

Case report

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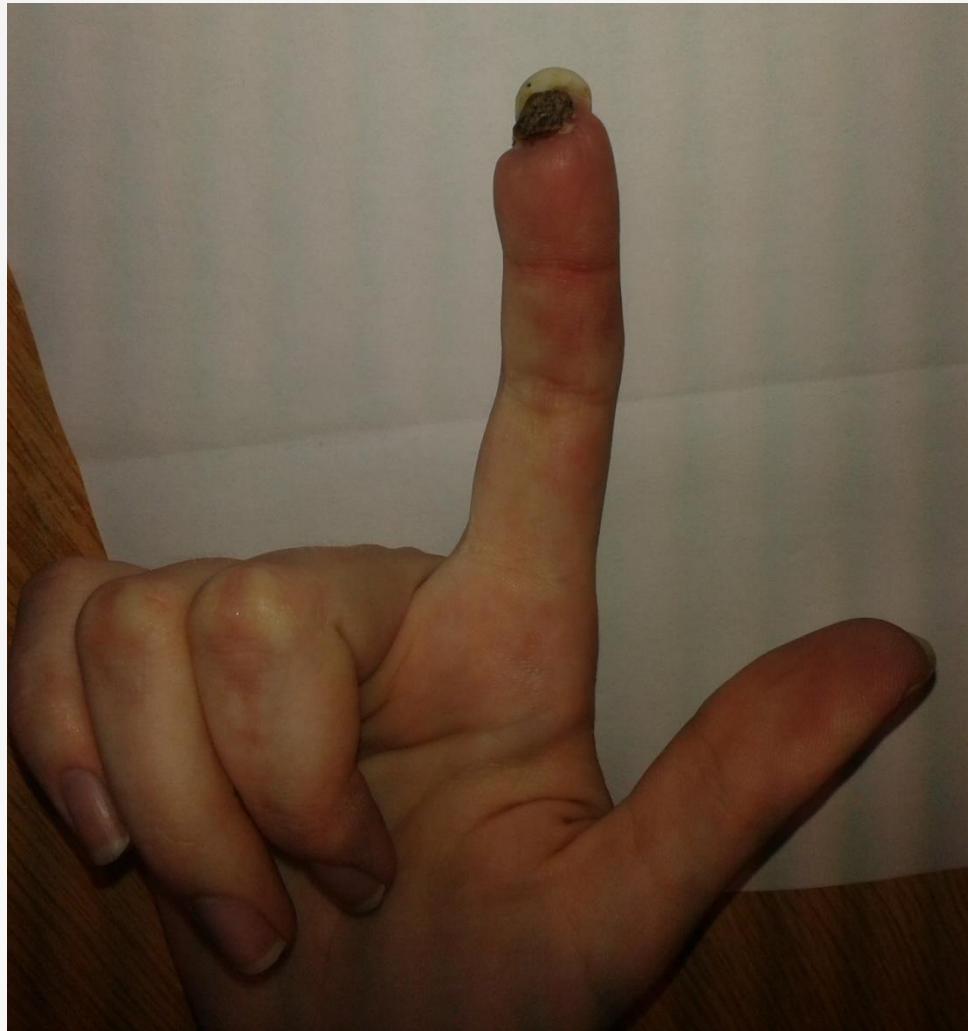
VL 1992

- Healthy young woman to 10/2014
- 10/2014 - acrocyanosis of hands
- 12/2014
 - ✓ necrosis of distal phalanx of II. right finger
 - ✓ acrocyanosis of other fingers on hands and foots too
 - ✓ problems with breathing by moving
- Christmas 2014
 - ✓ acute worsening
 - ✓ problems with visus - on both sides, worse on right - see only shadows
 - ✓ hospitalisation in general hospital in place of living
 - ✓ after two days hospitalisation in our clinic

VL 1992

- Admission report
 - ✓ no important information in history, non smoker, active sports
- Clinical examination:
 - ✓ dyspnoea at rest
 - ✓ quadruhyperreflections and neocerebellar symptomatology
 - ✓ livedo reticularis
 - ✓ amaurosis on right eye, see shadows on left eye
 - ✓ cyanosis - central and periferial
 - ✓ necrosis of II. right finger

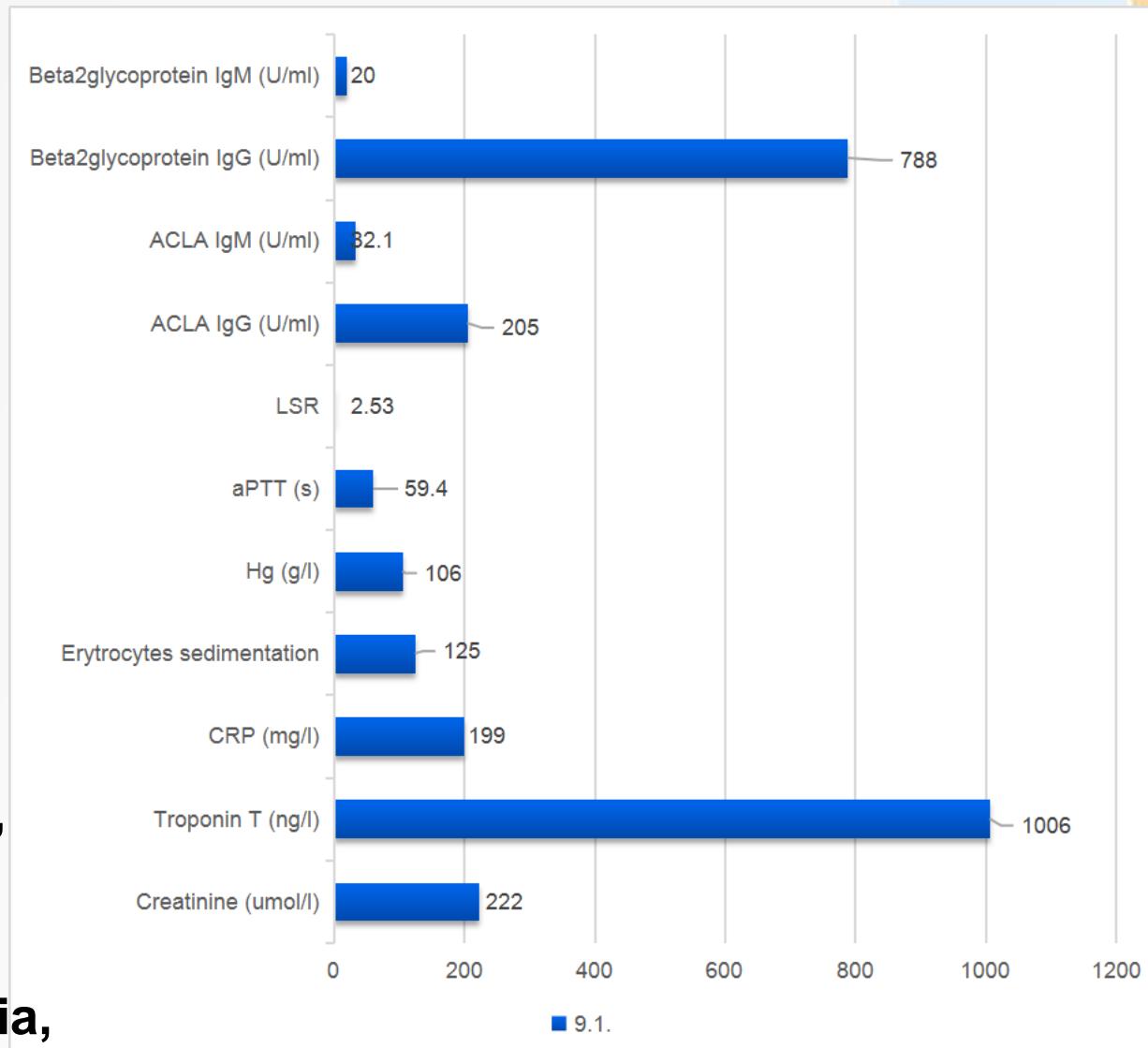
VL 1992



VL 1992 - Laboratory

- **Pathological:**
 - ✓ Myoglobin - 125ug/l
 - ✓ NTproBNP - >35 000
 - ✓ Proteinuria - 1g/day
 - ✓ Cultivation of candida albicans - tongue

- **Negative:**
 - ✓ Cryoglobulins, creatinkinase
 - ✓ Immunology - ANA, ENA, ANUC, anti ds DNA, ANCA, C3, C4
 - ✓ Thyroids hormones, antibodies
 - ✓ Serology (CMV, EBV, borrelia, hepatitis, listeria, herpes viruses, mycoplasma, chlamydia), cultivations



Normal range of ACLA, beta2 glycoprotein - 20U/ml
Normal range of LSR - 1-1.25

VL 1992 - Examination

- **CT angiography**
 - ✓ excluded embolism
 - ✓ pleural effusion on right
 - ✓ possible ARDS (Acute respiratory distress syndrome)
- **Echocardiography**
 - ✓ heavy damage of contractility
 - ✓ ejection fraction of left ventricle 25%
 - ✓ pulmonary hypertension - PAPS 40mmHg
 - ✓ without effusion
- **Kidney ultrasound**
 - ✓ normal size, oedema of kidney

VL 1992 - Examination

- **Ophthalmologic examination**

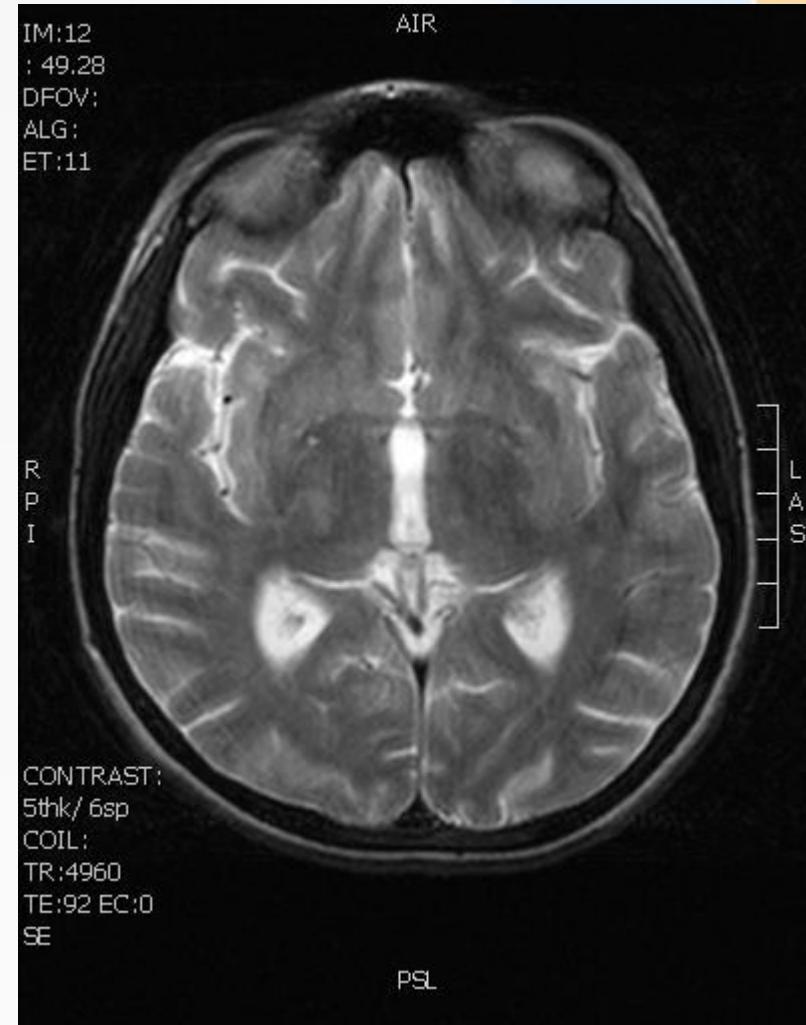
- ✓ right site - massive obliteration of vessels with total non perfusion on temporal site, laserphotocoagulation, lost of visus will be permanent
- ✓ left site - small vessel obliteration with collateral system
- ✓ possible vasculitis or obliteration aetiology

- **MRI+MRA**

- ✓ multi-local older lesion supratentorial bilateral
- ✓ acute supratentorial lesion bilateral
- ✓ possible postischemic, embolism aetiology?

- **USG+scintigraphy of thyroid gland**

- ✓ possible thyroiditis



VL 1992 - therapy

- **Hospitalisation on ICU (Intensive Care Unit)**
- **Plasmapheresis - 5x**
- **Pulses of high doses corticosteroid (methylprednisolon)**
 - ✓ 3days à 1g, than à 125mg (total 3,925g)
 - ✓ p.o. prednison 40 per day, after 3 months 5mg per day
- **Anticoagulant therapy**
 - ✓ **Low-molecular-weight heparin (LWHM), nadroparine 0.1/kg**
 - ✓ **Warfarin, INR 3-3,5**
- **Symptomatic therapy**
 - ✓ **Cardiac failure** - diuretic (i.v., p.o. furosemid 60mg), perindopril 4mg, carvediol 25mg
 - ✓ **Vasodilatation therapy** - i.v. alprostadil, p.o. naftidrofuryl, acidum acetylsalicylicum
 - ✓ **Others** - thiamazol, statins, substitution of kalium, calcium, magnesium, vitamin D, B
 - ✓ **Analgetics, antibiotics, antimycotisc**
- **Lasephototherapy** - repeatedly

VL 1992 - Diagnosis

- **Catastrophic antiphospholipid syndrome**
 - Myocarditis
 - Renal failure
 - Ischemic retinopathy
 - Ischemic brain lesions
 - Acrocyanosis with gangrene II. finger
 - Ischemic induces hyperthyroiditis
- High positive anticardiolipin antibodies

VL 1992 - 12/2016

- **Heart - EF 55%, dilatation of left ventricle**
- **Eyes - completed occlusion on right eye, on left side collateral vessel system, visus monocular, good adaptation to vision in space**
- **Kidney - normal**
- **Thyroid g. - normal**
- **Fingers - normal, without tissue changes**
- **Neurological symptoms - normal, old stationary lesions on MRI**

