EXANTEMA AGUDO EN VARÓN DE 34 AÑOS

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ANAMNESIS/HISTORIA ACTUAL

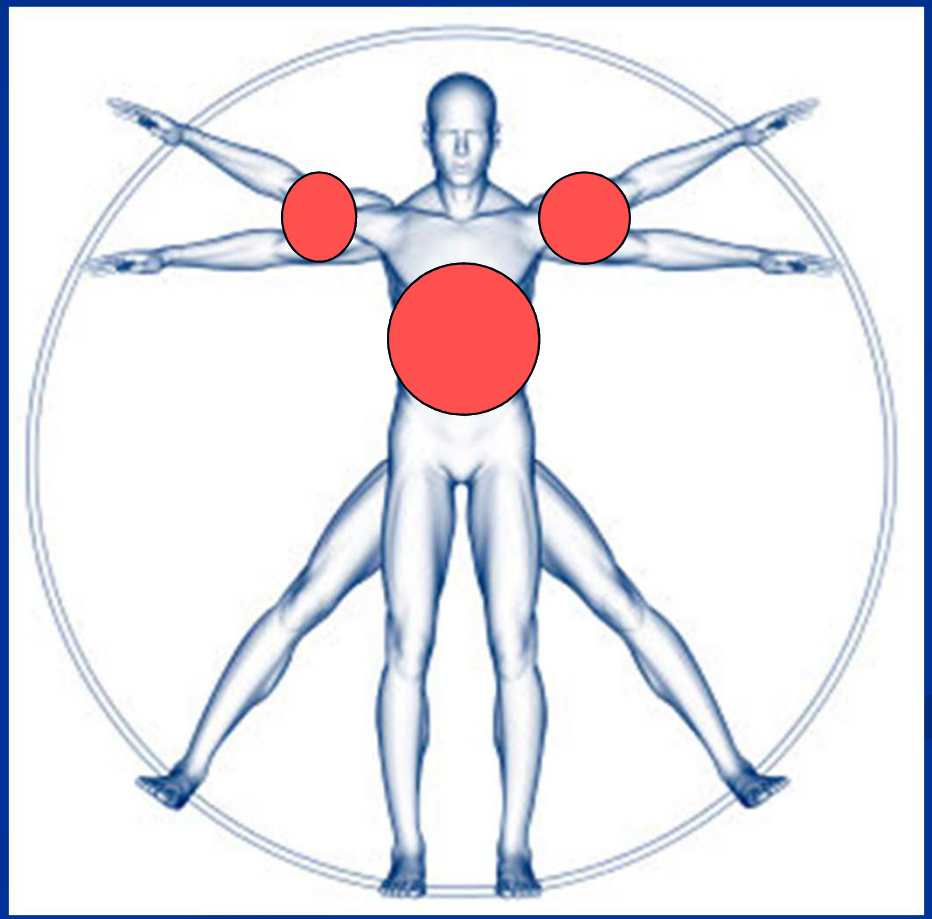
- Varón de 34 años , búlgaro:
  - Hepatitis C
  - Ex- consumidor de cocaína hace 4 años.
- Dolor torácico de características pleuríticas y disnea de 5 días:
  - Fiebre (los dos últimos días) y dolor en miembro inferior izquierdo.
- Eco-doppler de miembros inferiores y TAC torácico :
  - **TVP en MII**
  - **TEP bilateral**
  - **Nódulos pulmonares cavitados**
- Ingresa en UCI.

ANAMNESIS/HISTORIA ACTUAL

- Ingreso en Medicina Interna.
- Durante su estancia en planta:

EXANTEMA PUSTULOSO
FIEBRE
LEUCOCITOSIS

BIOPSIA



EXPLORACIÓN FÍSICA

- TA: 120/70 mmHg, FC: 90 lpm, Tª: 37,8ºC, Sat O2 100% con Vmask 50%.
- Cavidad Bucal: Lesión blanquecina en cara lateral izquierda de la lengua.
- CYC: Adenopatías retroauriculares izquierdas y axilares.
- AC/AP: normal.
- Abdomen: dolor a la palpación en hipocondrio derecho. RHA normales.
- MMII: aumento de perímetro de MII. Pulsos periféricos palpables.
  - Adenopatías inguinales bilaterales:
    - > izquierda, > 1 cm, adherida a planos profundos, no dolorosa.
- Lesiones pápulo-pustulosas (fondo eritematoso, consistencia firme y costra)

PRUEBAS COMPLEMENTARIAS

ANALÍTICA

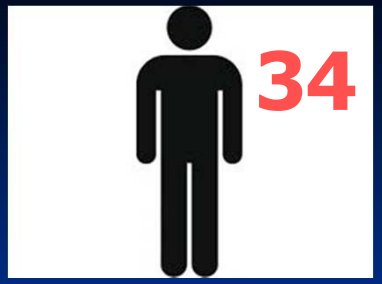
- Leucocitos 20.6 miles/mcL (PMN 86%), Hb: 12 g/dl, plaquetas: 233 miles
- Actividad de protrombina: 61%, ATTP: 32 seg. Fibrinógeno: 874 mg/dl.
- Gluc: 99 mg/dl, Urea:17 mg/d, Creatinina: 0.5 mg/d, Bilirrubina Total :0.5 mg/d, GOT:17 UI/L, GPT:33 UI/L, CPK:110 UI/L. Na:137 mmol/L, K:3.2 mmol/L. Ca:8.7 mmol/L, Fósforo:4.4 mmol/L, Magnesio:2.2 mmol/L.
- Fe: 8 mcg/dL, Ferritina: 851 ng/ml, Ácido Fólico: 3.1 pg/ml.
- VSG: 120 mm.

AUTOINMUNIDAD

- ANA, ANCA negativos, Complemento normal.

PRUEBAS COMPLEMENTARIAS

- **MANTOUX** : 0 mm de induración
- **SEROLOGÍA**
  - VHA (-), VHB (infección pasada), VHC (positivo) con carga viral indetectable.
  - VIH (-). *Mycoplasma, Chlamydia, Coxiella Burnetti y Brucella* Negativos.
- **HEMOCULTIVO**: positivo para *S. aureus* Meticilina sensible.
- **ECOCARDIOGRAMA TRANSTORÁCICO**
  - FEVI conservada. No se objetivan alteraciones en cavidades ni a nivel valvular.
  - No derrame pericárdico. No datos de endocarditis.
- **CULTIVO BIOPSIA ADENOPATÍA INGUINAL**: Aerobios (-). Tinción BAAR (-).
- **CULTIVO DE ESPUTO INDUCIDO**: BAAR (x3) Negativo.



EN RESUMEN:

TVP MMII

TEP BILATERAL

NÓDULOS PULMONARES CAVITADOS

+

FIEBRE

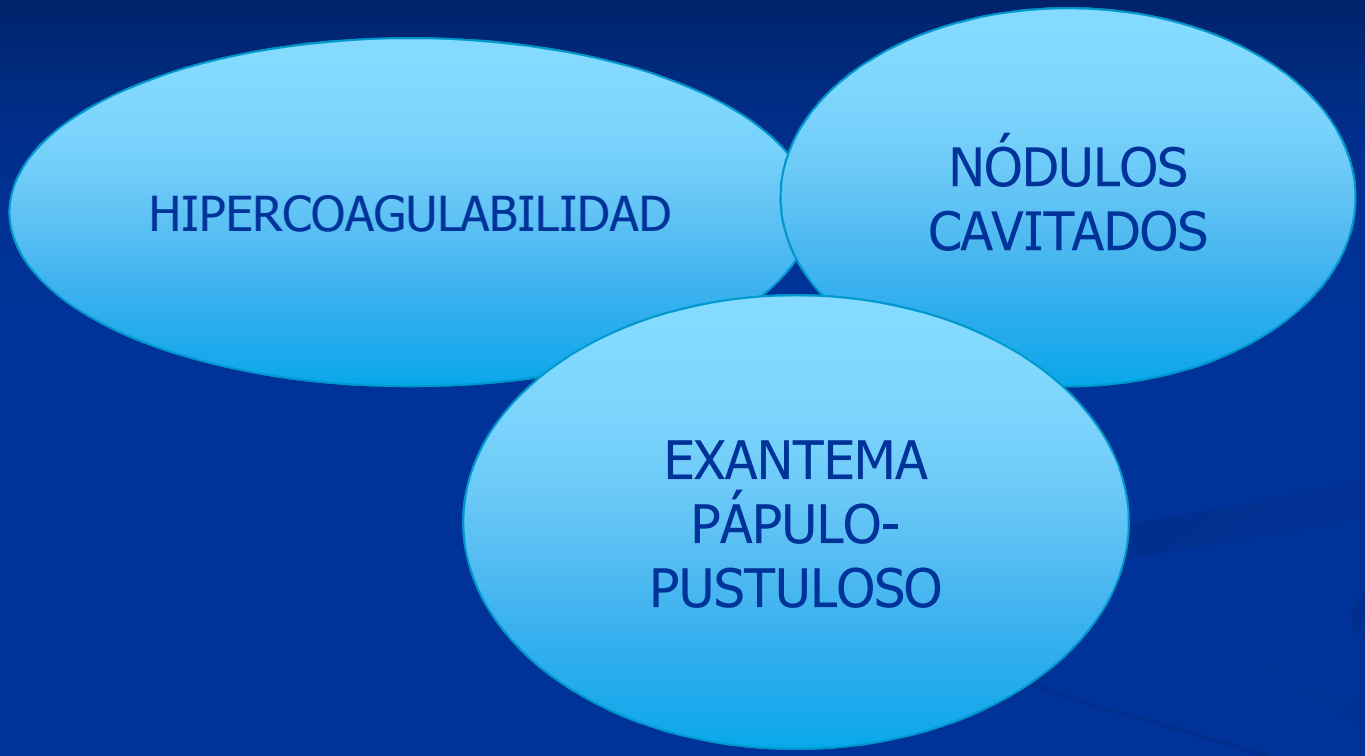
EXANTEMA PAPULO-PUSTULOSO

HIPERCOAGULABILIDAD

NÓDULOS CAVITADOS

EXANTEMA PÁPULO-PUSTULOSO

EN RESUMEN:



ADENOPATÍAS + LESIÓN BLANQUECINA LINGUAL

LEUCOS, FERRITINA, VSG + SAMS → ~~ENDOCARDITIS~~

DIAGNÓSTICO DIFERENCIAL

HIPERCOAGULABILIDAD

- CONGÉNITA

- ADQUIRIDA

Acquired disorders
Malignancy
Presence of a central venous catheter
Surgery, especially orthopedic
Trauma
Pregnancy
Oral contraceptives
Hormone replacement therapy
Tamoxifen, Thalidomide, Lenalidomide
Immobilization
Congestive failure
Antiphospholipid antibody syndrome
Myeloproliferative disorders
Polycythemia vera
Essential thrombocythemia
Paroxysmal nocturnal hemoglobinuria
Inflammatory bowel disease
Nephrotic syndrome

Anticuerpos??
Coagulación N

DIAGNÓSTICO DIFERENCIAL

HIPERCOAGULABILIDAD

- ASOCIADA CON ENFERMEDADES AUTOINMUNES.

Artritis Reumatoide
Lupus Eritematoso Sistémico
Vasculitis autoinmunes de pequeño vaso: -Granulomatosis de Wegener -Síndrome de Churg-Strauss -Poliangeítis microscópica
Enfermedad de Behçet

DIAGNÓSTICO DIFERENCIAL

NÓDULOS PULMONARES CAVITADOS

1.- INFECCIONES

Infecciones oportunistas
Embolismo séptico
Endocarditis

SAMS
ECO TT: N

Tuberculosis

BAAR -

2.- NEOPLASIAS

Carcinoma pulmonar primario
Metástasis
Mieloma múltiple
Linfoma

3.- INMUNOLÓGICAS

Granulomatosis de Wegener
Sarcoidosis
Artritis Reumatoide
Amiloidosis primaria
LES

ANA-
C3/C4 N

4.- VASCULARES

Infartos pulmonares → Sd. Behcet
Malformaciones A-V
Hamartomas

DIAGNÓSTICO DIFERENCIAL

EXANTEMA PÁPULO-PUSTULOSO

Cutaneous infection (bacterial, fungal, and mycobacterial) * Chickenpox *
Urticaria and urticarial vasculitis
Neutrophilic dermatoses
Drug eruptions (lithium, isoniazid)
Halogenoderma (eg, bromoderma, iododerma)

DIAGNÓSTICO DIFERENCIAL

EXANTEMA PÁPULO-PUSTULOSO

¿ DERMATOSIS NEUTROFÍLICA ?

Involving predominantly the dermis
Sweet's syndrome
Pyoderma gangrenosum
Behçet's disease
Bowel-associated dermatosis-arthritis syndrome
Inflammatory bowel disease (may also have small vessel vasculitis)
Neutrophilic eccrine hidradenitis
Rheumatoid neutrophilic dermatitis
Neutrophilic urticaria
Still's disease
Erythema marginatum
Hereditary periodic fever syndrome

Data from: Moschella, SL, Davis, MDP. Neutrophilic dermatoses. In: Dermatology, 2nd ed, Bolognia, JL, Jorizzo, JL, Rapini, RP (Eds), Mosby Elsevier, Spain 2008.

DIAGNÓSTICO DIFERENCIAL

NÓDULOS PULMONARES CAVITADOS

Granulomatosis de Wegener
Enfermedad de Behçet
Linfoma

HIP

Granuloma
Enfermedad de Behçet

EXANTEMA PÁPULO-PUSTULOSO

Síndrome de Sweet
Enfermedad de Behçet

¿ENFERMEDAD BEHÇET?

¿¿BIOPSIA TÁNEA??

TAS?
RENTES?

¿ADENOPATÍAS?

A FAVOR

Tabla 4. *Criterios de clasificación revisados del* International Study Group for Behçet's Disease *(2006).*

Aftosis oral	1 punto
Manifestaciones cutáneas	1 punto
Lesiones vasculares	1 punto
Prueba de la patergia positiva	1 punto
Aftosis genital	2 puntos
Lesiones oculares.	2 puntos

El diagnóstico de enfermedad de Behçet se establece con una puntación ≥ 3 puntos.

Sensitivity	Specificity	Accuracy
96.1 %	88.7%	93.8%

A FAVOR

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Mucocutaneou

Erkan Alpsoy,[1] Chri

Author information ▶ Arti

Cutaneous lesi

Cutaneous lesions
Group for Behçet'
mainly include eryt
of the skin to need

Papulopustular lesions

Go to:

PPL, the most common skin lesions, are cutaneous, sterile, folliculitis - or acne -like lesions on an erythematous base which appear as a papule and in the course of 24-48 hours become pustule[20,32] (Fig. 5). Although four of the five new international criteria for the diagnosis of BD relate to mucocutaneous lesions, disagreement exists as to the exact nature of the cutaneous lesions and the inclusion of cutaneous follicular or acneiform lesions as a major criterion. Many authors[33] believe that these lesions should not be included, since acneiform and folliculitis-like lesions are nonspecific and clinically, may not be differentiated from ordinary acne, particularly in adolescents. In a randomized and controlled study,[20] we counted PPL, including acneiform and folliculitis- like lesions, in a blind protocol according to the 7 different anatomic locations. The frequency of PPL in patients with BD was 96%, and the most common location was the trunk, followed by the extremities, whereas in the control group the frequency was 89% and the most common location was the face (the frequency was 100% and 86.1% in acne and non-acne patients, respectively). Our study suggests that PPL is very sensitive but not specific when acneiform or follicle-based lesions were also included. To differentiate acne lesions from the PPL of BD can be very important, especially in those patients for whom the diagnosis depends on this criterion. In a recent study, on the basis of the above studies,[20,33] we excluded follicle-based acneiform lesions and those lesions over the face since the clinical differentiation from acne vulgaris is nearly impossible at this anatomic location. Instead we investigated whether PPL can be a useful tool for the diagnosis of BD when non-follicular lesions over the trunk or extremities were selected, and were correlated with histologic and/or immunofluorescence study. Our results confirm that the selection of non-follicular lesions and combination with histopathologic and/or immunofluorescence studies increase the specificity of these lesions to establish the diagnosis.[34] On the contrary, the definitive histological diagnosis from the cutaneous lesions is also often difficult without clinical information. Therefore, the clinician should consider correlating the diagnosis of PPL for BD by histological examination and only vessel-based histopathology (leukocytoclastic vasculitis or a neutrophilic vascular reaction) should be included as fulfilling a diagnostic criterion.

A FAVOR

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PMCID: PMC2628050

Mucocutaneous Lesions of Behcet's Disease

Erkan Alpsoy,[1]
Author information

Thrombophlebitis

Go to:

The prevalence of vascular involvement in BD has been reported to be 7.7-60% in different patient populations.

Vascular disease in 728 patients with Behcet's disease

	Number of patients
Venous disease	
Deep venous thrombosis	221
Subcutaneous thrombophlebitis	205
SVC occlusion	122
IVC occlusion	93
Cerebral sinus thrombosis	30
Budd-Chiari syndrome	17
Other venous occlusion*	24
Arterial disease	
Pulmonary artery occlusion or aneurysm	36
Aortic aneurysm	17
Extremity arterial occlusion or aneurysm	45
Other arterial occlusion or aneurysm•	42
Right ventricular thrombus	2

* Other veins included subclavian, iliac, portal, renal, innominate, brachiocephalic.
• Other arteries included iliac, subclavian, renal, carotid, cerebral, coronary, innominate, mesenteric, aorta, basilar, splenic.

EN CONTRA

Tabla 3. *Criterios diagnósticos para la enfermedad de Behçet (Grupo Internacional de Estudio de la Enfermedad de Behçet)**

- **Úlceras orales recurrentes**

  Aftas menores (< 1cm), aftas mayores o úlceras herpetiformes observadas por el médico o el paciente, con un mínimo de 3 episodios, durante un periodo de 12 meses.

Más dos de los siguientes:

- **Úlceras genitales recurrentes**

  Úlceras o cicatrizaciones aftosas observadas por el médico o el paciente

- **Lesiones oculares**

  Uveítis anterior o posterior, o presencia de células en el vítreo al examen con lámpara de hendidura; o bien vasculitis retiniana diagnosticada por un oftalmólogo.

- **Lesiones cutáneas**

  Eritema nodoso observado por un médico o por el paciente, pseudofoliculitis, o lesiones pápulo-pustulosas; o nódulos acneiformes observados por el médico en pacientes post-adolescentes no tratados con corticoides.

- **Fenómeno de patergia positivo**

  Evaluado por un médico a las 48 horas.

* *International Study Group for Behçet's disease. Criteria for diagnosis of Behçet's disease. Lancet 1990; 335: 1078-1080.*

PROPUESTA DIAGNÓSTICA

DIAGNÓSTICO PRINCIPAL

ENFERMEDAD DE BEHÇET

SÍNDROME DE SWEET

DIAGNÓSTICO SECUNDARIO

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MUCHAS GRACIAS