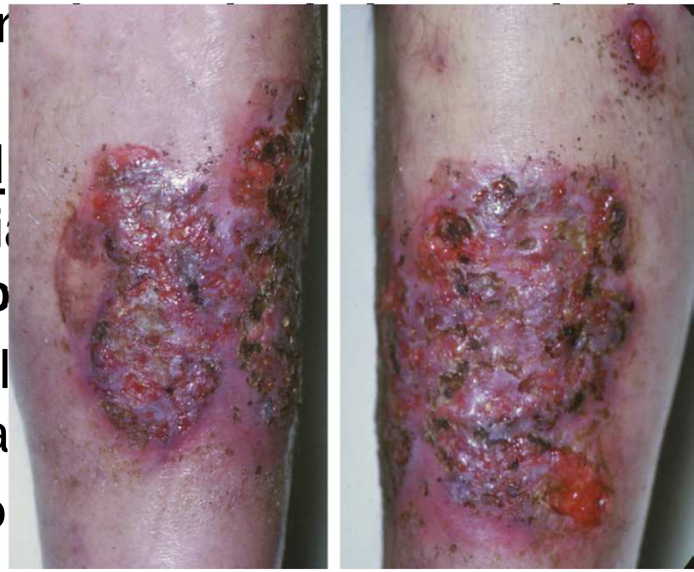
[a]Servicio de Dermatología. Fundación Jiménez Díaz. Área 7. Madrid. España.
[b]Medicina Familiar y Comunitaria. Área 7. Madrid. España.

SEMERGEN. 2009;35(8):406-9

Tabla 1. Formas clínicas de pioderma gangrenoso

Morfología clínica	Características	Localización	Enfermedades asociadas
Ulcerativa	Ulceración con rápida evolución en profundidad y extensión	Indiferente	Ninguna conocida
Pustulosa	Pápula o grupo de pápulas que confluyen, se transforman en pústulas y se ulceran	Tronco y superficies extensoras de los miembros	Enfermedad inflamatoria intestinal
Ampollosa	Ampolla superficial que se ulcera	Miembros superiores y cara	Enfermedades hematológicas
Superficial granulomatoso	Tejido vegetante, erosionado, de evolución crónica	Tronco	Ninguna conocida





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- …, co … …

DERMATOSIS PARANEOPLÁSICAS FACULTATIVAS

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

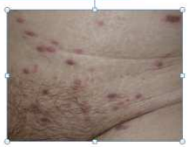
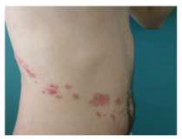
Tumores virilizantes:
- ováricos y adrenales

Tumores hipofisarios:
- prolactinomas, acromegalia

HERPES ZOSTER

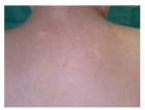
- Frecuente en el grupo de población con riesgo de cáncer
- Neoplasia relacionada con mayor frecuencia: PROCESOS LINFOPROLIFERATIVOS

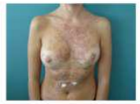
- Sospechar cáncer ante lesiones más severas: hemorrágicas, necróticas, multimetaméricas, diseminadas.

OTRAS ENFERMEDADES AMPOLLOSAS

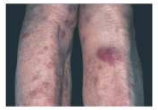
- PENFIGOIDE: edad avanzada?
- DERMATITIS HERPETIFORME: Posible asociación con linfomas intetinales
- HERPES GESTATIONIS Posible asociación con coriocarcinoma
- PÉNFIGO SEBORREICO Descrita asociación con timoma y miastenia gravis



NECROSIS GRASA NODULAR PANICULITIS PANCREÁTICA

- Paniculitis lobulillar, sobre todo EEII
- Necrosis grasa, con células fantasma
- Etiopatogenia: lipasa y amilasa pancreáticas
- Posible asociación a carcinoma páncreas
- Otras causas: pancreatitis aguda o crónica




Eur J Dermatol. 2010 May-Jun;20(3):373-7. doi: 10.1684/ejd.2010.0943. Epub 2010 Apr 19.

Erythroderma: A clinical-etiological study of 82 cases.

Yuan XY[1], Guo JY, Dang YP, Qiao L, Liu W.

Author information

Abstract

Erythroderma is an uncommon skin disorder characterized by generalized reddening and scaling of over 90% of the skin. It represents a maximal stage of skin irritation induced by several skin diseases such as psoriasis, contact dermatitis, drug reactions, and mycosis fungoides. Data including the clinical symptoms, laboratory examinations and skin biopsies were collected from 82 erythroderma patients admitted to our hospital in the period between Jan.1st, 2003 and Dec.31st, 2008. According to clinical findings, laboratory findings and biopsy results, the most common causative factors were pre-existing dermatoses (72.0%), followed by drug reactions (17.0%), idiopathic causes (6.1%) and malignancies (4.9%). Among the pre-existing dermatoses, psoriasis is the most c
causes of erythroderma. In the dru
series, with 9 of the 14 cases (64.3
treatment. In our series we found a
secondary to malignancy. Among c
study, the prognosis of most patien

J Eur Acad Dermatol Venereol. 2012 Jun;26(6):710-3. doi: 10.1111/j.1468-3083.2011.04150.x. Epub 2011 Jun 25.

Erythroderma as a paraneoplastic cutaneous disorder in systemic anaplastic large cell lymphoma.

Hanafusa T[1], Igawa K, Takagawa S, Yahara H, Harada J, Tani M, Sawada Y, Katayama I.

Author information

Abstract

BACKGROUND: Paraneoplastic cutaneous disorders (PCDs) or dermadromes are skin conditions that have an association with internal malignancies but are not themselves malignant. We report the first two cases of systemic anaplastic large cell lymphoma (s-ALCL) accompanied by erythroderma and multiple leg ulcers as PCDs. CASE 1: A 52-year-old Japanese man presented with disseminated itchy papular erythemas which be
had over his entire body for the prec
legs. Superficial lymph nodes (LNs)
contrast, lymphocytes infiltrating into
negative in skin biopsy specimens. T
to chemotherapy. CASE 2: A 71-ye
mastectomy. At her initial hospital vi
negative in skin samples with norma
it was considered a PCD.

Paraneoplastic erythroderma: an unusual manifestation of diffuse large B-cell lymphoma

To the Editor,

Diffuse large B-cell lymphoma (DLBL) is the most common type of adult non-Hodgkin lymphoma. In most patients, cutaneous DLBL occurs as a single nodule on the scalp, face, or neck. Erythrodermic manifestations of DLBL are very rare, and there have been no previous reports of erythroderma as a paraneoplastic syndrome of DLBL.

A 36-year-old Korean man presented with a 2-month history of generalized erythema and right neck mass. Skin lesions first appeared on both hands, rapidly progressing throughout his entire body. He began to experience severe scaling and generalized edema about 10 days before visiting our clinic. He complained of severe itching

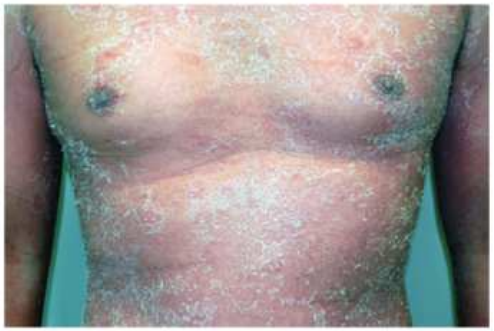


Figure 1 Patient photograph, showing generalized erythroderma with scaling

Case Rep Med. 2014;2014:351065. doi: 10.1155/2014/351065. Epub 2014 Sep 11.

Breast cancer presenting as paraneoplastic erythroderma: an extremely rare case.

Protopsaltis I, Drossou A, Katsantonis I, Roussos N, Manoludaki K, Arvanitis M, Papazafiropoulou A, Antonopoulos S.

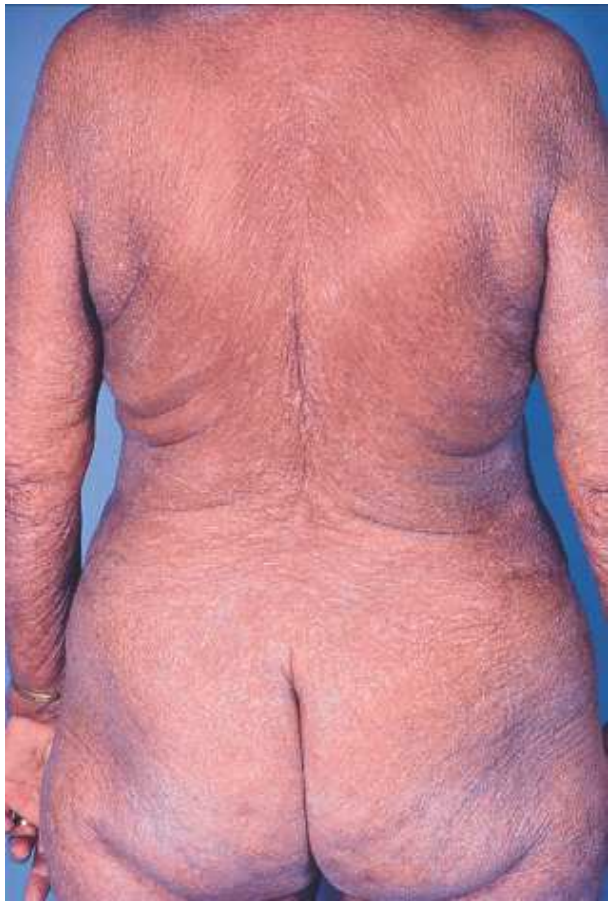
Author information

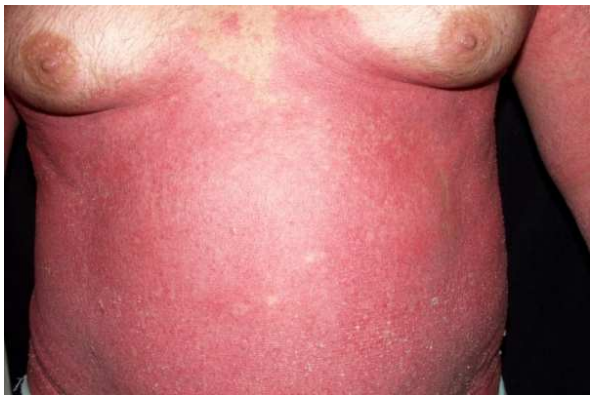
Abstract

The skin may exhibit the first clinical evidence of a systemic disease and may provide the first clues to a diagnosis in malignancies. Erythroderma is defined as generalized redness and scaling and it is a clinical manifestation of a variety of underlying diseases including, rarely, solid tumors. Breast cancer is associated with a variety of skin paraneoplastic manifestations like acanthosis nigricans, erythromelalgia, thrombotic thrombocytopenic purpura, acrokeratosis paraneoplastica, dermatomyositis, systemic sclerosis, and scleroderma. However, in the literature, the correlation of erythroderma with breast cancer is quite infrequent. Here, we describe a case of a 76-year-old woman who presented with a paraneoplastic manifestation of erythroderma due to breast cancer.

ERITRODERMIA

- La dermatitis exfoliativa generalizada, 90%) de aparición brusca en un paciente, generalmente adulto, sin antecedentes de dermatosis (atopia, psoriasis) ni toxicodermia, debe siempre **sugerir malignidad subyacente**.
- 4-21 % de eritrodermias se asocia con malignidad. Linfoma-leucemia
- A veces es por el propio proceso (síndrome de Sézary)





AMILOIDOSIS SISTÉMICA

doi: 10.1111/j.1346-8138.2008.00487.x *Journal of Dermatology* 2008; **35**: 371–372

LETTER TO THE EDITOR

Systemic amyloidosis associated with multiple myeloma presenting as periorbital purpura

Asociación con mieloma múltiple en el 15% de

Ann Oncol. 2014 Feb;25(2):511-8. doi: 10.1093/annonc/mdt544. Epub 2014 Jan 9.

Cancer risk in amyloidosis patients in Sweden with novel findings on non-Hodgkin lymphoma and skin cancer.

Hemminki K[1], Li X, Försti A, Sundquist J, Sundquist K.

Author information

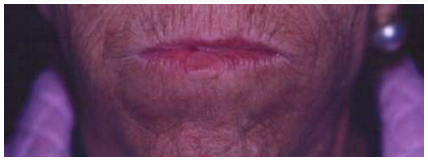
Abstract

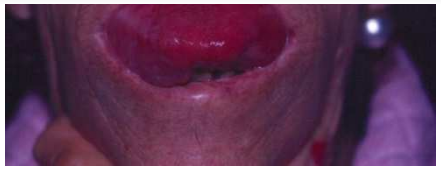
BACKGROUND: Systemic amyloidoses include immunoglobulin light chain (AL) amyloidosis, serum amyloid (AA)-related amyloidosis and senile systemic amyloidosis (SSA). AL amyloidosis is associated with myeloma, and we showed recently that transthyretin-related hereditary amyloidosis was related to non-Hodgkin lymphoma (NHL). In SSA, amyloids constitute wild-type transthyretin. We wanted to analyze cancer risks in amyloidosis, particularly in SSA.

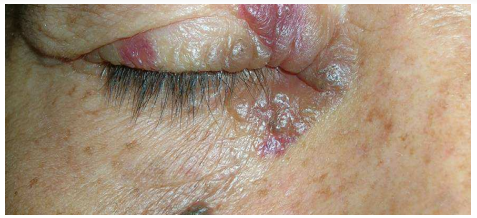
PATIENTS AND METHODS: Nonhereditary amyloidosis patients were identified from the Swedish Hospital Discharge and Outpatients Registers from years 1997 through 2010. Their cancer risk was assessed based on the Swedish Cancer Registry using standardized incidence ratio (SIR) between amyloidosis patients and the remaining population. To gain information about amyloidosis subtypes, we used the Swedish Prescribed Drug Register from years 2005 through 2010 to find out the specific medication prescribed.

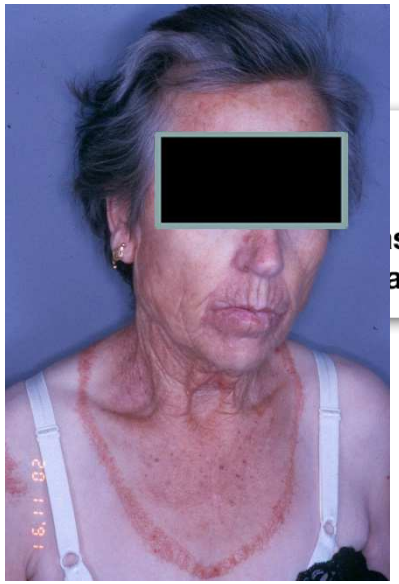
RESULTS: Among 1400 identified amyloidosis patients, cancer risk was increased for myeloma, NHL and squamous cell skin cancer. Myeloma and skin cancers were diagnosed 7-8 years earlier than in the population, whereas NHL was diagnosed in elderly patients. The SIR was 204 for myeloma in patients who received AL amyloidosis medication, and it was 17.22 in patients receiving rheumatoid arthritis medication, suggesting AA amyloidosis. In remaining patients, including SSA, NHL risk was 14.78, including lymphoplasmacytic lymphoma and Waldenstrom macroglobulinemia (51.41) and diffuse large B-cell lymphoma (18.69). In these patients, endometrial cancer (7.04) and cancer of unknown primary site (6.56) were also increased.

CONCLUSIONS: SSA is likely to be a main cause of NHL in the elderly population. The present findings suggest a novel mechanism for amyloidosis-related cancer, highlighting the role of chronic stimulation by amyloid.









Journal of Dermatology 2008; **35**: 371–372

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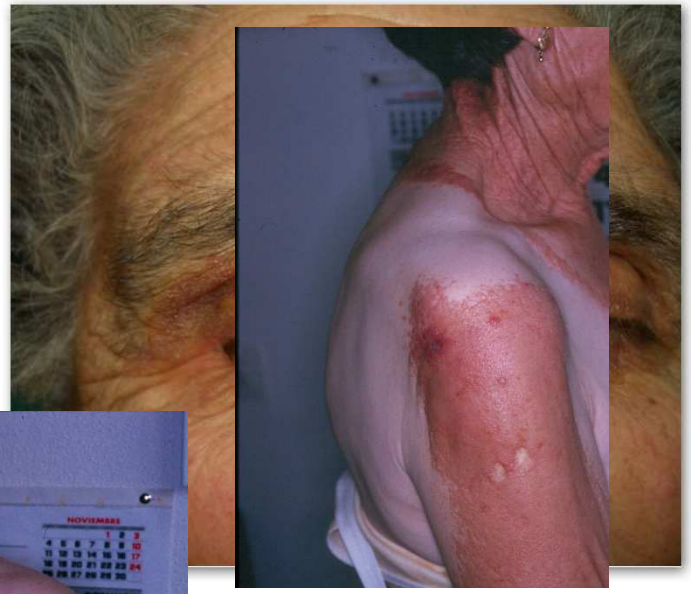

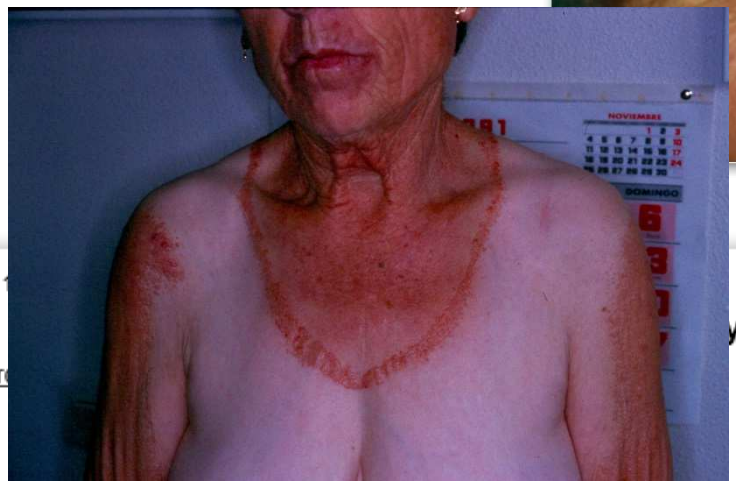

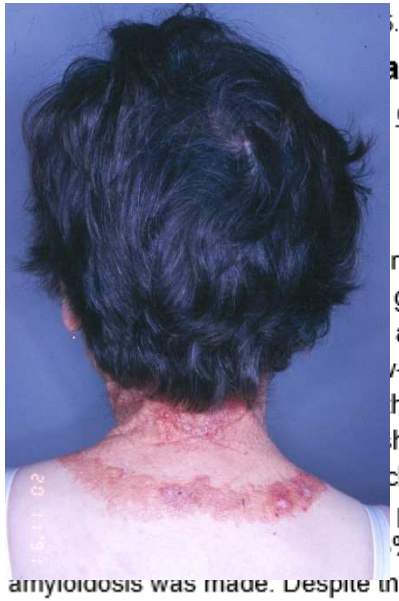

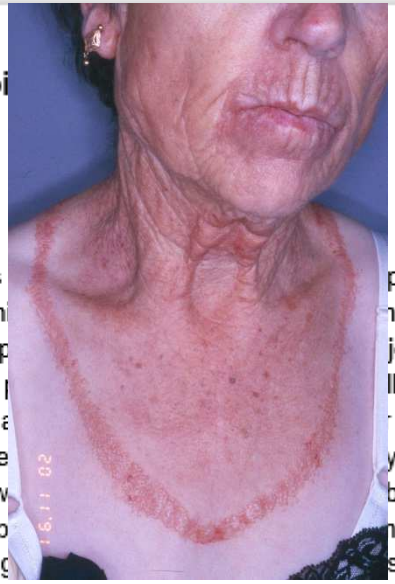

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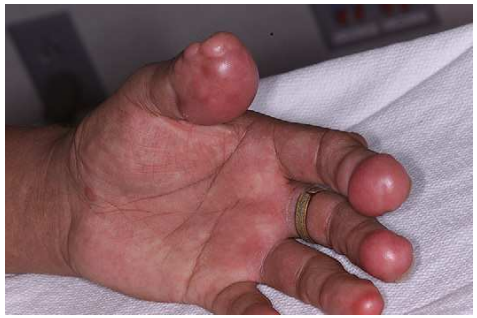
ry of bilateral carpal tunnel surgery complained of worsening hand pains presented generalized arthralgias, symmetric polyarthritis of the hands and feet, sh ning, tender and photosensitivity. The only laboratory abnormality found then was a p joint -dose prednisone and hydroxychloroquine. However, the skin tightness p lly and hat point, more specific autoimmune work-up showed negative relevant a antibody he reported dysphagia and hoarseness, and ecchymotic rashes appeare y of the clastic vasculitis. Staining for amyloid was negative. Subsequently, she w bulinemia progression of her skin symptoms provoked a repeat skin biopsy with sp yloidosis. % plasma cells, skeletal survey revealed multiple lytic lesions, and a diag ssociated amyloidosis was made. Despite the initial features of connective tissue disease in this young woman, a steadfast workup revealed the source of her problem.

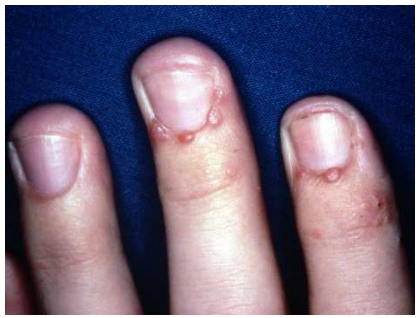
DERMATOSIS PARANEOPLÁSICAS FACULTATIVAS

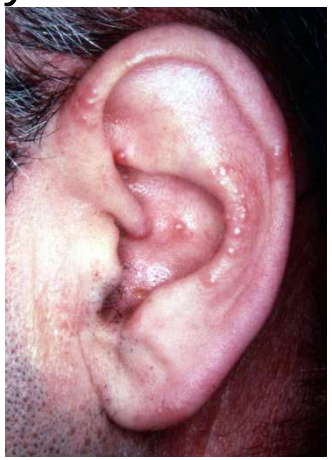
- Queratosis seborreicas eruptivas. S. de Lesser Trelat
- Reticulo histiocitosis multicéntrica
- Escleromixedema
- Xantogranuloma necrobiótico y otros xantomas
- Calcinosis cutánea tumoral
- Vasculitis paraneoplásicas
- Síndrome acral vascular paraneoplásico

RETICULOHISTIOCITOSIS MULTICÉNTRICA

- La reticulohistiocitosis multicéntrica (RHM) es una enfermedad de etiología desconocida que afecta principalmente a mujeres en la cuarta década de la vida.
- La afección articular se caracteriza por la presencia de sinovitis simétrica de miembros superiores especialmente en manos
- afección de piel presentando lesiones de diferente morfología más comúnmente nódulos y pápulas localizadas principalmente en cara y miembros superiores.
- Puede existir además compromiso de órganos internos como pulmón, corazón, tracto gastrointestinal y glándulas salivales.
- Esta patología se ha relacionado con la aparición de neoplasias malignas: como mama, gástrico, ovario, cérvix y linfomas, en el 25% de los casos

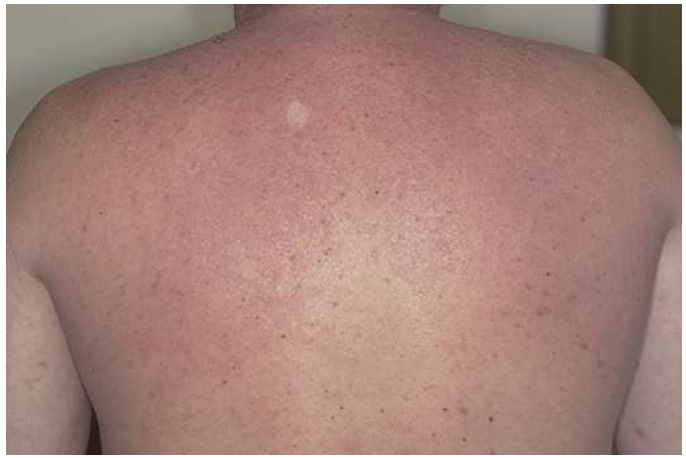


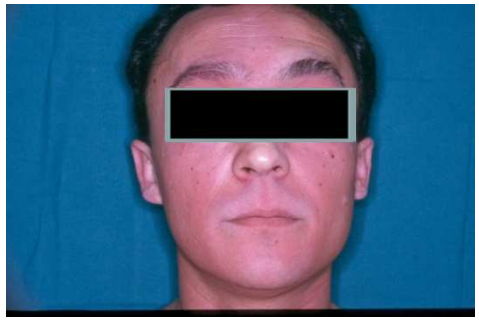


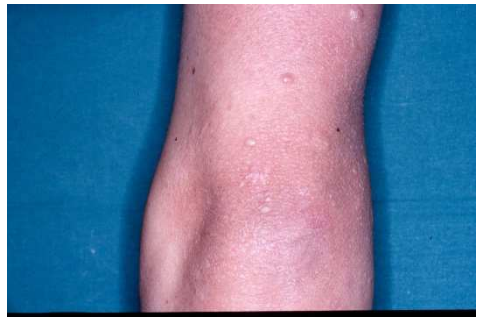


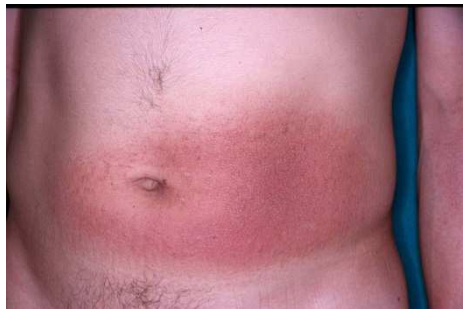
Ann Dermatol Venereol. 2011 May;138(5):405-8. doi: 10.1016/j.annder.2011.01.030. Epub 2011 Feb 26.
[Paraneoplastic multicentric reticulohistiocytosis].

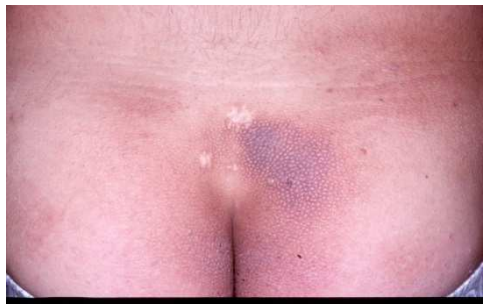
ESCLEREDEMA Y ESCLEROMIXEDEMA









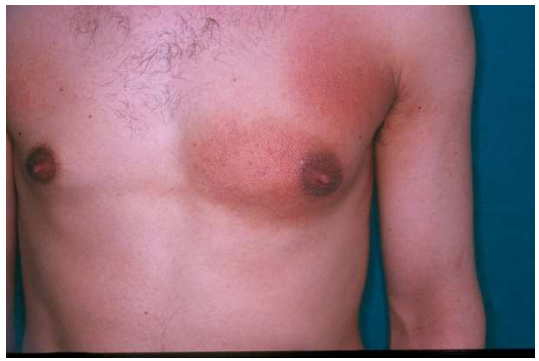


- Mucinosis cutánea
- Paraproteinemia

(gammapatía monoclonal IgG).

- Leucemias,Linfomas

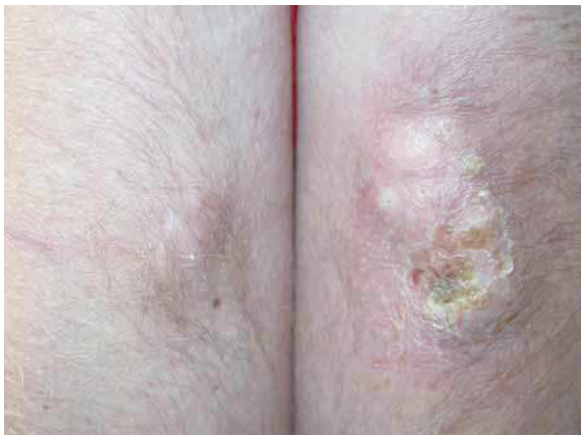
Y Ca. Timo

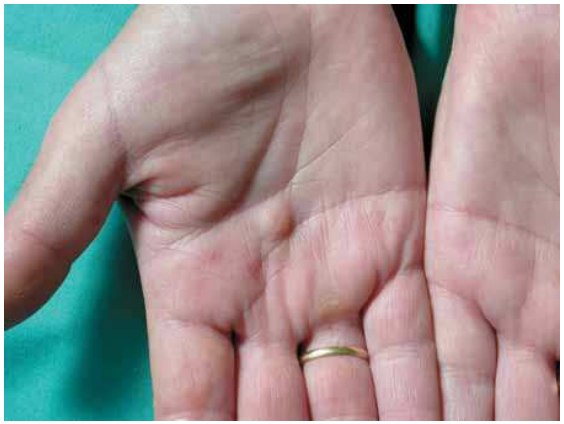


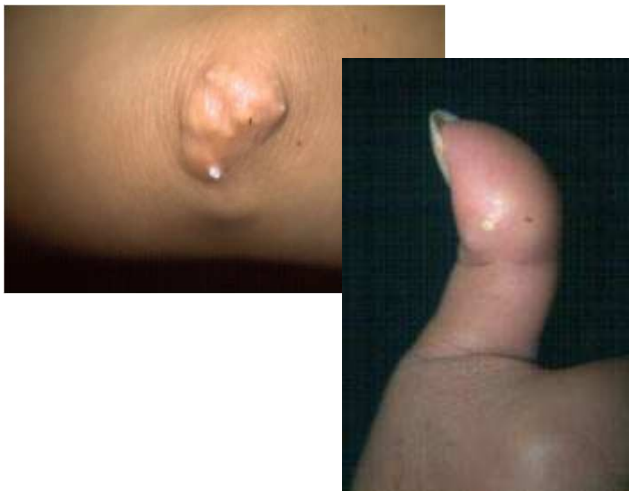
J Clin Oncol. 2012 Jan 20;30(3):e27-9. doi: 10.1200/JCO.2011.38.3935. Epub 2011 Dec 12.

Scleromyxedema: a cutaneous paraneoplastic syndrome associated with thymic carcinoma.

Chan JC[1], Trendell-Smith NJ, Yeung CK.







CALCINOSIS CUTÁNEA TUMORAL

- **Carcinoma de esófago**
- **Cáncer de mama**
- **Mieloma múltiple**
- **Linfoma**
- **Metástasis osteolíticas**

Journal of General Internal Medicine
January 2014, Volume 29, Issue 1, pp 246-247

Date: 14 Aug 2013

Calcinosis in Adult-Onset Dermatomyositis: Metastatic or Dystrophic?

Barry Ladizinski MD, Ali Khan MD, MPP, Christopher Sankey MD

Causes and mechanisms of paraneoplastic calcium deposition of the dermis and subcutis

Tumor product	Serum Ca	Serum PO_4	PTH	Tumor type
PTH secreting tumors	↑	↓	↑	Parathyroid adenoma, other tumors
PTHrP secreting tumors	↑	↓	nl/↓	Breast cancer, SCC, lymphoma, myeloma
Osteolytic metastases	↑	↓	nl/↓	Tumor secretion of cytokines and PTHrP
Vitamin D secreting tumors	↑	↑	nl/↓	Lymphomas

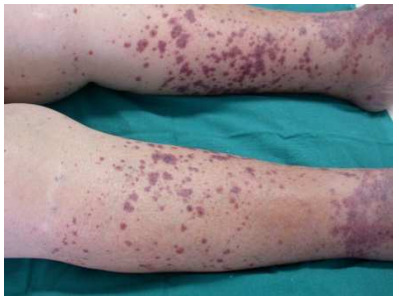
Abbreviations: Ca, Calcium; nl, normal; PO_4, phosphate; PTH, parathyroid hormone; PTHrP, parathyroid hormone–related peptide; SCC, squamous cell carcinoma.

The Spectrum of Paraneoplastic Cutaneous Vasculitis in a Defined Population

Incidence and Clinical Features

Javier Loricera, MD, Vanesa Calvo-Río, MD, Francisco Ortiz-Sanjuán, MD, Marcos A. González-López, MD, PhD, Hector Fernández-Llaca, MD, Javier Rueda-Gotor, MD, Maria C. Gonzalez-Vela, MD, PhD, Lino Alvarez, MD, Cristina Mata, MD, Domingo González-Lamuño, MD, PhD, Victor M. Martínez-Taboada, MD, PhD, Miguel A. González-Gay, MD, PhD, and Ricardo Blanco, MD, PhD**

(*Medicine* 2013;92: 331–343)



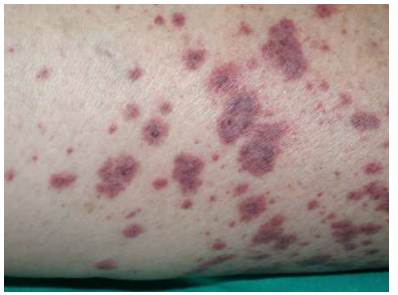
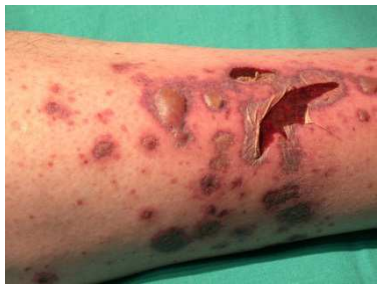
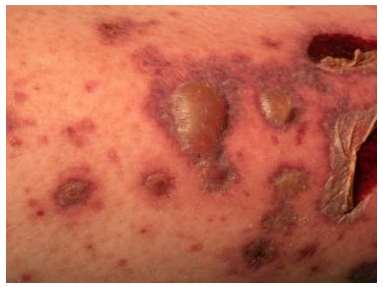


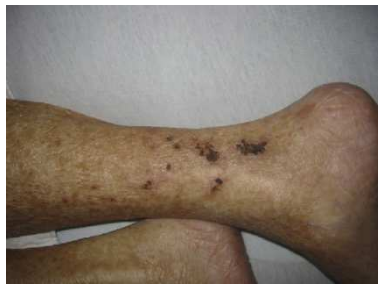
TABLE 1. Main Clinical Features of 16 Patients Presenting With Cutaneous Vasculitis, Confirmed by a Skin Biopsy Showing Leukocytoclastic Vasculitis, Who Were Finally Diagnosed as Having a Malignancy

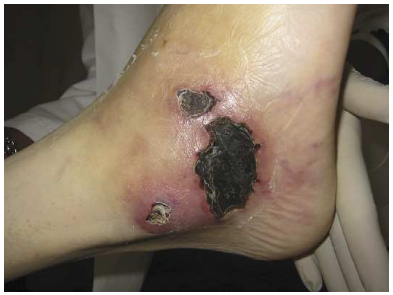
Patient	Age/Sex (yr)	Main Clinical Feature	Peripheral Blood Smear	Neoplasia
1	83/M	Palpable purpura, necrotic ulcer, constitutional symptoms	Anemia, leukopenia	Myelodysplastic syndrome
2	52/W	Palpable purpura, constitutional symptoms	Pancytopenia, immature cells	Myelodysplastic syndrome
3	56/M	Palpable purpura, constitutional symptoms, fever	Anemia, leukopenia, immature cells	Myelodysplastic syndrome
4	70/M	Palpable purpura, constitutional symptoms, polyarthritis	Anemia, leukopenia, immature cells	Non-Hodgkin lymphoma
5	78/M	Palpable purpura, constitutional symptoms, fever, arthralgia, abdominal pain	Anemia	Waldestrom macroglobulinemia
6	61/W	Palpable purpura, hematuria, polyneuropathy	Anemia	Waldestrom macroglobulinemia
7	76/M	Palpable purpura, urticarial lesions, constitutional symptoms, fever	Pancytopenia, immature cells	Hairy cell leukemia
8	81/W	Palpable purpura, erythema, constitutional symptoms, fever	Anemia, immature cells	Mantle cell lymphoma
9	40/W	Urticarial lesions, fever, polyarthritis	Anemia, immature cells	Megakaryocytic leukemia
10	49/W	Palpable purpura, constitutional symptoms, fever	Anemia	Infiltrating breast carcinoma
11	80/M	Palpable purpura, constitutional symptoms	Anemia	Lung adenocarcinoma
12	85/W	Palpable purpura, ulcers	Normal	Breast carcinoma
13	53/M	Palpable purpura, arthralgia	Normal	Pyriform sinus squamous cell carcinoma
14	71/M	Palpable purpura, constitutional symptoms	Normal	Bladder carcinoma
15	70/M	Palpable purpura	Normal	Glottic squamous cell carcinoma
16	82/M	Palpable purpura, abdominal pain, fecal occult blood, hematuria	Normal	Oropharyngeal squamous cell carcinoma

- 3.80%
- 100% 1º signo
- 56% Hematológicos
- 44% T. sólidos
- Comparando con Vasculitis no Paraneo.
  - ❖ Edad elevada
  - ❖ Sin factores precipitantes
  - ❖ Clínica mas intensa y persistente
  - ❖ Mayor clínica constitucional y menor clínica digestiva o renal
  - ❖ Citopenia, anemia y VSG elevada









Paraneoplastic acral vascular syndrome: Epidemiologic features, clinical manifestations, and disease sequelae

Ewa Poszepczynska-Guigné, MD,[a] Manuelle Viguier, MD,[a] Olivier Chosidow, MD, PhD,[b] Brigitte Orcel, MD,[c] Joseph Emmerich, MD, PhD,[d] and Louis Dubertret, MD[a] *Paris, France*

Background: Acral vascular syndromes associated with malignancy have rarely been reported.

Objective: Our purpose was to assess the clinical and evolving features of paraneoplastic acral vascular syndromes.

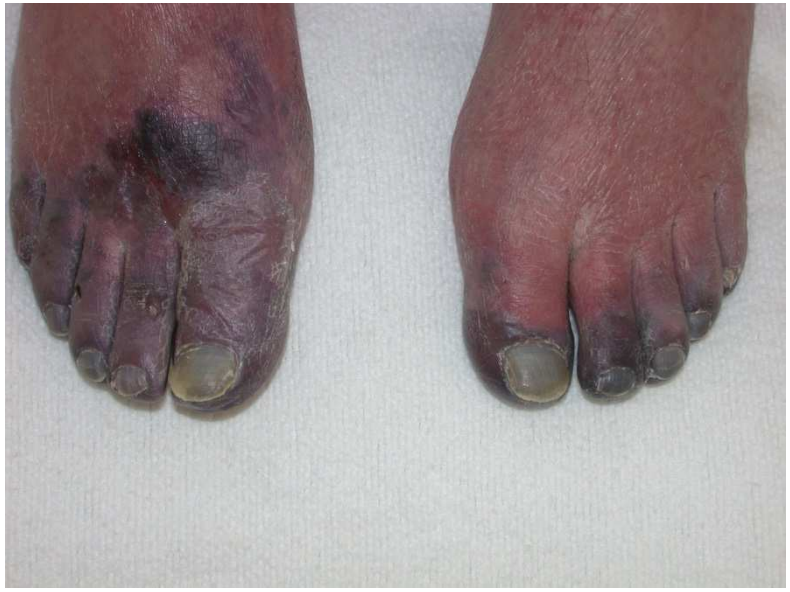
Patients and Methods: Two cases of paraneoplastic gangrene are described and analyzed together with previously reported cases identified by a MEDLINE search.

Results: Among the 68 patients identified, 40 had gangrene, 16 had acrocyanosis, and 12 had Raynaud's phenomenon. The male to female ratio was 0.89; median age was 59 years. Fingers were affected in 94%. Adenocarcinomas were the predominant associated malignancies (41%), and metastases were observed in 41%. The acral vascular syndromes in 48% of the patients definitively regressed after tumor treatment. Forty-four percent of the patients died within 2 years. A favorable cutaneous outcome was obtained with prostacyclin infusions in 6 patients.

Conclusion: A neoplastic origin of acral vascular syndrome should be considered in elderly patients, especially men, in the absence of usual causative conditions. (J Am Acad Dermatol 2002;47:47-52.)

Conclusion: A neoplastic origin of acral vascular syndrome should be considered in elderly patients, especially men, in the absence of usual causative conditions. (J Am Acad Dermatol 2002;47:47-52.)

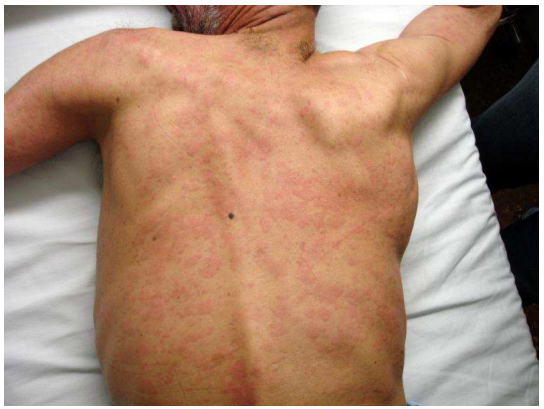
- **Clínica**
- Gangrena digital (40 pacientes, 59%)
- Fenomeno de Raynaud precediendo a la gangrena(21 pacientes 53%)
- Acrocianosis (16 pacientes, 24%)
- Fenómeno de Raynaud no complicado (12 pacientes, 18%)

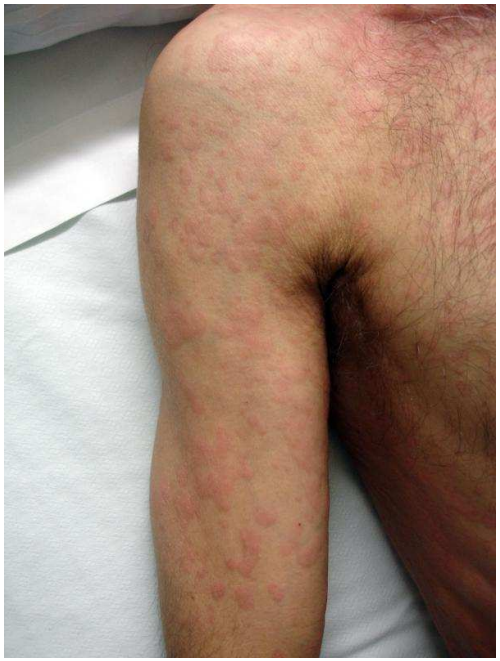


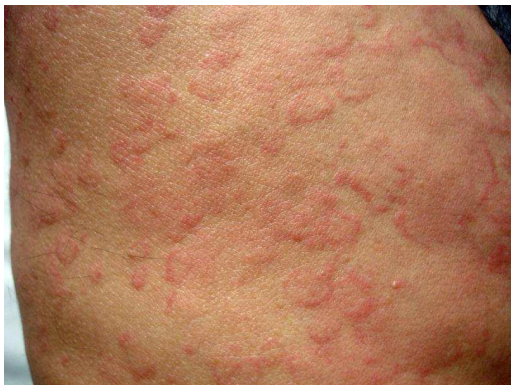
- **Epidemiología**
- 28 (41%) tenían adenocarcinomas, sobre todo pulmón (6) y ovario (6)
- 13 (19%) tenían neoplasias hematológicas (linfomas, distintos tipos de leucemia…)
- 12 (18%) asociaban carcinomas anaplásicos y epidermoides

Síndromes Urticariales

- **S. de Schnitzler: Urticaria crónica no pruriginosa, fiebre recurrente, dolor óseo y otoesclerosis, adenopatías, megalias, aumento VSG, asociado a gammapatía monoclonal Ig M con o sin cambios de enfermedad linfoproliferattiva.**







Jimenez Puya R y cols. Urticarias y exantemas urticariales. Dermatologia 2007; vol 20; nº 2; 78-91