

**SAP**

ASOCIACIÓN ARGENTINA DE PEDIATRÍA



Semana de Congresos y Jornadas Nacionales 2017

# Paciente con acné e hidrosadenitis

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Dr. R. Cano.

**A** Hospital  
Alemán  
Deutsches Hospital

Servicio de Dermatología  
Hospital Alemán

# Historia clínica:

masculino, 16 años de edad

Antecedentes familiares: niega.

Antecedentes personales:

-IMC: 30

-**Acné** con compromiso en

o.



# il de 2015:

dro interpretado como  
**ermitis a nivel axilar:**  
cotrimoxazol 160mg/800mg  
12 horas VO.

**Internación:**

Clindamicina EV.

Debridamiento quirúrgico: sin rescate  
microbiológico.



- Alta hospitalaria con clindamicina 300 mg/6 hs  
VO.

yo de 2015:

**Novo episodio:** internación y  
tratamiento con clindamicina EV.

**Acrosadenitis**

**purpurativa:**

isotretinoína 40 mg/día.

Metilprednisona: 40 mg/día.

- Alta con cotrimoxazol  
160/800 mg cada 12  
hs.



io de 2015:

a respuesta terapéutica: **Acitretín** 25 mg/12 hs +

prednisona

compromiso en región glútea e interglútea:

- Clindamicina + rifampicina



Inicia

**ciclospor**

# re DLQI: 28

	MUCHISIMO	MUCHO	UN POCO	NO
LOR, PICOR, ESCOZOR	X			
RGUENZA	X			
ERFERENCIA EN SALIR	X			
ERFERENCIA EN VESTIRSE	X			
TIVIDAD SOCIAL	X			
TIVIDAD FISICA	X			
BAJO/ESTUDIO	X			
MILIA/AMIGOS			X	
TIVIDAD SEXUAL	X			
MPO DISPONIBLE	X			

# Modificación de Hurley:

DIO	ABSCESOS	TRACTOS FISTULOSOS/CICATRIZACIÓN
	Uno o más	No
	Separados en el espacio y recurrentes	Escasa afectación
	Múltiples	Múltiples



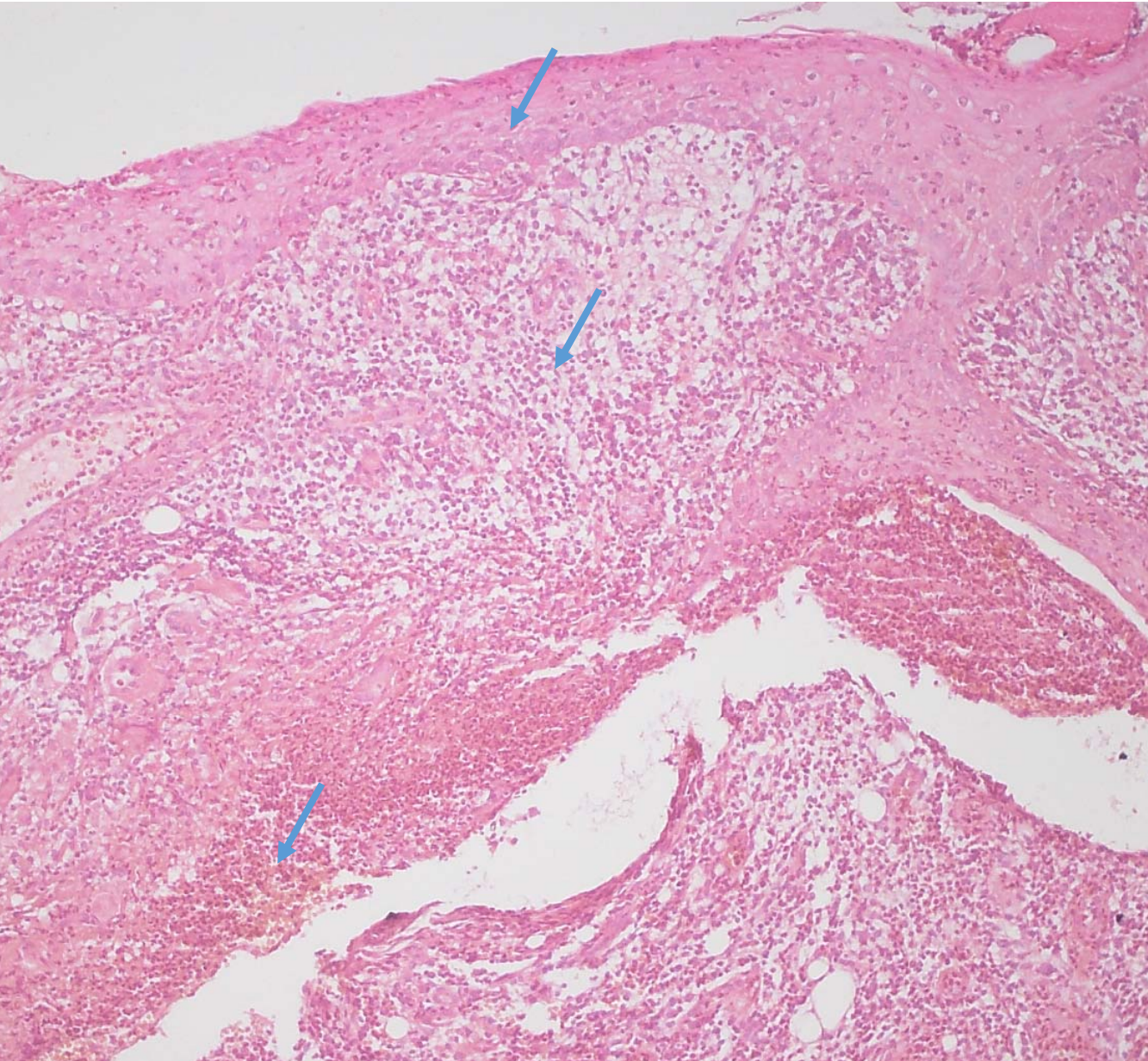
# Tratamiento con ciclosporina:

## Acta de respuesta:





# Estudio histopatológico:



- Epidermis:
  - Acantosis leve.
  - Exocistosis neutrofil
- Dermis:
  - Infiltrado neutrofilic
  - Microabscesos.

Compatible con  
pioderma gangren

# ordaje terapéutico:

útiples  
ternaciones.  
LQI28.  
urley III.  
ntibióticos,  
tinooides,  
cospolina.

Inicia

## Adalimuma

- 160 mg SC
- 80 mg SC a los 15 días.
- 40 mg/sem SC a a partir de  
mes.

evolució evolutiva: 1 er. mes.



evolutivo: 2 do. mes.



evolutivo: 6 to. mes.



Conclusión:

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Acné.  
Hidrosadenitis  
purpurativa  
calcitrante.

- ¿Pioderma gangrenoso?

¿Enfermedad  
autoinflamatori

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Grupo heterogéneo de síndromes clínicos secundarios  
actividad aumentada del sistema inmune  
ato, con una predisposición genética significativa.

# Immunomodulatory autoinflammatory diseases:

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## Comparison Chart of Systemic Autoinflammatory Diseases (SAID)

SAID	FCAS	MWS	NMDC/JCFA	SCHMITZLER	FMF	TRAPS	HIDS	MAJED	CEM/Q/SAPHO	DIRTRAPORP	CAMPS/PSORS2	PAPA	BLAU/PA/GEOS	NLRP2/FCAS2	CANDLE/PRAAS	BEHÇETS/BD	SOJA/PA/JIA	AOSD	1° HLH/FHL	PLD2/FCAS3	APLAID	SLCS2AS	AD-AD
<b>Classification</b>	Crystallin-Associated Periodic Syndromes (CAPS)	Muckle-Wells Syndrome*	Neonatal-Onset Multisystem Autoinflammatory Disease -aka Chronic Infantile Neurological Arteritis Syndrome (CINCA)	Schnitzler Syndrome	Familial Mediterranean Fever*	Tumour Necrosis Factor (TNF)-Associated Periodic Syndrome -aka Familial Fibrous Fever*	Hyperimmunoglobulinemia D with Periodic Fever Syndrome (HIDS)	Mevalonic Aciduria (MVA)	Deficiency of Interleukin-18 (IL-18) Receptor Antagonist (DIRA)	Majed Syndrome -aka Chronic Recurrent Multifocal Osteomyelitis (CRMO)	Deficiency of Interleukin-36 Receptor Antagonist (DIRA)	Pyogenic Sterile Arthritis, Pyoderma Gangrenosum, and Acro Syndrome	Granulomatous	Monarch-1	Proteasome	Sahlgrenska Disease	Idiopathic	Macrophage Activation Diseases	PLD2-associated	Autoinflammation & PLD2-associated	SLCS2AS related	AD-AD	AD-AD
<b>Genetics</b>	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive	Autosomal recessive
<b>Age of Onset</b>	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood	Infancy to childhood
<b>Key Features</b>	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis	Large febrile episodes, skin rashes, conjunctivitis

**AD-AD** (Autoinflammation & PLD2-associated) is a newly identified SAID characterized by recurrent febrile episodes, skin rashes, and conjunctivitis. It is caused by a mutation in the PLD2 gene. Treatment with IL-1 inhibitors is effective. **SLCS2AS** (Sendai-like Crystallin-Associated Syndrome) is a newly identified SAID characterized by recurrent febrile episodes, skin rashes, and conjunctivitis. It is caused by a mutation in the SLCS2AS gene. Treatment with IL-1 inhibitors is effective. **AD-AD** (Autoinflammation & PLD2-associated) is a newly identified SAID characterized by recurrent febrile episodes, skin rashes, and conjunctivitis. It is caused by a mutation in the PLD2 gene. Treatment with IL-1 inhibitors is effective. **SLCS2AS** (Sendai-like Crystallin-Associated Syndrome) is a newly identified SAID characterized by recurrent febrile episodes, skin rashes, and conjunctivitis. It is caused by a mutation in the SLCS2AS gene. Treatment with IL-1 inhibitors is effective.

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# Enfermedades autoinflamatorias:

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## Pyoderma gangrenosum, acne, and suppurative hidradenitis (PASH)—a new autoinflammatory syndrome distinct from PAPA syndrome

Markus Braun-Falco, MD,<sup>a</sup> Oleksandr Kovnerystyy, MD,<sup>a</sup> Peter Lohse, MD,<sup>b</sup> and Thomas Ruzicka, MD<sup>a</sup>  
*Munich, Germany*

“PASH”

**P:** Pyoderma gangrenosum  
**A:** Acné  
**SH:** Suppurative Hidrosadenitis

Aumento del número de repeticiones  
microsatelites CCTG del gen **PSTPIP1**





# Ermedades autoinflamatorias:

## “PAPASH”

2013

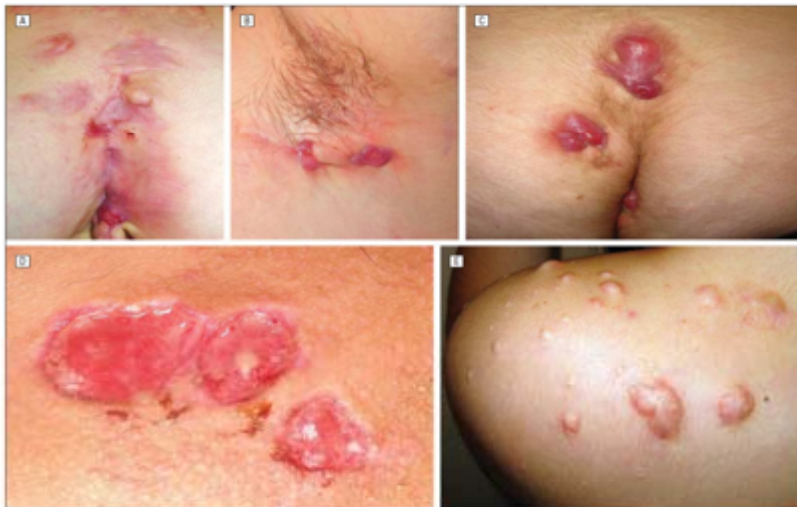
**Pyogenic Arthritis, Pyoderma Gangrenosum, Acne, and Hidradenitis Suppurativa (PAPASH): A New Autoinflammatory Syndrome Associated With a Novel Mutation of the *PSTPIP1* Gene**

PA: Pyogenic Arthritis

P: Pyoderma gangrenosum

A: Acné

SH: Suppurative Hidrosadenitis



2014

## “PAPASH”

Pyoderma gangrenosum, acne, psoriasis, arthritis and suppurative hidradenitis (PAPASH)-syndrome: a new entity within the spectrum of autoinflammatory syndromes?

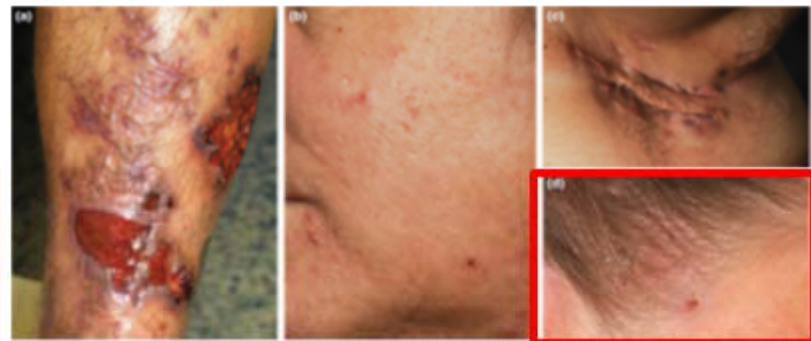
P: Pyoderma gangrenosum

A: Acné

P: Psoriasi

A: Arthritis

SH: Suppurative Hidrosadenitis



# Después de la presentación:

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paciente con dermatosis inflamatoria no infecciosa recalcitrante que requirió el uso de agentes biológicos.

Las características y curso clínicos atípicos junto con la biopsia con cambios compatibles con pioderma gangrenoso nos motivó a considerar a las enfermedades autoinflamatorias como diagnóstico diferencial.

Recordar que el acné puede formar parte del espectro de las enfermedades autoinflamatorias, debiendo sospecharse en casos recalcitrantes, con episodios febriles y cuando asocian otras manifestaciones clínicas.

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Muchas grac