Other vascular diseases

**1. Arteriosclerosis obliterans (ASO)**

**Concept**

As a result of arteriosclerosis in the extremities, distal ischemia, skin pallor, pain and ulceration occur. The patients often have diabetes and high blood pressure as primary diseases.

**Clinical features**

Symptoms caused by ischemia and poor circulation appear. The Fontaine classification of arteriosclerosis obliterans (ASO) is well known: In Grade I, a cooling sensation appears at the ends of the extremities and there is mild numbness; in Grade II, there is intermittent claudication (inability to walk more than a certain distance); in Grade III, there is pain even during bed rest; and in Grade IV, ulceration or necrosis occurs at the ends of the extremities, which are prone to ulceration. The lesion becomes enlarged and white or purplish-red from ischemia. Pulse in the peripheral arteries in the lesion is impalpable.

**Diagnosis, Differential diagnosis**

Pain and ulceration are present at the ends of the extremities in both Buerger’s disease and ASO; nevertheless, Buerger’s disease occurs mostly in young male smokers. Pulse is often palpable in Buerger’s disease (Table 11.4). Lumbar spinal canal stenosis is also known to cause intermittent claudication.

**Treatment**

Conservative therapies using vasodilators and thrombosis inhibitors, and physical therapies such as exercise and hot spring bathing are the main treatments for mild symptoms of Fontaine Grades I and II. For severe cases of ASO, surgical treatments such as stent placement and revascularization are performed. Amputation of the extremities may be necessary at the terminal stages.

**2. Diabetic gangrene**

With a microvascular disorder or arterial sclerosis as the primary disease, ulcers form in toes, soles and fingers. They are accompanied by sharp pain (Chapter 17).

**3. Raynaud’s phenomenon**

**Synonym:** Raynaud’s disease

**Definition**

Fingers and toes suddenly become bluish. After several min-
utes, they progress to a purplish-dark-blue hue from cyanosis and then return to normal color through a diffuse flushing phase. Raynaud’s phenomenon may be induced by frigidity without a primary disease (primary Raynaud’s phenomenon), or it may accompany a primary disease, such as a collagen disease, particularly systemic sclerosis (Raynaud’s syndrome, secondary Raynaud’s phenomenon).

**Clinical features**

Fingers and toes become white and there is a cool sensation, sharp pain, numbness, edematous sensation, and hypoesthesia (Fig. 11.21). Cyanosis heals with a diffuse flushing and burning sensation. The ends of the extremities are constantly cold, with or without an attack.

**Pathogenesis**

The potential causes of Raynaud’s phenomenon are shown in Table 11.7. Circulatory disorders are closely associated with it. A bluish tinge is caused by reduced blood flow resulting from arterial constriction. Cyanosis results from dilation of capillaries or small veins and stasis. Diffuse flushing occurs as reactive congestion.

**Examination**

Antinuclear antibody test is performed to rule out the involvement of secondary Raynaud’s phenomenon. Other useful tests for Raynaud’s phenomenon are a cold provocation test (soaking fingers and toes in 4°C water for 10 seconds) and thermography.

**Treatment**

Causative factors should be eliminated, and the body must be kept warm. A vasodilator (calcium channel antagonists are the first choice), prostaglandins, or sympatholytic agents are applied. Smoking cessation is effective.

---

**Table 11.7 Causes of Raynaud’s phenomenon.**

<table>
<thead>
<tr>
<th>Disease, Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Primary Raynaud’s phenomenon</strong></td>
</tr>
<tr>
<td>Raynaud’s disease</td>
</tr>
<tr>
<td><strong>Secondary Raynaud’s phenomenon</strong></td>
</tr>
<tr>
<td>Physical stimulation in people who work with vibrating machinery (vibration syndrome or vibration white finger), pianists, typists, and food industry workers (meat industry, fresh fish industry)</td>
</tr>
<tr>
<td>Drugs: ergotamine, tryptamine, β-blockers, oral contraceptives</td>
</tr>
<tr>
<td>Collagen diseases: e.g., mixed connective tissue disease (MCTD), systemic sclerosis (SSc)</td>
</tr>
<tr>
<td>Blood disease: cryoglobulinemia, cold agglutinin disease</td>
</tr>
<tr>
<td>Neurovascular disorders: arteriosclerosis, Buerger’s disease, thromboembolism, thoracic outlet syndrome</td>
</tr>
<tr>
<td>Other diseases: malignant tumor, hypothyroidism</td>
</tr>
</tbody>
</table>

---

**4. Stasis dermatitis**

**Synonym:** Chronic venous insufficiency (CVI)

**Clinical features**

Stasis dermatitis usually occurs in the medial aspects of the lower legs or ankles of obese elderly women, but it may be widespread. It may develop as a complication of impaired venous return from the lower legs. Superficial varicose veins are a frequent predisposing factor. These lesions are often seen around chronic stasis ulcers, and they are itchy, scaly, often swollen, and hyperpigmented. Superficial veins of the lower legs including the greater saphenous veins and lesser saphenous veins enlarge in hose shape, nodular shape, or saclike shape and take on a serpentine appearance. The skin surface appears dark blue (varicose veins in lower legs, Fig. 11.22). As the varicosity progresses, subjective symptoms appear. Fatigue of the lower extremities,
sharp pain, edema, pigmentation, eczematous lesions and skin induration (sclerosing panniculitis) occur. In the last stages, a minor external injury in the lower legs may induce ulceration.

Stasis dermatitis shows the epithelial changes of spongiotic dermatitis and a characteristic lobular pattern of superficial and/or deep dermal neovascularization.

**Pathogenesis**

The pressure in the superficial veins is elevated, and veins are dilated and serpentine from congenital fragility of the venous walls and venous peripheral supporting tissues, stenosis and obstruction of deep veins, hyperplasia of superficial veins, pregnancy, standing for long periods, and venous valve dysfunction.

**Diagnosis**

Stasis syndrome is easily diagnosed by varicosity. History-taking on occupation and pregnancy is helpful. When surgical treatment is conducted, venography is also performed.

**Treatment**

Standing or walking for long periods should be avoided. The patients should be protected from extrinsic injury and infection, and they should keep the lower extremities elevated and use elastic bandages. When the condition progresses to stasis syndrome, besides these treatments, topical steroids, oral antihistamines, and vasodilator drugs are administered. Surgical therapy including removal of the superficial vein, ligation and sclerotherapy may also be performed.

**5. Livedo reticularis**

Livedo reticularis is a generic term for purplish-red to purplish-black discoloration with a characteristic network pattern caused by stenosis in the veins of the dermis and subcutaneous fat tissue junctions (Fig. 11.23). Although it can be divided into several subtypes, the classification is not standardized. The color deepens with exposure to cold. The condition is largely divided

<table>
<thead>
<tr>
<th>5. Livedo (reticularis)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cause</strong></td>
</tr>
<tr>
<td>Physiological</td>
</tr>
<tr>
<td>Secondary</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Unknown</td>
</tr>
</tbody>
</table>
into cutis marmorata, in which there is no particular primary disease, and secondary livedo reticularis, in which there is a primary disease such as collagen disease. The former transiently appears in infants and adult women without subjective symptoms and disappears when the body warms. The sympathetic nerve is thought to be involved. Livedo that is found at birth and that is often accompanied by combined malformation is thought to be caused by angiomatous abnormality; this condition is called cutis marmorata telangiectasia congenita (Chapter 21). When cutis marmorata occurs intermittently, the involvement of underlying diseases such as blockage of vascular cavity, circulatory disorder or vascular disorder is suspected (Table 11.8).

6. Erythromelalgia

Synonym: Erythralgia

Erythromelalgia occurs in the extremities. It is characterized by three main symptoms: paroxysmal burning sensation, flushing and skin temperature increase. The pathogenesis is unknown. It is extremely painful and tends to appear when the body is warmed by exercise or bathing. It may also be caused by a primary disease such as erythrocytosis, primary thrombocytosis, or collagen disease.

7. Lymphangitis

**Concept, Pathogenesis**

Lymphangitis is an inflammatory change in the lymphatic vessels, usually those in the extremities. The spread of various infections (e.g., various secondary infections, cellulitis, trichophytic infection), malignant tumor (e.g., breast cancer), and parasitic infestation (e.g., filariasis) are known to cause lymphangitis.

**Clinical features**

Painful, linear, soft, palpable, cord-like reddening accompanied by tenderness occurs in the primary lesion and the nerve area. Systemic symptoms such as fever (may reach 40°C), fatigue, and poor appetite are seen in many cases. As chronic lymphangitis recurs, lymphatic vessels become obstructed, which may result in lymphatic edema or elephantiasis.

**Treatment**

Antibiotics and analgesic drugs should be promptly applied systemically. In the event that an abscess forms, an incision may be required.

8. Lymphedema

Lymph fluid volume increases locally from lymphatic vessel dysfunction. Soft edema is often produced in the lower extremities and the external genitalia.
and it gradually moves upwards. Soft tissues become fibrotic or stiff. Papillary thickening of the epidermis and follicular dilation are seen in the terminal stages (Figs. 11.24-1 and 11.24-2). The causes of lymphatic vessel dysfunction are largely divided into congenital (e.g., hypoplasia of lymphatic vessels) and acquired (e.g., metastasis of a tumor into a lymph node, lymph node dissection, filariasis, deep-lying thrombophlebitis).

### 9. Ataxia telangiectasia (AT)

**Synonym: Louis-Bar syndrome**

**Outline**

- AT is an autosomal recessively inherited disease involving functional abnormality of the DNA repair mechanism.
- The three main symptoms are progressive cerebellar ataxia, telangiectasia in the auricular region and bulbar conjunctiva, and T-cell immunodeficiency.
- IgA deficiency and high α-fetoprotein level are found in the serum.

**Clinical features**

Telangiectasia in the skin and bulbar conjunctiva occurs in patients 4 to 6 years old, and it extends to the auricular region, eyelids, cheeks and extremities. Ataxia telangiectasia (AT) may produce lesions that resemble xeroderma pigmentosum. As the patients age, severe bronchiectasis and malignant lymphoma occur as complications.

**Pathogenesis**

Disconnection or translocation of the AT gene on chromosome 11 (11q22.3) influences T cells and the Ig gene region, leading to T-cell immunodeficiency and decreased production of immunoglobulin.

**Pathology**

Peripheral blood T cells are reduced in number and function, and serum IgA and IgE (sometimes IgG2 and IgG4) are absent or markedly reduced. The serum α-fetoprotein value is elevated. Brain CT and MRI show significant atrophy in the cerebellar vermis. Histopathologically, denaturation of Purkinje cells is observed.

**Diagnosis**

AT is diagnosed by the pathological symptoms and laboratory findings. Diagnosis can also be made by AT gene analysis.

**Treatment**

Symptomatic therapy is the main treatment.