REVIEW ARTICLE

Vascular Tumours of the Skin

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THE subject of vascular tumours as related to the skin can be a maze of contradictory terms, confusing titles and considerable tension to the physician attempting to attach a "pigeonhole" diagnosis to a vascular skin lesion.

The following classification is offered as a guide in the situation and is followed by comments on the individual disorders. To help resolve some of the confusion, the alternative names attached to these diseases are included in parentheses. To denote the frequency with which the various lesions are encountered, the following symbols are used:

[C] Commonly seen tumours.
[O] Occasionally seen tumours.
[R] Rarely seen tumours.

In most instances, the more rarely seen entities will be discussed at greater length because practising physicians may be less familiar with such lesions.

CLASSIFICATION:

I. TELANGIECTASIA

(1) Arterial spider [C]
(2) Venous star [C]
(3) Senile angioma [C]
(4) Venous lake [C]
(5) Petechial angiomata [O]
(6) Angioma serpiginosum [O]
(7) Hereditary hemorrhagic telangiectasia [R]
(8) Telangiectasia macularis eruptiva perstans [R]
(9) Ataxia telangiectasia [R]
(10) Microtelangiectasis essential progressive unilateral [R]
(11) Thrombosed capillary aneurysm [R]

II. ANGIOMATA

A *Hemangiomata*

(1) Nevus flammeus [C]
variants:
(a) Sturge-Weber syndrome [R]
(b) Hypertrophic angiectasis of Klippel-Trenaunay-Parkes Weber [R]
(c) Brushfield-Wyatt syndrome [R]
(2) Nevus vasculosus [C]
variants:
(a) Granuloma pyogenicum [C]
(b) Thalidomide hemangioma [R]
(3) Cavernous hemangioma [C]
variants:
(a) Maffucci's syndrome [R]
(b) Aldrich's syndrome [R]
(c) Kast's syndrome [R]
(d) Hemangioma associated with retrolental fibroplasia [R]
(e) Von Hippel-Lindau's syndrome [R]
(f) Blue rubber bleb nevus syndrome [R]
(g) Gorham's disease [R]
(h) Kassabach-Merritt syndrome [R]
(i) Riley-Smith syndrome [R]

B *Lymphangiomata*

(1) Lymphangioma circumscriptum [R]
(2) Lymphangioma cavernosum [R]
(3) Cystic hygroma [R]
(4) Lymphangiosarcoma [R]

III. MIXED VASCULAR TUMOURS

(1) *Angiokeratomata*
(a) Angiokeratoma scroti of Fordyce [C]
(b) Angiokeratoma of Mibelli [O]
(c) Nevoid angiokeratoma of Fabry [R]
(d) Diffuse angiokeratoma of Fabry [R]
(2) Sclerosing hemangioma [C]
(3) Multiple idiopathic hemorrhagic sarcoma [O]
(4) Angioleiomyoma [O]
(5) Hemangioendothelioma
(a) Benign [R]
(b) Malignant [R]
(6) Hemangiopericytoma [R]
(7) Glomus tumour [R]
(8) Angiosarcoma [R]

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(9) Hemolymphangioma [R]
(10) Lymphangioendothelioma [R]

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I. TELANGIECTASIA (dilatation of previously existing terminal small vessels)

(1) *Arterial Spider* (Nevus araneus, stellar nevus, spider nevus, telangiectasia)

This lesion is most commonly seen on the face, neck and upper chest. It is an arborescent erythematous macule with its branches radiating out from a central red dot much like a spider's legs. It blanches with pressure. Often there is an increase in skin surface temperature in an involved area owing to increased blood flow. Spider nevi may be seen in normal people as well as in the following states: pregnancy, liver disease and hyperestrogenic states, during steroid treatment, in Cushing's syndrome, vitamin deficiency, rheumatic fever and cardiac disease, chronic rheumatoid arthritis, lupus erythematosus, polymyositis, scleroderma, periarteritis nodosa, syphilis, xeroderma pigmentosum, radiodermatitis, keloid, adenoma sebaceum, rosacea, hyperthyroidism, arteriosclerosis, polycythemia, metastatic carcinoid syndrome, cutis marmorata, erythema ab igne, livedo racemosa, livedo reticularis, Bloom's syndrome and Cockayne's syndrome.

Microscopic examination reveals a central arteriole with a muscular wall which widens into a subepidermal ampulla. Capillaries branch out from this centre.

This lesion may be left alone or lightly electrodesiccated. Management of the underlying disease process (when present) may induce regression of some lesions.

(2) *Venous Star*

This red 1-2 mm. nodule is caused by increased venous pressure. It appears on the lower extremities overlying the larger veins and is similar in structure to an arterial spider. It is obliterated by pressure. No associated underlying pathology is found.

No treatment is necessary, but light electrodesiccation may be used.

(3) *Senile Angioma* (Campbell de Morgan spot, ruby spot, cherry angioma, capillary angioma, senile hemangioma)

This lesion is due to weakening of the capillary wall with age and is seen in 75% of people over 30. It is initially a soft 1-5 mm. red papule or nodule which is sharply circumscribed and without branches. It gradually becomes more solid in consistence. The trunk is most commonly involved, but the lesions may be seen elsewhere. They are obliterated by pressure. No associated underlying pathology is found.

Microscopic examination in the early stage reveals numerous newly formed capillaries; later these capillaries dilate.

No treatment is necessary, but light electrodesiccation may be used.

(4) *Venous Lake*

This lesion is due to weakening of the venule wall with age. It is a 1-5 mm. blue nodule and occurs primarily on the face and ears. Ninety-five per cent of these lesions are seen in men. They may obliterate with pressure. No associated underlying pathology is found.

Microscopic examination reveals dilated venules.

No treatment is necessary, but light electrodesiccation may be used.

(5) *Petechial Angiomata*

These are multiple petechiae in an adult with no associated pathology or bleeding tendency. They are due to capillary wall dilatation similar to senile angiomata, but the lesions are smaller and clinically resemble petechiae. They are obliterated by pressure.

They may be removed by light electrodesiccation.

(6) *Angioma Serpiginosum* (essential telangiectasia)

This eruption of asymptomatic red punctate macules may show linear, mottled or netlike patterns. Peripheral extension may eventually result in a serpiginous outline. Women are affected more commonly than men, and the lower legs are the site most frequently involved. The oral mucosa is unaffected. They partially blanch with pressure.

Microscopically there are dilated and tortuous capillaries in the papillary and subpapillary regions of the dermis.

Pressure gradient supportive hose and/or light electrodesiccation may be used.

(7) *Hereditary Hemorrhagic Telangiectasia* (Rendu-Osler-Weber disease, Osler's familial hereditary telangiectasia)

This autosomal (incomplete) dominant familial disease usually presents with frequent nose bleeds in childhood. Telangiectatic lesions of the skin develop by the third decade of life. These red macules are from a pin point to 3 mm. in diameter and are sharply outlined. They may

appear on any portion of the body and are commonly seen on mucous membranes, e.g. nasal mucosa, tip of tongue, palate and lips. The distal aspect of the fingers is frequently involved. The macules usually blanch with firm pressure. The skin temperature is normal. The patient may bleed from stomach, bowel, lung, brain, bladder, kidney or liver lesions. About one-half of pulmonary arteriovenous fistulas are due to this disease. The bleeding tendency may either lessen or increase with age. There may be a severe anemia due to repeated hemorrhages, and death has been known to result from bleeding.

The microscopic picture is of capillaries and venules with thinned, ballooned (aneurysmal) walls in the papillary and subpapillary layers.

Light electrodesiccation with an electrolysis needle, using a low amperage, will obliterate the telangiectatic vessels on the skin surface. Transfusions may be necessary when there is marked or steady internal bleeding.

(8) *Telangiectasia Macularis Eruptiva Perstans*

This is an adult type of urticaria pigmentosa. Telangiectasia may be seen over the entire skin surface. One may demonstrate urticaria along the line of skin stroking (dermographism), but this is not as marked as in the other varieties of urticaria pigmentosa.

Microscopic examination, using metachromatic staining techniques, reveals numerous tissue-mast cells about cutaneous vessels.

No treatment is applicable, as the process is too diffuse to permit elimination of the telangiectatic vessels.

(9) *Ataxia-Telangiectasia* (Louis-Bar syndrome, cephalo-oculo-cutaneous telangiectasia syndrome)

This autosomal recessive familial disorder begins with cerebellar ataxia noticeable between 12 and 18 months of age. At 4 to 5 years of age, telangiectasia is first noted on the bulbar conjunctivae; then symmetrical telangiectatic lesions develop on the butterfly region of the face, about the ears, neck flexures, V of the chest, dorsa of the hands and feet, and the popliteal and cubital fossae. Peculiar eye movements are noted. These children suffer from frequent sinopulmonary infections and growth retardation. Gamma A globulin is decreased and the incidence of lymphoma and leukemia is increased. The disorder is progressive without reversibility.

Histologic studies reveal telangiectatic vessels in the subpapillary venous plexus. Venules of the leptomeninges are dilated and engorged.

Management of the disease is symptomatic, with treatment of infections as they occur.

(10) *Microtelangiectasis Essential Progressive Unilateral (Tommasi's syndrome)*

This name is given when there is a progressive unilateral development of small telangiectatic lesions in later life.

Light electrodesiccation may be used as lesions develop.

(11) *Thrombosed Capillary Aneurysm*

This is a soft, dark-blue, tender nodule which may have an erythematous or brown surrounding ring. It measures 0.5 to 1 cm. in diameter and is more often seen on the face.

The histologic appearance is of one or more dilated capillaries in the upper dermis. The lumen is occluded by a thrombus. Early thrombus organization, extravasated red cells and hemosiderin may be present.

This lesion should be excised and examined microscopically as clinically it may resemble a malignant melanoma.

II. ANGIOMATA

This term is applied to a swelling or tumour formed by dilatation and/or proliferation of new blood- or lymph-carrying vessels. They are usually congenital or developmental and are ordinarily present at birth or appear in early childhood.

A. HEMANGIOMATA

(1) *Nevus Flammeus* (port-wine stain, nevus telangiectaticus)

One or more dull pink to bluish red patches are seen, usually on the head and neck. There may be more widespread distribution and involvement of up to one-half of the body has been reported. Frequently it is unilateral, and the oral mucosa may be involved. One-half of newborns have a nuchal stain (known as angioma simplex) which is completely benign and usually fades in later life. Other port-wine stains that may fade in later life are pale lesions seen around the eyes and above the nose.

The microscopic picture is one of capillary dilatation in the dermis at any level without endothelial cell proliferation.

Various syndromes demonstrate this skin lesion; these include:

(a) *Sturge-Weber syndrome* (encephalotrigeminal angiomatosis, Sturge-Kalischer-Weber syndrome, Sturge-Weber-Dimitri syndrome, nevoid dementia).

This familial condition is inherited as an irregular autosomal dominant pattern. There is a unilateral nevus flammeus in the trigeminal region (usually of the first two branches) with ipsilateral angiomatosis of the pia mater and choroid. There is calcification of the underlying cortex beginning around the blood vessels and giving a "railroad track" appearance on roentgen examination. The neurological symptoms include convulsions, paralysis, mental retardation and visual disturbances.

Seizures usually appear first in infancy or early childhood. Paralysis develops later, with hemiplegia on the side opposite to the facial nevus. Mental retardation is present in over half the cases and is variable in degree. The visual disorders include congenital glaucoma, homonymous hemianopsia, optic atrophy and other bulbar changes.

The buccal mucosa may be involved by the hemangioma. It may be bilateral or midline; parts of the body other than the trigeminal region may be affected and there may be a cavernous hemangioma component.

Cover-up make-up may be used on the hemangioma region. The results from tattooing are unpredictable.

(b) *Hypertrophic angiectasis of Klippel-Trenaunay-Parkes Weber* (Klippel-Trenaunay osteohypertrophic varicose nevus, congenital dysplastic angiopathy, Weber-Klippel syndrome).

This condition is a nevus flammeus of an extremity with or without a cavernous hemangioma component. There is developmental hypertrophy of the underlying bone and soft tissue structures resulting in a larger, longer and warmer limb. Less often, bone hypoplasia with a resultant smaller limb develops owing to vascular congestion, disuse and pressure atrophy. Varicose veins may or may not be present.

Pressure gradient supports may be used on the involved extremity.

(c) *Brushfield-Wyatt syndrome.*

This eponym has been used when there are extensive port-wine staining of the skin, hemiplegia, mental retardation and various stigmata of degeneration. It is best included under the Sturge-Weber syndrome.

Cover-up make-up may be used over the hemangioma.

(2) *Nevus Vasculosus* (capillary hemangioma, strawberry mark, immature angioma)

This is a raised, bright red, soft and often lobulated (resembling a strawberry) tumour. Fifty per cent are present at birth and 85% are present by 1 month of age. There may be a blanched macule in the first few weeks before an angioma develops on that site. There is an initial period of growth in early infancy, reaching maximal size about the age of 6 months, and then the lesion usually involutes spontaneously over a few years. An early sign of impending involution is the development of blanched spots over the surface of the red angioma.

The histologic appearance is that of capillary dilatation and early proliferation, with fibrosis during the phase of involution.

Since most of these hemangiomata regress spontaneously, the cosmetic result is usually better without active therapy (surgery, irradiation or dry ice).

Reassurance of the parents and periodic observation of the infant is the best course to follow unless the tumour involves a vital structure or interferes with function. If complications such as infection, hemorrhage, very rapid growth or thrombocytopenia develop, surgical intervention may be contemplated.

Two variants of nevus vasculosus include:

(a) *Granuloma pyogenicum* (septic granuloma, Botryomycosis hominis)

The etiology of this condition remains unknown, but it likely represents a vasculo-proliferative response to a focus of trauma and/or infection. It is most frequently seen in children. This lesion develops rapidly from a small red macule to a 0.5-2 cm. firm, pedunculated, vascular nodule which may crust and which bleeds easily. It often develops at a site of injury, inflammation or infection. Most often it is seen on the fingers and face.

Microscopy reveals numerous newly formed, dilated capillaries and young connective tissue. In the early stage there is a sparseness of inflammatory infiltrate, but a mixed infiltrate is seen later. Moderate endothelial cell proliferation is seen. Acanthosis of the epidermis at the base gives an epidermal collarette or buttress appearance.

A variant of granuloma pyogenicum is granuloma gravidarum (pregnancy tumour) which arises on the gums during pregnancy and involutes after delivery. The histologic appearance is that of granuloma pyogenicum.

This lesion may be undercut with an iris scissors and the base desiccated. Histologic examination of the specimen should be done to rule out a neoplastic process.

(b) *Thalidomide hemangioma*

This facial hemangioma is seen in children of women who took thalidomide during pregnancy.

The moustache area is a frequent site of involvement.

Corrective plastic surgery may be helpful.

(3) *Cavernous hemangioma* (mature angioma, angioma cavernosum)

This soft pink to purple subcutaneous mass is often located on the face. The overlying skin may be normal or the site of a nevus flammeus or nevus vasculosus. Regression may be slower and less constant than in the superficial capillary lesions.

The histologic appearance is that of large, blood-filled, endothelial-lined spaces in the lower dermis. There is associated adventitial cell overgrowth.

Surgical excision may be performed on cavernous lesions which have not regressed completely.

Syndromes that demonstrate this lesion include:

(a) *Maffucci's syndrome* (Dyschondroplasia with hemangiomas syndrome)

This non-familial disorder presents with multiple cavernous hemangiomata of the skin, mucosae and internal organs. Absent at birth, the onset is before puberty. Dyschondroplasia, ossification defects and poor musculature are evident. There are 1-2 cm. nodules (phleboliths) on the small bones of the hands and feet. Other bones may be affected: the long bones, ribs, scapulae and vertebrae. The process is usually unilateral or asymmetrical. These children have normal intelligence, are short in stature and have an increased susceptibility to fractures. The disease is stationary after full growth is reached.

About 20% of patients with this disorder develop chondrosarcoma.

Pressure gradient supports may be used when an extremity is involved.

(b) *Aldrich's syndrome* (Wiskott - Aldrich syndrome)

This is a sex-linked recessive disorder thereby affecting male infants only. Giant cavernous hemangiomata may be present with a thrombocytopenic purpura. Chronic eczema, beginning at the age of 2 to 3 months, and frequent infections (furuncles, pyoderma, otitis media) are seen. These children lack natural isoagglutinins and show abnormal delayed hypersensitivity reactions. Rheumatoid factor, as well as antibodies to goat or bovine protein, may be present. The liver and spleen are usually enlarged.

The course is usually fatal by the age of 4 years in spite of splenectomy and steroids. Female relatives may carry the "thrombocytopenic trait".

No successful therapy is known.

(c) *Kast's syndrome*

This eponym has been used when the hemangiomata are associated with chondromata.

No successful therapy is known.

(d) *Hemangioma associated with retrolental fibroplasia*

This disorder occurs in premature infants one week to three months after birth. Retinal hemangiomata extend into the vitreous, with resultant cloudiness and hemorrhages. The latter may either regress or proceed to cause retinal detachment with subsequent blindness. The retrolental fibroplasia is directly related to hyperoxia (over 40% oxygen) exposure. The skin hemangiomata (usually cheeks and eyelids) are incidental in this condition.

Excision of lesions on the skin is possible if they are not too large.

(e) *Von Hippel-Lindau syndrome* (angiomatosis retinae et cerebelli syndrome)

This disorder is transmitted as an autosomal dominant trait. It is characterized by hemangiomata of the retina and cerebellum, tumours, and cysts of various organs (including the medulla, spinal cord, liver and kidney) and, rarely, hemangiomata of the face. Retinal lesions develop about the age of 25, and cerebellar symptoms begin after 30 years of age.

Skin lesions when present may be excised.

(f) *Blue rubber bleb nevus syndrome*

This autosomal dominant abnormality presents with soft blue swellings which resemble phlebectasia in appearance and which refill after release of compression. They are variable in number and are .1 to 4 cm. in diameter. These lesions are present at birth or appear in childhood and are found primarily on the trunk and arms. Many of them are tender on palpation and distinctively are the source of spontaneous nocturnal pain. This pain may not occur until after puberty and may be due to smooth muscle contracture in the actual lesions. The attacks of pain often initiate sweating over the lesions. There are multiple cavernous hemangiomata on the mucosa of the gastrointestinal tract, particularly of the small intestines, which can be the source of hemorrhage.

No satisfactory treatment is available.

(g) *Gorham's disease* (massive osteolysis, phantom bones, disappearing bones)

This disorder is a replacement of bone by hemangiomata, with resultant bone dissolution and fibrotic replacement. Cutaneous hemangiomata may be present. The osteolysis may be due to the local hyperemic state induced by the presence of hemangiomata. The process is self-limited.

No satisfactory treatment is available.

(h) *Kassabach-Merritt syndrome*

This eponym has been applied when thrombocytopenia and bleeding tendencies are found in association with hemangiomata. The falling platelet count is related to the size of the vascular tumours.

Radiation of the hemangiomata may be necessary and fresh whole-blood transfusions may be required.

(i) *Riley-Smith syndrome*

This disorder includes macrocephaly without hydrocephalus, pseudopapilledema and multiple cutaneous and subcutaneous cavernous hemangiomata.

No satisfactory treatment is available.

B. LYMPHANGIOMATA

(1) *Lymphangioma circumscriptum* (superficial)

This eruption is composed of grouped small thick-walled vesicles which may resemble frog spawn and is usually a pale skin colour. It may be dark blue if there is mingling of small blood vessels. The surface may be verrucous. It becomes prominent at puberty and is usually localized to one region. The sites of predilection are the thighs, upper arms, axillae and oral mucosa.

The microscopic picture is that of dilated lymphatic vessels in the upper dermis.

If it is small enough, the lesion may be excised. If larger, full-thickness skin grafting may be necessary.

(2) *Lymphangioma cavernosum* (deep)

The enlargement of the affected region in this case is more diffuse, owing to formation of large cysts. When the oral mucosa is involved, macrochilia or macroglossia is seen. The trunk or distal extremities may be involved.

The histologic appearance is that of dilated cyst-like lymph spaces in the dermis and subcutaneous tissue, together with connective tissue hypertrophy.

Treatment is generally unsatisfactory; the lesions are not usually susceptible to radiotherapy.

(3) *Cystic Hygroma*

The malformed lymphatic channels form blind loops, resulting in multiloculated cysts which present as a swelling or tumour of the neck. This is a variant of lymphangioma cavernosum.

Surgical excision is the treatment of choice; these tumours are usually resistant to radiotherapy.

(4) *Lymphangiosarcoma* (Stewart-Treves syndrome)

This is a lymphangiosarcoma of the upper extremity following chronic lymphedema and is usually seen following an ipsilateral mastectomy. There may be metastases to the lung. The histologic appearance is that of endothelial cell proliferation with cells that are highly atypical.

Surgical removal of the affected region is possible if detected early.

III. MIXED VASCULAR TUMOURS

(1) *Angiokeratomata*

(a) *Angiokeratoma scroti of Fordyce*

These common dark red papules, 1 to 5 mm. in diameter, are found on the scrotum in the elderly. The penis is not involved (in contradistinction to diffuse angiokeratoma of Fabry). They are usually not hyperkeratotic and are likely more closely related to senile angioma than angiokeratoma. Venous occlusive disorders in the region have been implicated, such as varicocele, tumour of the epididymis, hernia, hernioplasty, etc.

These lesions do not require treatment.

(b) *Angiokeratoma of Mibelli*

These small (1 to 3 mm.) vascular papules begin in childhood or adolescence and are seen in both sexes. They are numerous, dark red, and situated over bony protuberances (dorsa of fingers and toes, elbows and knees). Later they darken, become more verrucous in character and enlarge to 6 to 8 mm. New lesions may develop after exposure to cold.

Microscopic examination reveals dilated capillaries in the upper dermis with epidermal hyperkeratosis and papillomatosis.

The lesions may be undercut and the bases electrodesiccated.

(c) *Nevoid Angiokeratoma of Fabry* (angiokeratoma neviforme, angiokeratoma circumscriptum)

These vascular papules are seen early in life and in both sexes. They usually are localized to one lower extremity. They may become large and more keratotic with age.

Histologically they resemble angiokeratoma of Mibelli except that the rete ridges are not as elongated.

The lesions may be undercut or left alone.

(d) *Diffuse Angiokeratoma of Fabry* (angiokeratoma corporis diffusum)

This sex-linked recessive disorder (essentially limited to males) is actually a lipoid storage disease rather than a hemangiomatous process. The dermal, cardiovascular, ocular, renal and pulmonary systems are involved. It is important to differentiate this disease from the other (benign) angiokeratomas because of its poor prognosis.

There are three phases to this disorder. In childhood (8 to 12 years), multiple red macules appear on the trunk and genitalia. They are 1 to 8 mm. in diameter, and are red to black in colour. They may be slightly hyperkeratotic and may blanch with pressure. Other sites may be involved, including the labial and buccal mucosa. Ocular manifestations include aneurysmal dilatations of the conjunctival and retinal vessels. Corneal opacities which do not impair vision are present; these are also seen in female carriers of the disorder who are otherwise unaffected and who can be identified by slit lamp examination. The child has episodic crises of fever, abdominal pain and burning pains in his hands. Lipid granules are found in the urine.

The second phase occurs between the ages of 20 and 30 years. The crises of childhood are fewer, but the skin lesions are more extensive. The renal function deteriorates. Because of diminished sweating ability, these patients cannot adjust well to changes in temperature.

The third phase occurs at about 40 years of age with further diminution of renal function (low fixed specific gravity of urine, presence of casts and lipids), renal hypertension, cardiomegaly, congestive heart failure and retinopathy. Patients die usually by the early forties from uremia, cerebrovascular accident, coronary thrombosis or cardiac failure.

Histologically there is a deposition of a PAS-positive, Sudan-positive, birefringent glycolipid (ceramidetrihexoside) in the blood-vessel walls of the involved organs.

The management of renal function is most important in this condition. Skin lesions may be left alone, or some may be undercut and the bases electrodesiccated.

(2) *Sclerosing Hemangioma* (dermatofibroma, histiocytoma, nodular subepidermal fibrosis)

These are single or multiple firm nodules. They are most commonly located on the extremities, and usually measure less than 1 cm. in diameter. They may be red, yellow or brown in colour. Their histogenesis remains unknown; the view most widely held is that they represent a post-inflammatory fibrotic reaction but other authorities claim that they are benign histiocytic tumours. It is generally agreed that the vascular element is incidental.

Histological examination reveals many spindle-shaped cells in young collagen. Most lesions show scattered small capillaries with prominent endothelial cells. Many have acanthosis of the overlying epidermis. Lipid or hemosiderin may be seen in the histiocytic cells.

Characteristic lesions may be left alone. If there is doubt about the diagnosis, they should be excised and examined microscopically.

(3) *Multiple Idiopathic Hemorrhagic Sarcoma* (sarcoma of Kaposi, multiple pigmented hemorrhagic sarcoma)

This disorder is usually seen in middle-aged or older men of Central European descent. Lesions are usually found on the legs but may be elsewhere. They begin as blue-purple macules which become nodular and coalesce into plaques which may ulcerate, hemorrhage and crust. They are firm (occasionally cystic) and tender. There may be telangiectasia of the surrounding skin.

There may be associated lymphedema, stasis changes, and verrucous thickening of the feet. Ten per cent show systemic involvement (gastrointestinal tract, liver, lung, heart and abdominal lymph nodes). In these cases the disorder is multicentric rather than metastatic. Rarely is visceral involvement found without cutaneous lesions. Sarcomatous change may develop.

Microscopic features in the early stages are a chronic inflammatory or granulomatous appearance with dilated new blood and lymph vessels and edema, hemorrhage, hemosiderin deposits and a dense mononuclear infiltrate. Later there is fibrous connective tissue proliferation.

Pressure gradient supports, excision and microscopic examination of localized lesions, and irradiation are useful palliative forms of therapy.

(4) *Angioleiomyoma*

These are usually tender nodules located intracutaneously and subcutaneously, most commonly on the legs.

Microscopically the lesions are similar in structure to pure leiomyomas, but are encapsulated and contain numerous blood vessels.

Excision and microscopic examination are called for.

(5) *Hemangioendothelioma*

(a) *Benign.*—Most are congenital or appear in early life. They are firm red plaques and have a predilection for the scalp.

Microscopically they are similar to a nevus vasculosus with capillary dilatation, but with large multilayered endothelial cells.

These lesions should be excised and examined microscopically to rule out a neoplastic process.

(b) *Malignant.*—This condition is seen mostly in the elderly and usually involves the face and scalp. The lesion is a raised, soft, dark red tumour and is highly malignant.

Microscopically there are numerous vascular lumina, lined by large atypical endothelial cells, some of which are multinucleated. Numerous mitotic figures are present. Foot's silver stain shows that this cell proliferation lies between the vascular lumina and the surrounding reticulum fibres.

Wide excision should be done, and the tissue should be submitted to microscopic examination.

(6) *Hemangiopericytoma*

This firm, nodular, sometimes painful tumour is usually benign and occurs in the skin and subcutaneous tissue. About 20% become malignant and metastasize.

Microscopic examination shows proliferation of pericytes (contractile cells, glomus cells) surrounding the capillary lumina. When Foot's silver stain is used, these cells are found to lie outside the reticulum fibres. This feature helps to differentiate a hemangiopericytoma from a hemangioendothelioma.

Wide excision is indicated, along with microscopic examination of the tumour.

(7) *Glomus Tumour*

Glomus tumours occur as painful single or non-tender multiple blue nodules, usually on the fingers (especially subungually). They are well localized. They are similar in nature to the hemangiopericytoma, but proliferate about the cutaneous glomus (neuromuscular organ which controls patency of the arteriovenous anastomoses of the arterial bed). When pain is present, it is due to the neural component. The solitary type is not inherited but the multiple type is dominantly transmitted.

Multiple glomus tumours may be associated with hypoplasia and osteoporosis of the bones of the affected forearm and are then known as the glomangiomatous-osseous malformation syndrome.

The microscopic picture is that of numerous small vascular lumina lined by a single layer of endothelial cells and several layers of glomus cells. Solitary lesions have a fibrous capsule which is absent from the multiple lesions. The multiple tumours are also much more vascular.

Complete excision and microscopic examination should be done. Recurrence takes place if the lesion is incompletely excised.

(8) *Angiosarcoma*

This erythematous nodule microscopically shows a spindle-cell sarcoma with vascular channels in its midst. The endothelial cells show variation in size and shape and numerous mitotic figures.

Wide excision with microscopic examination of the tissue is indicated.

(9) *Hemolymphangioma*

This term is used when the hemorrhagic pseudovesicular lesion is made up of a combination of telangiectatic blood and lymph elements.

Local excision with microscopic examination should be carried out.

(10) *Lymphangioendothelioma* (lymphangiosarcoma)

The existence of this lesion as an entity is still in dispute. Local excision with microscopic examination should be performed.