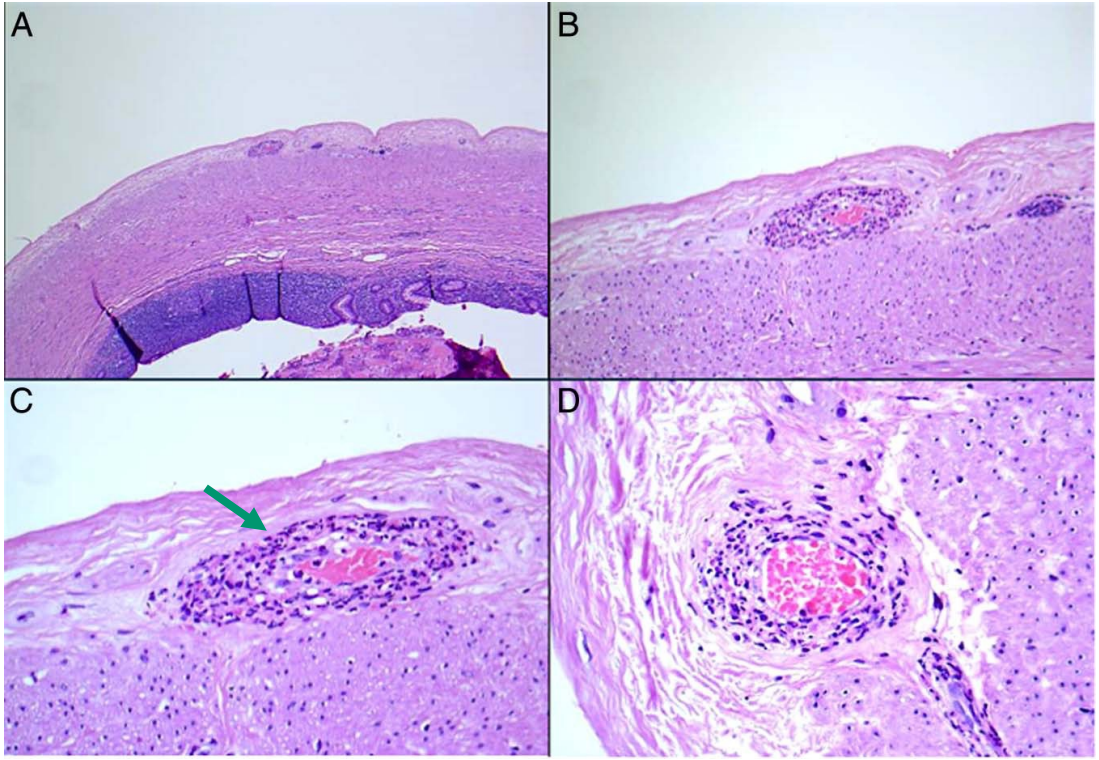
INSELGRUPPE

GI manifestations of vasculitis

Bible class, 17. November 2021, Niklas Krupka

Background



Chetty R, Serra S. J Clin Pathol 2017;70:470–475

"Presence of leukocytes in vessel walls with reactive damage to mural structures"

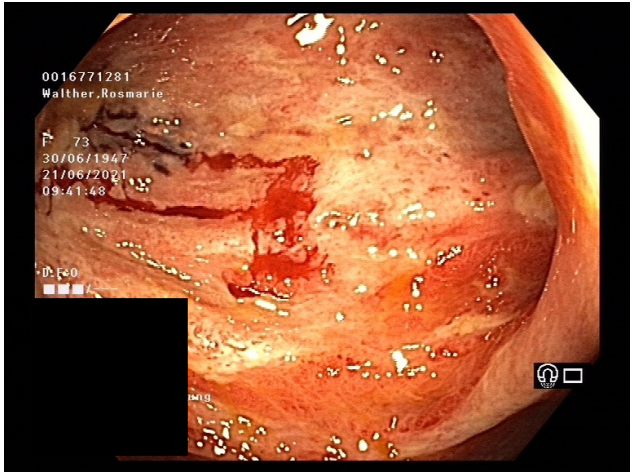
Vasculitis is almost always a multi-organ disease

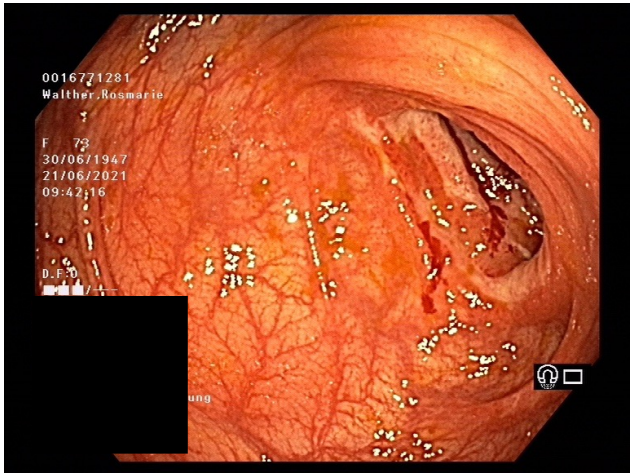
When to consider vasculitis?

Frequent symptoms:
- Diarrhea
- GI bleeding
- Pain
- Perforation

Endoscopic findings:
- Unexplained signs of ischemia

Problems:
- No specific signs & symptoms for vasculitis
- GI biopsies are only diagnostic in 5% of cases



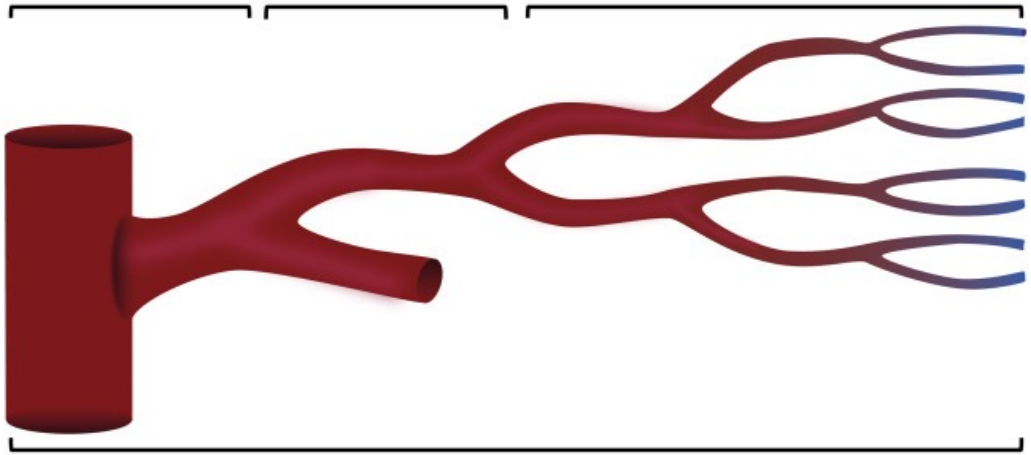


Classification of vasculitis

Large vessel vasculitis:
- Takayasu´s arteritis
- Giant cell arteritis

Medium vessel vasculitis:
- Kawasaki arteritis
- Polyarteritis nodosa

Small vessel vasculitis:
- *ANCA-associated vasculitis:*
  - Microscopic polyangiitis
  - Granulomatosis with polyangiitis
  - Eosinophilic granulomatosis with polyangiitis
- *Immune complex vasculitis:*
  - Anti–glomerular basement membrane
  - Cryoglobulinemic vasculitis
  - IgA vasculitis
  - Hypocomplementemic urticarial vasculitis



Variable vessel vasculitis:
- Behçet disease
- Cogan´s syndrome

We will **not** discuss management of individual vasculitis

Aim: Overview for gastroenterologists

2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides

INSELGRUPPE

Large vessel vasculitis

- Takayasu arteritis
- Giant cell arteritis

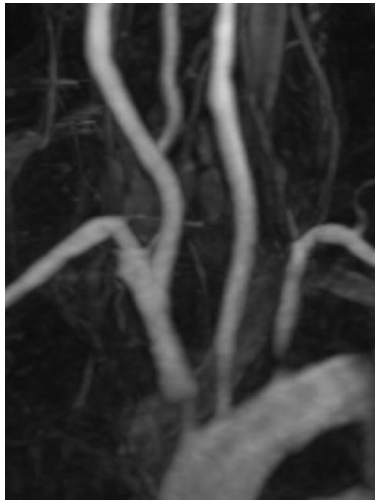
Takayasu arteritis

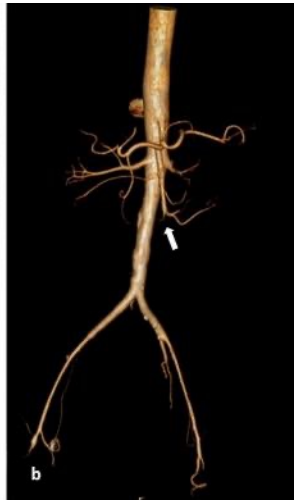
- “Pulseless disease”
- Typically ♀, 20–40yo
- Prevalence: Japan, SE Asia, India, Mexico ↑
  North America / Europe ↓

GI involvement:
- Rare
- Ischemia in SI, colon, liver and spleen
- Possible association with IBD

Management:
- Prednisone. Immunosuppressants
- Vascular surgery or endovascular treatments

Mikito Takayasu





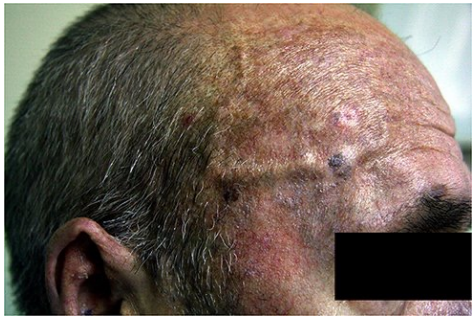
Radiopaedia.com
doi:10.14740/jmc2945w

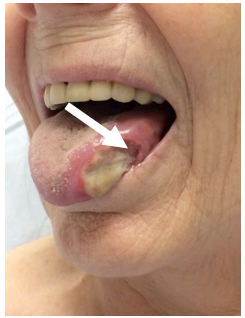
Giant cell arteritis

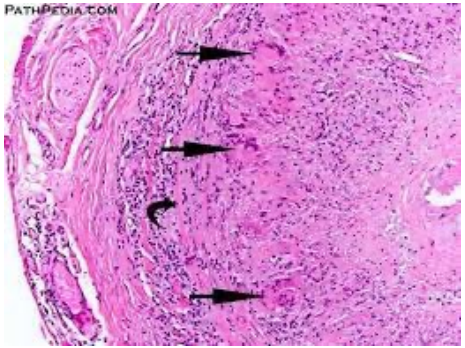
- Typically elderly patients (70–80y), never <50y
- Prevalence: West > East

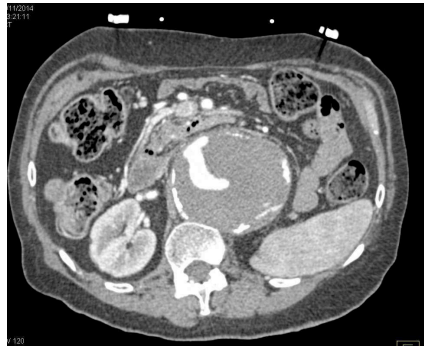
GI involvement:
- Rare
- SI/colon, liver
- Lab abnormalities (AP, transaminases) frequent

Management:
- Prednisone. Immunosuppressants
- Always screen for aortic aneurysm









doi:10.2147/CCID.S284795
Pathpedia.com

INSELGRUPPE

Medium vessel vasculitis

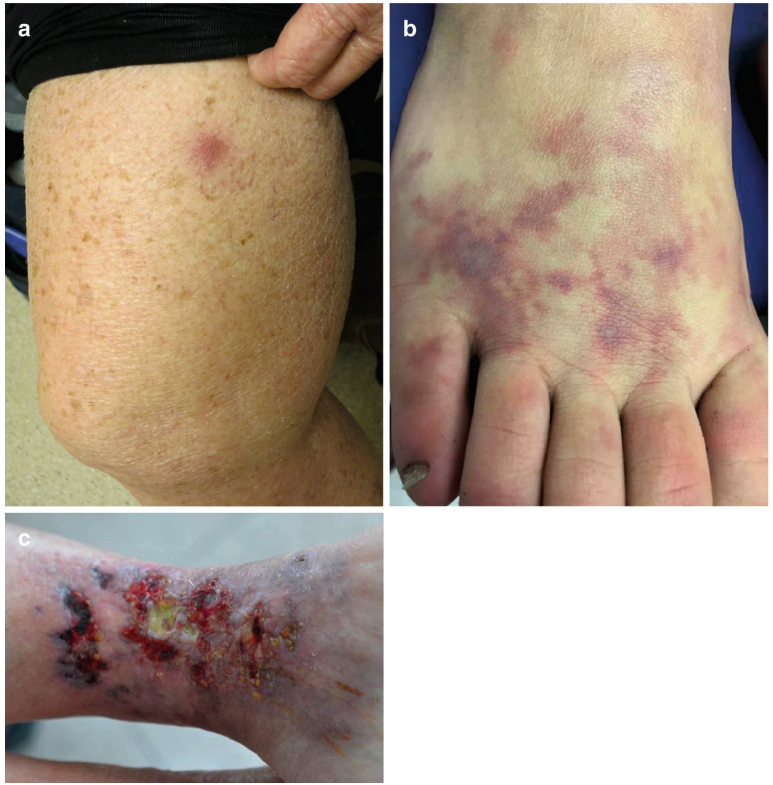
- Polyarteritis nodosa
- Kawasaki disease

Polyarteriitis nodosa (PAN)

- Typically ♂ in their 50s
- Strong association with HBV or HCV

GI involvement:
- Abdominal angina
- Small-bowel ulcers and bleeding (ischemia)
- Liver infarcts, Budd-Chiari syndrome

Management:
- Prednisone, immunosuppressants
- HBV/HCV treatment



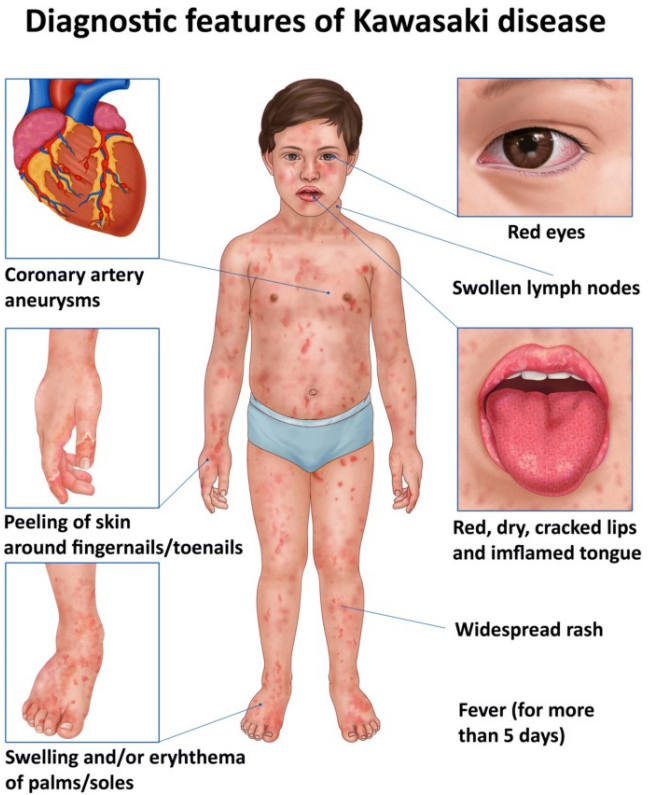
doi:10.1007/978-3-319-18449-4_51

Kawasaki disease

- Typically young children, rare in adults
- East >> West
- Acute onset, always fever

GI:
- Small-bowel ischemia
- Gallbladder hydrops (GB wall vasculitis)

Management:
- Aspirin & IVIG



INSELGRUPPE

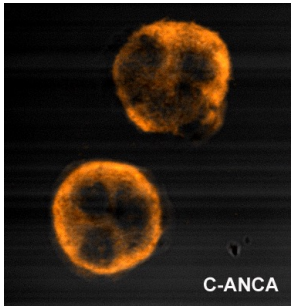
Small vessel vasculitis

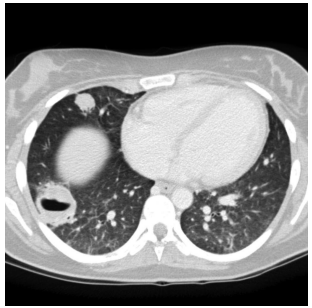
- **ANCA-associated**
  - Granulomatosis with polyangiitis (GPA)
  - Microscopic polyangiitis (MPA)
  - Eosinophilic granulomatosis with polyangiitis (EGPA)

- **Immune complex-associated**
  - Cryoglobulinaemic vasculitis

Granulomatosis with polyangiitis (GPA)

- Peak: 55–65 years
- Organs: upper & lower respiratory tract, kidney
- Serology: c-ANCA

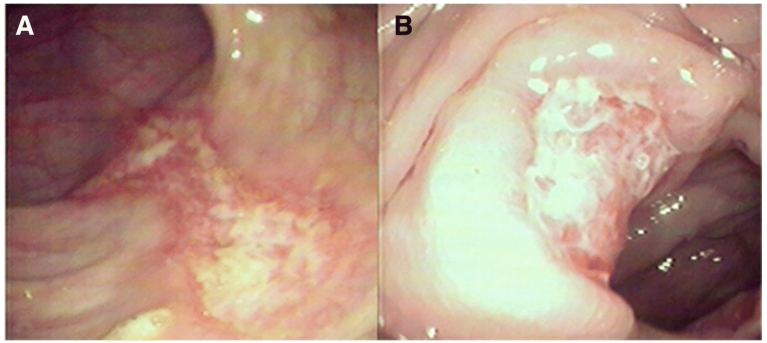




GI (5–10%):

- Mostly SI and colon with granulomatous ulcers (mimics **Crohn's**)

- Rare: granulomatous cholecystitis, granulomatous pancreatic mass, liver granulomas (mimics **malignancy**)

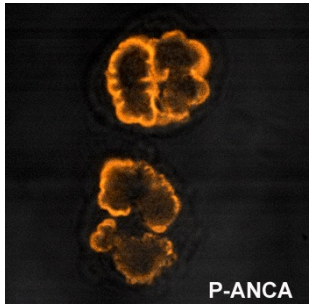


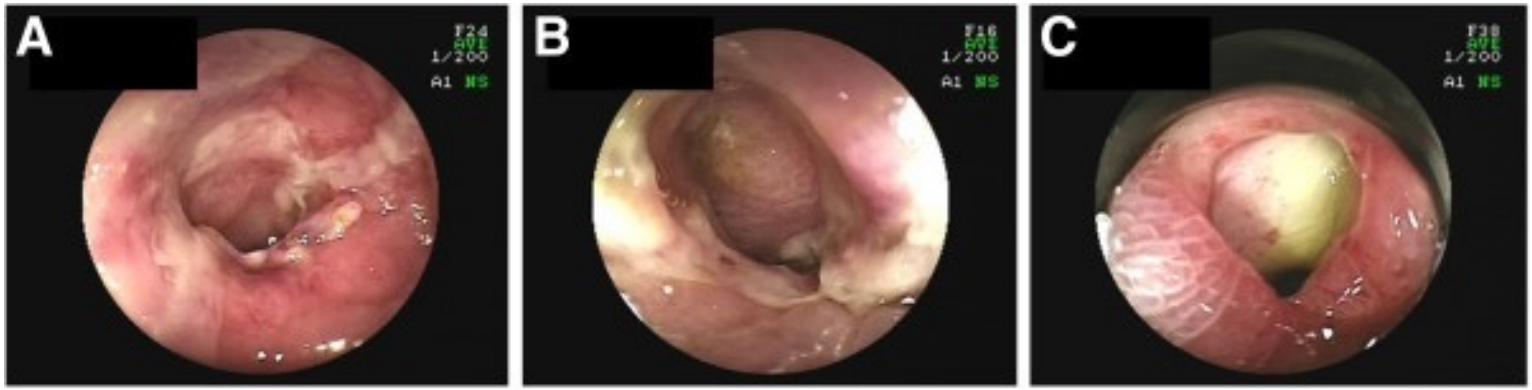
doi:10.1186/s12876-018-0885-9

Microscopic polyangiitis (MPA)

Almost the same signs & symptoms as GPA, but:

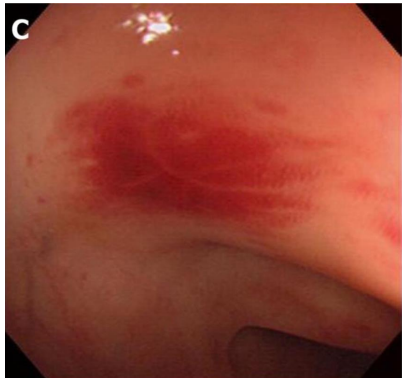
- Upper respiratory tract affection **rare**
- Serology: p-ANCA
- No granuloma
- GI tract more commonly affected





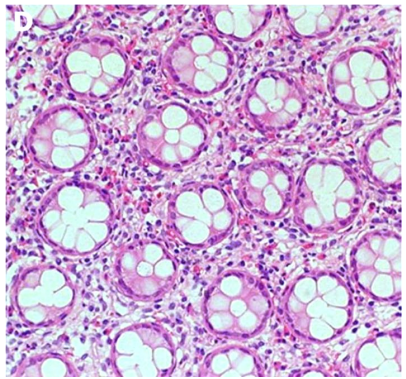
Eosinophilic granulomatosis with polyangiitis (EGPA)

- Often late-onset asthma and eosinophilia
- Skin purpura, cardiac affection
- ANCA only in 30–40%



GI:

- More commonly affected than in GPA and MPA
- Mesenteric artery affection (pain, diarrhea, ischemia)
- Eosinophilic infiltration (motility disorders, bleeding).

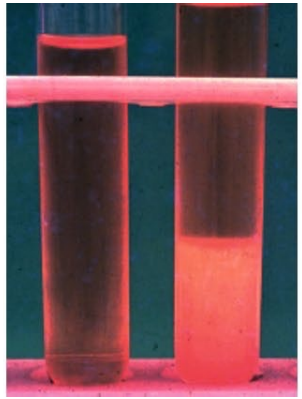


In EGPA, GI biopsies may be diagnostic

doi:10.4253/wjge.v4.i3.50

Cryoglobulinaemic vasculitis

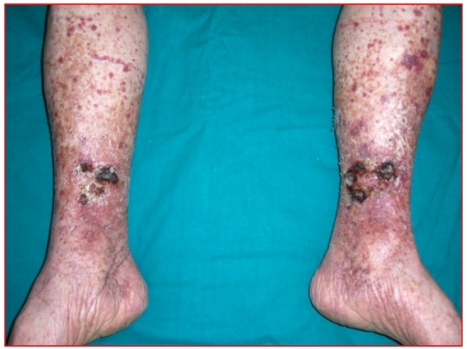
- **Cryoglobulins:** Ig's that precipitate below 37°C, difficult to detect
- **Labs:** Low C4, normal C3, positive rheuma factor
- Mixed cryoglobulinemia very common in HCV- (HBV/HIV-) infected patients (← exam question!)



GI involvement:

- Rare, if affected often severe bleeding or perforation

Treatment: underlying infection, steroids or rituximab in non-viral cases



doi:10.1155/2012/502156

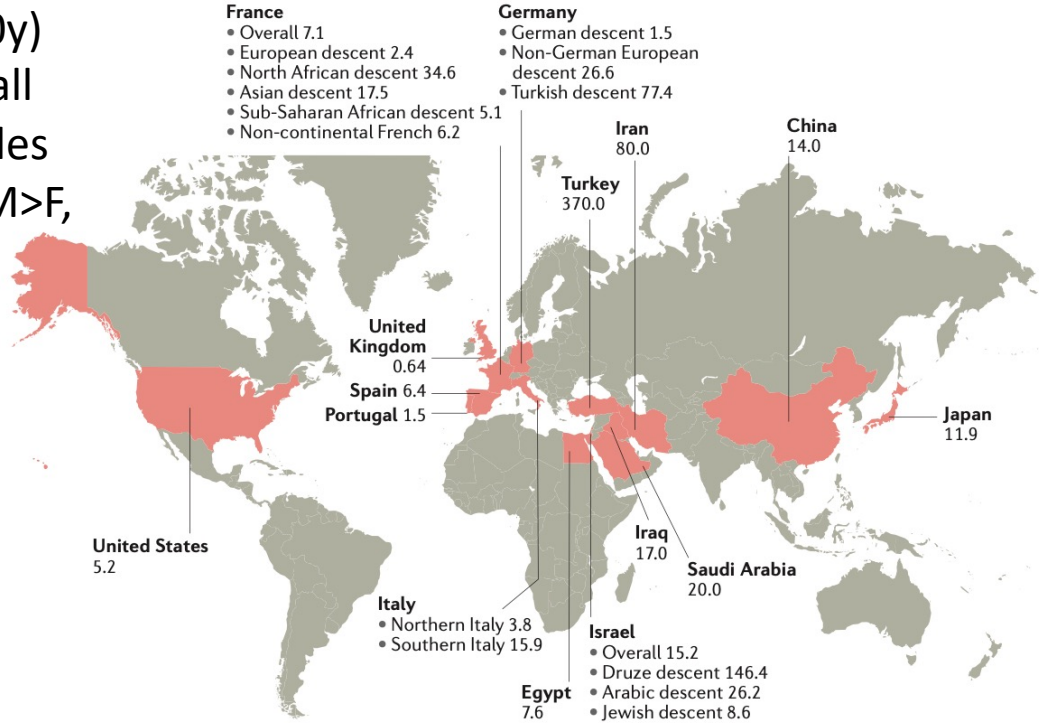
INSELGRUPPE

Variable vessel vasculitis

- Behçet’s disease

Behçet's disease

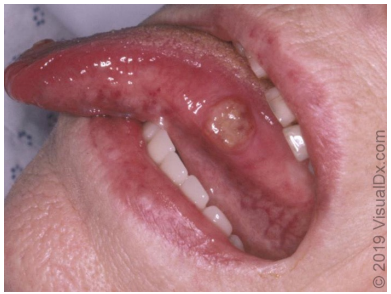
- Typically young adults (20–40y)
- Can involve blood vessels of all sizes, most affected are venules
- In Mediterranean countries M>F, in the United States F>M
- Association with HLA-B51

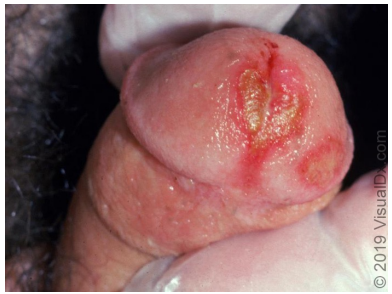


10.1038/nrrheum.2017.208

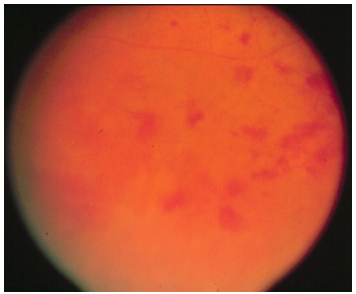
Behçet's disease – Signs and symptoms

Ulcerations

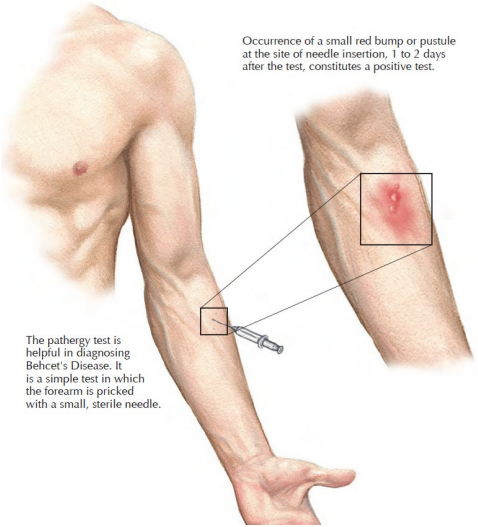




Ocular disease



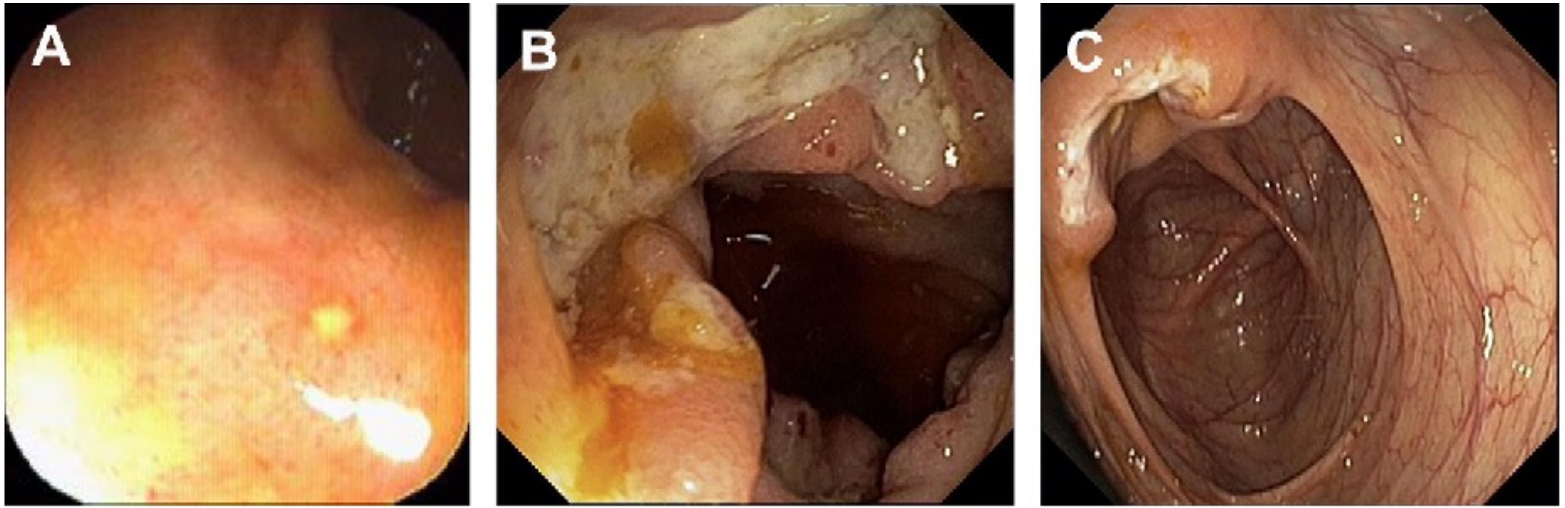
Positive pathergy test



Plus:

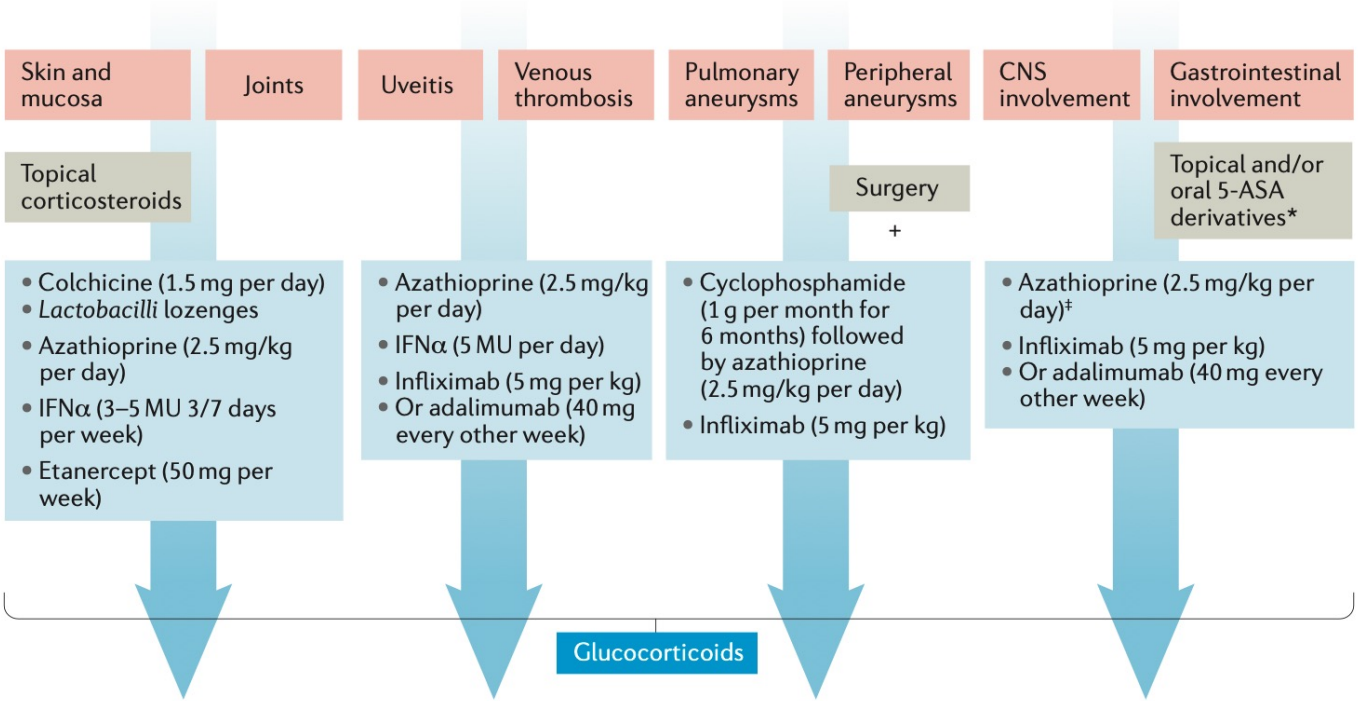
- Skin lesions (acne-like, erythema nodosum)
- Neurologic disease
- Arthritis

Behçet’s disease – GI involvement



- Mimics Crohn’s (location, ulceration)
- Can also cause ischemia / infarction

Behçet's disease – Management



10.1038/nrrheum.2017.208

Summary

- GI vasculitis is rare, diagnosis is often difficult
- Think of it in patients with signs of ischemia that are unexplained
- GI vasculitis can look like IBD (esp. Behçet's)
- Biopsies mostly non-diagnostic

doi:10.1016/j.gie.2010.02.033

Multiple choice questions

MCQ 1

A 64-year-old Caucasian male is admitted to the hospital with fatigue, cough, hemoptysis, difficulties with nasal breathing and abdominal pain. At admission, his vital signs are as follows: temperature of 38.1° C, HR 114 bpm, BP of 145/95 mm Hg, and RR of 22 breaths/min. Physical examination reveals moderate puffiness of the face and dullness on percussion and reduced breathing over the left lung. Chest x-ray examination reveals several cavitating opacities in the left lung. Urinalysis detects hematuria and mild proteinuria. Which of the following laboratory tests is most likely to be positive in this patient?

A. Anti-HIV antibodies
B. C-ANCA
C. Blood and urine Histoplasma antigen
D. Anti-glomerular basement membrane antibodies

MCQ 1

A 64-year-old Caucasian male is admitted to the hospital with fatigue, cough, hemoptysis, difficulties with nasal breathing and **abdominal pain**. At admission, his vital signs are as follows: temperature of **38.1° C**, HR 114 bpm, BP of 145/95 mm Hg, and RR of 22 breaths/min. Physical examination reveals moderate puffiness of the face and dullness on percussion and reduced breathing over the left lung. Chest x-ray examination reveals several **cavitating opacities** in the left lung. Urinalysis detects **hematuria and mild proteinuria**. Which of the following laboratory tests is most likely to be positive in this patient?

A. Anti-HIV antibodies
B. C-ANCA → Suspected GPA
C. Blood and urine Histoplasma antigen
D. Anti-glomerular basement membrane antibodies

MCQ 2

A 56-year-old Hispanic male presents to the outpatient clinic with reddish-blue lesions on his lower extremities, fever, muscle pain, and weight loss. He reports a history of acute viral hepatitis B three months ago. Physical examination reveals multiple red-purple nodules on the skin of both legs. Laboratory results for P- and C-ANCA are negative. Urinalysis reveals hematuria and proteinuria. Abdominal ultrasound reveals few 1 – 2 cm fluid-filled cavities in both kidneys. Which of the following is the most likely diagnosis?

A. Polyarteritis nodosa
B. Microscopic polyangiitis
C. Eosinophilic granulomatosis with polyangiitis (EGPA)
D. Giant cell arteritis

MCQ 2

A 56-year-old Hispanic male presents to the outpatient clinic with reddish-blue lesions on his lower extremities, fever, muscle pain, and weight loss. He reports a history of **acute viral hepatitis B** three months ago. Physical examination reveals multiple **red-purple nodules on the skin** of both legs. Laboratory results for **P- and C-ANCA are negative**. Urinalysis reveals hematuria and proteinuria. Abdominal ultrasound reveals few 1 – 2 cm fluid-filled cavities in both kidneys. Which of the following is the most likely diagnosis?

A. Polyarteritis nodosa
B. Microscopic polyangiitis
C. Eosinophilic granulomatosis with polyangiitis (EGPA)
D. Giant cell arteritis

MCQ 3

Behcet’s syndrome (BS) is characterized by recurrent _____

A. Oral ulcers
B. Genital ulcers
C. Cutaneous lesions
D. Ocular lesions
E. All of the above

MCQ 3

Behcet’s syndrome (BS) is characterized by recurrent _____

A. Oral ulcers
B. Genital ulcers
C. Cutaneous lesions
D. Ocular lesions
E. All of the above

MCQ 4

The differential diagnosis for Behcet's disease does **not** include which of the following?

A. Herpes simplex
B. Inflammatory bowel disease
C. Celiac disease
D. Psoriasis

MCQ 4

The differential diagnosis for Behcet's disease does **not** include which of the following?

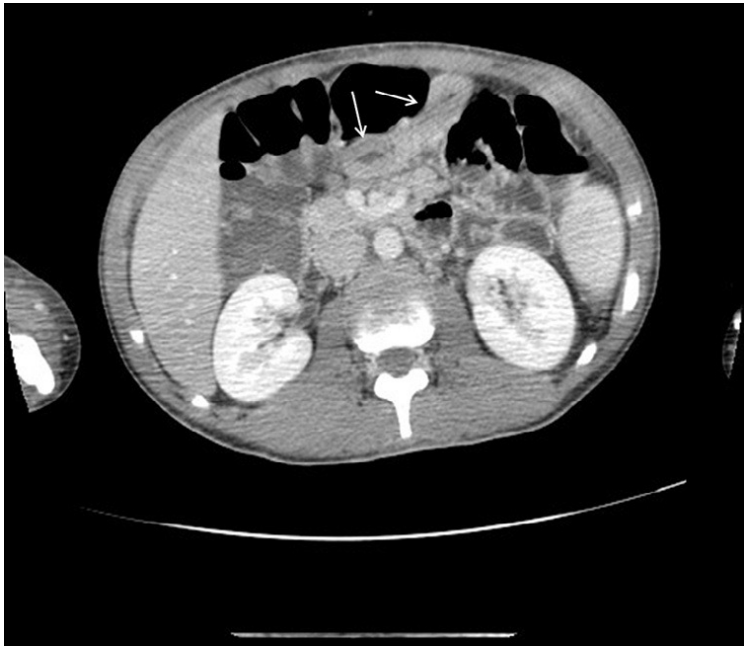
A. Herpes simplex
B. Inflammatory bowel disease
C. Celiac disease
D. Psoriasis

MCQ 5

A ten-year-old boy presents with fever, conjunctivitis and abdominal pain. Physical examination is significant for oral erythema along with a generalized maculopapular rash and cervical lymphadenopathy. A CT scan is performed:

What is the most likely diagnosis:

A. Eosinophilic granulomatosis with polyangiitis
B. Polyarteritis nodosa
C. Kawasaki disease
D. Takayasu's arteritis

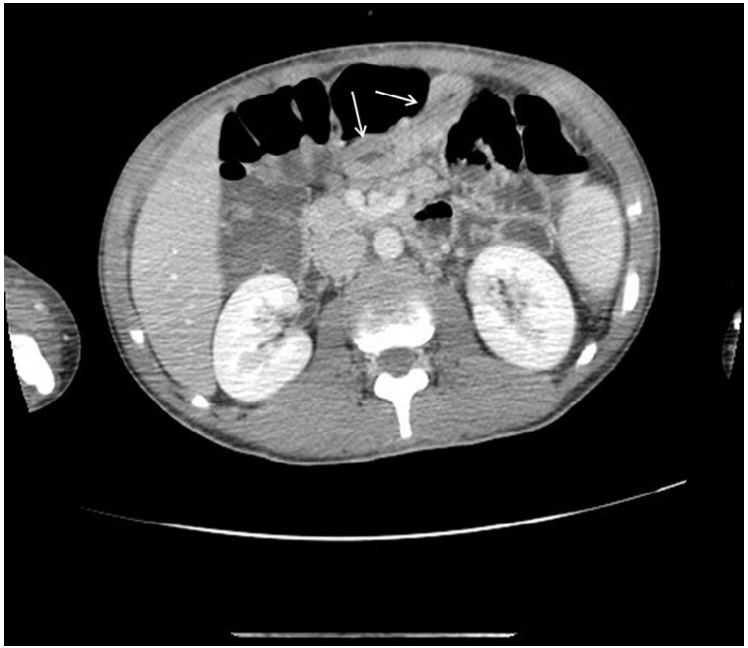


MCQ 5

A **ten-year-old** boy presents with **fever**, conjunctivitis and abdominal pain. Physical examination is significant for oral erythema along with a generalized maculopapular **rash** and cervical lymphadenopathy. A CT scan is performed:

What is the most likely diagnosis:

A. Eosinophilic granulomatosis with polyangiitis
B. Polyarteritis nodosa
C. Kawasaki disease
D. Takayasu's arteritis



MCQ 6

A 48-year-old man visiting from Japan presents to your office with several months of diarrhea and right lower quadrant abdominal pain. On further questioning, he reports a history of cluster of oral ulcers, arthritis and uveitis. Physical exam is remarkable for mild right lower quadrant tenderness and multiple erythematous nodules on his shins. Ulcerations are also found on the glans penis. A colonoscopy is performed, which shows multiple deep ulcers in the terminal ileum and cecum.
Which of the following is true about this disease?

A. GI involvement is uncommon
B. Recurrences rarely occur after surgery
C. It primarily affects small veins and venules, leading to ulcers
D. It frequently causes bowel strictures and perianal disease
E. The vasculitis mainly affects the gastric region

MCQ 6

A 48-year-old man visiting from Japan presents to your office with several months of diarrhea and right lower quadrant abdominal pain. On further questioning, he reports a history of cluster of **oral ulcers, arthritis and uveitis**. Physical exam is remarkable for mild right lower quadrant tenderness and multiple **erythematous nodules on his shins**. Ulcerations are also found on the **glans penis**. A colonoscopy is performed, which shows multiple **deep ulcers in the terminal ileum and cecum**.
Which of the following is true about this disease?

A. GI involvement is uncommon
B. Recurrences rarely occur after surgery
C. It primarily affects small veins and venules, leading to ulcers
D. It frequently causes bowel strictures and perianal disease
E. The vasculitis mainly affects the gastric region