# Clinicopathological Conference
# A woman with chronic peritonitis

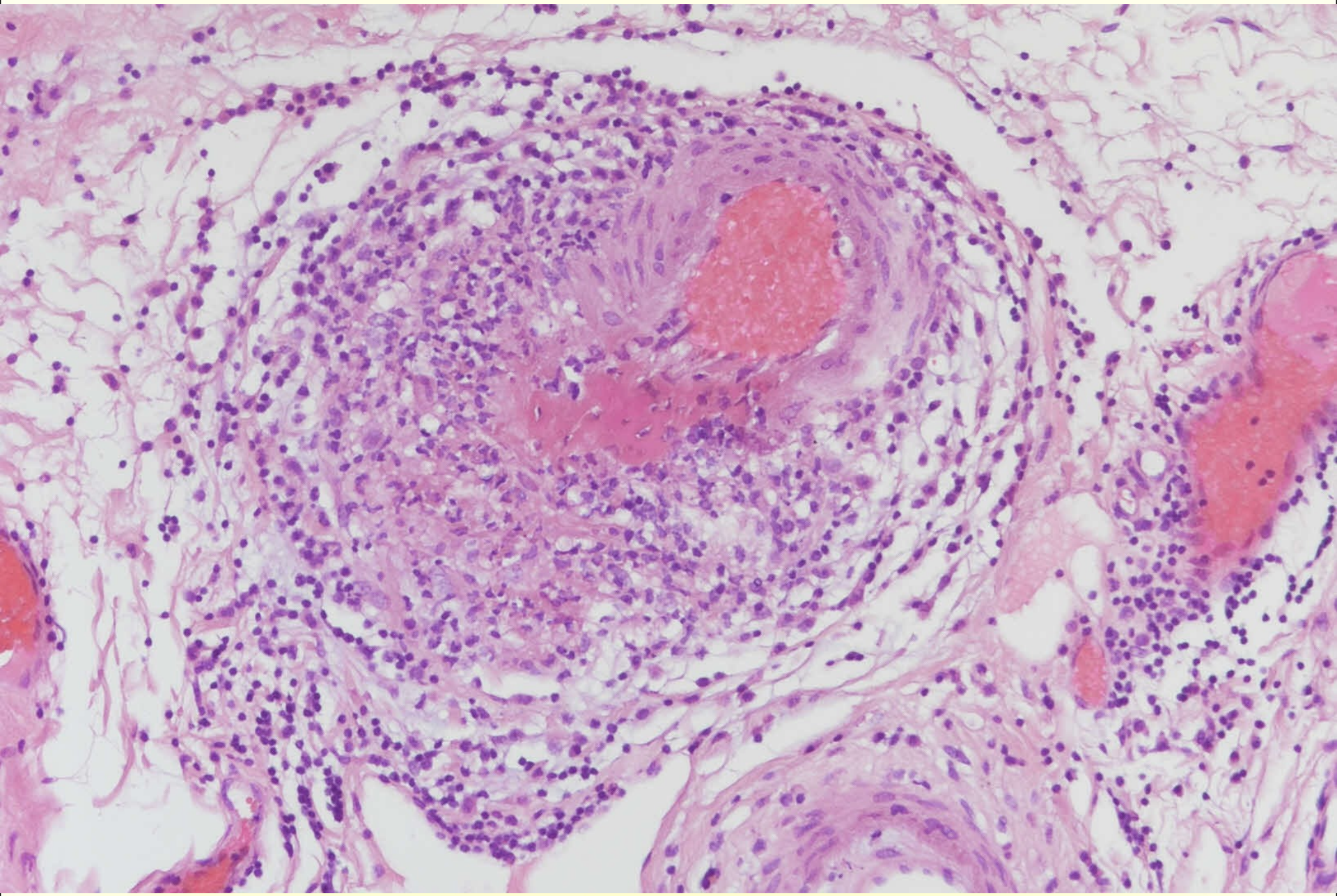
20160602

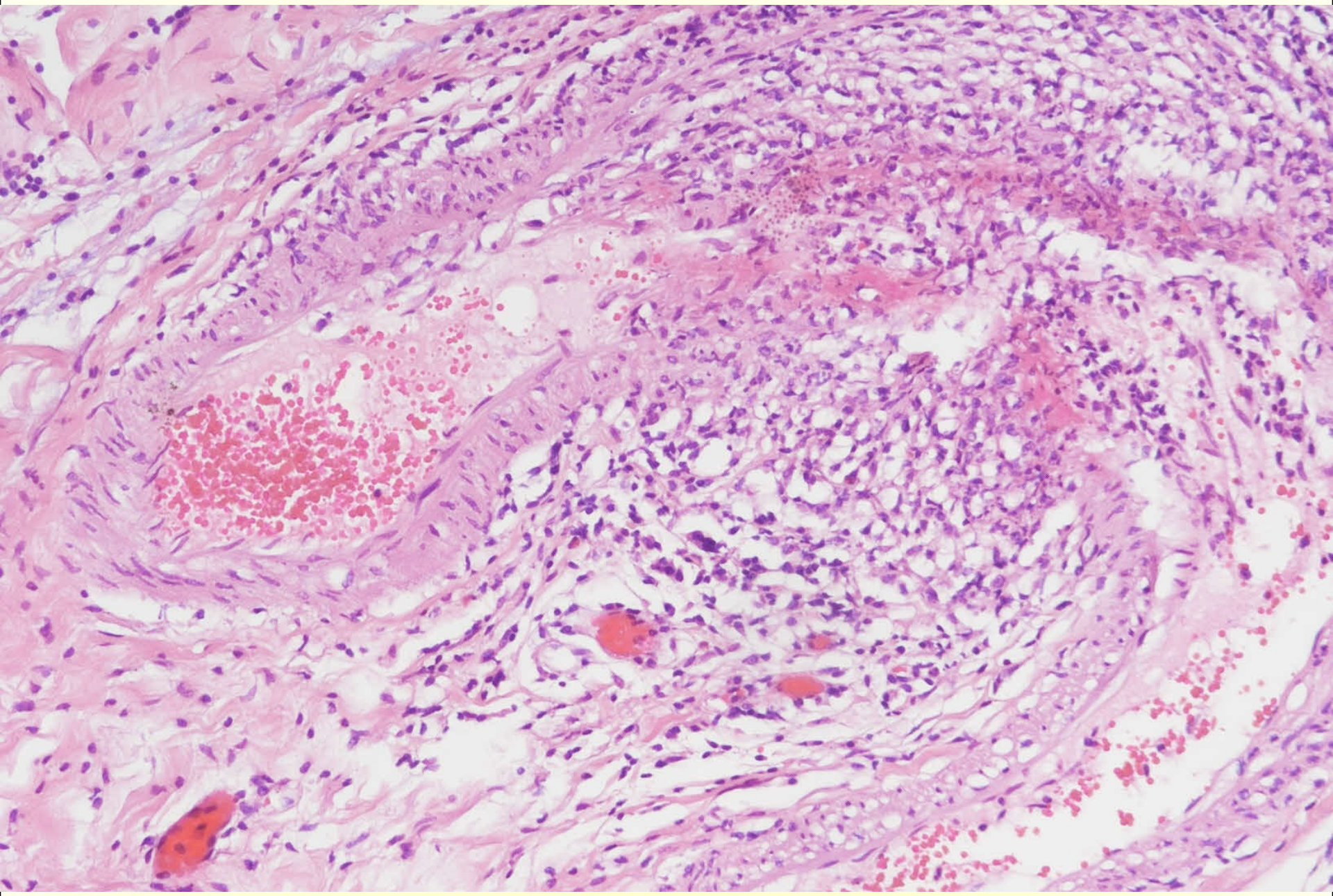
病理檢驗部

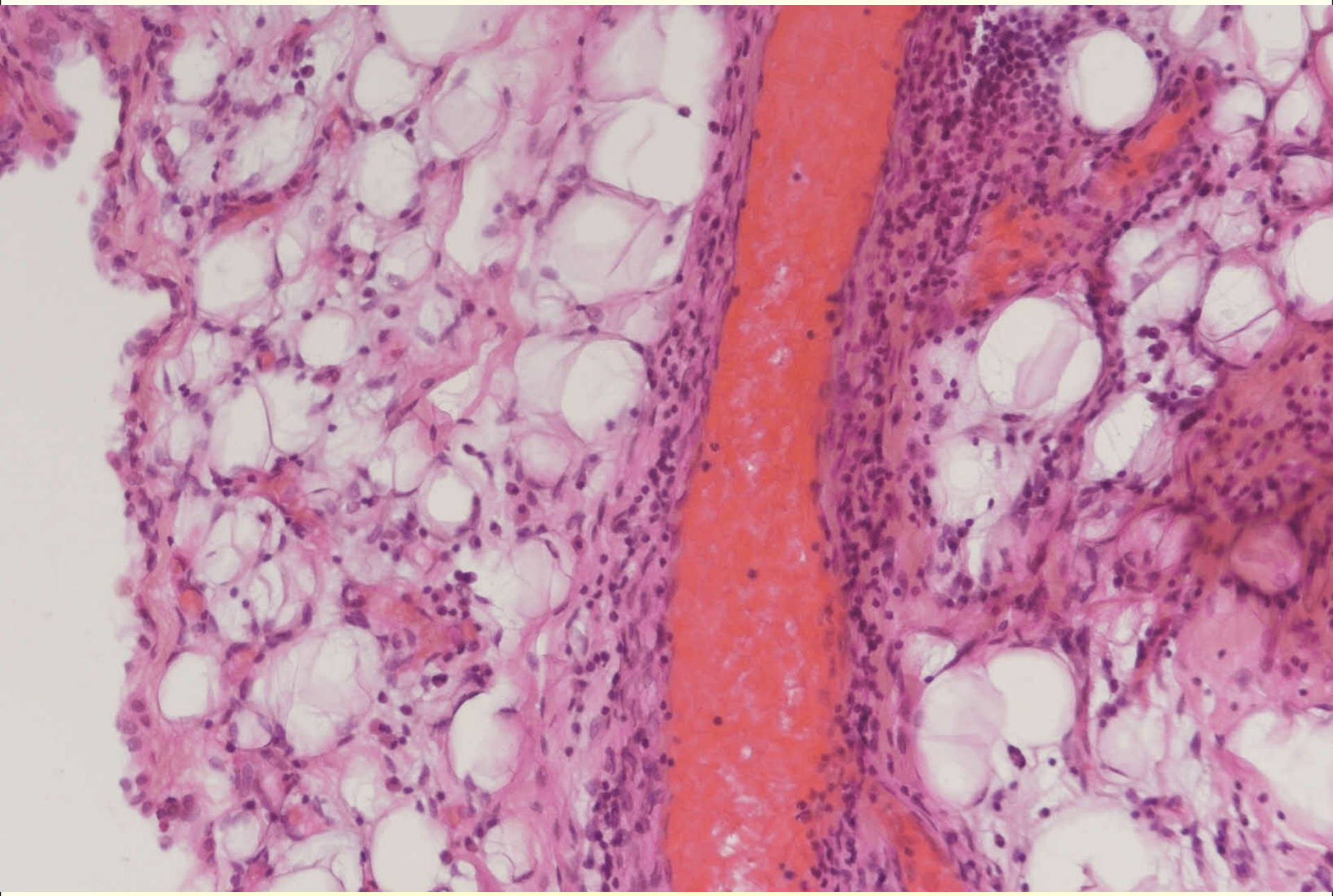
李懷寶， 王志生

# bilateral partial ovarian and fallopian tube resection + omentum biopsy

Microscopic examination







# Summary of microscopic findings

- Small blood vessel vasculitis in the mesosalpinx soft tissue
- Infiltration of blood vessel walls by neutrophils and nuclear dusts
- Fibrinoid deposition
- Thrombosis

- Omentum: hemorrhage, congestion

# Leukocytoclastic vasculitis

- The most common form of vasculitis
- Not a disease entity
- Vascular reaction pattern due to circulating immune complexes
- Idiopathic or caused by underlying disorders.

# Pathogenesis-based differential diagnoses of vasculitis

- Infection
- Immunologic
- Unknown

# Pathogenesis of vasculitis

- Infection
  - Bacterial
  - Rickettisial
  - Spirochetal
  - Fungal
  - Viral
- Immunologic
- Unknown

# Pathogenesis of vasculitis

- Infection
- Immunologic
  - Immune complex-mediated
  - ANCA-medicated (Wegener granulomatosis, Churg-Strauss syndrome, microscopic polyangiitis)
  - Direct Ab mediated (Goodpasture syndrome, Kawasaki disease, allograft rejection)
  - Cell mediated (allograft rejection)
  - Inflammatory bowel disease
  - Paraneoplastic vasculitis
- Unknown

# Pathogenesis of vasculitis

- Infection
- Immunologic
- Unknown
  - Giant cell arteritis
  - Takayasu arteritis
  - Polyarteritis nodosa

# Pathogenesis of vasculitis

- Infection
- Immunologic
  - Immune complex-mediated (leukocytoclastic vasculitis)
  - ANCA-medicated (microscopic polyangiitis)
- Unknown

# Histochemistry

Neither mycobacteria nor fungi can be identified in the acid fast and PAS stains.

# Leukocytoclastic vasculitis

**TABLE 16.2** Possible causes of allergic **vasculitis**

- Infection
  - bacterial: *Streptococcus*
  - mycobacterial: Mycobacterium tuberculosis
  - viral: hepatitis, influenza
  - cytomegalovirus
  - HIV infection
  - leprosy
- Drugs
  - aspirin, phenacetin, sulfonamides, penicillin, iodides, phenothiazines
- Chemicals
  - insecticides, weed killers, petroleum products
- Foreign proteins
  - serum sickness
  - hyposensitization antigens
- Associated diseases
  - autoimmune diseases: systemic lupus erythematosus, inflammatory bowel disease
  - hemolytic anemia
  - Hodgkin's lymphoma, carcinoma
  - rheumatoid arthritis
  - mixed connective tissue disease
  - dermatomyositis
  - relapsing polychondritis
  - Sjögren's syndrome
  - Henoch-Schönlein purpura
  - cryoglobulinemia
  - polyarteritis nodosa
  - Wegener's granulomatosis
  - Churg-Strauss disease
  - granuloma faciale
  - erythema elevatum diutinum
  - Waldenström's hypergammaglobulinemia
  - sarcoidosis

# Discussion

## Vasculitis in Sjögren's syndrome (SS)

- Vasculitis develops in about 10% of patients with primary SS, and affects the skin in half of them
- Vasculitis usually occurs several years after diagnosis of SS

http://www.dermnetnz.org/immune/sjogren.html

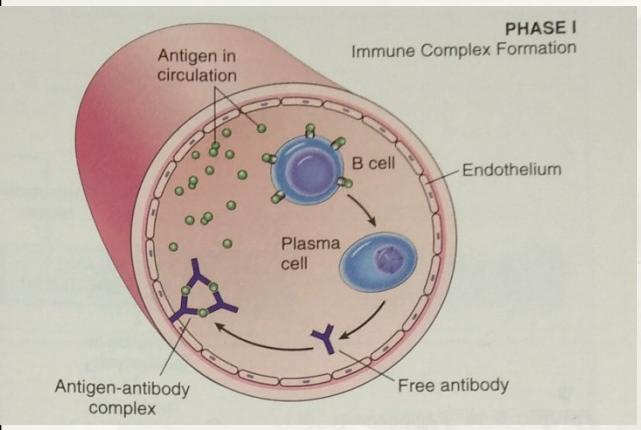
- SS pts affected by vasculitis are mainly those with more widespread extraglandular diseases: Raynaud phenomenon, enlarged LNs/ liver/spleen; and those with kidney and lung involvement;
- commonly detected serum cryoglobulins

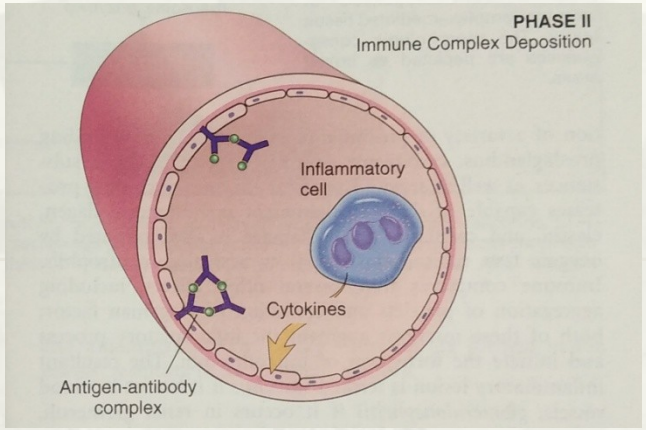
*The New Sjogren's syndrome Handbook, pp.74-83*

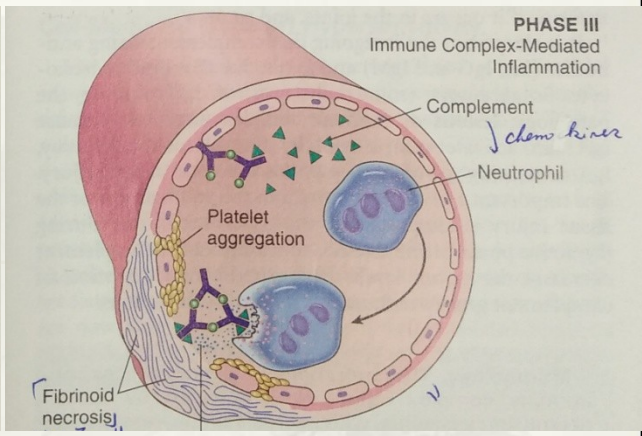
- Blood vessels in primary SS can be affected in two ways:
  - Vasculitis
  - Raynaud phenomenon
- Several sub-types of vasculitis occur with considerable frequency in SS:
  - **Small vessels** such as Leukocytoclastic angiitis
  - **Medium Vessel**s such as Granulomatous arteritis

*The New Sjogren's syndrome Handbook, pp.74-83*

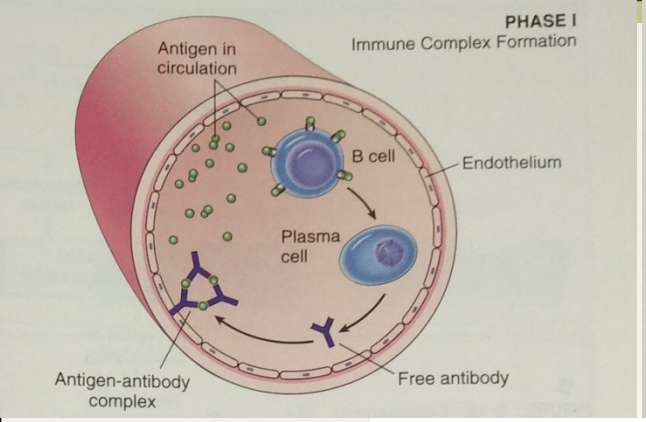
# Development of vessel wall injury

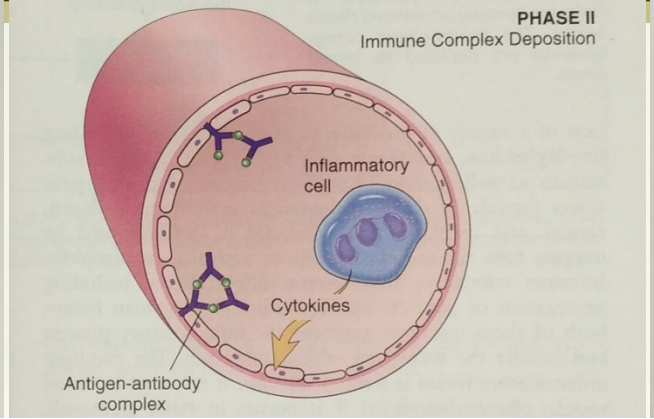


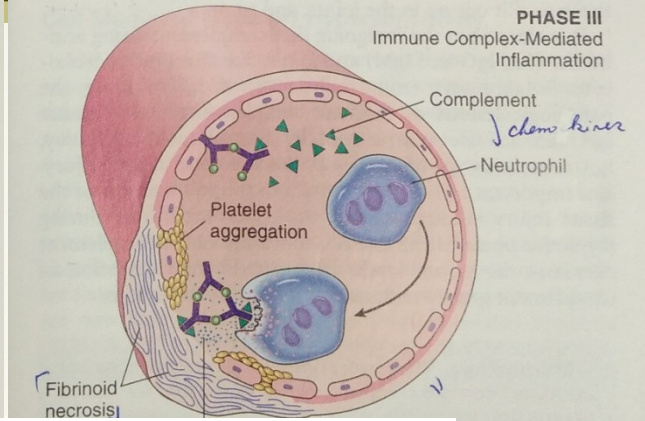



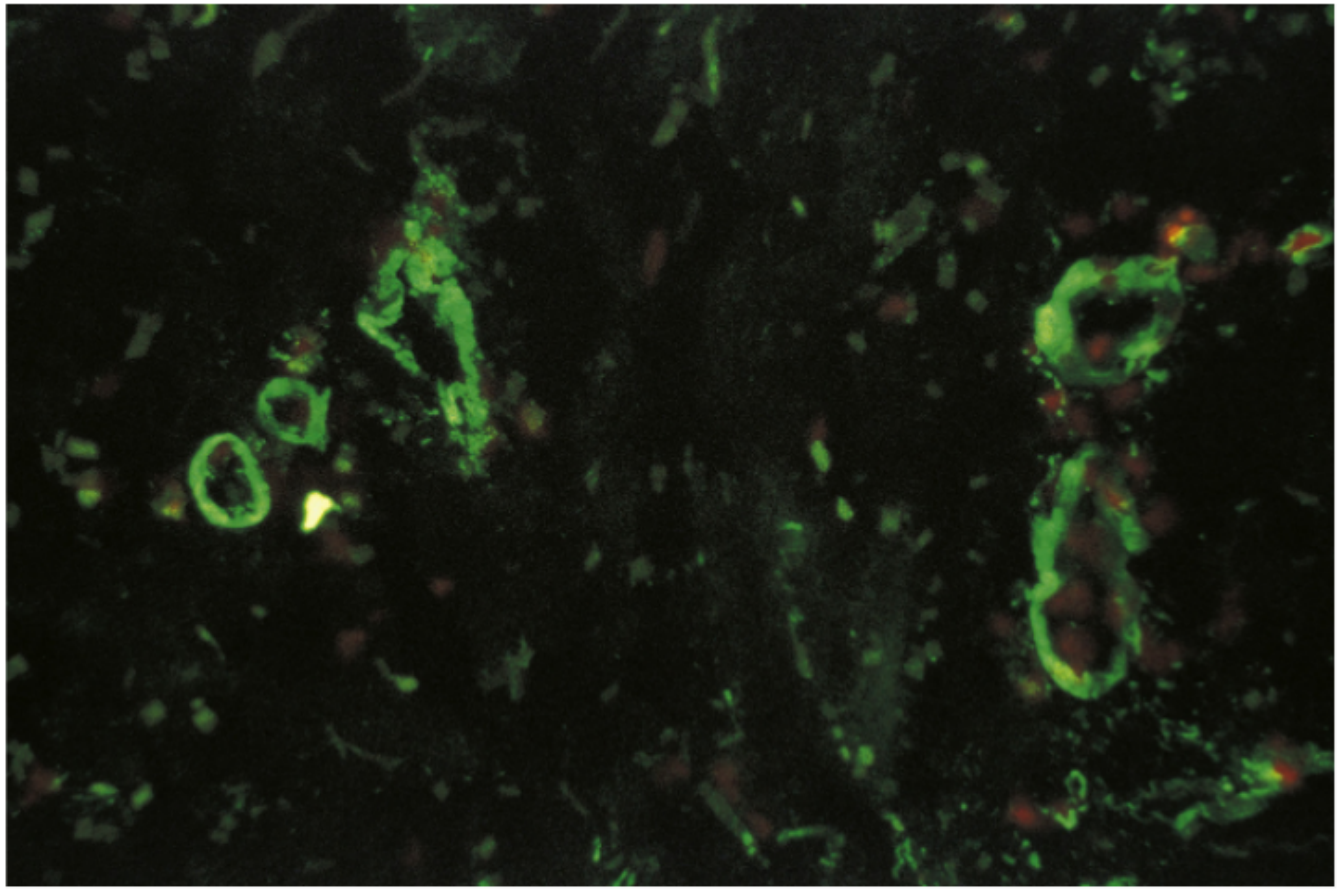




Robbins, Pathology Basis of Diseases, p.213



MacKee, Dermatopathology, p.663

- Primary SS vasculitis can affect the small blood vessels supplying the skin(most commonly), nerves, muscles, intestines, kidneys, lungs.
- Involvement of internal organs is more likely if the vasculitis is associated with cryoglobulins.
- Some patients with primary SS and vasculitis have an underlying B-cell lymphoma.

http://www.dermnetnz.org/immune/sjogren.html

**TABLE 6 PREVALENCE OF LABORATORY ABNORMALITIES IN NINE PRIMARY SJOGREN'S SYNDROME PATIENTS WITH VASCULITIS**

| Laboratory Abnormality | Number of Patients |
|---|---|
| Increased gamma globulins | 7 |
| Positive RF | 9 |
| Positive ANA | 9 |
| Cryoglobulins in serum | 9 |
| Low serum complement | 7 |

*The New Sjogren's syndrome Handbook, pp.74-83*

# Primary SS presenting as pleural effusion, pericardial effusion and ascites

- Among a total of 221 cases, 30 patients (5.7%) were identified with **pleural effusion**
- In the 352 Chinese pSS patients, **pericardial effusion** was detected in 52 patients (14.8%)
- **Ascites** can occur in SS when combined with
  - PBC(primary biliary cirrhosis): jaundice, ascites, abn liver function tests, increased anti-mitochondrial Ab
  - 6% of SS pts: autoimmune liver disease
- No previous reports have described pSS with peritonitis due to autoimmunological inflammation.

*Eastern Journal of Medicine 19 (2014) 54-57*
*The New Sjogren's syndrome Handbook, pp. 47-48*

# Ascites and peritonitis in autoimmune diseases

- Primary: (Lupus) Mesenteric Vasculitis
  - A leukocytoclastic vasculitis of mesenteric vessel walls, presents with abdominal pain, impaired intestinal motility, signs of peritonitis, ascites
- Secondary: nephrotic syndrome, congestive heart failure, constrictive pericarditis, protein-losing enteropathy, intestinal obstruction and perforation due to vasculitis or thrombosis

Ross E Petty et al. Textbook of Pediatric Rheumatology. P. 309

# Prognosis of SS

- Worse prognostic factors are the presence of glomerulonephritis, decreased blood levels of C4 complement, vasculitis and cryoglobulinaemia.
- SS pts with vasculitis usually respond well to appropriate treatment, better than do pts with other forms of idiopathic vasculitis.

*Information on Vasculitis in Sjögren's Syndrome excerpted from an article by Steven C. Carsons, MD, Winthrop University Hospital, Vol. 28, Issue 2,* The Moisture Seekers, *published by the Sjögren's Syndrome Foundation.*

# Treatment

- When vasculitis occurs, it often requires treatment with drugs that suppress the immune system. Medications such as cyclophosphamide, azathioprine, or mycophenolate mofetil may be prescribed by clinicians experienced in their use.

http://www.uptodate.com/

# Treatment

- A more severe vasculitis involving the peripheral nerves and or muscles, the kidney, GI tract and central nervous system:
  - moderate to high doses of corticosteroids and immunosuppressive agents such as azathioprine and cyclophosphamide,
  - Plamapheresis (blood cleansed of circulating immune complexes and cryoglobulines)
  - Intravenous immunoglobulin,
  - Rituximab

http://www.uptodate.com/

- physicians caring for these patients should be aware of this as early diagnosis and prompt therapy leads to rapid recovery and decreased morbidity and mortality.

# Thanks for your listening !!