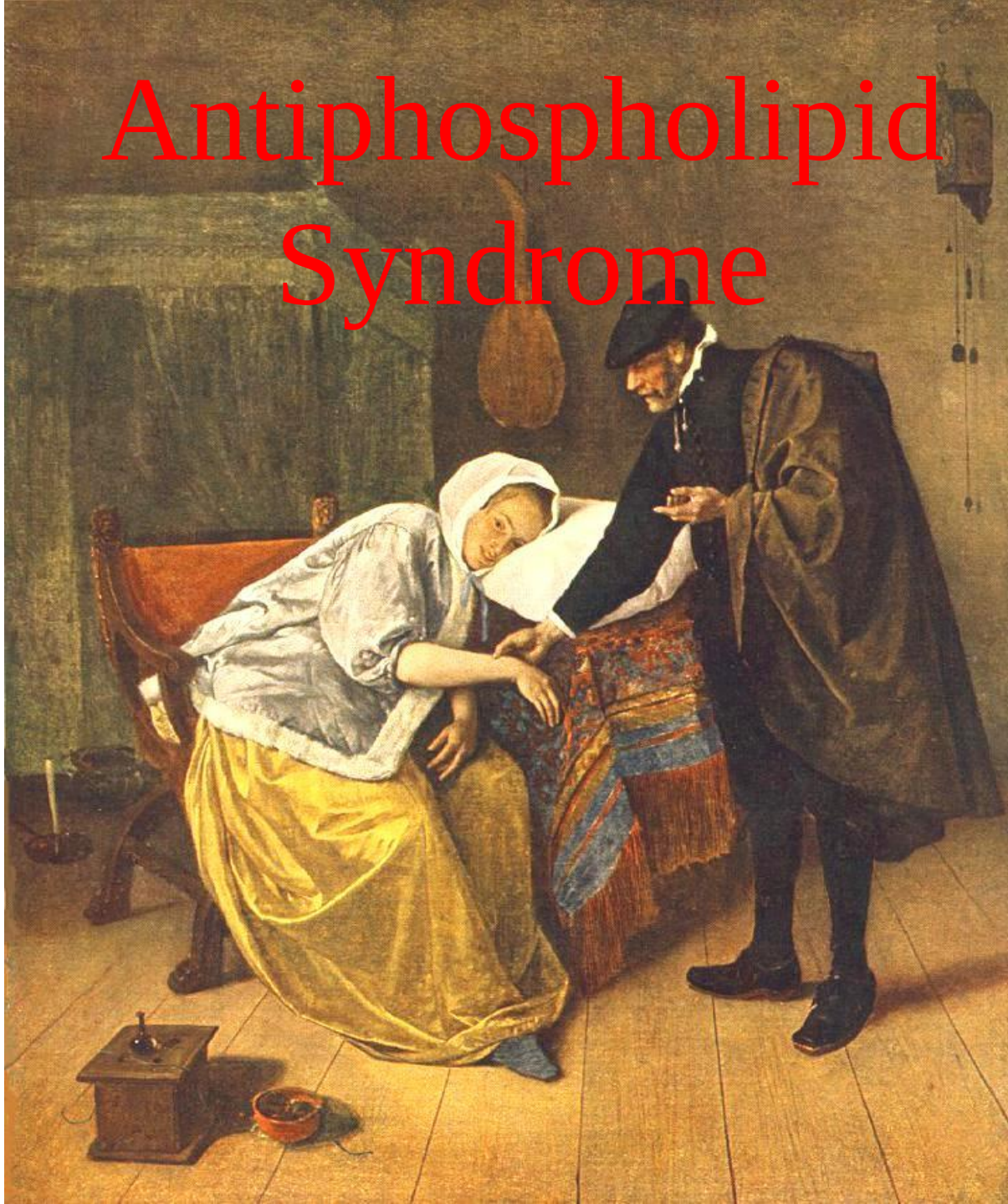


Antiphospholipid Syndrome



The antiphospholipid syndrome is an autoimmune disease that is characterized clinically by *vascular thrombosis* and *pregnancy morbidity*, and serologically by the presence of *antiphospholipid Abs*

EPIDEMIOLOGY

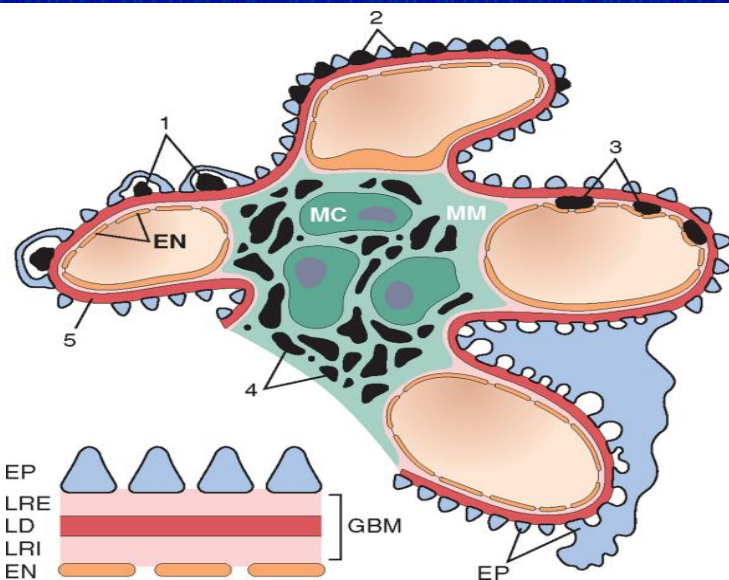
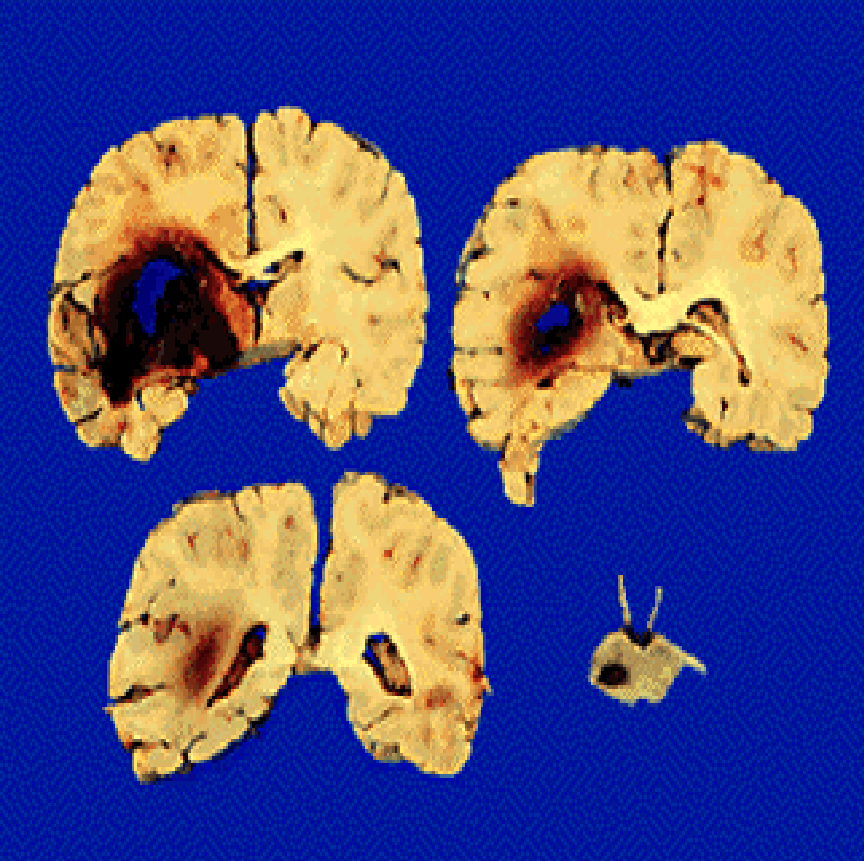
- 10% of normal population have transient low-titer anticardiolipin; 1% have moderate-high titerspatients
- 50% of APS patients have SLE (defined as *secondary APS*)
- Upto 40% of SLE patients have positive aPL tests and 10-20% have APS
- Asymptomatic aPL-positive patients have a 0% to 4% annual risk of thrombosis
- 14% of patients with recurrent venous thromboembolic disease have aPLs



MANIFESTATIONS: THROMBOSIS

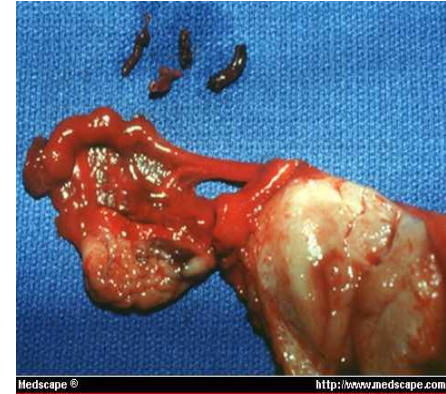
- Venous and/or arterial occlusion may occur in any organ
- Thromboses may be severe, recurrent, and occur in unusual sites and in young peoples
- DVT and stroke are the most common clinical manifestations of APS
- Severe HTN, renal failure and proteinuria without casts may occur due to renal microangiopathy, glomerular capillary injury, and thrombosis of renal vessels





PREGNANCY COMPLICATIONS

- Losses in patients with aPLs typically occur after *10 weeks'*
- *Fetal growth* slows and amniotic fluid volume decreases in second trimester
- Severe, early *preeclampsia* or *HELLP* syndrome may develop
- Prior late pregnancy losses predict future losses



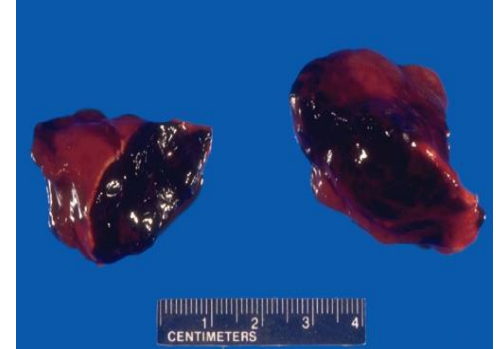
NON-THROMBOTIC MANIFESTATIONS

- *Livedo reticularis* is common but is not specific for APS
- *Cardiac valve* disease (vegetations, thickening), is a late manifestation
- Accelerated *atherosclerosis* is not evident in recent studies
- Leg ulcers may develop



CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME (CAPS)

- Multiple thromboses of *medium and small arteries* occurring over a period of days
- Stroke, cardiac, hepatic, adrenal, renal, and intestinal infarction; *acute adrenal failure*, peripheral gangrene occur
- *Thrombocytopenia* is moderate; erythrocytes are less fragmented than in the HUS or TTP, and *fibrin splits* are not strikingly elevated
- Tissue biopsies show *noninflammatory* vascular occlusion



LABORATORY TESTS

- Repeatable positive *LA* test or a moderate- to high-titer *aCL IgG* or *IgM* test is required
- *IgA aCL* and *IgG*, *IgM*, or *IgA anti- β 2GPI* are helpful when *LA* and *aCL* tests are negative
- *LA* test is more specific but less sensitive predictor of thromboses



LABORATORY TESTS



- *ANA* and *anti-DNA* antibodies occur in 45% of patients with primary APS
- *Hypocomplementemia*, *erythrocyte casts*, and *pyuria* imply lupus nephritis rather APS
- *ESR*, *Hb*, and *WBC* count are normal in uncomplicated primary APS, except during acute thrombosis

DIAGNOSIS: CLINICAL CRITERIA

1-Vascular thrombosis:

A clinical episodes of arterial, venous, or small vessel thrombosis in any organ

2-Pregnancy morbidity :

a) An unexplained fetal deaths beyond the 10th week of gestation, **or**

b) One premature birth before the 34th week because of eclampsia, severe preeclampsia or placental insufficiency **or**

c) Three unexplained consecutive abortions before the 10th week (not with anatomic, hormonal or chromosomal causes)

DIAGNOSIS: LAB CRITERIA

- 1- *LA* on two or more occasions at least 12 weeks apart
- 2- *aCL* of immunoglobulin Ig G or IgM in medium or high titer, on two or more occasions at least 12 weeks apart, by ELISA
- 3- *Anti- β 2GPI* antibody of IgG or IgM on two or more occasions at least 12 weeks apart, measured by ELISA

Note: Definite APS is present if at least one of the clinical and one of the lab criteria are met

DIAGNOSIS:SUGGESTING FEATURES

Clinical:

- Livedoreticularis
- Thrombocytopenia
(usually 50,000-100,000/mm³)
- Autoimmune hemolytic anemia
- Cardiac valve disease
- Multiple sclerosis–like syndrome, chorea, or other myelopathy

Laboratory:

- IgA *aCL* antibody
- IgA *Anti-β2GPI*

DIAGNOSIS: CAPS

1. involvement of *three* or more organs or tissues
2. Development of in less than *1 wk*
3. Confirmation by *histopathology* of small vessel occlusion in at least one organ or tissue
4. Laboratory confirmation of the presence of aPL (*LA* or *aCL* or *Anti- β 2GPI*)

Definite Catastrophic APS: All 4 criteria

PROGNOSIS

- Prognostic factors are: pulmonary hypertension, neurologic involvement, myocardial ischemia, nephropathy, gangrene of extremities, and CAPS
- Renal failure due to microangiopathy is rare
- Thrombosis may cause loss of a transplanted kidney or other organ
- Valvulopathy may need valve replacement
- Perioperative complications may occur despite prophylaxis
- Mortality is 50% in CAPS



Name:
The
anemic
lady

Painter:
Van
Hoogst-
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