Disorders of the vessel wall

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Vascular disorders associated with purpura and bleeding

I. **Hereditary disorders of connective tissue:**
Osler-Weber-Rendu, Ehlers-Danlos, Marfan syndromes, osteogenesis imperfecta

II. **Allergic:** Henoch-Schönlein purpura

III. **Atrophic:** senile purpura, Cushing sy., steroid induced bruising, amyloidosis, paraproteinemias, scurvy

IV. **Infectious:** viral, bacterial, rickettsial

Osler-Weber-Rendu syndrome (hereditary hemorrhagic teleangiectasia)

Inheritance: **autosom dominant. Incidence: 1:50000**

Pathology: **vascular fragility, elastin deficiency.**

Symptoms: **widespread cutaneous, mucosal, visceral teleangiectasias.**

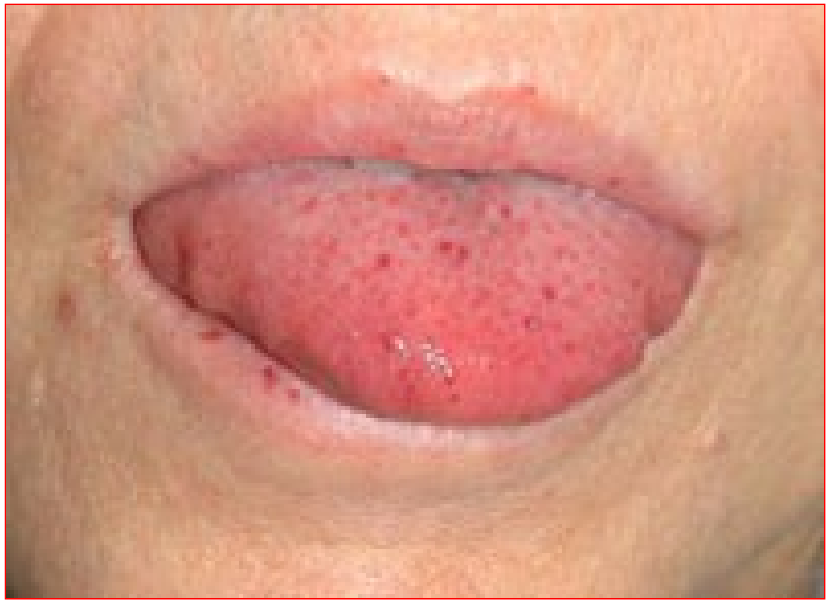
– predominantly on the tongue, lips, digit tips, perioral region and trunk. Epistaxis is the most common symptom. Iron deficiency anaemia.

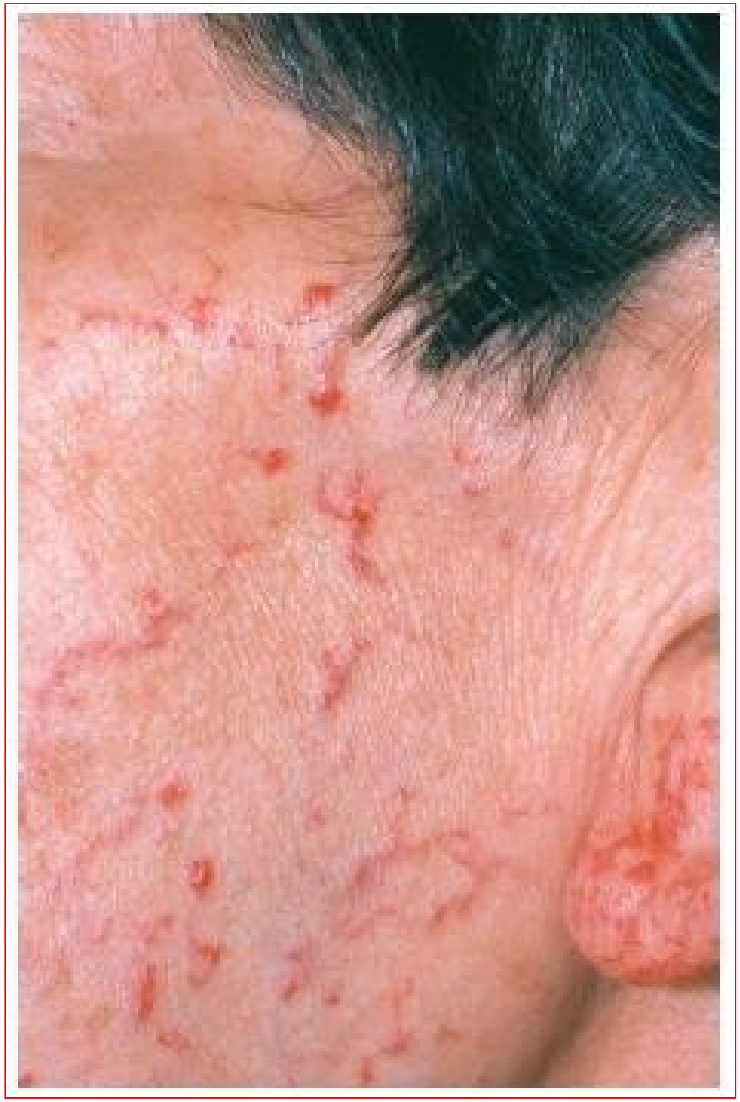
– gastric-, duodenal, pulmonary arteriovenous malformations ⇒ gastrointestinal bleeding, hypoxemia.

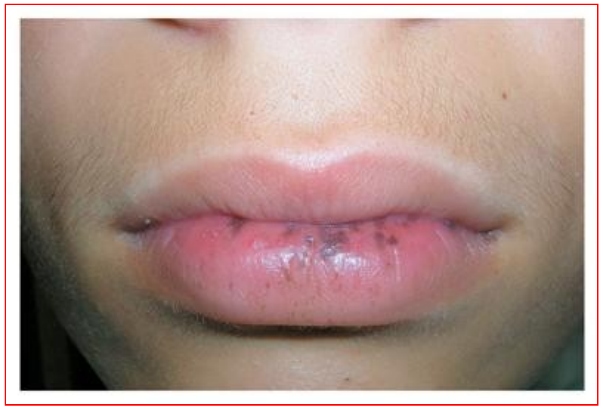
Sings and symptoms wores with aging.

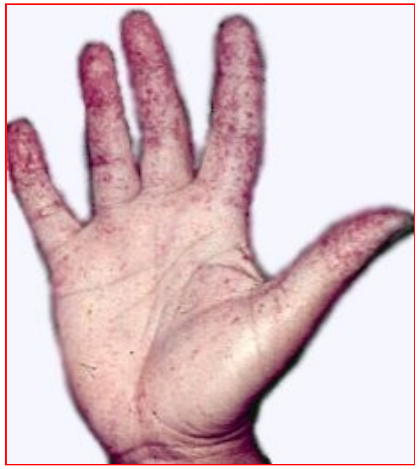
Treatment: **no specific therapy. Iron supportation, cauterize bleeding vessels, blood transfusion.**

Osler- Weber- Rendu syndrome



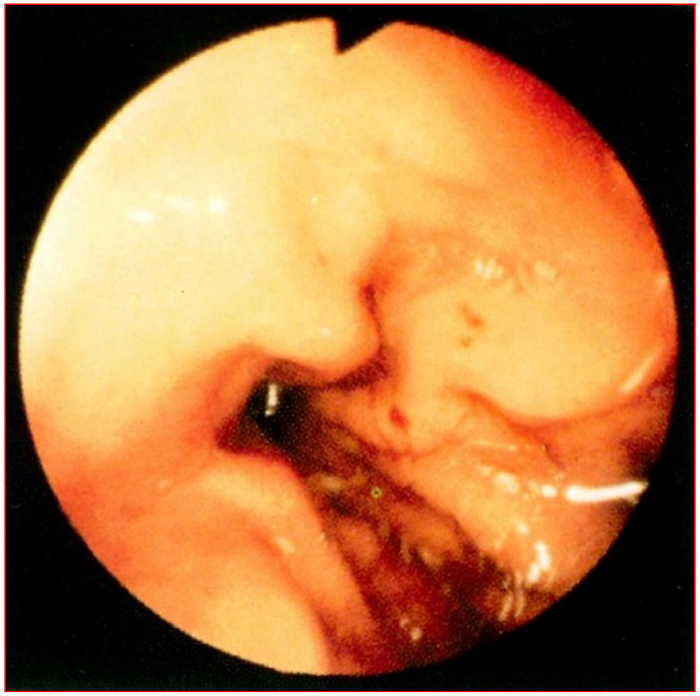




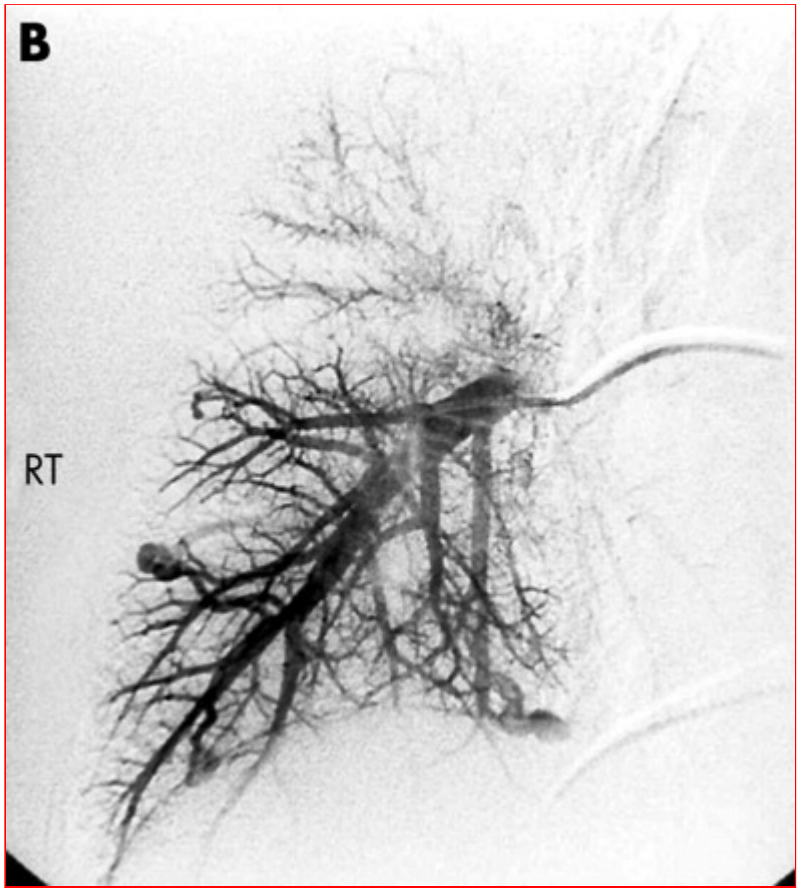


Osler- Weber- Rendu syndrome

Gastroscopy: telangiectases in the gastric mucosa



Pulmonary angiography: multiple arterial venous malformations



Ehlers-Danlos syndrome:

Hyperelasticity of the skin, hypermobile joints („rubber man”). Other features: mitral valve prolapse, hernias.

Incidence: **1:5000 births**

Pathogenesis: **mutations in structural genes for collagen or procollagen.**

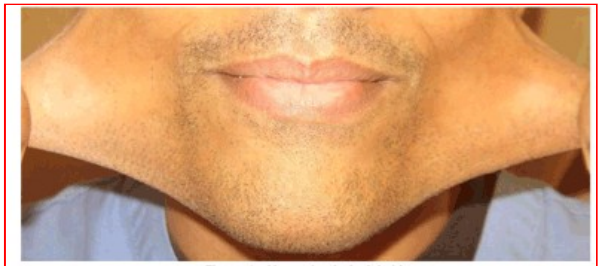


Figure 1—Hyperelastic facial skin.

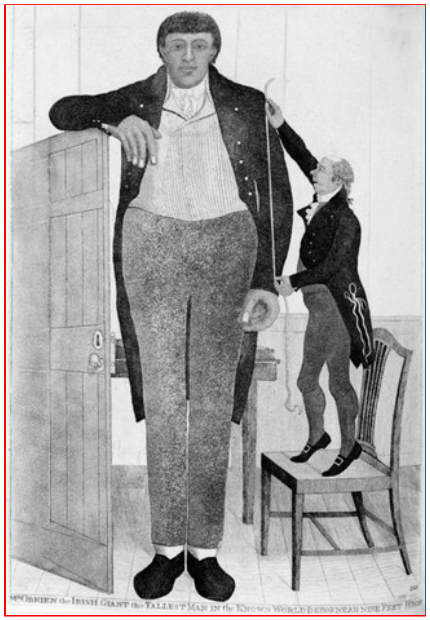
Marfan syndrome:

Incidence: **1:3000-5000,** ***inheritance:*** **autosomal dominant**
Pathogenesis: **fibrillin 1 (FBN1) mutation.**
Symptoms:

- Sceletal changes: long, thin extremities, loose joints

- Reduced vision (as a result of dislocation of the lenses: ectopia lentis)

- Aortic aneurisms



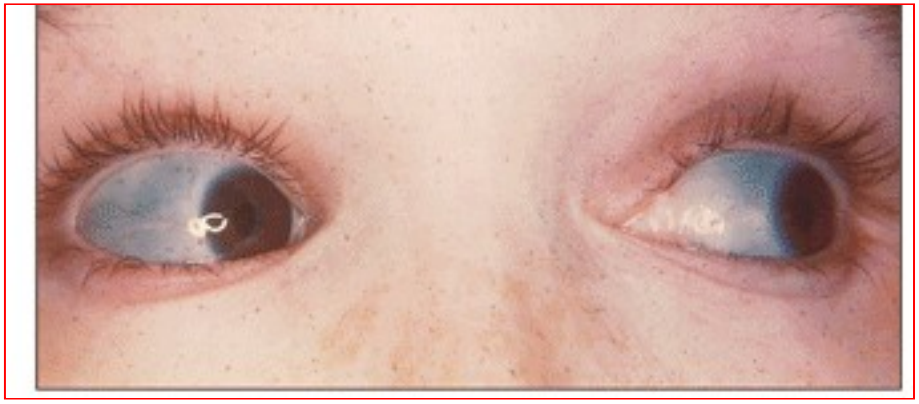
Osteogenesis imperfecta (Brittle Bone Disease):

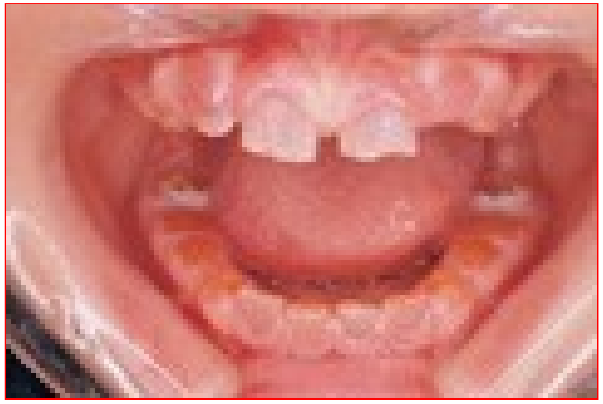
Is characterized by a generalized decrease in bone mass (osteopenia).

Incidence: **1:30000-60000,** ***inheritance:*** **autosomal dominant trait.**

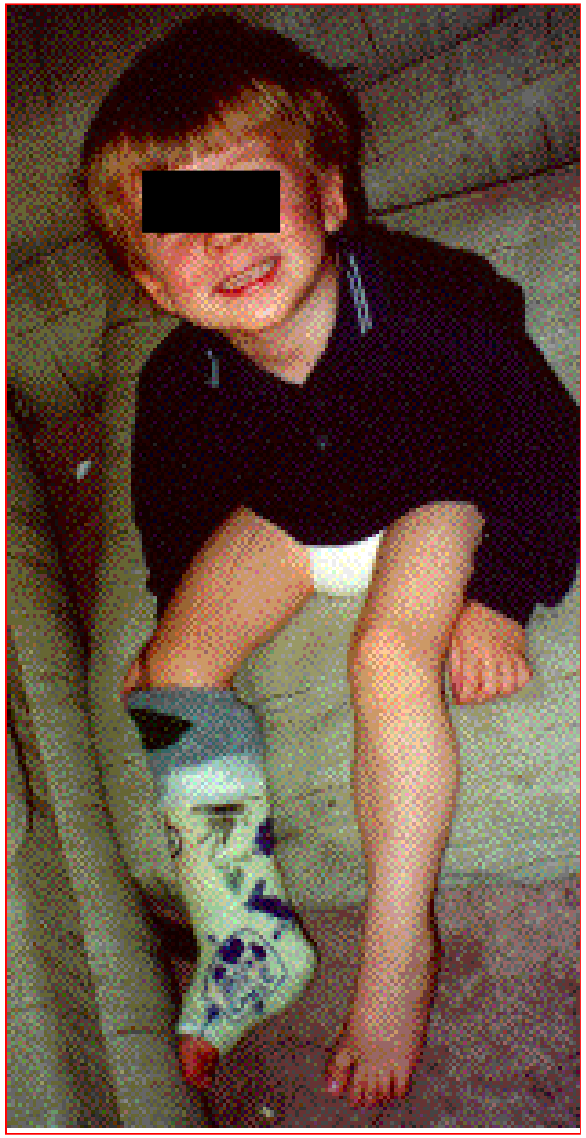
Pathogenesis: **deficiency of Type-I collagen.**

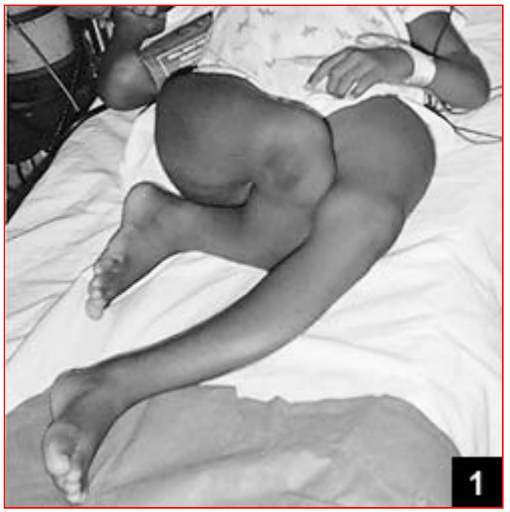
Symptoms: **bones fracture easily, discolouration of the sclera (blue sclera), dental abnormalities, progressive hearing loss, poor muscle tone, loose joints, slight spinal corvature.**

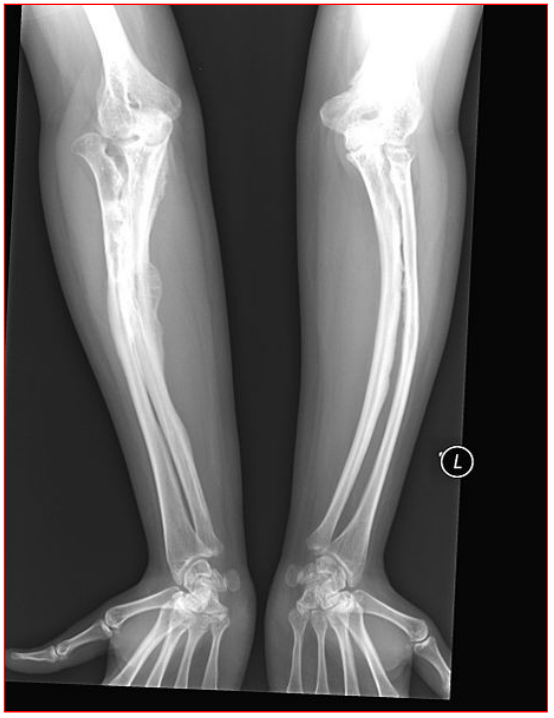




Osteogenesis imperfecta







Henoch-Schönlein anaphylactoid purpura:

Allergic vasculitis, most common in childhood (4-7 years)

Pathogenesis: **immun complex deposition (IgA antibody class) in the skin and kidney. It may occur after certain viral and bacterial infections, as well as an adverse drug reaction to some medications.**

Symptoms:

- palpable purpuric lesions (most commonly over the buttocks and lower extremities)

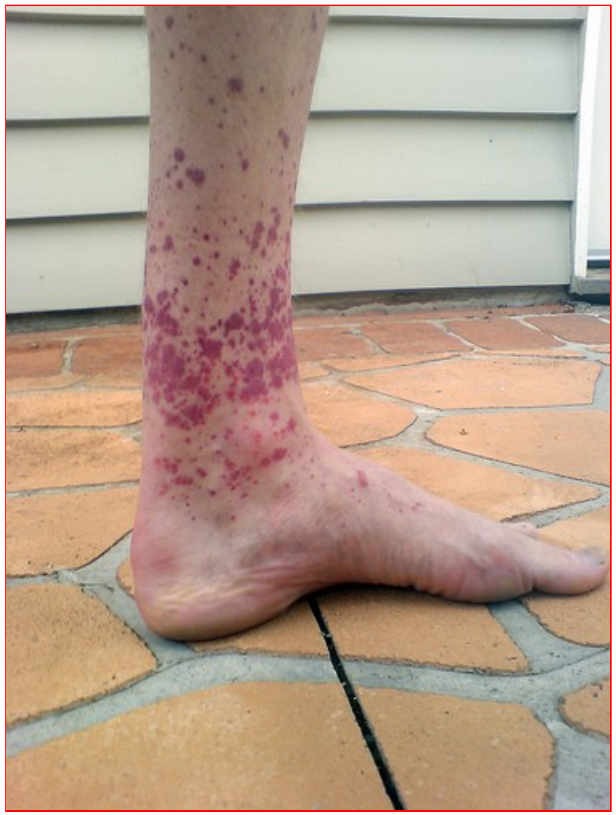
- pruritic papules and plaques. Lesions are frequently symmetrical.

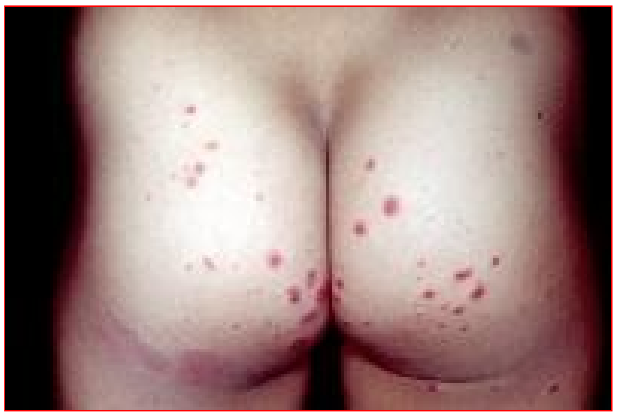
- Others: intestinal submucosal hemorrhage, hematuria (glomerulonephritis), arthritis.

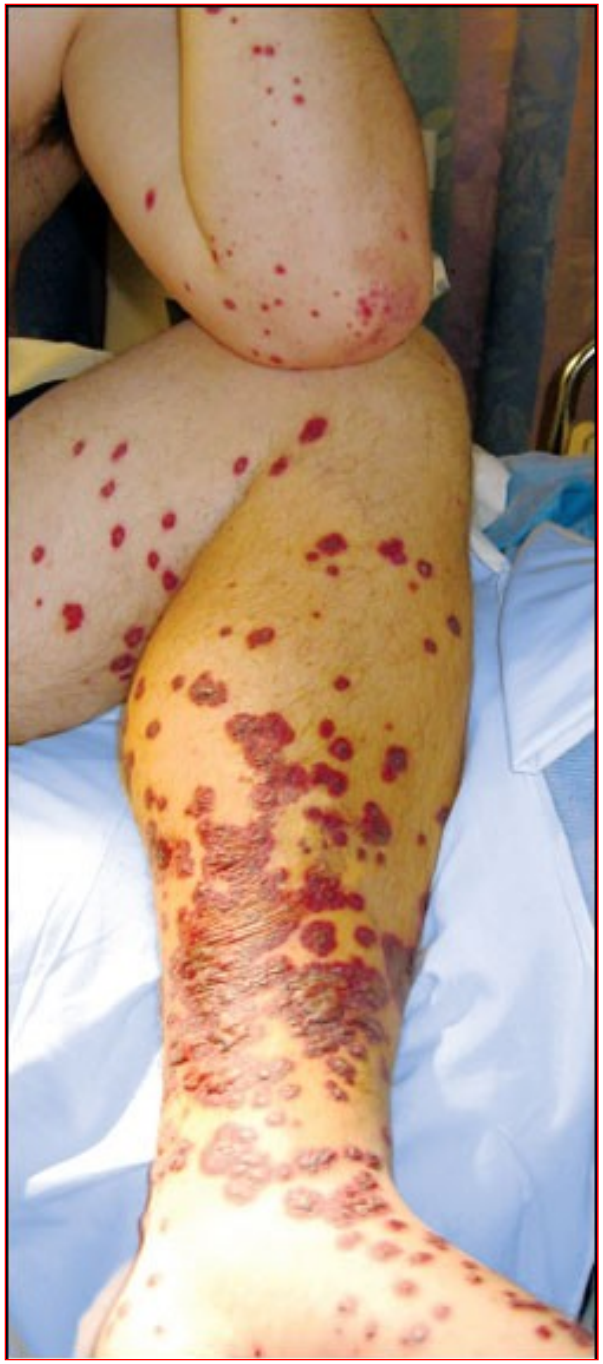
Treatment: **most cases do not require therapy, glycocorticoid.**

Prognosis: **excellent.**

Henoch- Schönlein purpura



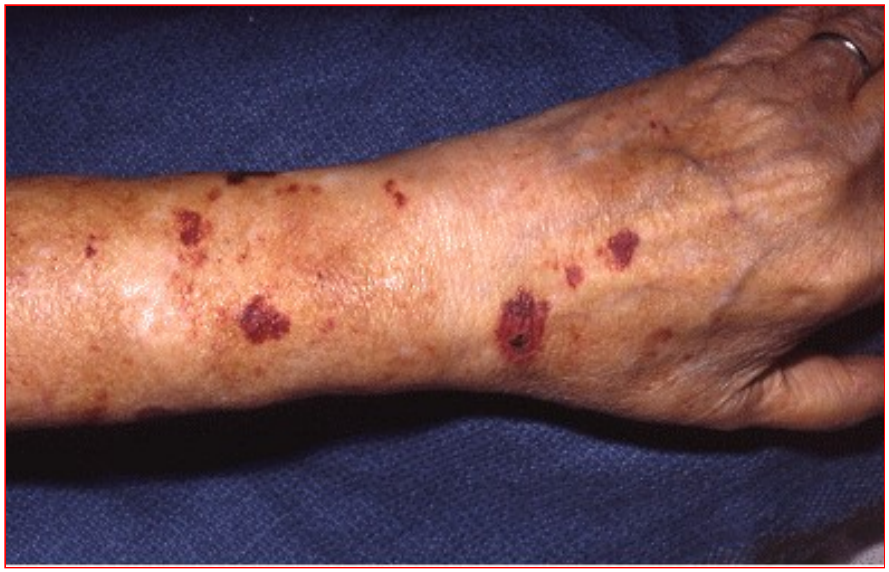




Senile purpura:

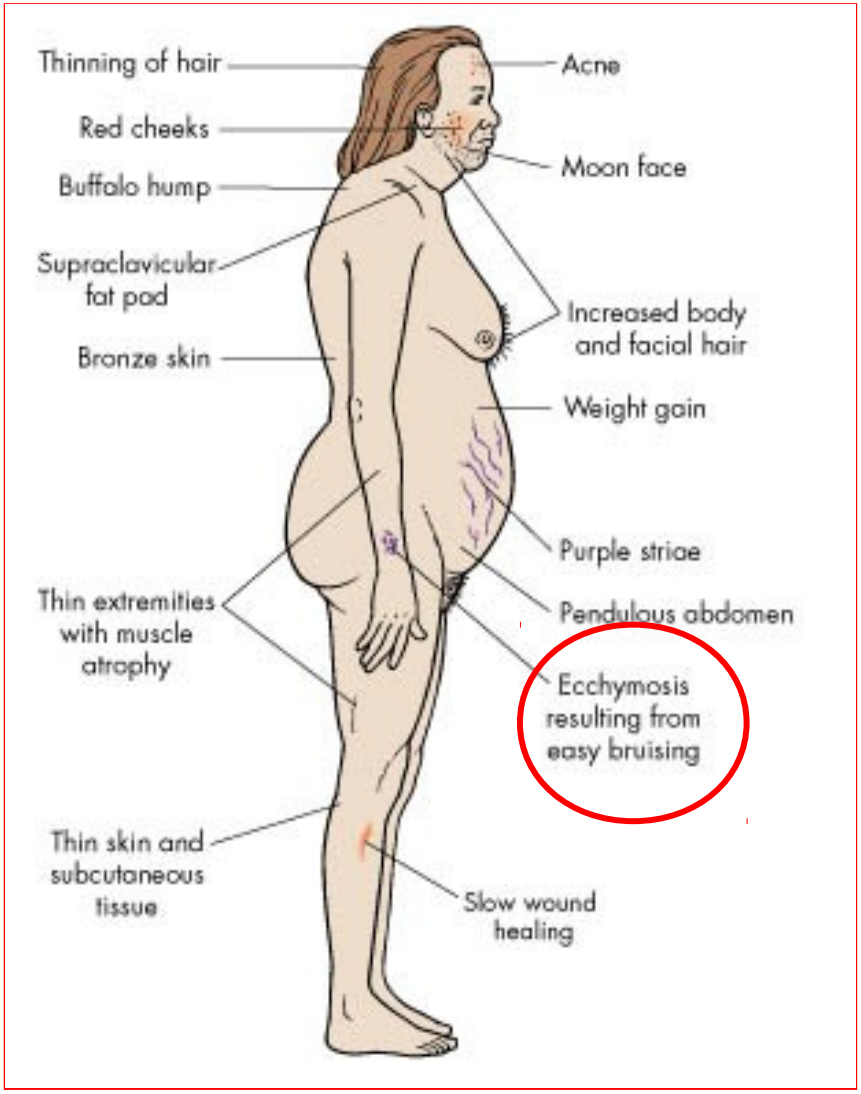
Pathogenesis: **atrophic origin**
– aging related decreases in collagen, elastin and subcutaneous fat combined with years of sunlight-induced damage.

Symptoms: **skin is thin, lacks elasticity, tears easily.**
Red and purple purpuric and ecchymotic pathes (due to extravasation of RBCs from fragile vessels), mainly on the extensor surface of the arms and hands.

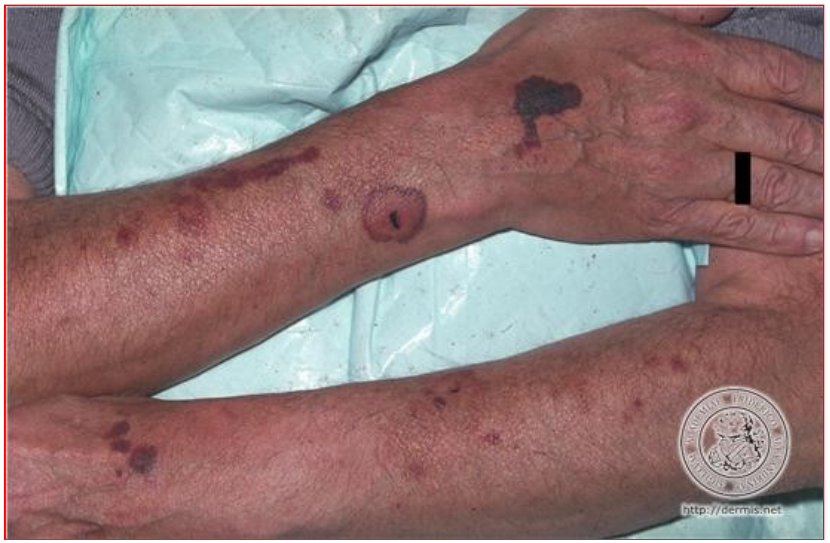


Although cosmetically displeasing, the disorder has no health consequences.

Cushing syndrome



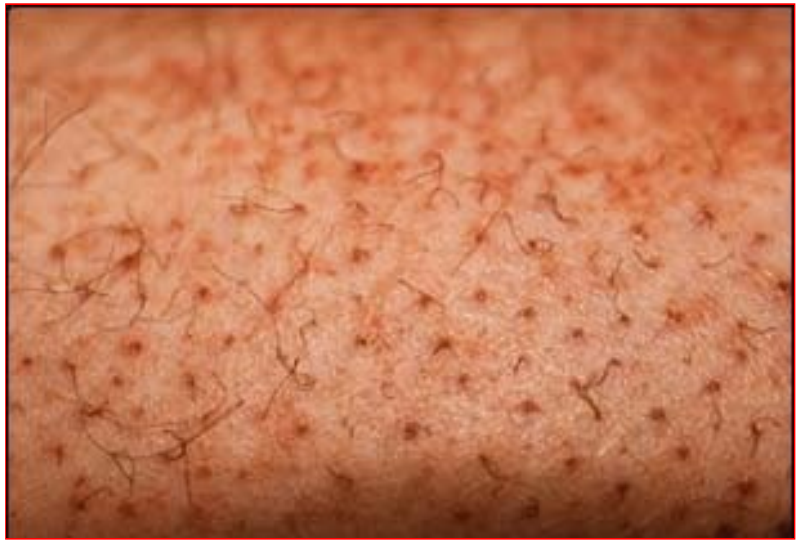
Purpura Steroidica



Scurvy:

Pathogenesis: **vitamin C deficiency (it is needed to sythesize hydroxyproline, an essential constituent of collagen)**

Symptoms: **painful episodes of perifollicular skin bleeding, gingival bleeding.**



Purpura associated with infections:

- Viral (smallpox, influenza, measles)

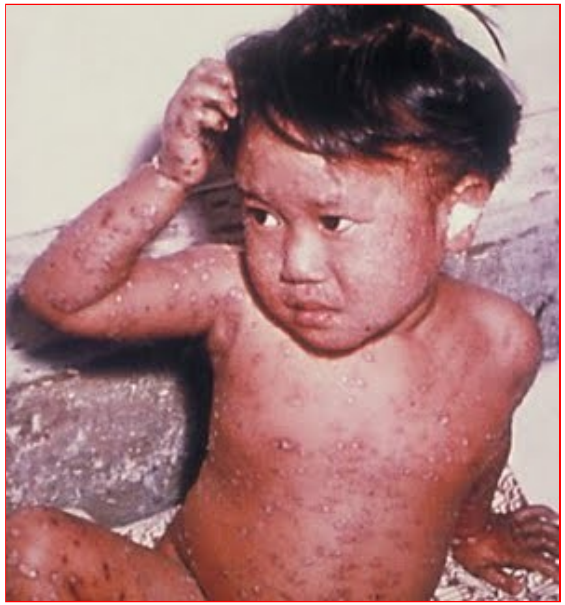
- Bacterial (meningococcus, typhoid fever, diphtheria, scarlet fever, tuberculosis, endocarditis, leptospirosis)

- Richettsial (Rocky Mountain spotted fever)

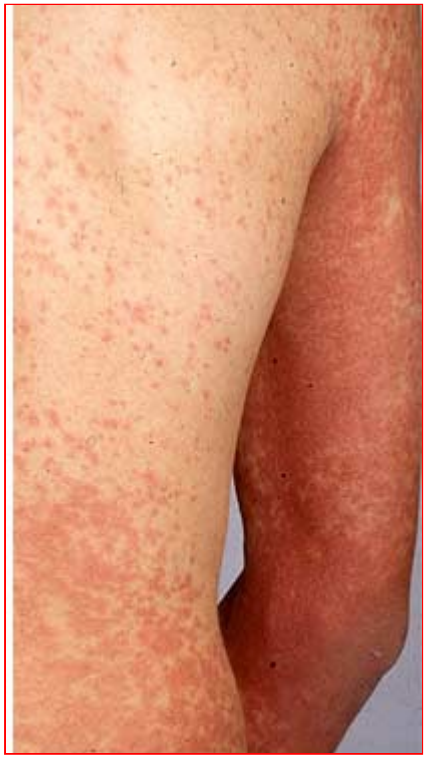
Pathogenesis:

direct vascular invasion by the organism, immune-complex vasculitis, septic emboli (e. g. bacterial endocarditis)

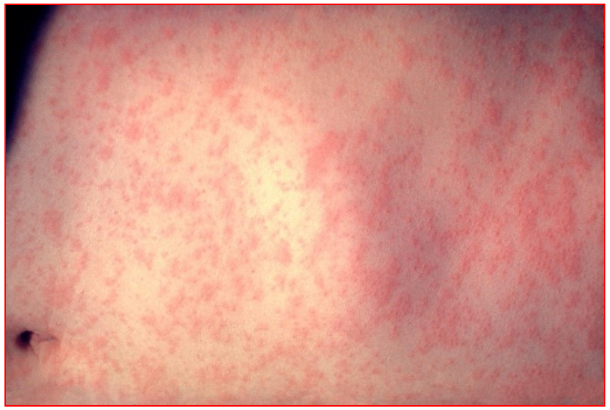
Smallpox



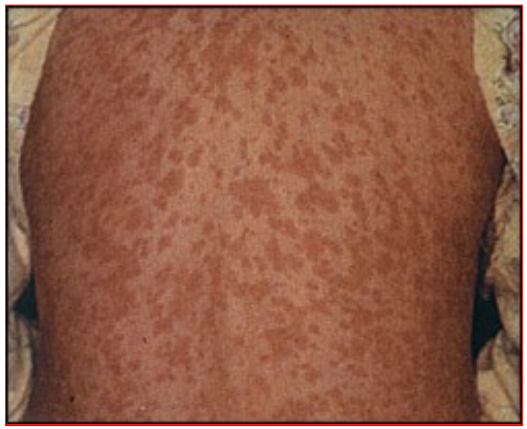
Scarlet fever



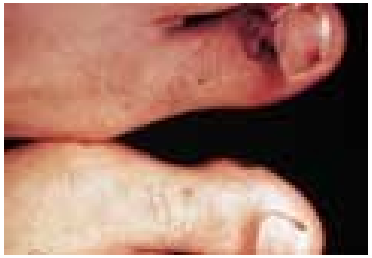
Measles



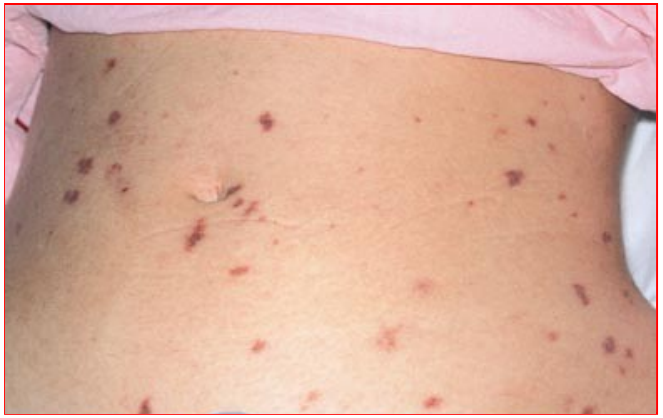
Rocky Mountain spotted fever



Septic emboli



Meningococcal infection



Autoerythrocyte sensitisation (Gardner Diamond syndrome):

Pathogenesis: autosensitisation to some component of the own red cells membrane.

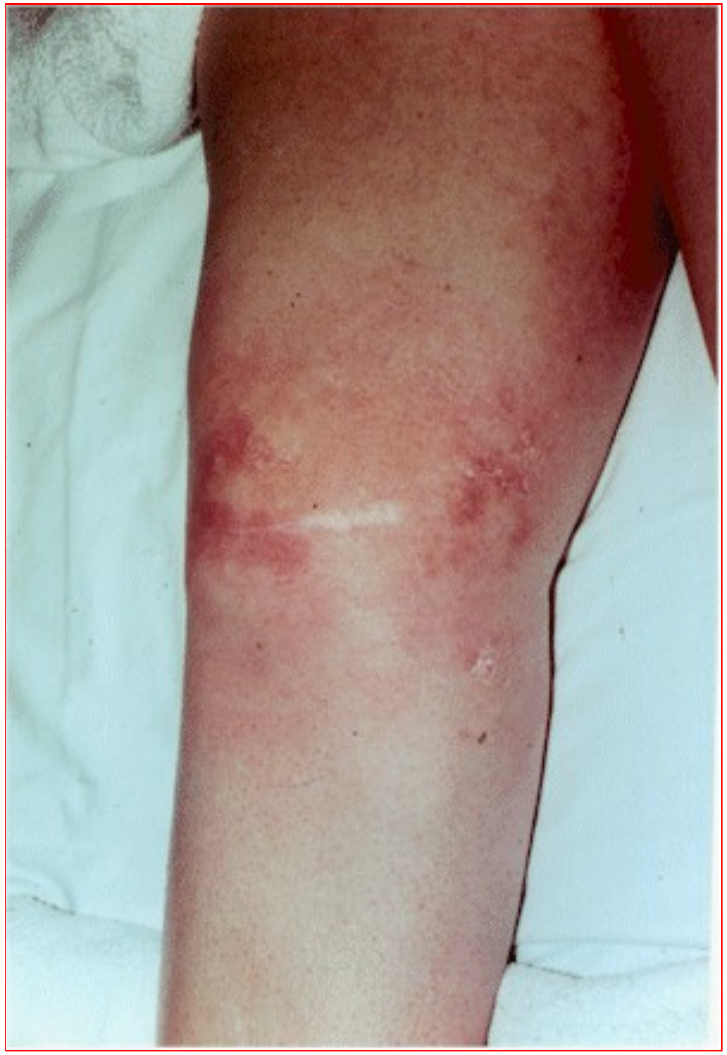
Rare disease.

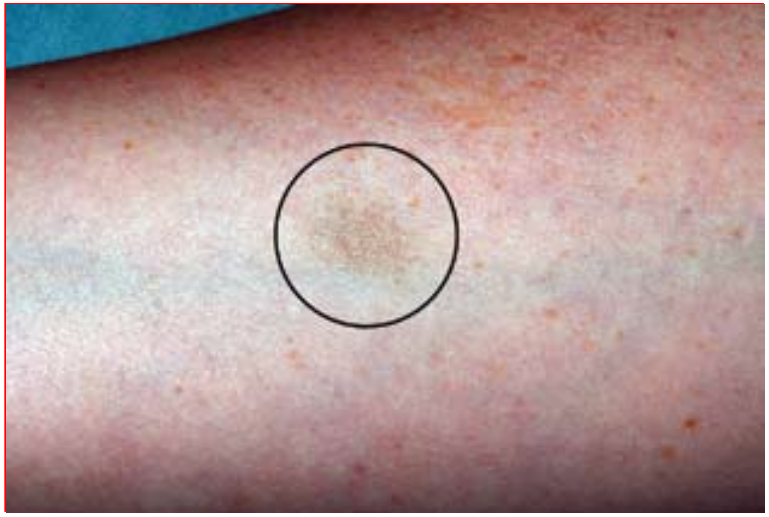
Typically occurs in white women who are experiencing emotional stress or who have concomitant psychologic illness.
Symptoms: spontaneous painful ecchymoses on different sites of the body. Commonly associated with headache, nausea, vomiting.

Tests of the coagulation system are normal.

Diagnosis: intradermal injection of 0.1 ml of autologous RBCs or RBC stroma may result in pain, swelling, and induration at the injection site.

Gardner Diamond syndrome





Following the injection of the autologous red cells,the patient noticed a tingling sensation similar to her earlier reports at the site, and within 24 hours of the initial test developed a bruise.