
VASCULITIS AND INFECTION

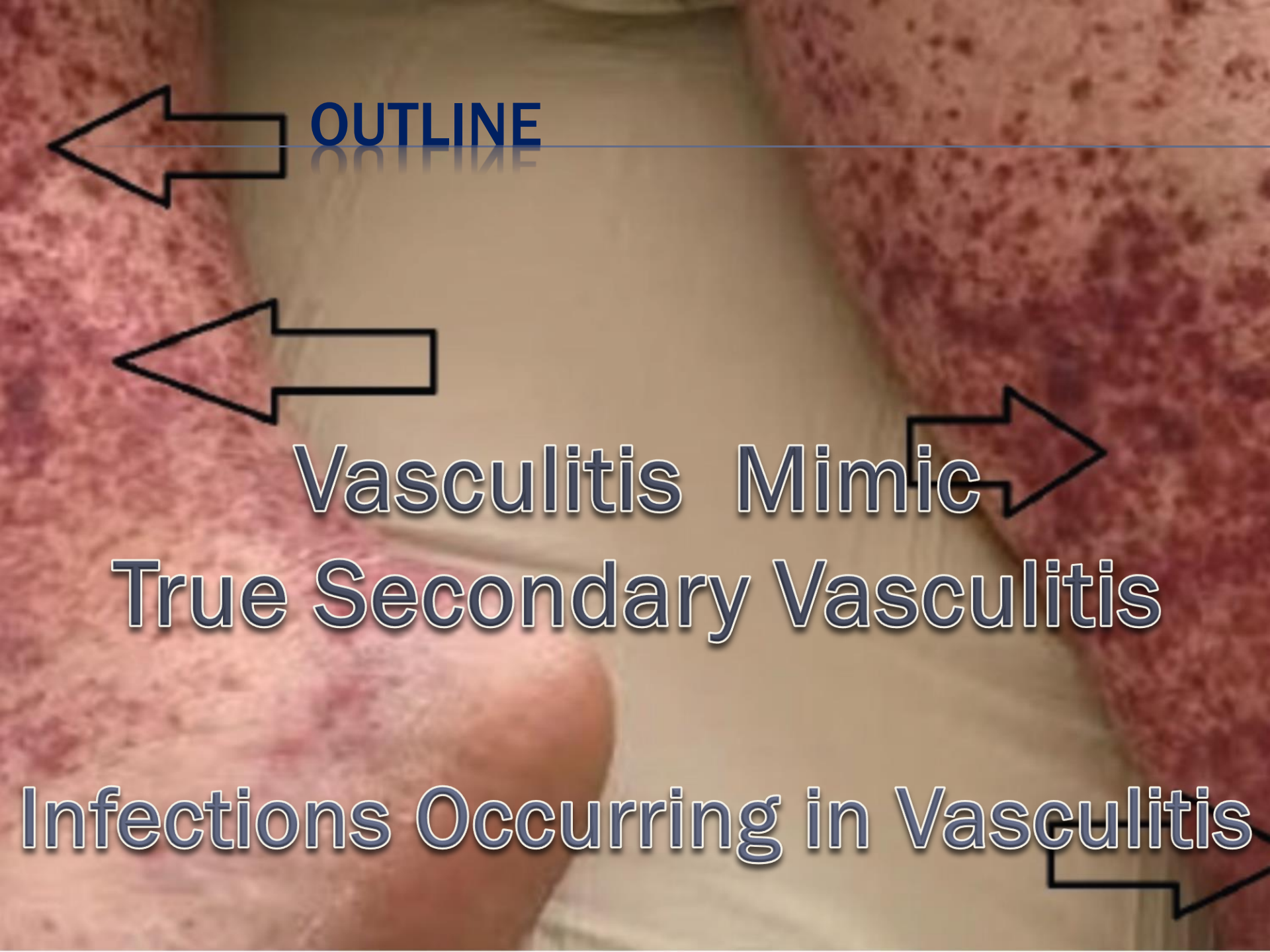
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OUTLINE

Vasculitis Mimic

True Secondary Vasculitis

Infections Occurring in Vasculitis

TRUE SECONDARY VASCULITIS OR VASCULITIS MIMIC PER SE

- ✗ Acute bacterial or viral infections or chronic infections with bacteria, viruses, mycobacteria, fungi or parasites may mimic vasculitis.
- ✗ A critical exclusion of an infectious origin in the evaluation of a patient with suspected primary systemic vasculitis should be done.
- ✗ Treatment strategies differ from those applied to non-infectious forms.

Syphilitic aortitis: diagnosis and treatment

Case report

Aortite sífilítica: diagnóstico e tratamento. Relato de caso

Roberto Santos SARAIVA¹, Claudio Albernaz CÉSAR², Marco Antonio Araújo de MELO



Scand J Rheumatol. 2006 Mar-Apr;35(2):147-51.

A large ulcer and cutaneous small-vessel vasculitis associated with syphilis infection.

Chao YC¹, Chen CH, Chen YK, Chou CT.

SYPHILIS

- ✗ Aortitis: The primary lesion of cardiovascular syphilis. In 70–80% of untreated cases after the primary infection,
- ✗ In 10% aortic aneurysm, aortic regurgitation and coronary ostia stenosis will occur.
- ✗ Cardiovascular syphilis is a late form of syphilis, in the 4th–5th decade of life, typically 5–40 years after the primary infection
- ✗ The ascending aorta most commonly affected (50%), the arch (35%) and the descending aorta (15%)
- ✗ In the presence of an aortic aneurysm, particularly in younger patients, syphilitic serological testing is advised
- ✗ The sensitivity of (VDRL) test is 73% and (FTA-ABS) test sensitivity is 96%.

Takayasu's or tuberculous arteritis?

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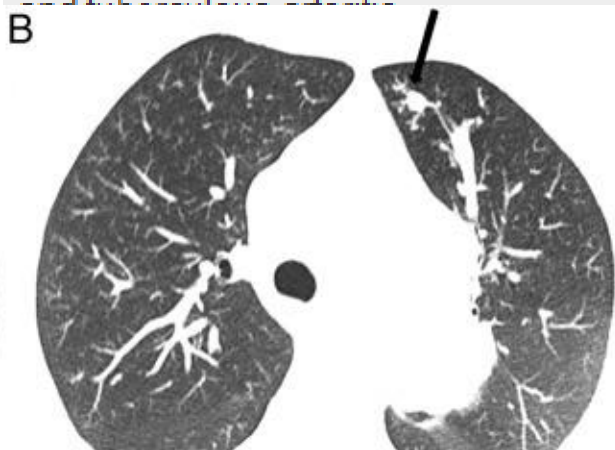
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Summary

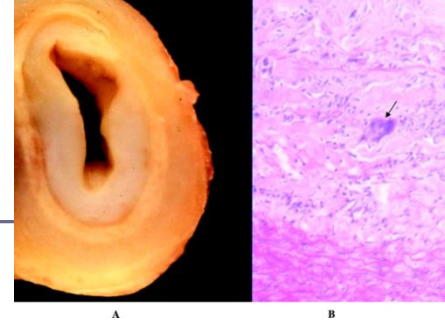
Takayasu's arteritis (TA) is a large vessel vasculitis of unknown aetiology characterised by involvement of the aorta and its major branches. Tuberculous arteritis of the aorta is an uncommon condition usually secondary to the dissemination of *Mycobacterium tuberculosis* infection from the mediastinum and/or lung to the adjacent aorta; this may mimic TA. We report a case of a 23-year-old woman with cutaneous granulomatous vasculitis and saccular aneurysmal dilation of the aorta and large vessels, and highlight the findings shared by TA



BMJ Case Reports 2015
Annals of pediatric cardiology 2016



TUBERCULOSIS (TB)



- ✗ TB can result in granulomatous arteritis leading to vessel wall thickening, aneurysm formation and stenoses that can affect the aorta and its branches.
- ✗ May occur as a result of direct seeding of the thoracic aorta from adjacent infected tissues such as infected lymph nodes or lung lesions or by miliary spread.
- ✗ Involvement of the descending aorta or renal artery may resemble Takayasu's arteritis (TAK)
- ✗ - TB : Erosion of the vessel wall with the formation of true or false aneurysms, particularly the descending thoracic and abdominal aorta
- TAK: Arterial stenoses
- ✗ Atypical mycobacteria have been reported to result in granulomatous and vascular pulmonary disease
- ✗ ANCA can be positive positive

www.medscape.org 2009

Esquivel et al, clin Exp rheumatol 2010



- ✕ The diagnosis may be suggested by the coexistence of extra or pulmonary TB

Tuberculosis should not be ignored in patients with peripheral gangrene

Yuan Yao, MM, Bin Liu, MD, Ji-bo Wang, MD, Hua Li, MM, and Hong Da Liang, MM, *Shandong Province, China*

Peripheral gangrene, characterized by distal ischemia of the extremities, is a rare complication in patients with tuberculosis (TB). We diagnosed a female patient with gangrene of her left toe caused by TB infection. She presented with fever, lymphadenectasis, and peripheral gangrene of the left toe. Lymph node biopsy confirmed tuberculous lymphadenitis and the computer tomography angiography showed vasculitis. The patient underwent antituberculous therapy and her condition was gradually improved. Although it is rare, TB should be considered as a possible cause of peripheral gangrene. (J Vasc Surg 2010;52:1662-4.)

INFECTIVE ENDOCARDITIS

SUBACUTE BACTERIAL ENDOCARDITIS WITH POSITIVE CYTOPLASMIC ANTINEUTROPHIL CYTOPLASMIC ANTIBODIES AND ANTI-PROTEINASE 3 ANTIBODIES

HYON K. CHOI, PETER LAMPRECHT, JOHN L. NILES,

[Indian J Nephrol.](#) 2012 Nov-Dec; 22(6): 469–472.

PMCID: PMC3573492

doi: [10.4103/0971-4065.106057](#)

Double ANCA-positive vasculitis in a patient with infective endocarditis

[I. Veerappan](#), [E. N. Prabitha](#), [A. Abraham](#),¹ [S. Theodore](#),² and [G. Abraham](#)

Leukocytoclastic Vasculitis with Infective Endocarditis Mimicking Henoch-Schönlein Purpura

Yuki Hashimoto* Kayo Enosawa Kanna Honmura

INFECTIVE ENDOCARDITIS (IE)

- ✗ As a cause of vasculitis is uncommon
- ✗ Of 172 adults with vasculitis, only 4 (2.3%)
- ✗ Mycotic (infected) aneurysms affecting the aorta, pulmonary artery, small vessel inflammation in various organ
 - Major branches of the aorta,
 - Femoral artery and abdominal aorta
 - thoraco–abdominal and thoracic aorta
- ✗ . Direct infection (septic vasculitis)
 - - Immune complex-mediated (purpura, [GN])
 - - Embolic mechanisms

Blanco R. Medicine 1998

www.medscape.org 2009

INFECTIVE ENDOCARDITIS (IE)

- ✗ The differentiation can be difficult sometimes especially if the heart murmur is absent
- ✗ Blood cultures are useful in identifying the responsible organism as well.
- ✗ GN observed in the setting of IE is typically associated with 'granular' immune complex deposits
- ✗ In patients with double ANCA positivity (PR3- and/ MPO-ANCA), SBE and HCV infection should be excluded

VASCULITIS-ASSOCIATED WITH PROBABLE CAUSE

- ✘ The cause of most vasculitides has not been fully elucidated.
- ✘ Infections have been suggested to contribute to the induction and reactivation of some forms.
- ✘ A causal relationship between infection and vasculitis has only been established in a few instances and many mechanisms remain hypothetical.

Mirjan M, Curr Opin Rheumatol 2014,

Cristina C, Infection and vasculitis, Rheumatology (Oxford) 2009

VASCULITIS-ASSOCIATED WITH PROBABLE CAUSE

Large-vessel vasculitis	
Takayasu arteritis	<i>Mycobacterium tuberculosis</i>
Giant cell arteritis	<i>Burkholderia</i>
Medium-vessel vasculitis	
Polyarteritis nodosa	Hepatitis B virus (HBV), hepatitis C virus (HCV), and HIV infection
Kawasaki disease	'New' to be identified virus; Bacteria such as <i>Staphylococcus aureus</i> and <i>Streptococcus pyogenes</i> that are capable of superantigen production
Small-vessel vasculitis	
Immune complex vasculitis	
Antiglomerular basement membrane disease	Increased incidence during influenza epidemics
Cryoglobulinemic vasculitis	Hepatitis C virus (HCV)
IgA vasculitis (Henoch-Schönlein purpura)	Many bacteria, viruses, and even protozoa
Hypocomplementemic urticarial vasculitis	Hardly any associations with infections
ANCA-associated vasculitis	
Microscopic polyangiitis	
Granulomatosis with polyangiitis (Wegener's granulomatosis)	<i>Staphylococcus aureus</i>
Eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome)	

- ✗ *Mirjin etal current opinion 2014*
- ✗ *Graiales etal Autoimmune disease 2015*

MAIN VIRUS ASSOCIATED VASCULITIS

- × HBVPAN
- × HCV.....Cryoglobulinemic Vasculitis, PAN
- × HIV.....PAN
 - Large, Medium and or Small size Vasculitis
 -Cerebral Vasculitis
- × VZV.....GCA, Retinitis, Meningoencephalomyelitis
- × CMV.....Retinitis, Colitis, PAN
- × HTLV1..... Necrotizing Retinitis, Cerebral Vasculitis

Vasculitides secondary to infections / C. Pagnoux et al.2006

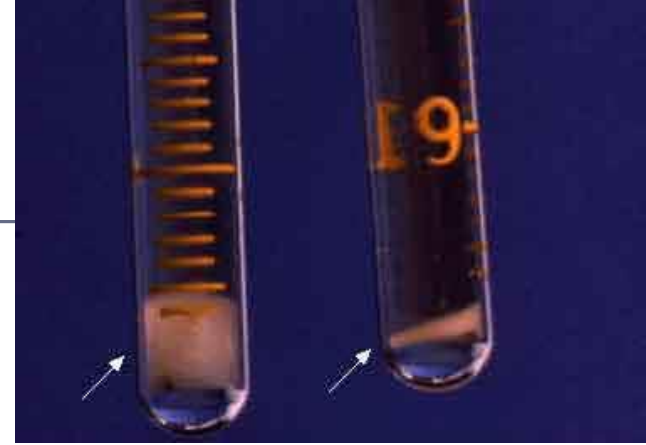
INFECTIOUS RELATED PAN

- ✗ HBV- PAN being the most severe and HIV-PAN being the mildest form.
- ✗ The occurrence of relapses, most frequent in HCV-related and least in HIV-related
- ✗ Duration of the viral infection prior to PAN:
 - For HCV.....20 y
 - For HBV.....<1 y
- ✗ Orchitis, Gastrointestinal and Renal artery involvement resulting in hypertension are common in HBV- PAN, whereas cutaneous and pulmonary involvement is rare.

INFECTIOUS PAN / NON INFECTIOUS PAN

- ✖ HBV-PAN compared with non-infectious PAN: a dramatic onset, a milder favorable clinical course and seroconversion followed by complete healing
- ✖ The association between PAN and HBV has been reported frequently (10–54%).
- ✖ Less than 10% of the cases of PAN are associated with HCV.

HCV -VASCULITIS



- ✗ Cryoglobulinemic vasculitis(CV)
 - 80% MC are + HCV
 - Mixed cryoglobulinemia (MC) in around 50% of HCV
 - CV in 5–10% of HCV
 - Low level of C4 and high titer RF

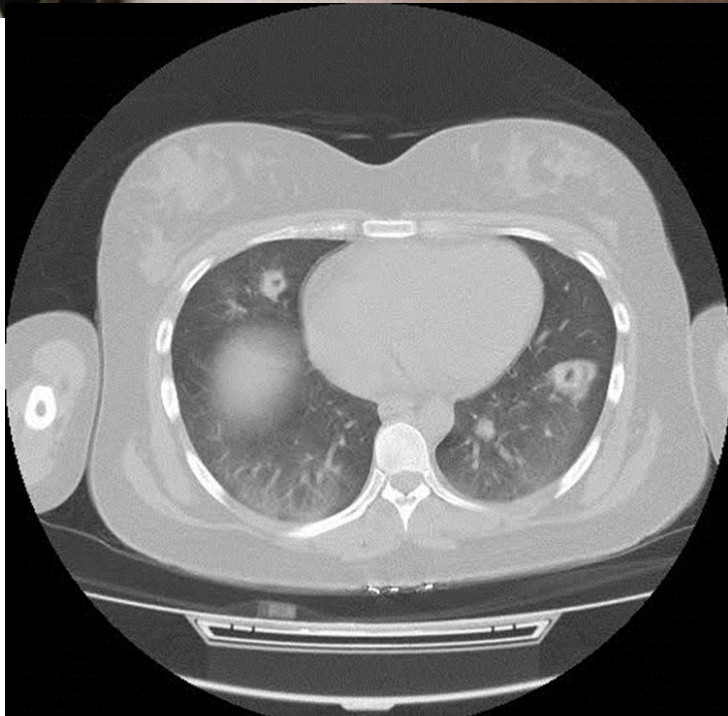
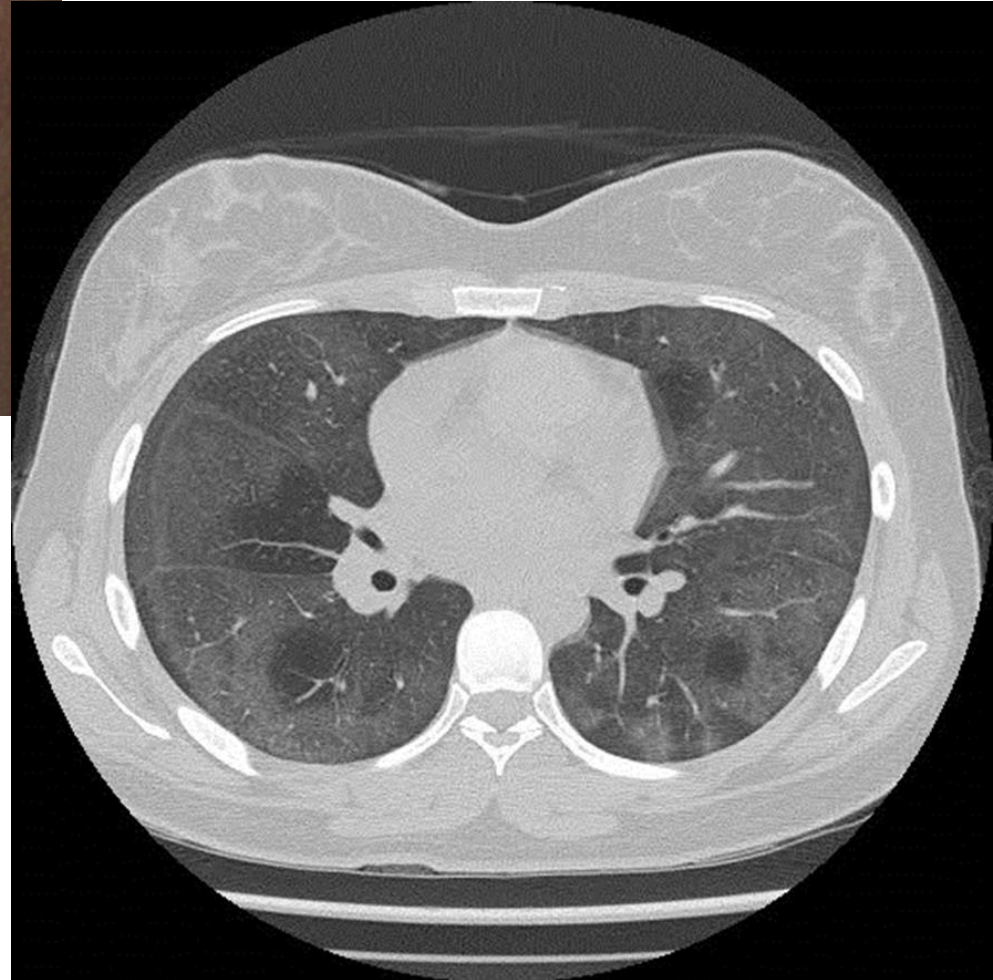
- ✗ HCV- PAN
 - More fever, weight loss, severe HTN, GI and CNS vasculitis, kidney and liver microaneurysms, and renal failure and increased ESR but good response to therapy
 - Saadoun et al, Arthritis Rheumatism 2006

INFECTIONS IN VASCULITIS

- ✗ Patients with vasculitis may develop infections, which sometimes mimic relapse.
- ,
- ✗ Patients, considered at higher risk for infections, should be followed closely and their immunological status monitored periodically.
- ✗ We recommend especially to monitor neutrophiles, lymphocytes and if needed CD3-, CD4- and CD8-cell counts in patients receiving steroids and cyclophosphamide or other cytotoxic agents.
- ✗ In patients treated with rituximab, CD19 and gammaglobulins should be monitored regularly.
- ✗ *Guillevin L, Infections in vasculitis. Best Pract Res Clin Rheumatol..2013*

A MISSED DIAGNOSIS

- ✖ A 21 y old woman known case of GPA based on arthralgia, proteinuria, high ESR and anti PR3 for 3y treated with azathioprine and steroid
- ✖ Presented with dyspnea, erythematous plaques and bullae in abdomen and thigh, anemia, high ESR, proteinuria and rising Cr
- ✖ She was doing well up to 2 months before admission who developed proteinuria, dyspnea, that Endoxan was administered.

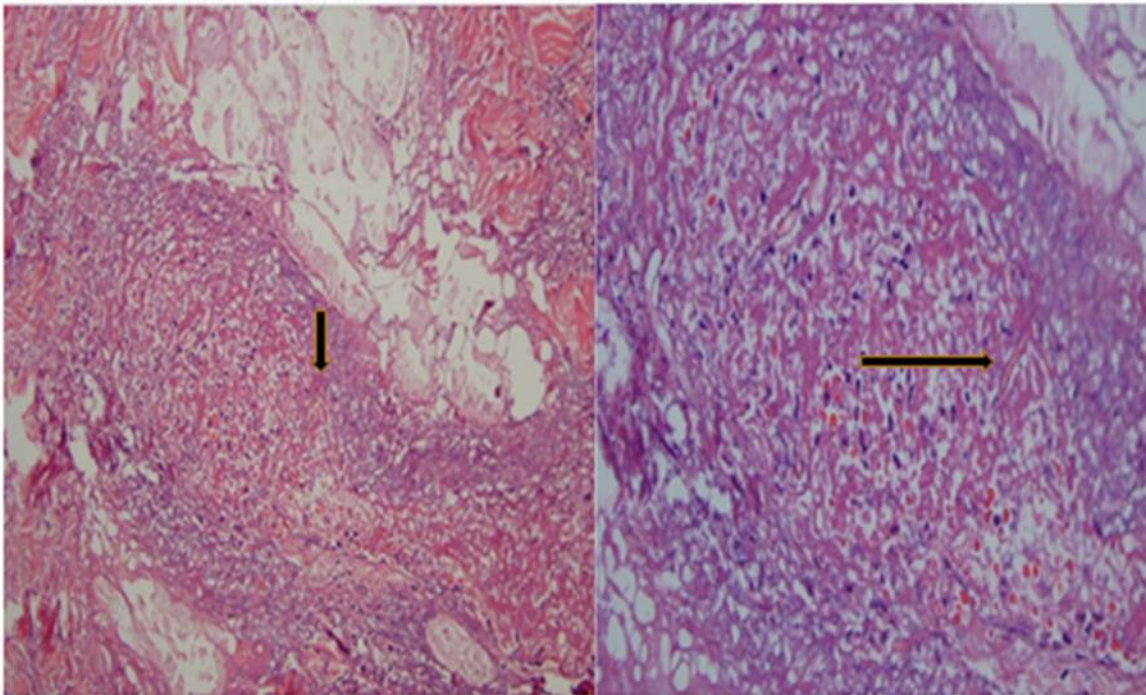


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- ✗ Pathology reported severe leukocytoclastic vasculitis
 - ✗ Antibiotic and then steroid pulse were administered with the impression of GPA flare and probable sepsis
 - ✗ BAL was planned
 - ✗ The condition deteriorated.



POST MORTEM SKIN PATHOLOGY REVIEW

× CUTANEOUS MUCORMYCOSIS



SUMMARY

Not Recognizing an infectious origin of vasculitides is of great importance because of different treatment strategies.



Thank You