Thrombocytopenic purpura is the general term for purpura that accompanies a decrease in platelet density. When that density is less than 100,000 per microliter, subcutaneous bleeding is easily produced by bruising. When it is less than 50,000 per microliter, bleeding becomes marked and causes purpura. Thrombocytopenic purpura is classified by pathogenesis into idiopathic thrombocytopenic purpura, which is caused by auto-antiplatelet antibodies; symptomatic thrombocytopenic purpura, which accompanies drug-induced purpura, leukemia, bone-marrow cancer, SLE, infectious diseases and DIC; and hereditary thrombocytopenic purpura, which accompanies Wiskott-Aldrich syndrome and Fanconi syndrome.

1) Idiopathic thrombocytopenic purpura (ITP)

Idiopathic thrombocytopenic purpura (ITP) occurs in children during recovery from infectious disease; in adults it develops without any particular pathogenesis. Its main symptoms are cutaneous petechia and ecchymosis, which are followed by bleeding in the oral mucosa, nasal mucosa and gingiva; hematuria; mele- na; and menorrhagia. Splenomegaly is not found.

Platelets are destroyed as a result of production of platelet-associated IgG (PAIgG), which leads to ITP. The mechanism of production of these autoantibodies is unknown.

 decreased platelet density (100,000 per microliter or less) and an extended duration of bleeding (3 minutes or longer) are observed. PAIgG is found in the blood in more than 90% of cases. In a bone-marrow biopsy, the megakaryocyte count is found to be elevated from consumption of platelets. The coagulation system is normal; the prothrombin time (PT), activated partial thromboplastin time (APTT) and fibrinogen value are normal.

Bone-marrow biopsy and detection of PAIgG are essential for diagnosis. ITP should be differentiated from symptomatic or hereditary thrombocytopenic purpura listed below (Table 11.5). Henoch-Schönlein purpura and hemophilia are also differentiated.

**Pathology**

**Diagnosis, Differential diagnosis**

Bone-marrow biopsy and detection of PAIgG are essential for diagnosis. ITP should be differentiated from symptomatic or hereditary thrombocytopenic purpura listed below (Table 11.5). Henoch-Schönlein purpura and hemophilia are also differentiated.
from ITP. Henoch-Schönlein is distinguished by relatively localized purpura in the lower extremities that results in systemic symptoms other than cutaneous symptoms, such as arthralgia and abdominal pain; hemophilia causes deep bleeding in the joints and elsewhere.

**Treatment**

Oral steroids are the treatment of choice. Immunoglobulin is administered in large doses for severe cases. Immunosuppressants and temporary platelet transfusion are also helpful. ITP subsides in 80% of cases by drug therapy. For chronic cases that do not respond to these treatments, splenectomy may be performed.

2) **Symptomatic thrombocytopenic purpura**

Drugs, leukemia, metastasis of a malignant tumor in the bone marrow, SLE, viral infectious diseases, or vasculitis (Kasabach-Merritt syndrome; see Chapter 21) cause reduced production and enhanced consumption of platelets, leading to decrease of platelets and the onset of purpura (Table 11.5).

3) **Hereditary thrombocytopenic purpura**

Wiskott-Aldrich syndrome (Chapter 7) and Fanconi anemia (congenital aplastic anemia accompanied by malformation) are classified as hereditary thrombocytopenic purpura.

### Table 11.5 Causes of symptomatic thrombocytopenic purpura.

<table>
<thead>
<tr>
<th>Decreased productivity of platelets</th>
<th>Aplastic anemia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disseminated intravascular coagulation (DIC)</td>
<td>Thrombotic thrombocytopenic purpura (TTP)</td>
</tr>
<tr>
<td>Viral infection</td>
<td>Hemolytic-uremic syndrome (HUS)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Causative therapy</th>
<th>Drugs, radiotherapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enhanced consumption and destruction of platelets</td>
<td>Drugs, blood transfusion</td>
</tr>
</tbody>
</table>

### Clinical features

Cryoglobulinemic purpura most frequently occurs in middle-aged women. The purpura may be accompanied by petechia, hemorrhagic papules, ecchymosis and subcutaneous nodules. There may be necrosis. It occurs mostly on the lower legs, and may spread to the thighs, lumbar region, and lower abdomen; it seldom appears on the trunk or face. Raynaud’s phenomenon is found in fingers. The disease typically causes renal symptoms such as proteinuria, erythrocyturia, nephrosis syndrome, high blood pressure, acute or chronic renal failure, arthralgia, liver disorder and polyneuritis.

### Pathogenesis

Cryoglobulinemic purpura is caused by an increase in the level of cryoglobulins in the blood. Infectious diseases (viral hepatitis in particular), multiple myeloma, macroglobulinemia, collagen diseases (e.g., SLE, rheumatoid arthritis) or malignant tumors may occur as complications. The elevated blood viscosity results in inadequate blood flow and in deposition of protein on the blood vessel walls to induce necrotizing vasculitis, which causes purpura. Cryoglobulinemic purpura may appear without a primary
Necrotizing vasculitis caused by cryoglobulin embolization in the lesion is found in the skin lesions of cryoglobulinemic purpura. However, when type I cryoglobulin is involved in the onset, vasculitis tends not to occur. Cryoglobulinemic purpura is characterized by membranoproliferative glomerulonephritis in the kidney.

**Laboratory findings**

Cryoglobulin is detected by processing the blood at 37 °C to isolate the serum. Rheumatoid factor and HBs antigen are often positive, and hypocomplementemia and hepatitis C virus antibodies may be found. M proteins in the serum and Bence-Jones proteins in the urine protein electrophoretogram may be observed.

**Treatment**

Systemic administration of steroids and removal and inhibited reproduction of cryoglobulin by plasma exchange therapy are the main treatments. Any underlying diseases are treated.

**3. Pigmented purpuric dermatoses**

**Synonyms:** Idiopathic pigmentary purpura, Purpura simplex, Chronic capillaritis, Purpura pigmentosa chronica

**Clinical features, Classification**

Pigmented purpuric dermatoses occur most frequently in the lower extremities of middle-aged men. They are characterized by orange/brown pigmentation (from hemosiderin deposition) that

**Table 11.6 Differential diagnosis of pigmented purpuric dermatoses.**

<table>
<thead>
<tr>
<th></th>
<th>Schamberg's disease</th>
<th>Majocchi’s disease</th>
<th>Pigmented purpuric lichenoid dermatosis of Gougerot-Blum</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex distribution</td>
<td>More common in men</td>
<td>Slightly more common in women</td>
<td>Slightly more common in men</td>
</tr>
<tr>
<td>Age of onset (yrs.)</td>
<td>20 to 59</td>
<td>30 to 49</td>
<td>30 to 49</td>
</tr>
<tr>
<td>Rate of Onset</td>
<td>Gradual</td>
<td>Gradual</td>
<td>Rather acute</td>
</tr>
<tr>
<td>Most common site</td>
<td>Lower legs</td>
<td>Lower legs</td>
<td>Upper and lower extremities, trunk</td>
</tr>
<tr>
<td>Symptom</td>
<td>Irregular coalescence of petechiae</td>
<td>Enlargement of petechiae, formation of circular lesion</td>
<td>Hemorrhagic brown papules, aggregated coalescence of lichenified papules</td>
</tr>
<tr>
<td>Venous congestion</td>
<td>+</td>
<td>+ (sporadic)</td>
<td>–</td>
</tr>
<tr>
<td>Pigmentation</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Itching</td>
<td>–/+ (mild)</td>
<td>–</td>
<td>+</td>
</tr>
</tbody>
</table>

Clinical images are available in hardcopy only.
occurs after the onset of petechia. Pigmentation shows both clinical and histological features. There are no significant systemic symptoms (Fig. 11.18). These dermatoses are classified by distribution of eruptions into three main types: Majocchi’s disease, Schamberg’s disease, and pigmented purpuric lichenoid dermatosis of Gougerot-Blum. However, the main three types are the same in essential (Table 11.6).

**Pathogenesis**

The etiology is unknown; however, there may be the involvement of venous circulatory disorder, focal infection, or drug-induced factor.

**Pathology**

Lymphocytic infiltration, vascular dilatation, and bleeding are found in the perivascular area in the upper dermal layer. Idiopathic pigmentary purpura is chronic hemorrhagic inflammation. Hemosiderin deposits are seen in old lesions (Fig. 11.19).

**Treatment**

Topical application of steroids and bed rest with the lower extremities raised is the main treatment. Agents to reinforce the blood vessels (e.g., vitamin C) are administered, as are hemostatic and antiplasmin agents.

### 4. Senile purpura

The vascular supporting tissues weaken from age, and purpura is easily caused even by stimulation so light the patient can scarcely feel it. Senile purpura occurs mostly in the dorsal hands and the extensor surface of forearms, producing sharply marginated subcutaneous hemorrhagic spots.

### 5. Steroid purpura

When the vascular supporting tissues are weakened by prolonged topical or oral use of steroids, the capillary blood vessels are readily broken by mechanical stimulation, leading to purpura (Fig. 11.20). Steroid purpura occurs most frequently in the elderly. Mechanical stimulation should be avoided, and steroids should be used appropriately.