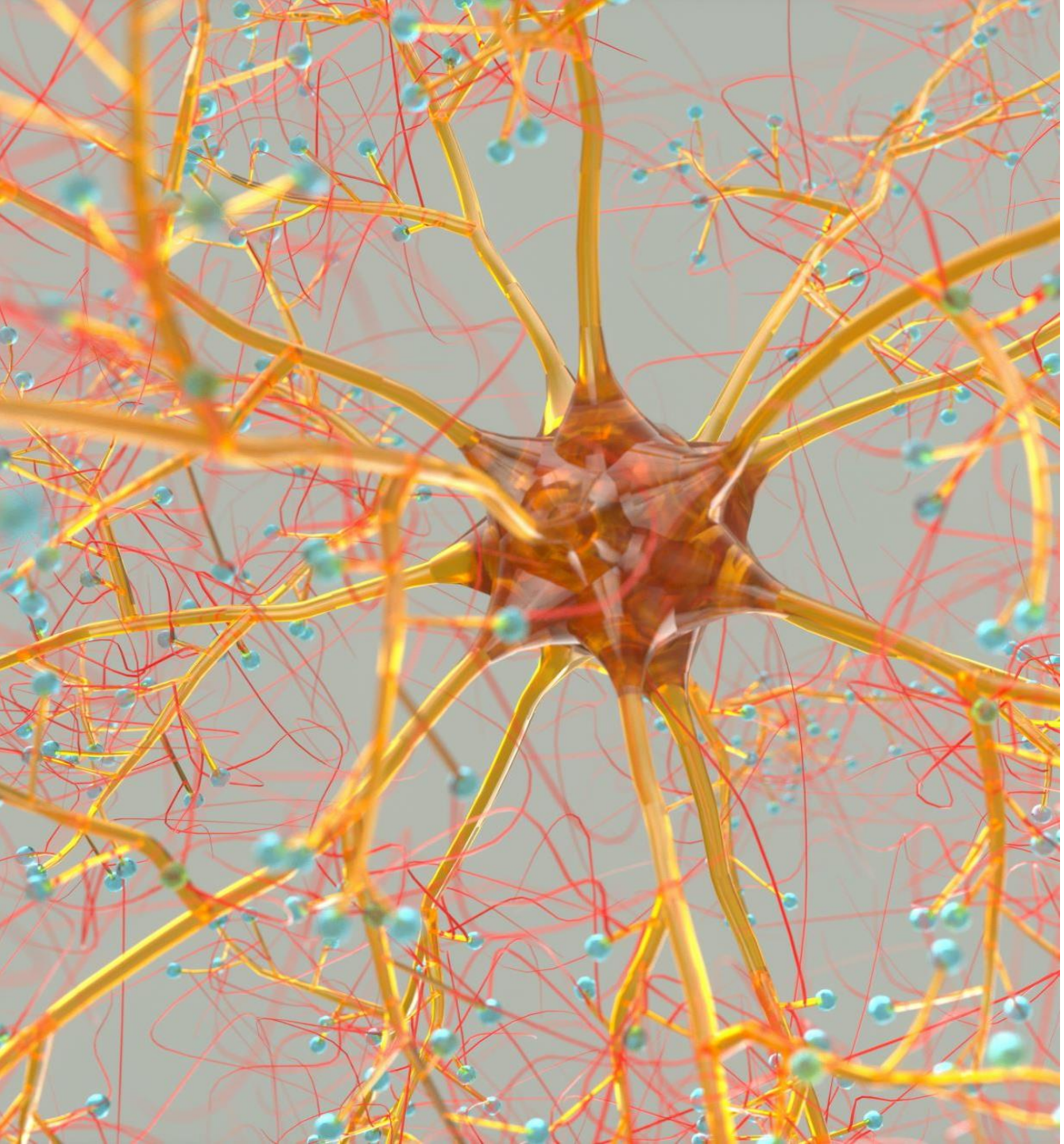


Vascular malformations

Dr. Sundus Abdull Wadood Aljazaeri[

Objectives

- Epidemiology
- Definition
- Aetiology
- Types
- Causes
- Diagnosis
- Treatments

Epidemiology

- Vascular malformations are believed to be due to errors of development of vessels that occur during the fourth and tenth weeks of intrauterine life.
- Most vascular malformations are sporadic, although several families with inherited forms have been identified.
- They are very heterogeneous and affect about 0.3% of the population.
- Vascular malformations are mostly congenital, even if they may be diagnosed only later in life.

Etiology

- Although several genes have been identified as the cause of inherited forms of vascular malformations, the etiology of the most common sporadic lesions is unknown.

- Histologically, vascular malformations consist of enlarged, tortuous vessels with quiescent endothelium.

- In contrast to hemangioma, there is neither parenchymal mass nor cellular proliferation.

Classification

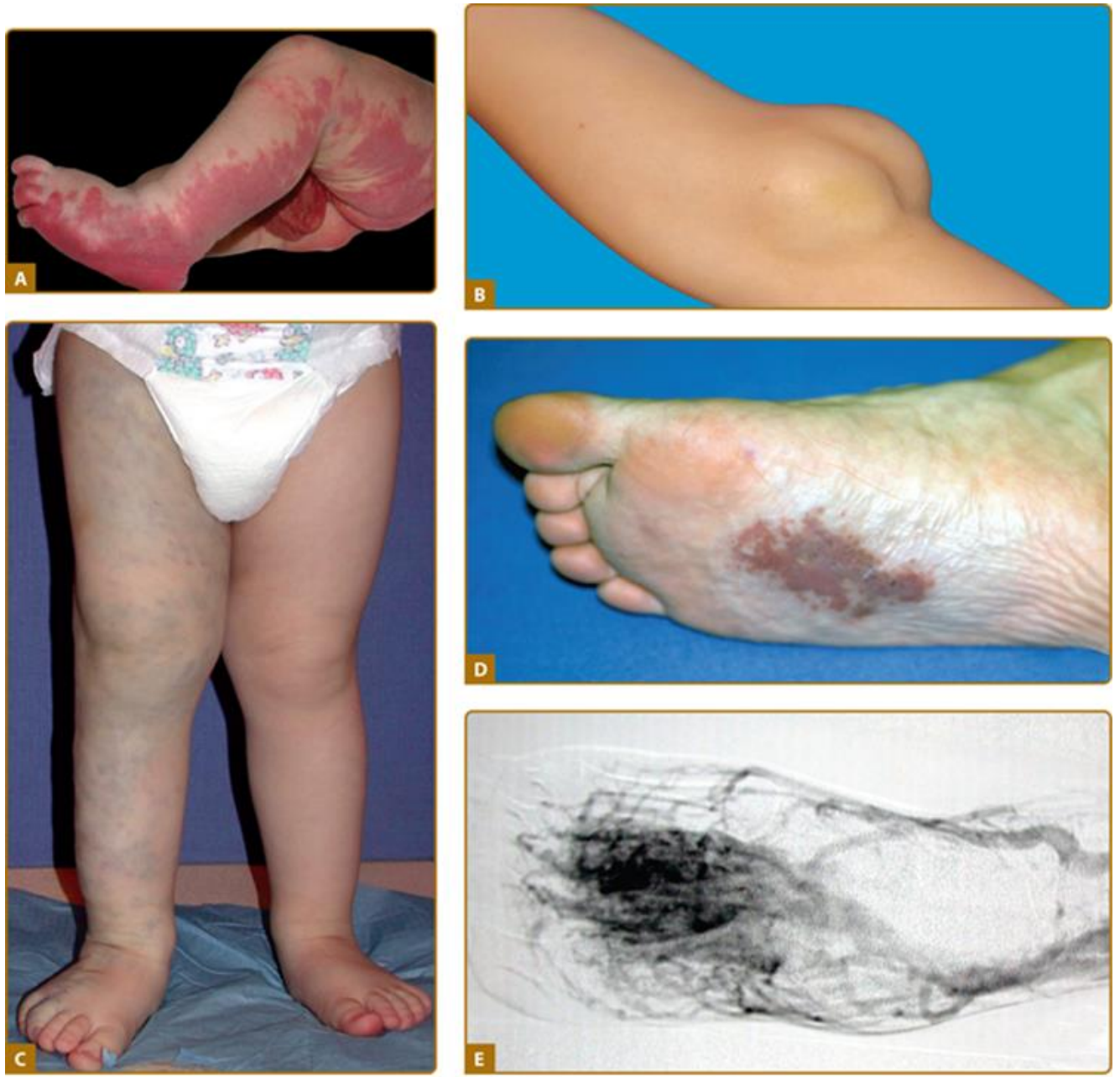
- For many years, vascular anomalies were grouped under the term ***angioma***, hampering precise classification and leading to incorrect diagnosis and improper management.
- For example, the term ***hemangioma*** has been used both for vascular malformations, often of venous type (cavernous hemangioma) as well as for vascular tumors (strawberry hemangioma).
- This nomenclature changed in 1982 with the development of a biologic classification by Mulliken and Glowacki. It divided vascular anomalies into two major categories:
- (1) vascular tumors (with cellular proliferation; hemangioma being the most common)
- (2) vascular malformations (structural anomalies of blood vessels) that are subsequently subdivided, depending on the affected vessel type, into arterial, capillary, lymphatic, or venous malformations.

	Capillary Malformation	Venous Malformation	Lymphatic Malformation	Arteriovenous Malformation
Skin color	Pinkish-red to purple	Normal-bluish-purple GVM: red (neonates) to dark-blue-purple (adult)	Normal-yellowish-purple	Normal-red
Aspect	Flat	Flat-raised	Raised-vesicular	Flat-raised
Temperature	Normal	Normal	Normal	Warm
Palpation	Normal	Compressible GVM: nodular, painful	Firm, non compressible	Thrill, bruit
Associations	Hypertrophy pyogenic granulation	Phleboliths, deformation	–	Ulceration, deformation
Radiology	No-flow	Slow-flow	Slow-flow, cyst	Fast-flow
Histology	Dilated capillaries D2–40 negative	Thin-walled venous channels Relative lack of smooth muscle cells D2–40 negative	Cystic lymphatic channels D2–40 positive	Arterialized veins D2–40 negative
Etiology	?	VMCM: TIE2 GVM: Glomulin Sporadic VM: 50% somatic TIE2 mutations	LM: ? Primary lymphedema: SOX18, VEGFR3, FOXC2, CBR2	AVM: ? CM-AVM: *RASA1* HHT: endoglin, Activin-like receptor tyrosine kinase, SMAD4 PTHS: PTEN
Treatment	Pulsed-dye laser	Sclerotherapy, surgery	Surgery, sclerotherapy	Embolization followed by surgical resection of nidus

Clinical findings

- Vascular malformations **grow proportionately with the patient**. Usually they **do not regress**.
- Most frequently, they are **well demarcated and localized** .
- In **rare instances**, they can be the **stigmata of deep lesions**.
- Vascular malformations are rheologically divided into
- *slow-flow* (capillary, lymphatic, venous, and combined)
- *fast-flow* (arterial, arteriovenous, and combined).
- **Doppler ultrasound** is the best noninvasive radiologic examination that provides clues into the differentiation of the various types.
- **Magnetic resonance imaging (MRI)** details the extension and precise location of the lesion .

Vascular malformations of various vessel types:
A. capillary malformation of left lower extremity and genitalia;
B. subcutaneous lymphatic malformation invading the cubital nerve;
C. extensive venous malformation of right lower extremity causing pain, swelling, and coagulopathy;
D. arteriovenous malformation of sole;
E. note nidus of the lesion on arteriography.



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Classification

- In 1996, this classification was adopted and further developed by the **International Society for the Study of Vascular Anomalies (ISSVA)** .
- Vascular malformations mostly affect only one vessel type, yet combined malformations also exist. They are named according to the affected vessel types, (e.g., **capillary-venous** or **venolymphatic malformation**).
- In addition to isolated forms, vascular malformations occur in syndromes such as
- Klippel–Trenaunay (KT) (capillary-lymphaticovenous malformation with limb hypertrophy),
- Maffucci syndrome (multiple enchondromas associated with multiple venous anomalies and high incidence of malignancy)
- Cloves syndrome (congenital lipomatous overgrowth with vascular malformations, epidermal nevi, and scoliosis)
- Parkes Weber syndrome (high-flow vascular malformation of the extremity with soft tissue hypertrophy).

Capillary malformation (CM)

- **Capillary malformation (CM), commonly called port-wine stain, is a slow-flow vascular malformation with an incidence of 0.3% (Fig. 1).**
- **They are located on the nape of the neck (81%), the eyelids (45%), or the glabella (33%).13 When located on the occiput, it is called Unna nevus (see Fig. 2).**
- **These patches have a much higher incidence (42%) in white infants than in black infants (31%).**
- **They are also present in various syndromes such as Beckwith–Wiedemann and Rubinstein–Taybi.**
- **These lesions disappear spontaneously around the ages of 1–4 years.**

Clinical findings

- CM is a red homogenous congenital lesion that is often unilateral, sometimes bilateral, but usually not median. CMs involve skin and subcutis, and sometimes mucosa.

- Their color varies from pinkish red to deep purple with a geographic contour or a dermatomal distribution. Lesions are flat and painless, do not bleed spontaneously, and are never warm on palpation.

- They can extend throughout the entire body. *Fifty percent of CMs are located on the face where they follow the distribution of the trigeminal nerve: ophthalmic branch V1 (front and upper eyelid), maxillary branch V2 (lower eyelid, cheek, and upper lip) (see Fig. 3B), or mandibular V3 (lower lip, chin, and mandibule).*

- In combination with another vascular malformation, CM can be part of a syndrome, such as Sturge–Weber (see Fig. 4),, phakomatosis pigmentovascularis (PPV) (see Fig. 5),, , Klippel–Trenaunay (KT) (see Fig. 6),, , Servelle–Martorell, etc.

- None of these syndromes is inherited. In rare instance, CM can be the cutaneous hallmark of occult spinal dysraphism, especially if located in the lumbosacral area.

Capillary Malformations

A. Extensive capillary malformation of upper extremity.
B. Histologically, capillary malformation is characterized by dilated capillaries of normal number on the papillary and upper reticular dermis in combination with areas of increased number of normal-looking capillaries (hematoxylin and eosin staining).

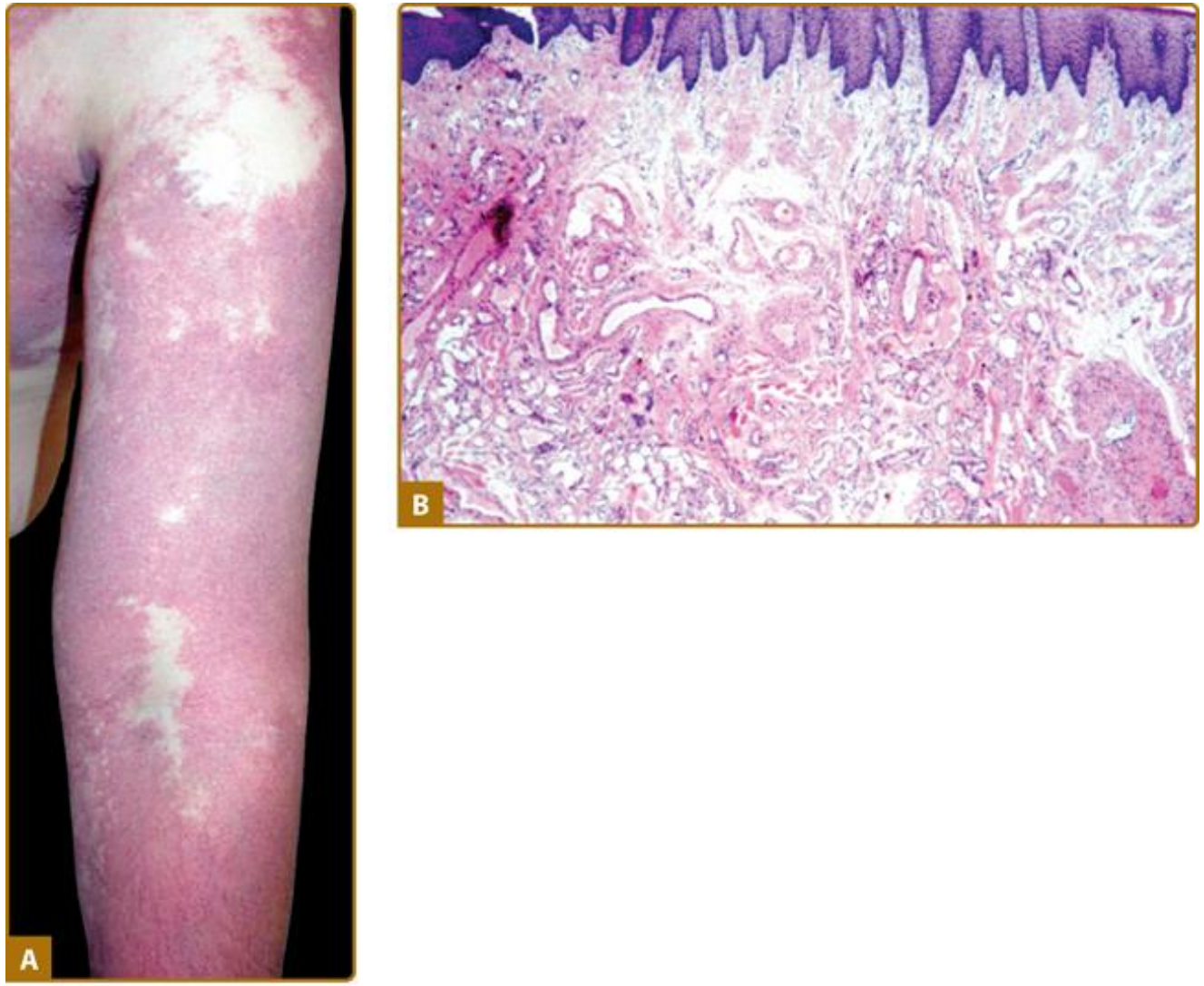
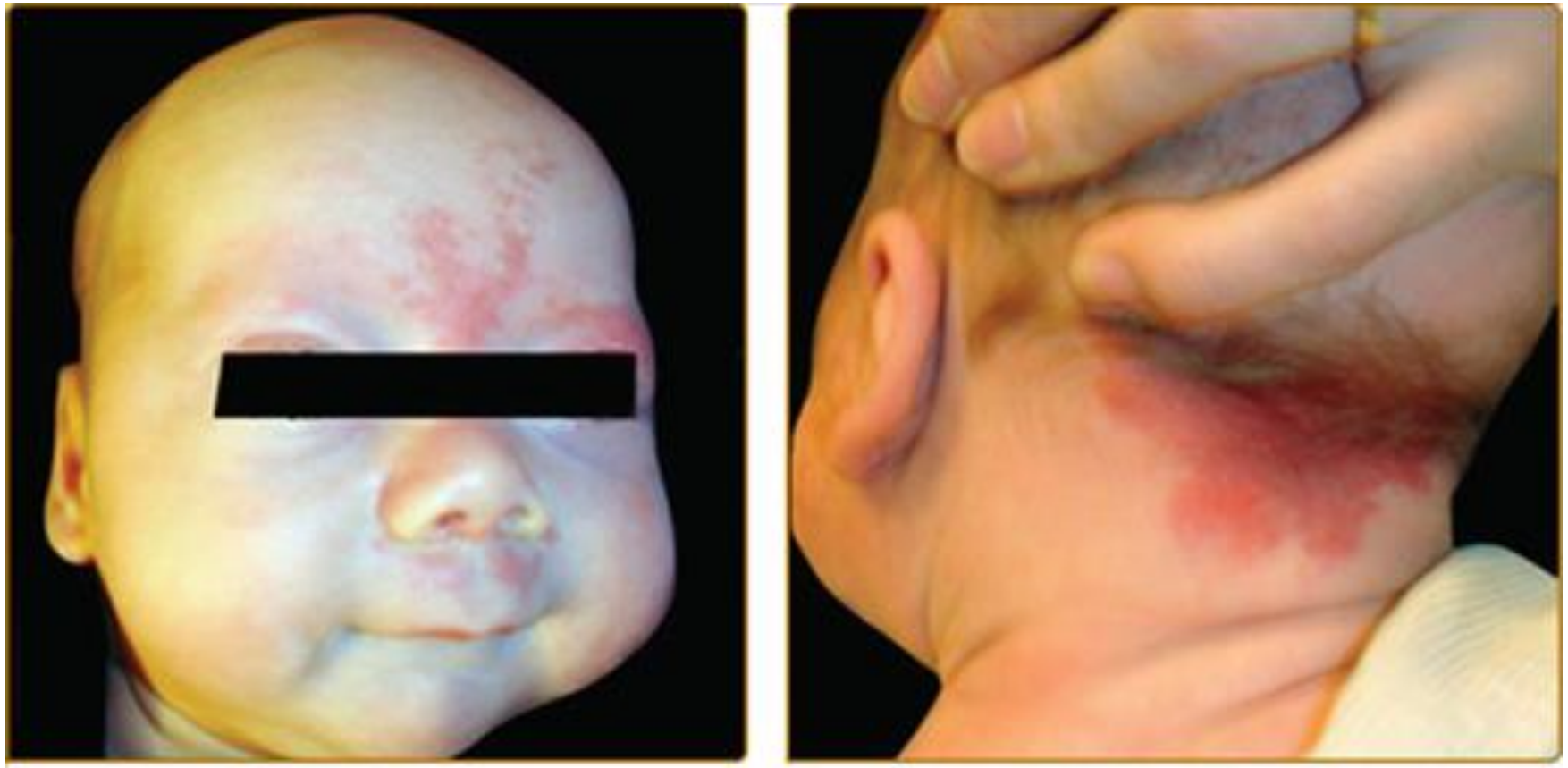


Fig 1

Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com



Nevus flammeus neonatorum on the glabella, upper eyelids, and upper lip **(A)** of a 6-week-old neonate who also has an Unna nevus on the occiput **(B)**.

Fig 2

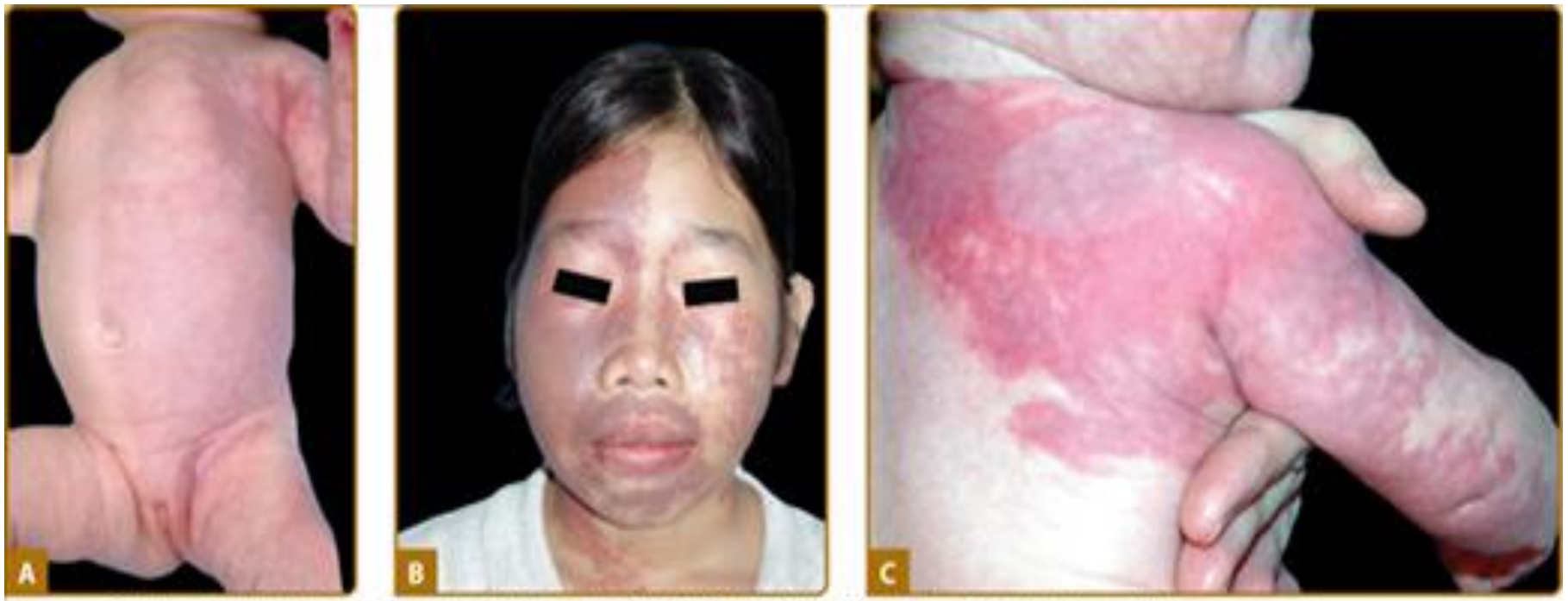
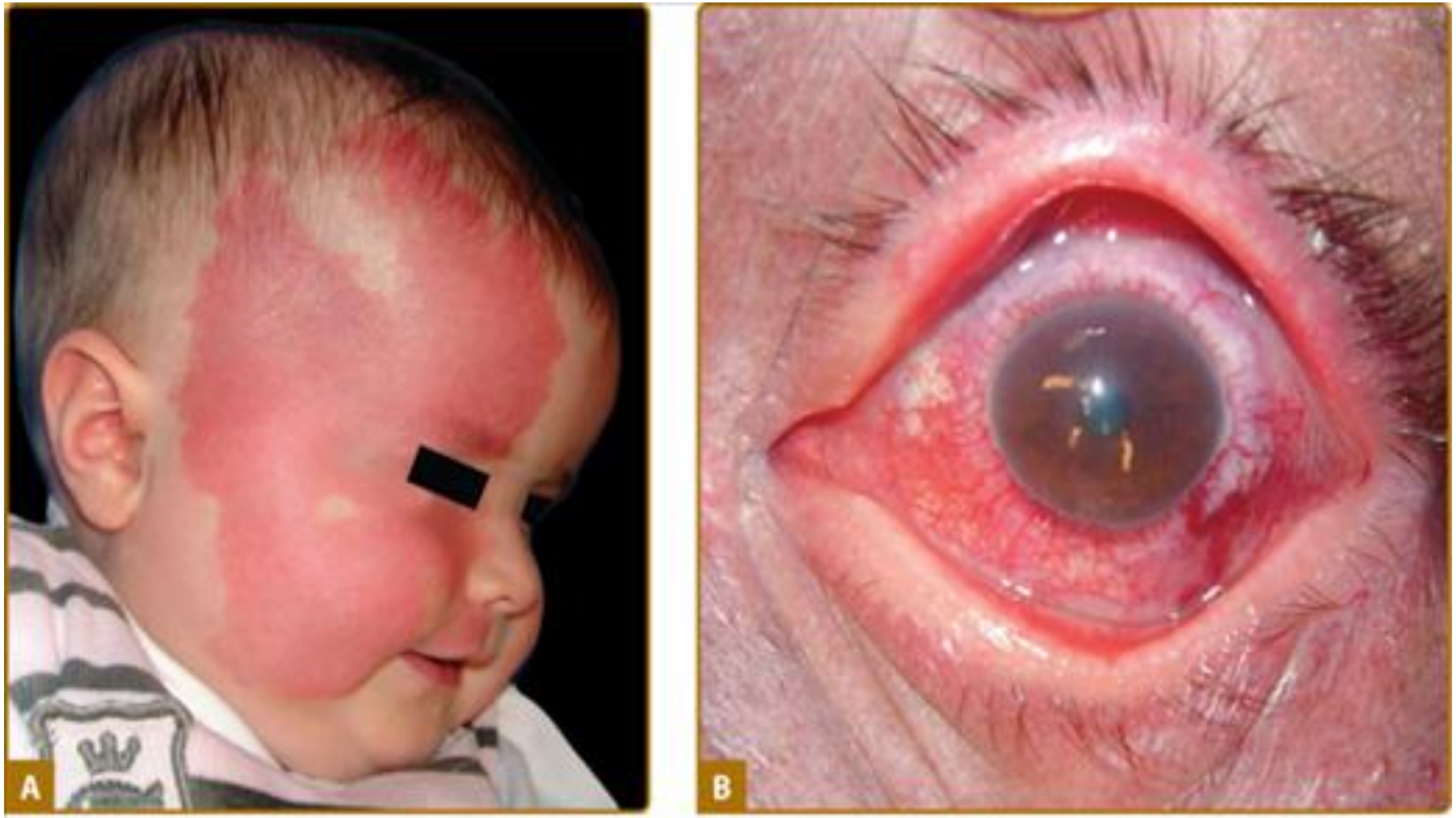


Fig 3

Several aspects of capillary malformations: light red lesions involving half of the body (**A**), dark red lesions on the face with mucosal involvement and soft tissue hypertrophy (**B**), and more localized on shoulder and arm (**C**).

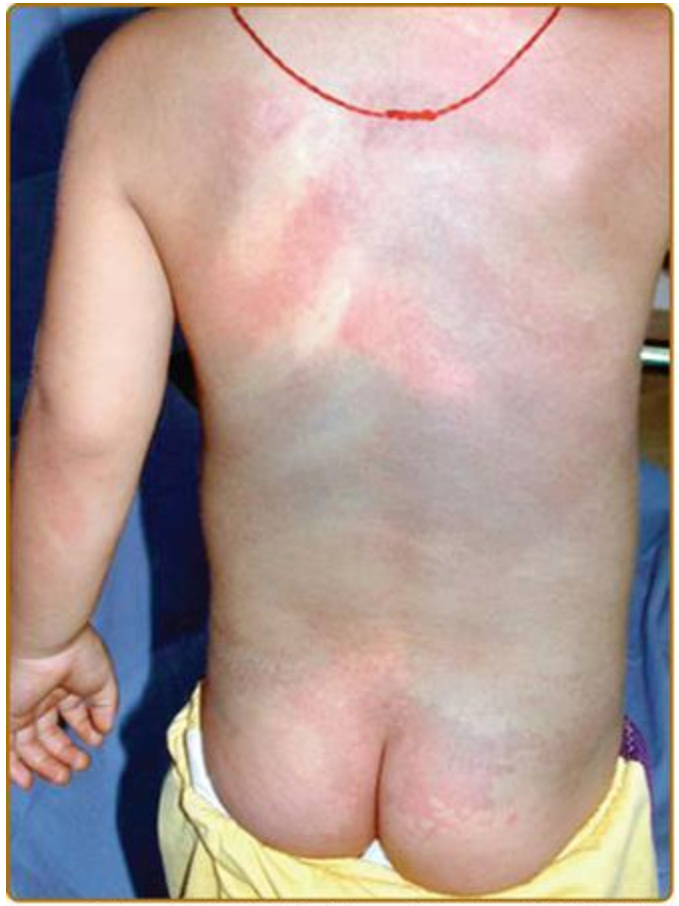


Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Fig 4

Extensive right facial capillary malformation (**A**) involving the ophthalmic and maxillary branches of the trigeminal nerve in a 6-month-old girl affected with Sturge–Weber syndrome. Choroid capillary-venous malformation (**B**) causing vision loss in a 50-year-old man with Sturge–Weber syndrome.

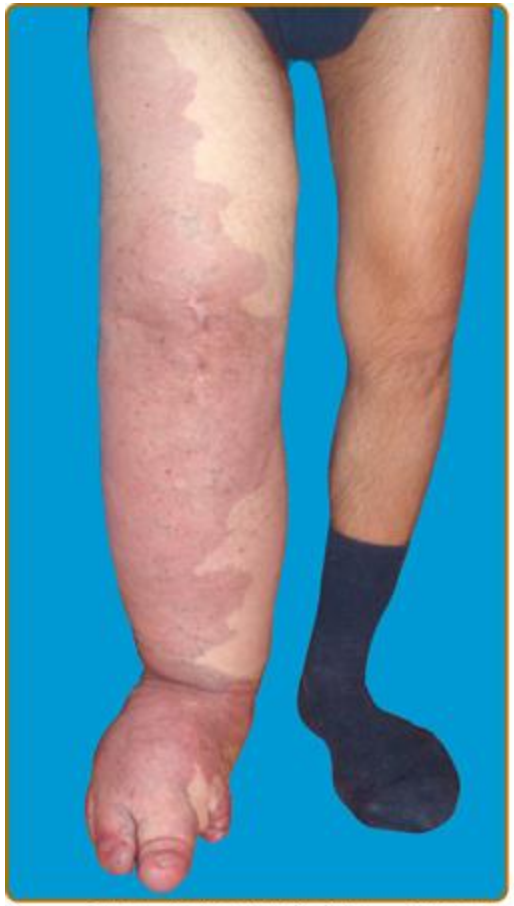
Fig 5



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Clinical characteristics of phakomatosis pigmentovascularis: large metameric capillary malformation with atypical extensive mongoloid patch located on the back.

Fig 6

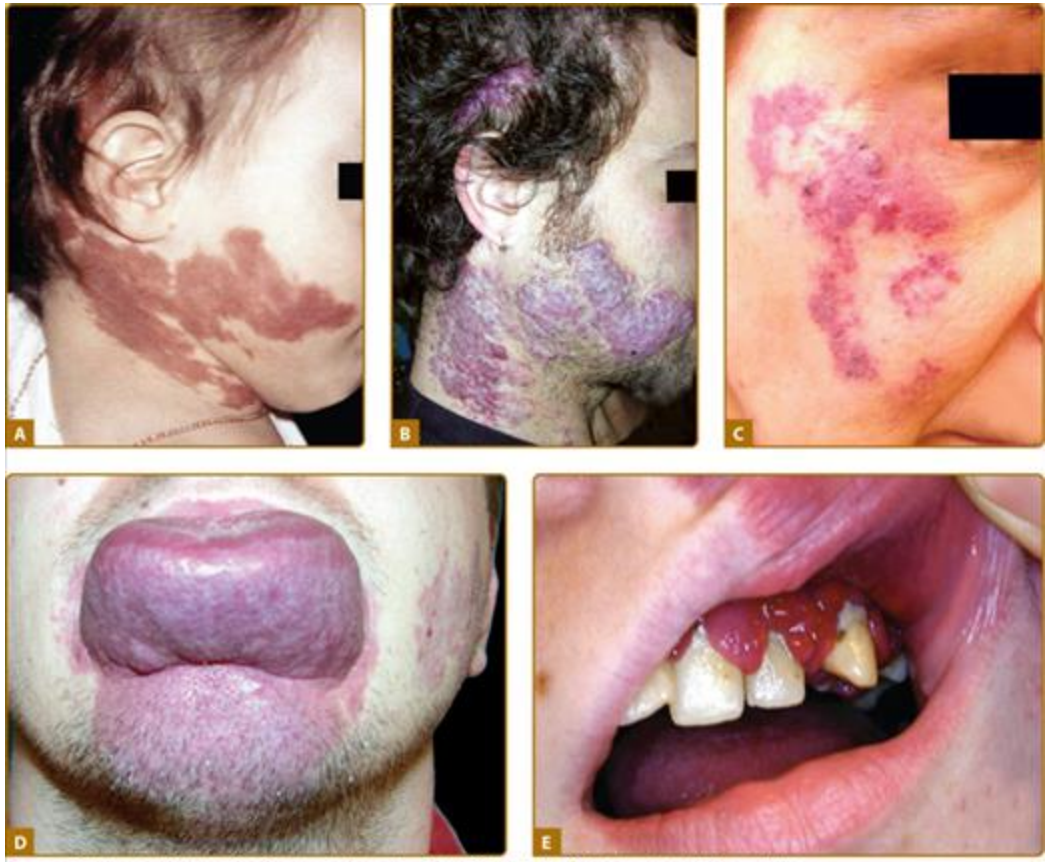


Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Capillary-lymphaticovenous malformation of the right lower extremity with soft tissue hypertrophy (Klippel–Trénaunay syndrome).

Prognosis and Clinical Course

CM never regresses. It is present at birth, and slowly thickens and darkens with time. It often becomes raised and nodular. Pyogenic granuloma can occur with time as well as soft tissue or bony hypertrophy.



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Clinical course of capillary malformation: darkening and thickening with time: at age of 6 months (**A**), and at age of 33 years (**B**). Development of pyogenic granuloma (**C**), soft tissue (**D**), and mucosal hypertrophy (**E**).

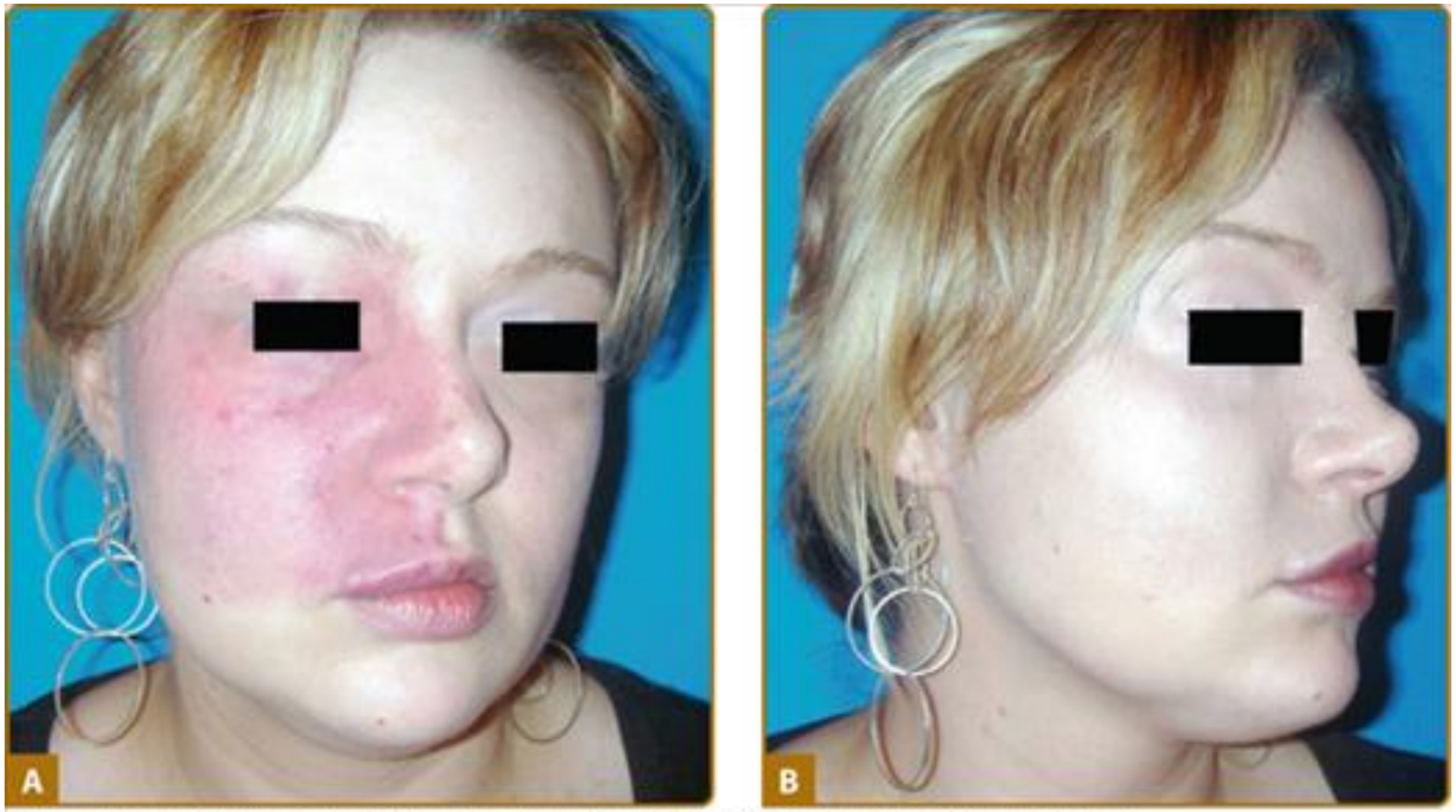
Treatment

Many therapeutical approaches have been used for CM, such as

- **electrocoagulation,**
- **tattooing,**
- **dermabrasion,**
- **cryosurgery,**
- **cosmetic makeup.**
- **Laser treatment is the gold standard for most CM. Pulsed-dye laser with its specific wavelength (585 nm) and a short pulse duration (400 ms) currently gives the best results in infants and children.**

There are few complications.

- ✓ **Multiple sessions (6–12) are needed,**
- ✓ **general anesthesia may be necessary as the procedure is painful.**
- ✓ **Laser treatment is more efficient on the cervicofacial and trunk area than on the extremities.**
- ✓ **Early treatment during childhood does not reduce the number of laser sessions.**
- ✓ **The use of a dynamic cooling system to avoid heating the epidermis allows increasing of laser fluences, resulting in optimal lightening of the lesion.**
- ✓ **Recurrence can occur after cessation of therapy.**
- ✓ **Laser has no impact on associated hypertrophy. Contour resection is needed mainly to treat complications such as pyogenic granuloma or lip hypertrophy.**



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Efficient noninvasive management of facial capillary malformation using cosmetic makeup.

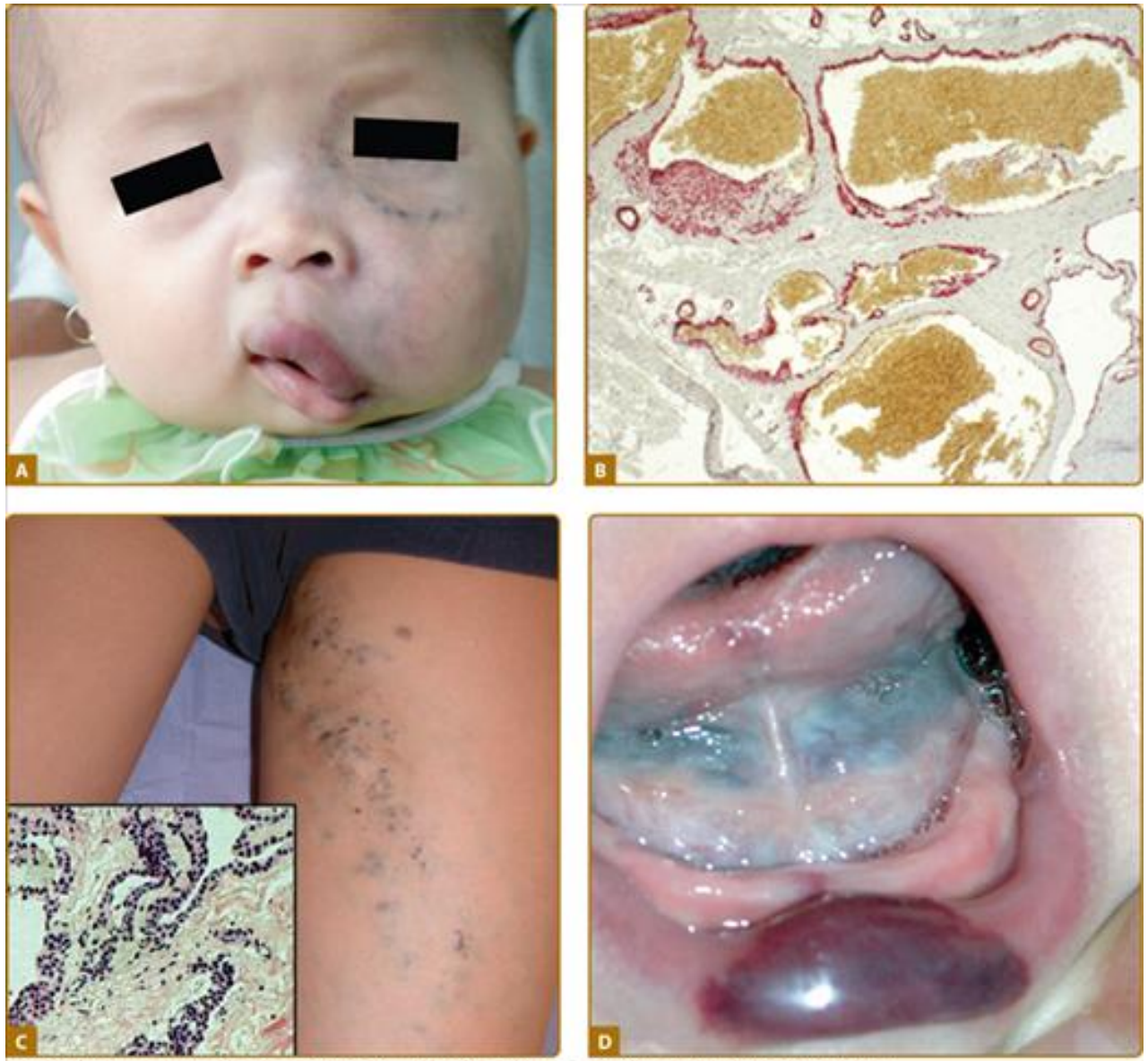
Venous Malformations

- VM is a congenital defect on the skin or mucosa, but can involve any structure (subcutis, muscles, bones, and nerves) and any organ [central nervous system, gastrointestinal (GI) tract].

- Fifty percent of VMs are located in the cervicofacial area and 37% on the extremities.

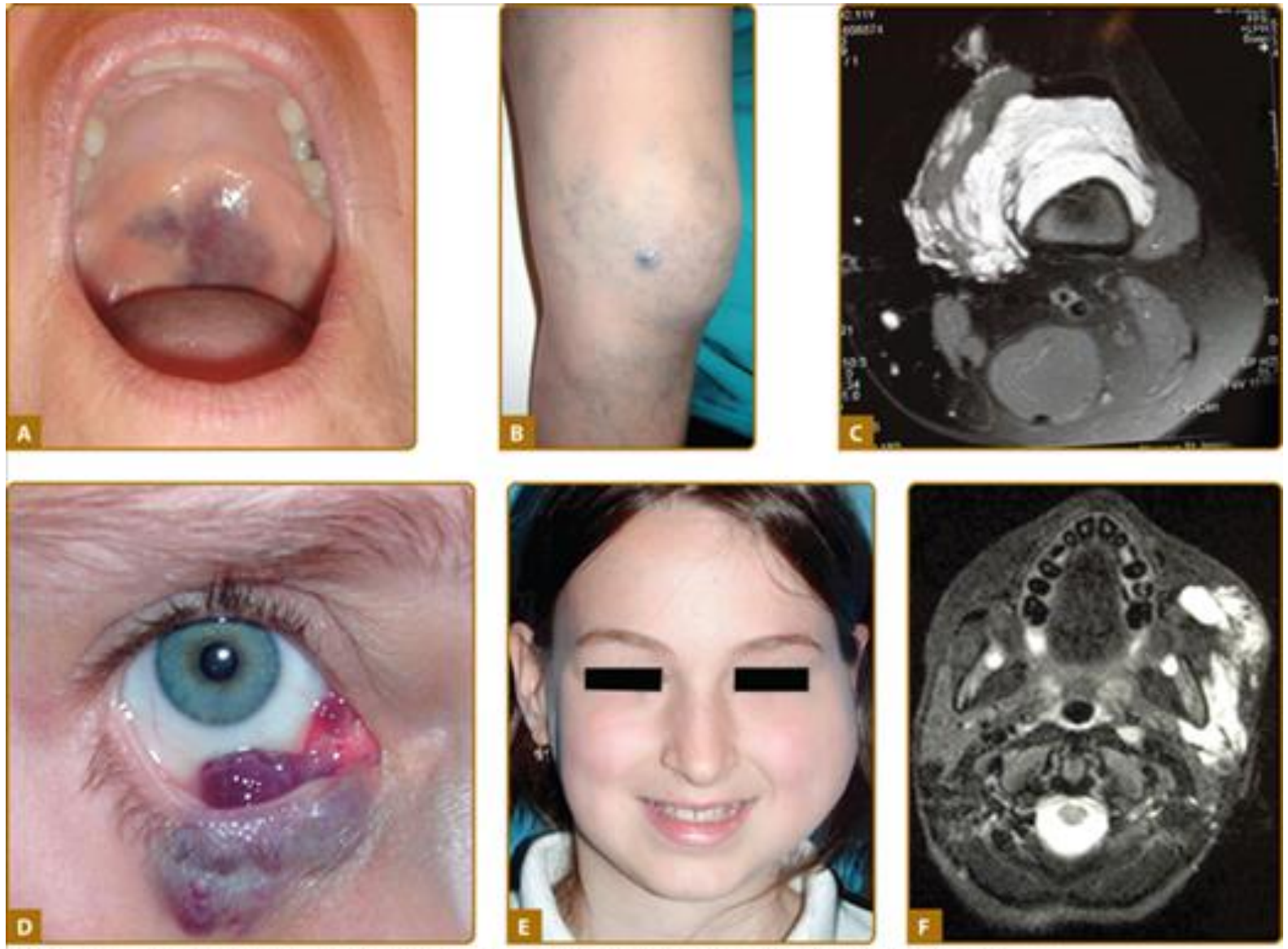
- VMs are mainly isolated, but can be part of complex vascular disorders of unknown etiology, such as KTS, and Maffucci and blue rubber bleb nevus (BRBN) syndromes.

	VM	VMCM	GVM glomuvenous malformations	BRBN blue rubber bleb nevus	Maffucci Syndrome
Location	50% head and neck	Extremities, mucosa	70% extremities	Especially palms and soles	Extremities
Skin color	Normal-bluish-purple	Bluish	Bluish-red-purple	Bluish-dark-blue	Normal-bluish
Aspect	Flat-raised	Raised dome-shaped	Raised, cobblestone Plaque like, hyperkeratotic	Dome shaped, nipple-like	Exophytic
	Single large	Multiple, small	Multiple	Multiple, small often + 1 large lesion	Multiple, important deformation of hands and feet
Palpation	Phleboliths	Compressible	Non compressible Painful	Firm, rubbery, Spongy	Firm
Associated	Coagulopathy	–	–	Visceral venous malformation, coagulopathy	Multiple enchondromas High risk of cancer
Radiologic	Phleboliths	Venous channels	No phlebolith	Venous channels	Venous channels Enchondromas
Tissue involved	Often deep location	Cutis and subcutis	Cutis and subcutis	Often deep component	Deep location
Histology	Thin-walled venous channels	Thin-walled venous channels	Glomus cells	Thin-walled venous channels	Spindle cells Hemangioendothelioma
Inheritance	Sporadic	Autosomal dominant	Autosomal dominant	Sporadic	Sporadic
Cause	50% somatic TIE2 mutations	Gain of function mutation in TIE2	Loss of function mutation in Glomulin	?	?



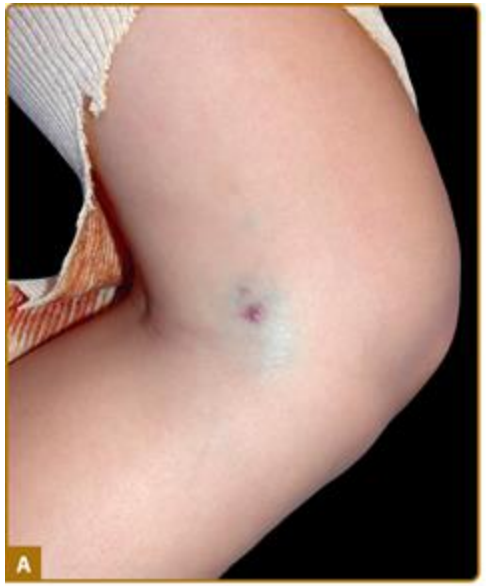
Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

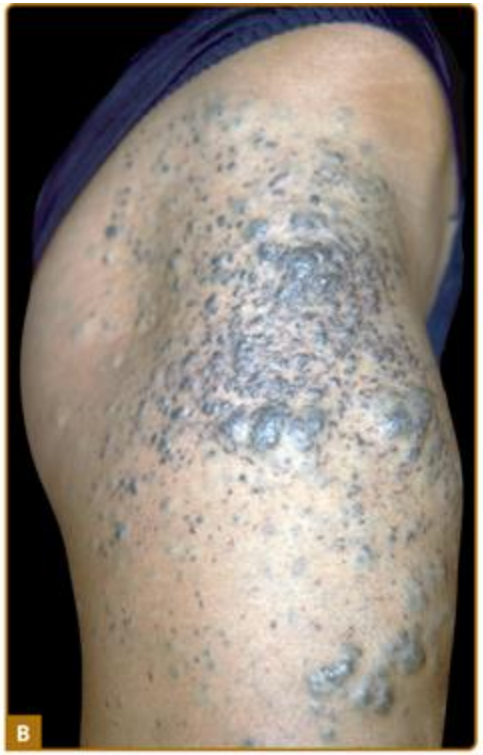
A. Venous anomalies: extensive hemifacial venous malformation (VM) causing soft-tissue hypertrophy and lip deformation in a 6-month-old boy. **B.** Histologically, VM consists of ectatic venous-like channels with anomalies in mural cells (hematoxylin and eosin). **C.** Superficial glomuvenous malformation of the inner thigh. Note the presence of mural glomus cells on histology (hematoxylin and eosin). **D.** Mucosal VM involving the lower lip and the floor of the mouth.

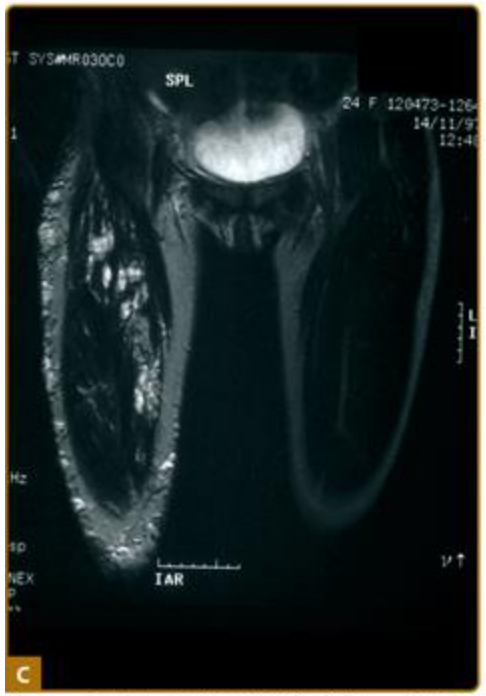


Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Various venous malformations.
A. Of soft palate, causing sleeping apnea.
B and **C.** Of knee, causing hemarthroses; confirmed by T2-weighted images.
D. Of mucosal eyelid.
E and **F.** Of cheek, causing facial asymmetry and pain.





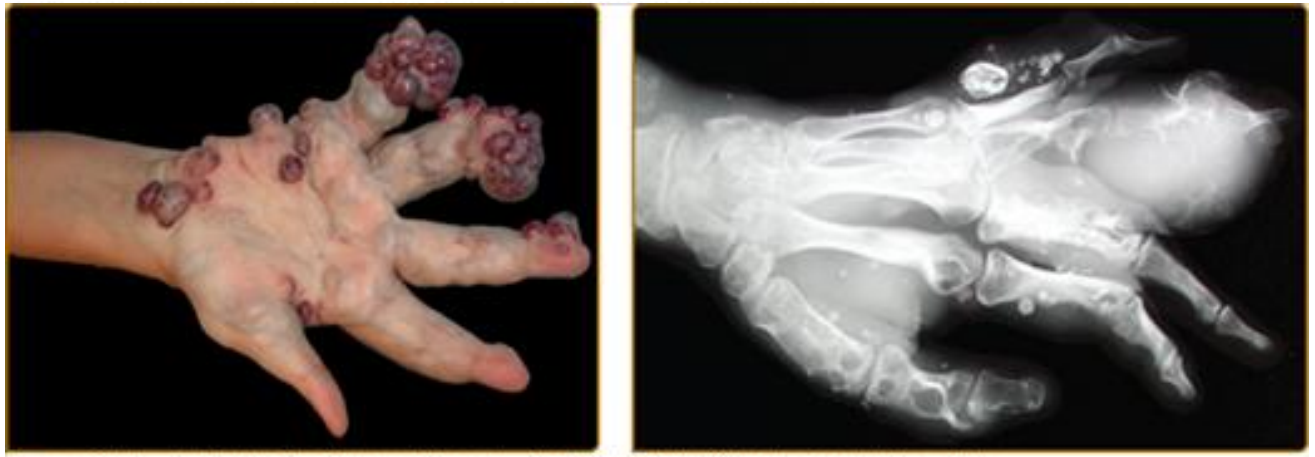


Two glomuvenous malformations on the extremity: small nodule (**A**), extensive but superficial with cobblestone appearance (**B**), and as seen on T2-weighted image (**C**).

Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

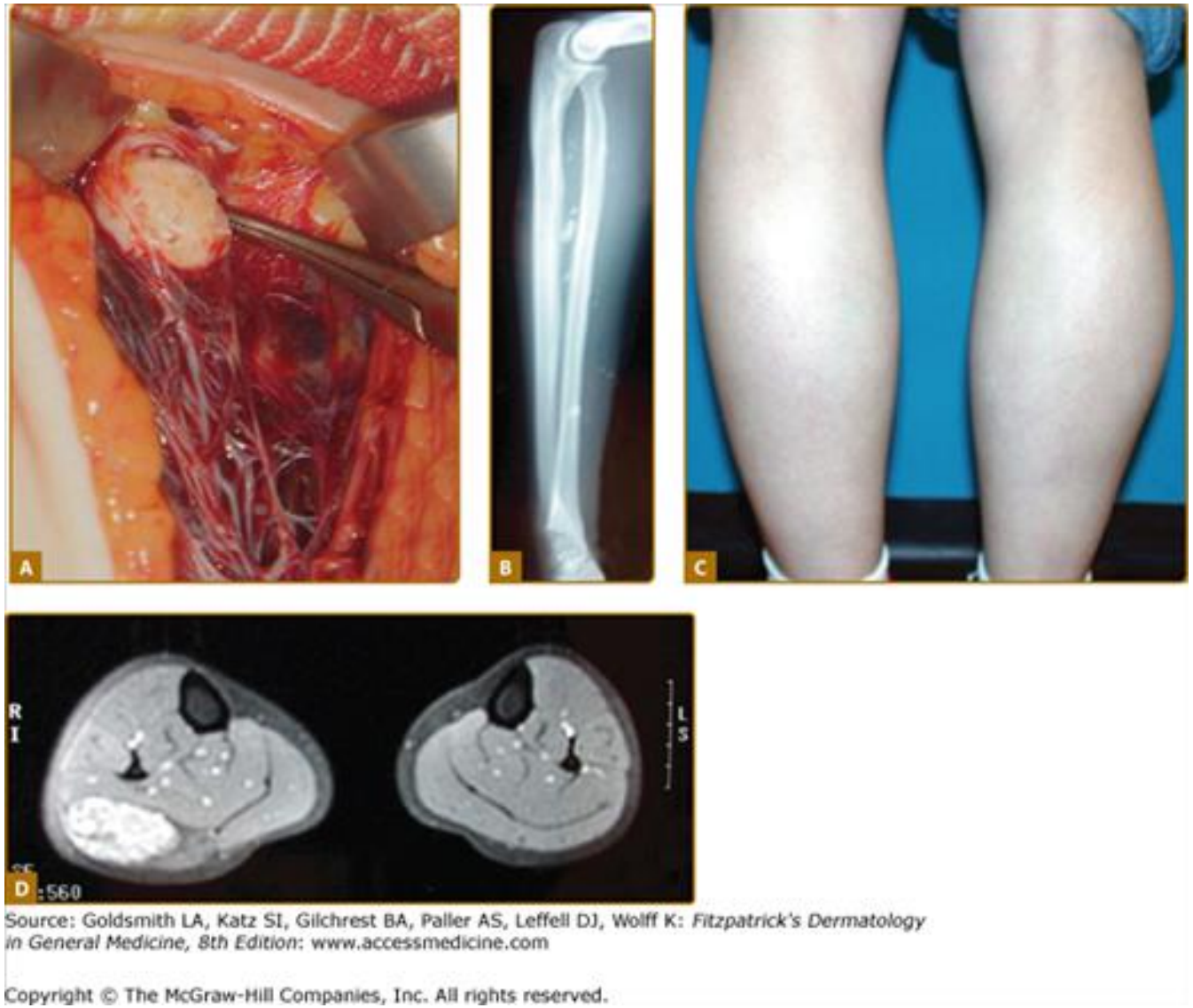
Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Hyperkeratotic, small, rubbery blebs characteristic of blue rubber bleb nevus syndrome.



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Exophytic venous-like lesions of hand associated with multiple enchondromas (as seen on plain radiography) specific of Maffucci syndrome.

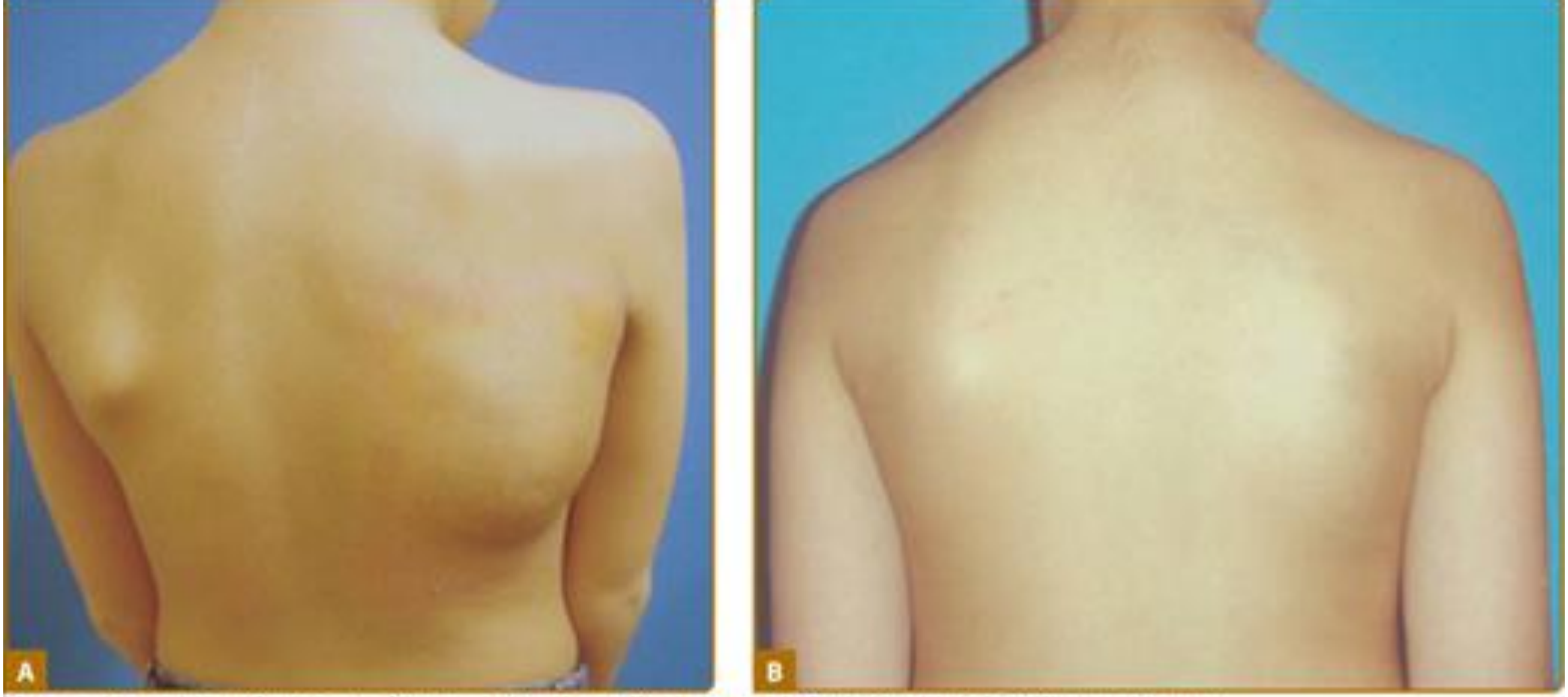


Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Diagnosis of venous malformation (VM):
the presence of phleboliths, as seen perioperatively (**A**)
or on plain radiography (**B**) is the hallmark of VM.
Bright on T2-weighted images, VM often involves deep muscles
(**C** and **D)**.

Treatment

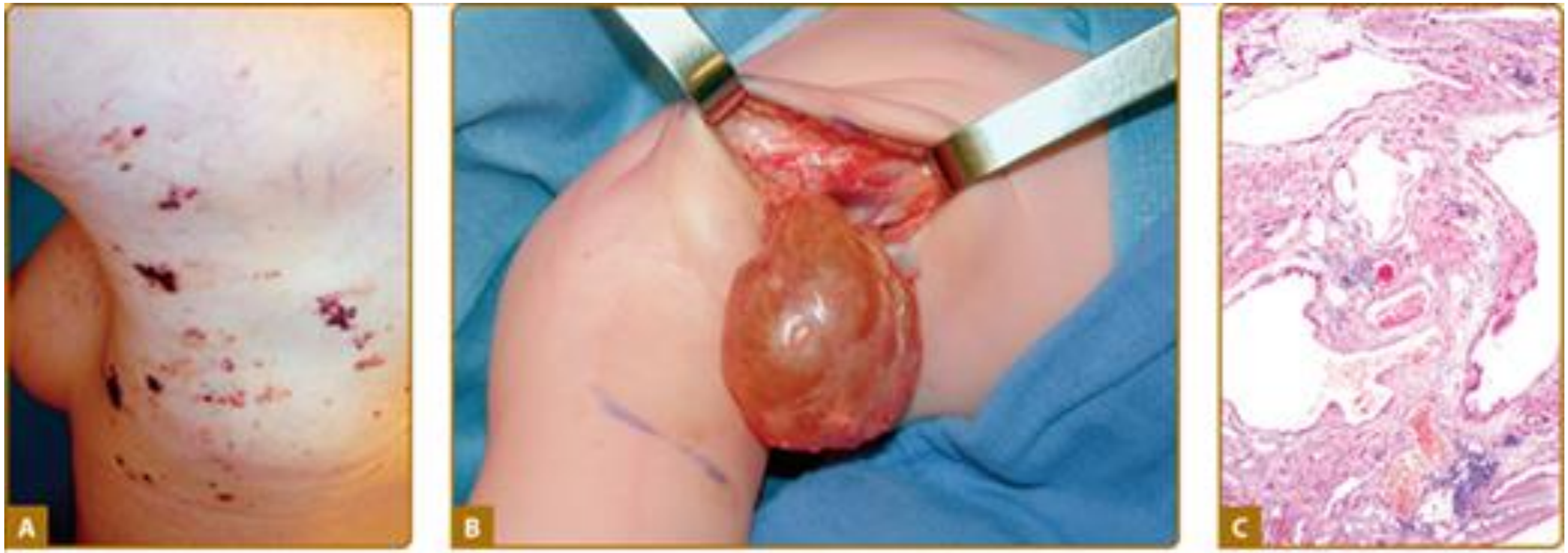
- Indications for treatment include esthetic preference, pain or functional impairment.
- VMs can be treated conservatively or medically and, more aggressively with either sclerotherapy and/or surgical resection.
- VM in an extremity is conservatively managed by a custom-made elastic compressive garment. Aspirin (80 mg/day) can alleviate chronic pain due to local and painful thrombosis. Low-molecular-weight heparin is much more efficient in case of LIC (high d-dimer).
- The commonly used dosage is 100 anti-Xa/kg/day for adults and 30 anti-Xa/kg/day in children. Reduction of pain and d-dimers are seen within 10 days.
- The treatment can be continued for several months and is mandatory before any surgery to avoid decompensation into DIC during or after surgery.
- It will also improve sclerotherapy results. Small cutaneous VMs can be injected, at any site, with a sclerosant.
- For large VMs, fluoroscopic guidance and general anesthesia are mandatory during percutaneous sclerotherapy.
- Sclerotherapy is potentially dangerous and requires the skills of an experienced interventional radiologist. Several sessions are commonly necessary. For GVM, sclerotherapy is less effective .



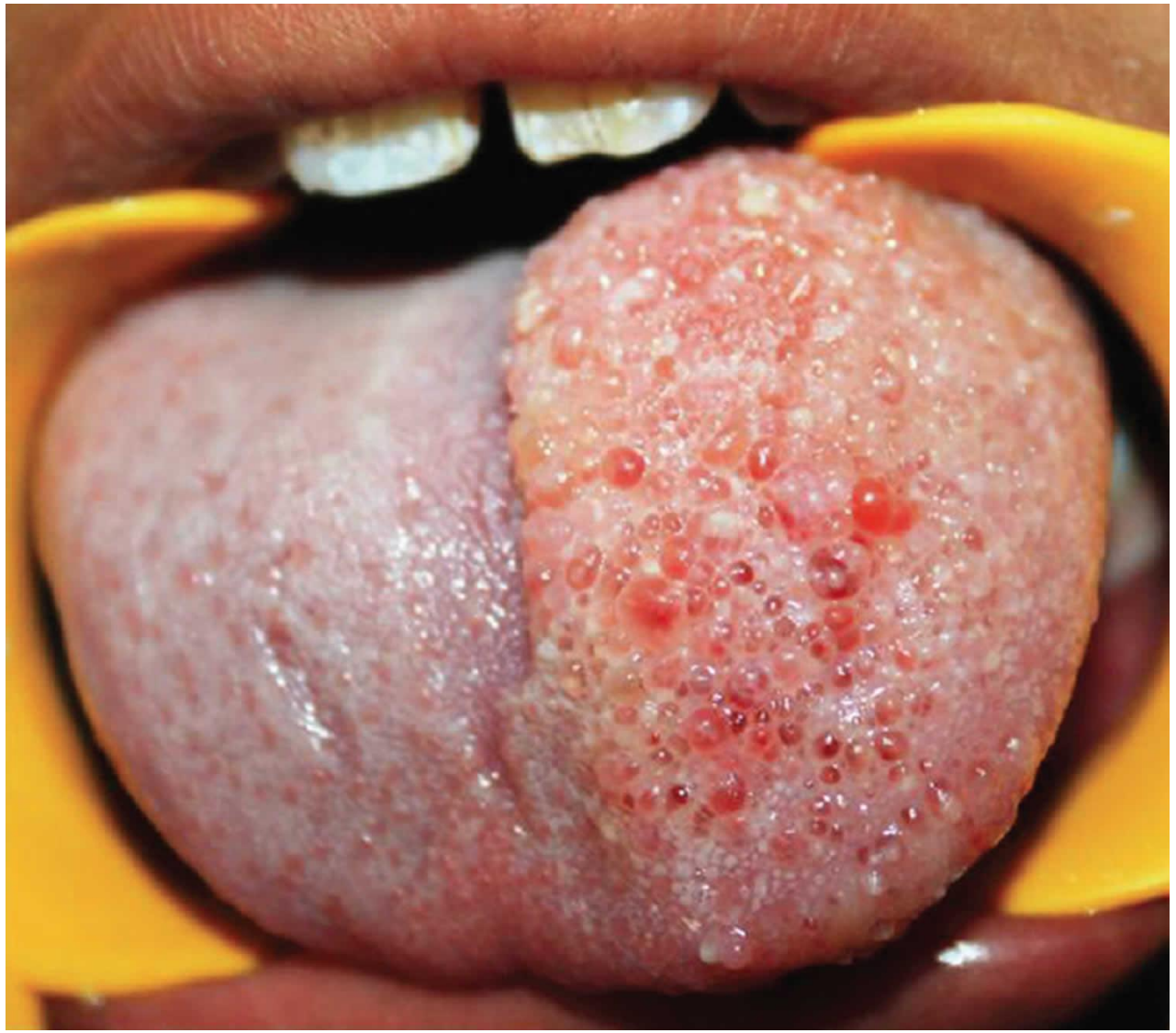
Extensive venous malformation of back before (**A**) and 2 years (**B**) after two ethanol sclerotherapies.

Lymphatic Malformations

- Lymphatic malformation (LM) is a congenital disorder of unknown incidence.
- It consists of vesicles or large cysts filled with lymphatic fluid.
- Macrocystic LM can be diagnosed in utero as early as the first trimester of pregnancy. However, most LMs are diagnosed during infancy, before the age of 2 years. Some can manifest only at puberty or during adulthood.
- Macrocystic LMs are often located on the neck, axilla, chest wall, or groin, whereas microcystic lesions are commonly seen in the face.
- LMs occur sporadically in contrast to primary lymphedema that can be inherited as an autosomal dominant trait in up to 20% of cases. Rare families with recessive inheritance have also been identified.
- Primary lymphedema is divided into congenital (present at or soon after birth) and late onset (present at puberty) .
- It can be isolated or part of a syndrome, such as Turner and Noonan syndromes



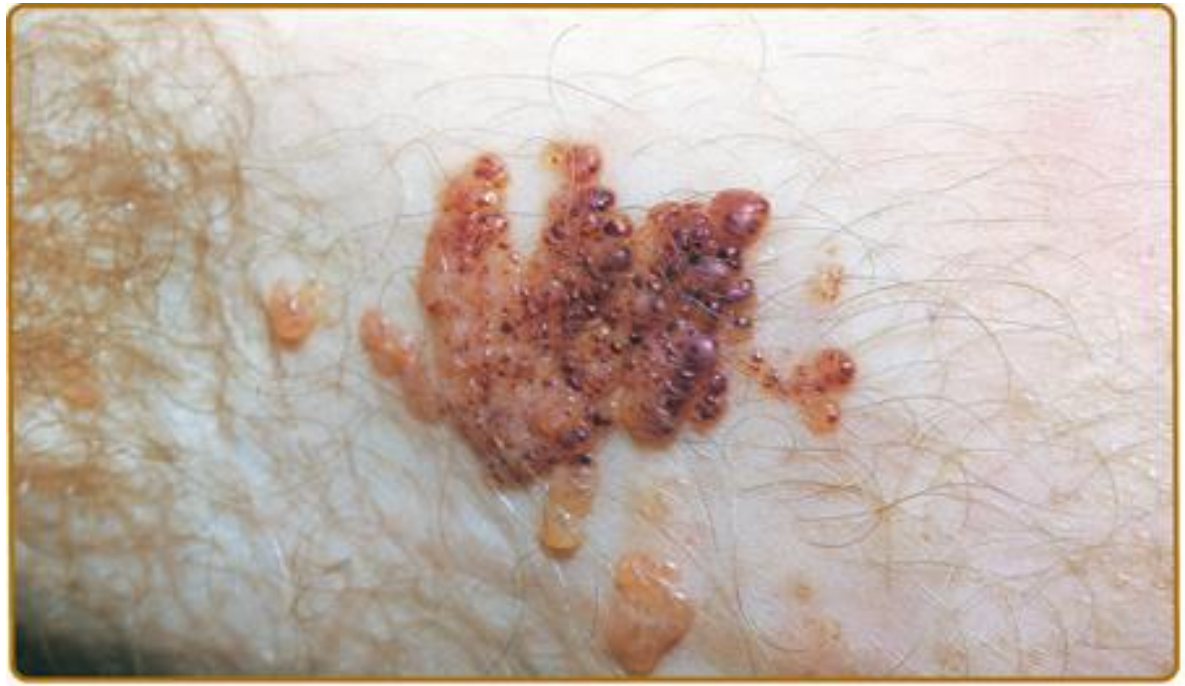
A. Microcystic, dermal lymphatic malformation (LM) consisting of clear and dark-red vesicles. **B.** Intraoperative view of a macrocystic LM of the axilla showing a well-delineated cyst filled with clear lymph fluid. **C.** Histologically, LM consists of dilated lymphatic channels with flat endothelium (hematoxylin and eosin).



Lymphangioma of the tongue

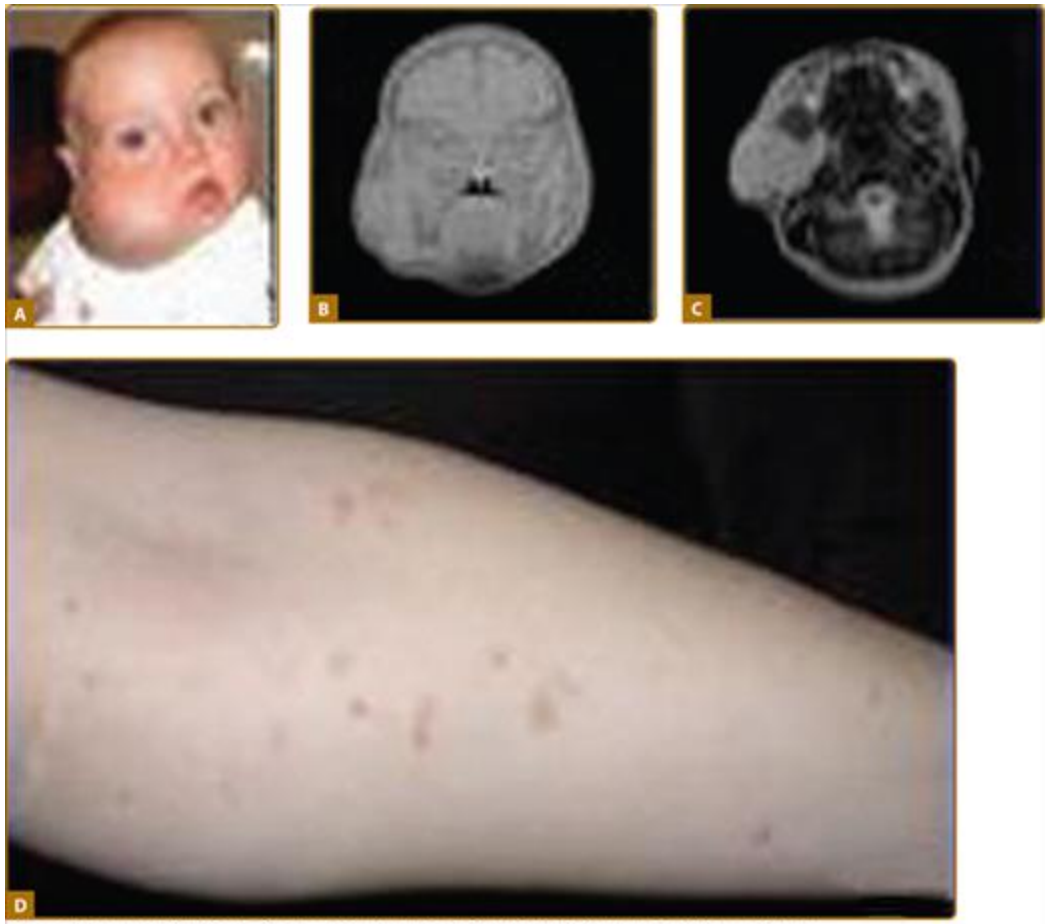
Clinical findings

- LMs are composed of microcysts (previously lymphangioma circumscriptum; Fig. 6)
- and/or macrocysts (larger than 1-cm diameter, previously known as cystic hygroma) that grow proportionally with the child. They are filled with clear or serosanguineous fluid (see Fig. 7).
- Macrocystic LM manifests as a soft, multilobulated, well-defined mass, whereas microcystic LMs are ill defined and often invade adjacent structures. Skin can be normal in color or, due to intracystic bleeding, have a bluish discoloration (see eFig. 7 a).
- Dermal LMs can manifest as small millimeter-sized vesicles, clear (see Fig. 7 d) or dark red (if intracystic bleeding) that can bleed and become purple and nodular.



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Fig. 6: Lymphatic malformation. Frog-spawn-like confluent grouped “vesicles” filled with a serosanguineous fluid ***lymphangioma*** .

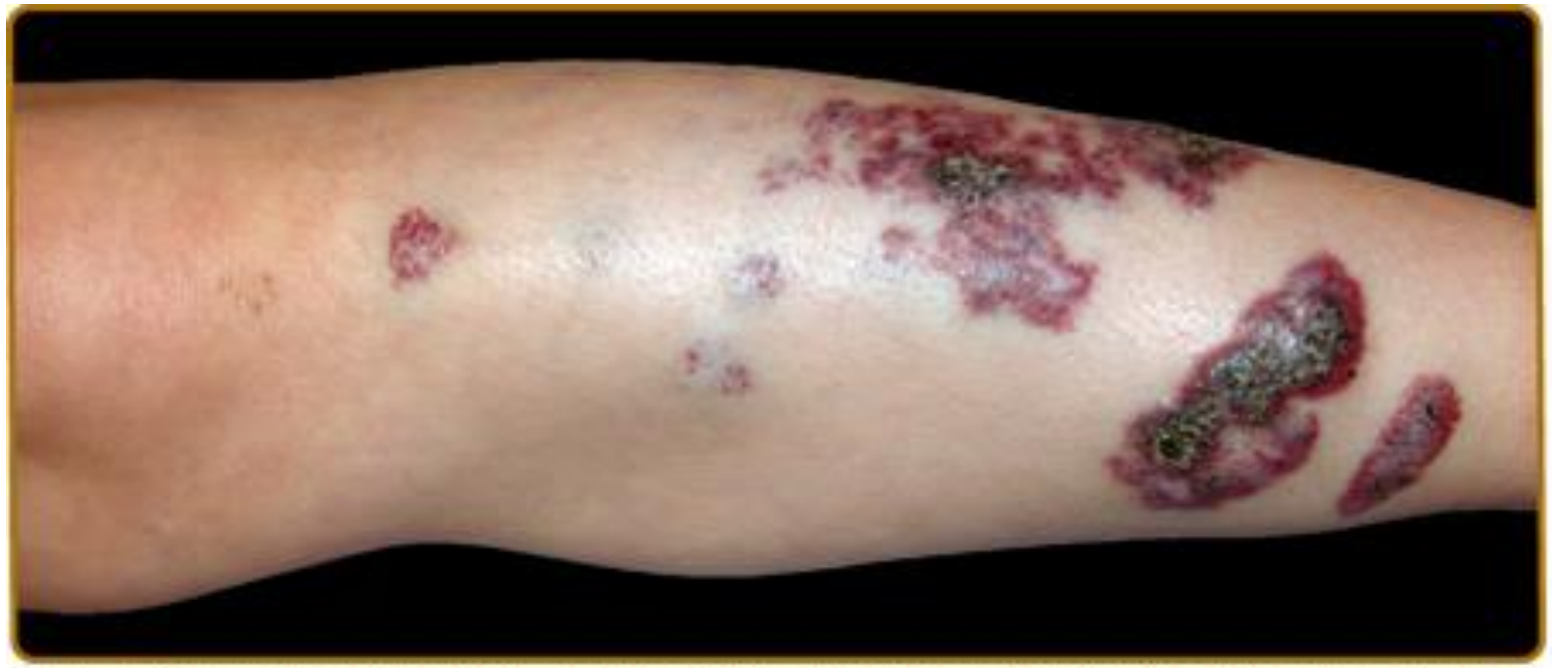


Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Fig. 7: Various aspects of lymphatic malformation (LM): bluish discoloration (**A**) of extensive LM of ***cheek cystic hygroma,*** as seen on computed tomography scan (**B** and **C**). Small clear vesicles specific of dermal LM (**D**).

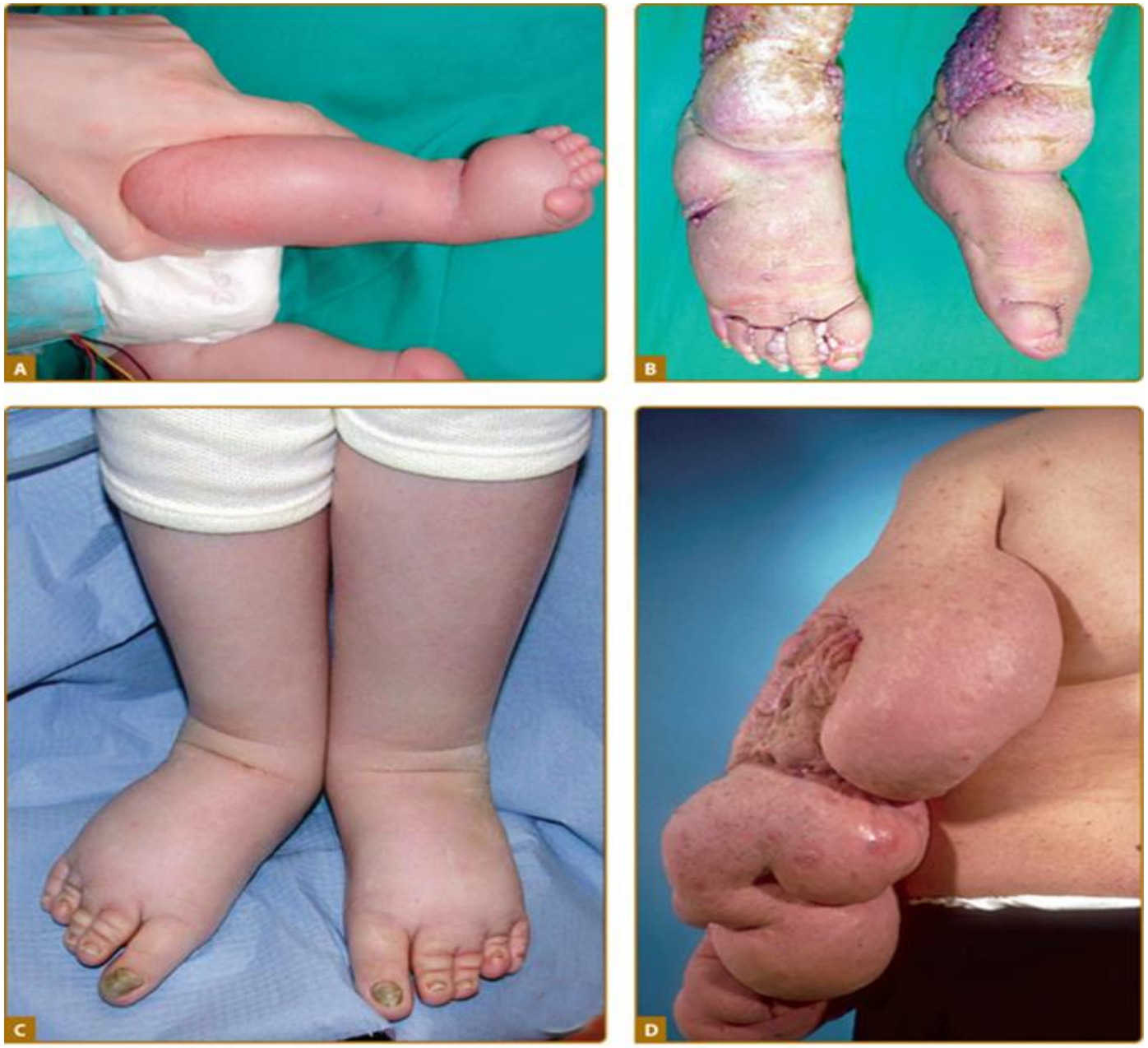
Angiokeratoma Circumscriptum

Angiokeratoma circumscriptum, or capillary-lymphatic malformation (CLM), is a combined, well-demarcated lesion often located on the extremity. This lesion is pink to bluish red in color, slightly raised, and usually hyperkeratotic



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Capillary-lymphatic malformation (angiokeratoma circumscriptum) on the lower extremity, causing oozing. It is purple–red in color and slightly raised, with hyperkeratosis

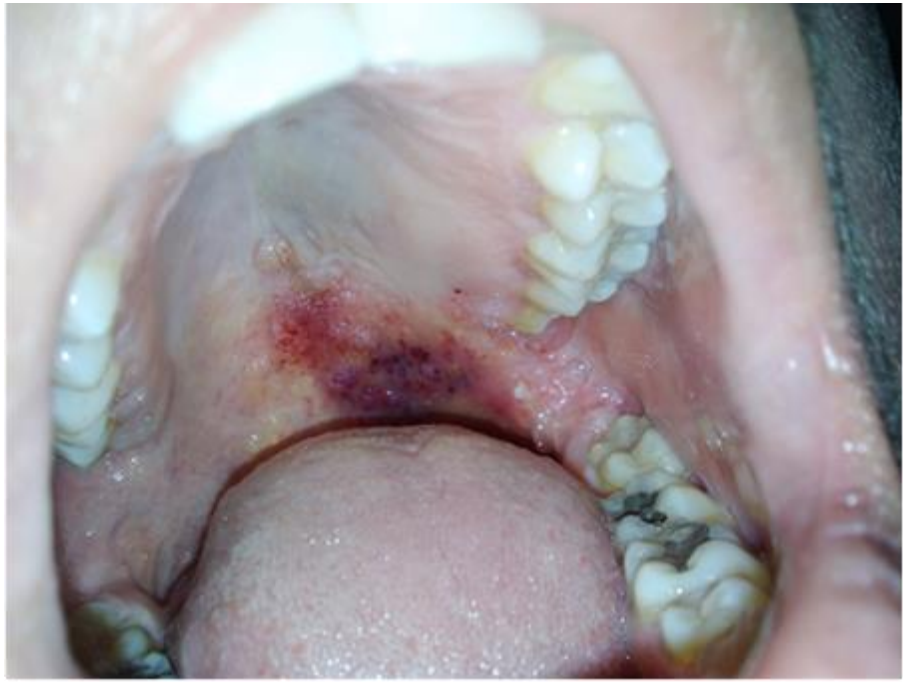


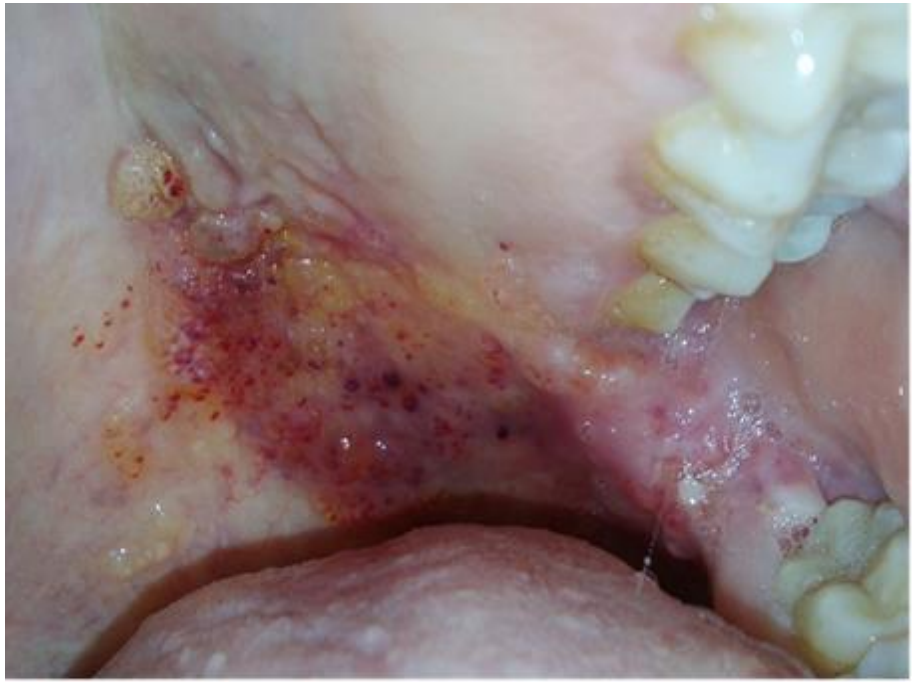
Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Various types of congenital primary lymphedema characterized by lymphedema of lower limb, below the knees (**A**), with papillomatosis (**B**), and with up slanting toenails (**C**), that can evolve into elephantiasis (**D**).

Treatment

- Intralesional bacterial infection as well as early cellulitis should be monitored with prolonged antibiotics first given intravenously.
- Pain medication and anti-inflammatory drugs may be required.
- Immune-modifying medications, such as interferon and corticosteroids, (see Fig .8).
- Neodymium:yttrium-aluminum-garnet laser or carbon dioxide laser photocoagulation has been used to treat dermal LM vesicles that cause oozing (see fig. 9).
- Macrocystic LM is treated with fluid aspiration followed by percutaneous, intralesional injection of sclerosing agents performed by an interventional radiologist.
- Several sclerosing agents have been used, such as sodium tetradecyl sulfate, pure ethanol, OK432 (extract from a killed strain of group A *Streptococcus pyogenes*: picibanil), doxycycline, or bleomycin. Side effects include fever, erythema, and oedema.
- Surgocal resaction (see Fig. 10)





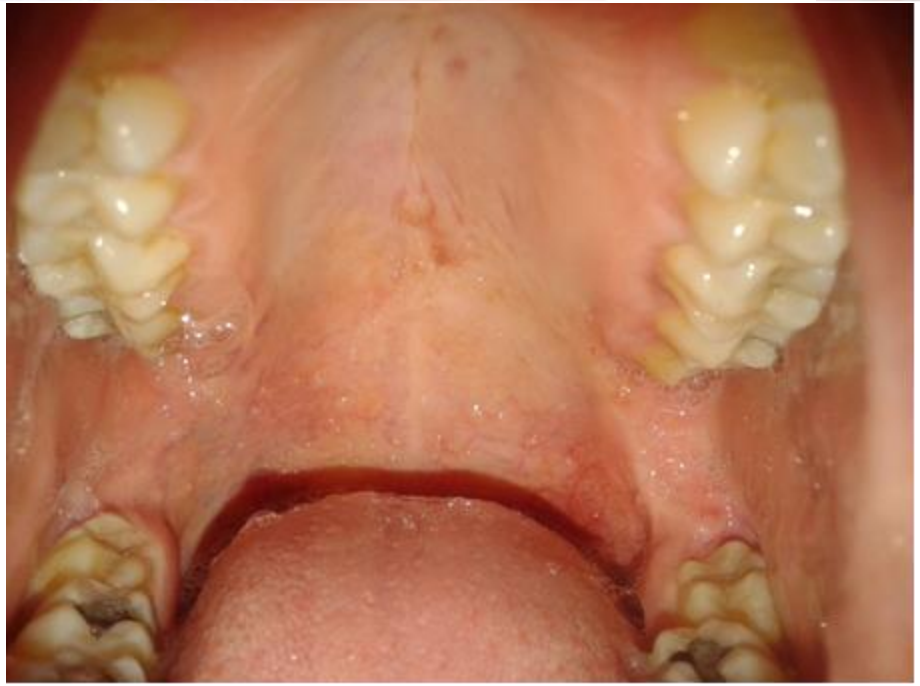
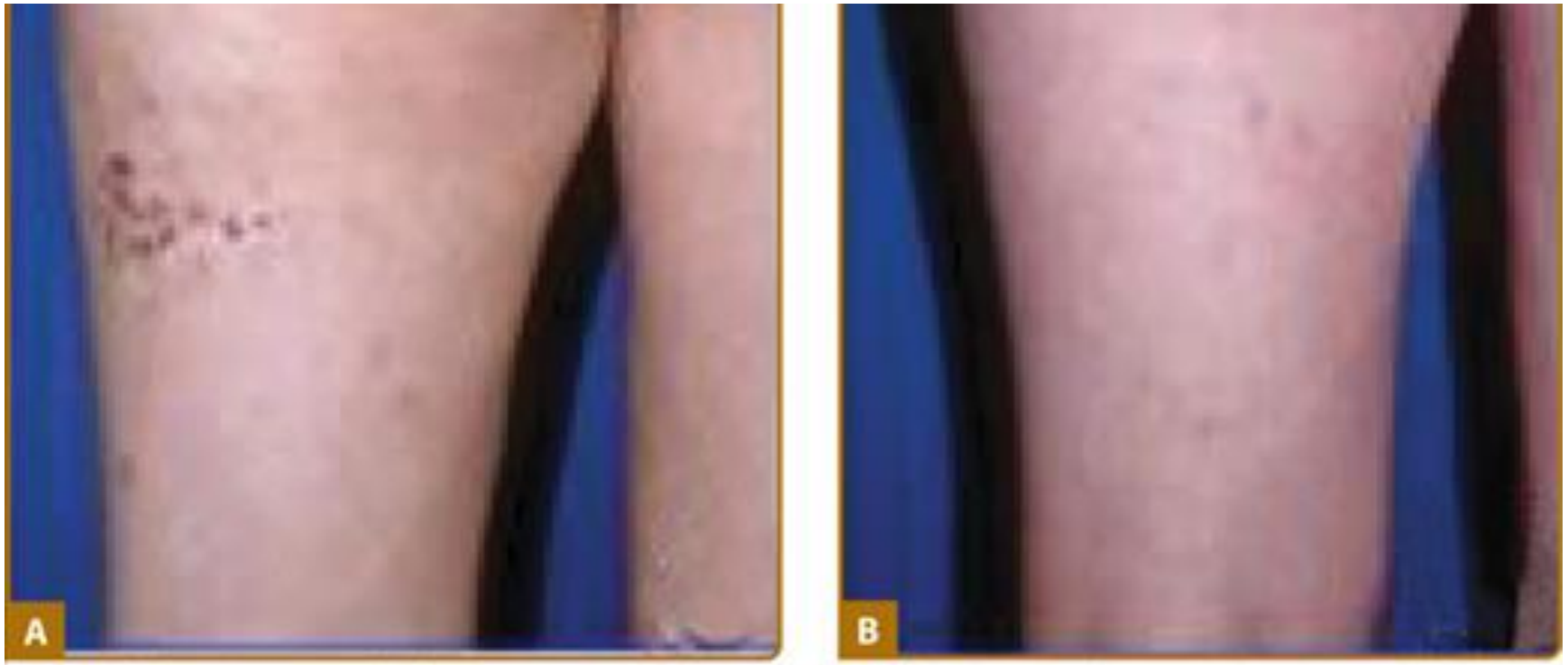


Fig 8: Lymphangia of soft palate treated by intralesional injection of cortisone



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Dermal lymphatic malformation (**A**) efficiently treated with carbon dioxide laser (**B)**.

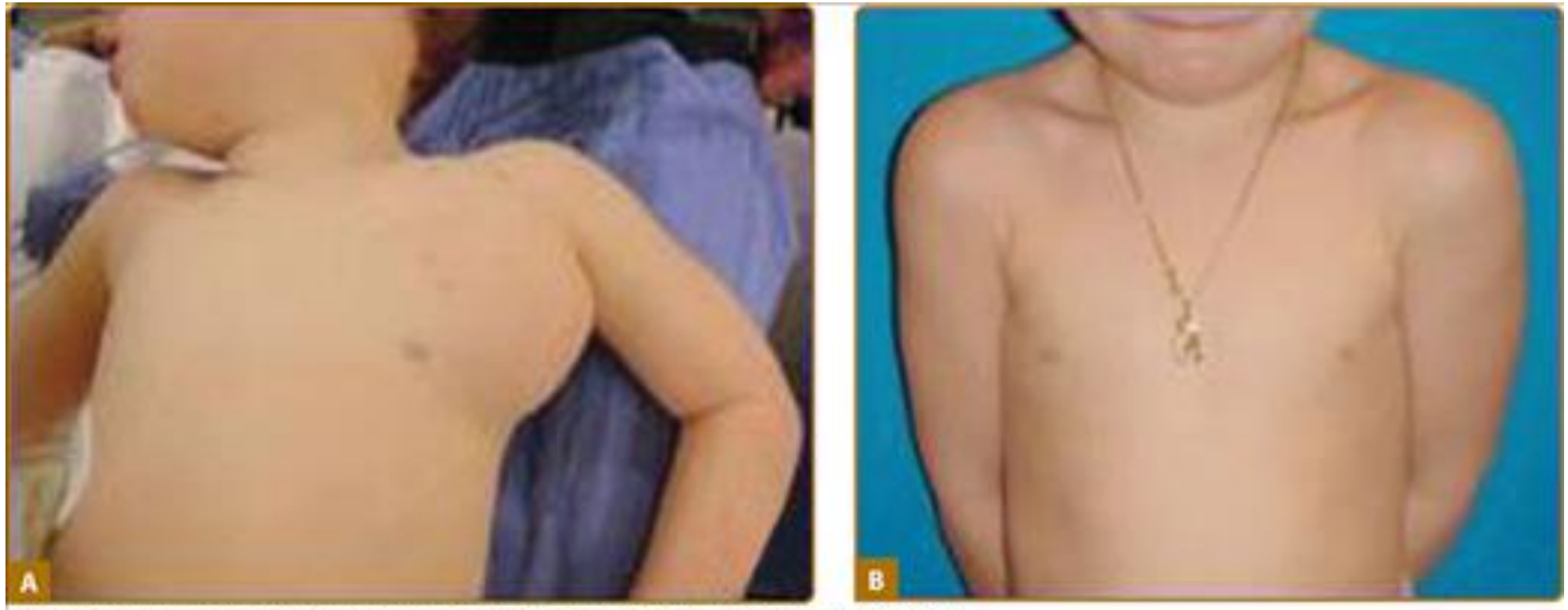
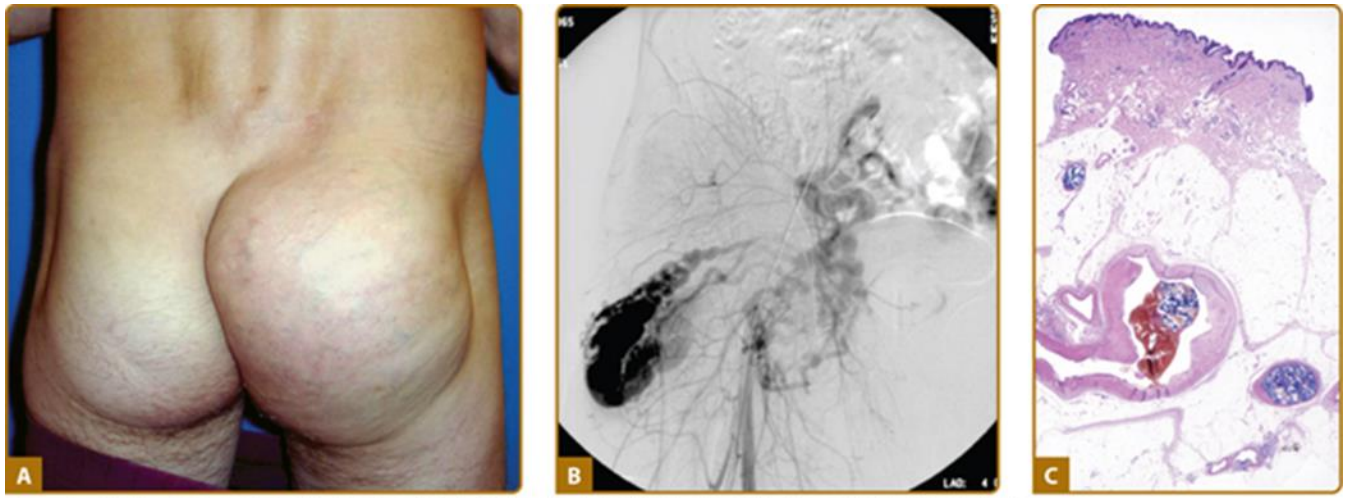


Fig. 10: Lymphatic malformation of the axilla (**A**) treated with surgical resection with 2 years follow-up (**B)**.

Arteriovenous Malformations

Fast-flow cutaneous vascular malformations are mainly AVM and rarely arteriovenous fistula (AVF). AVF is usually traumatic. AVM is characterized by the presence of a "nidus," the epicenter of the lesion that is composed of direct communications between multiple feeding arteries and draining veins without intervening the normal capillary bed. Histologically, AVM consists of distorted arteries and veins with thickened muscle walls due to arteriovenous shunting and fibrosis

Extensive arteriovenous malformation deforming the right buttock (**A**). Arteriography showing the nidus of the malformation (**B**). Histology shows a direct connection between artery and vein, with thrombotic material secondary to the embolization procedure (**C**).

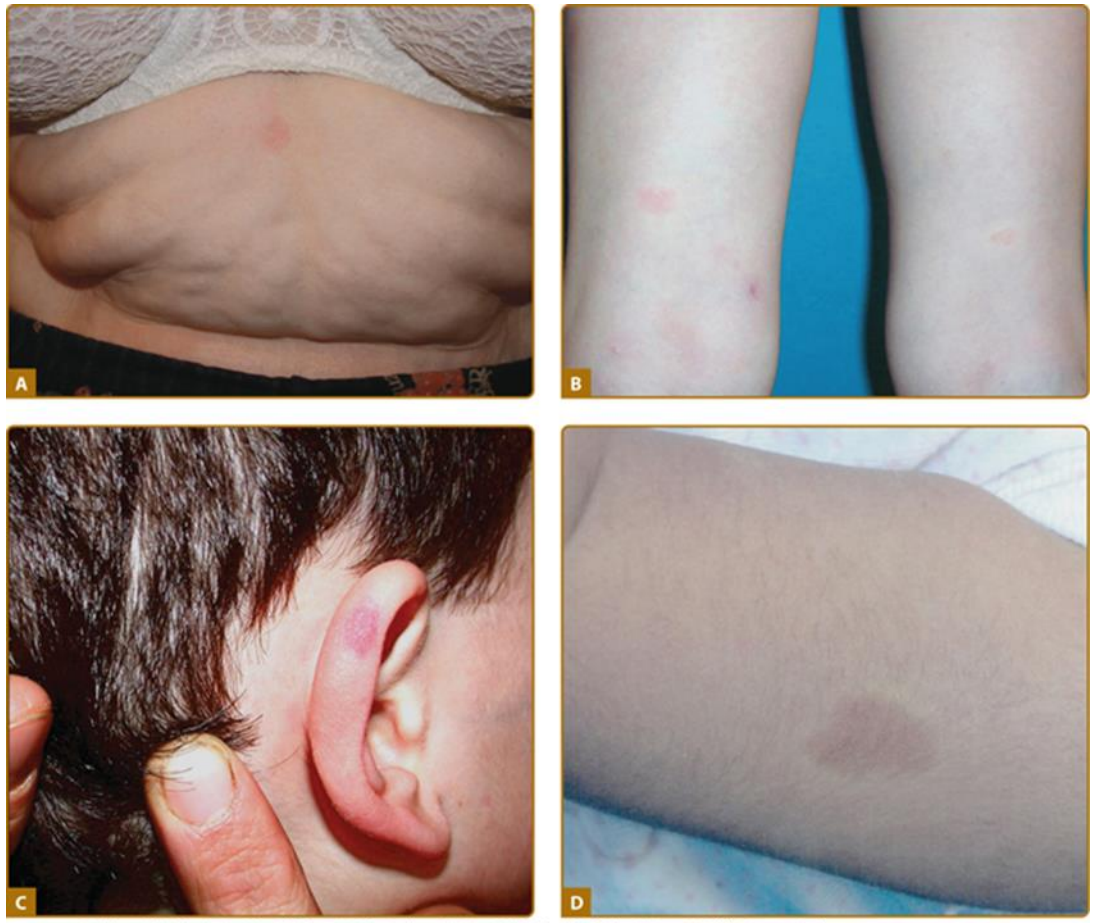


Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Clinical findings

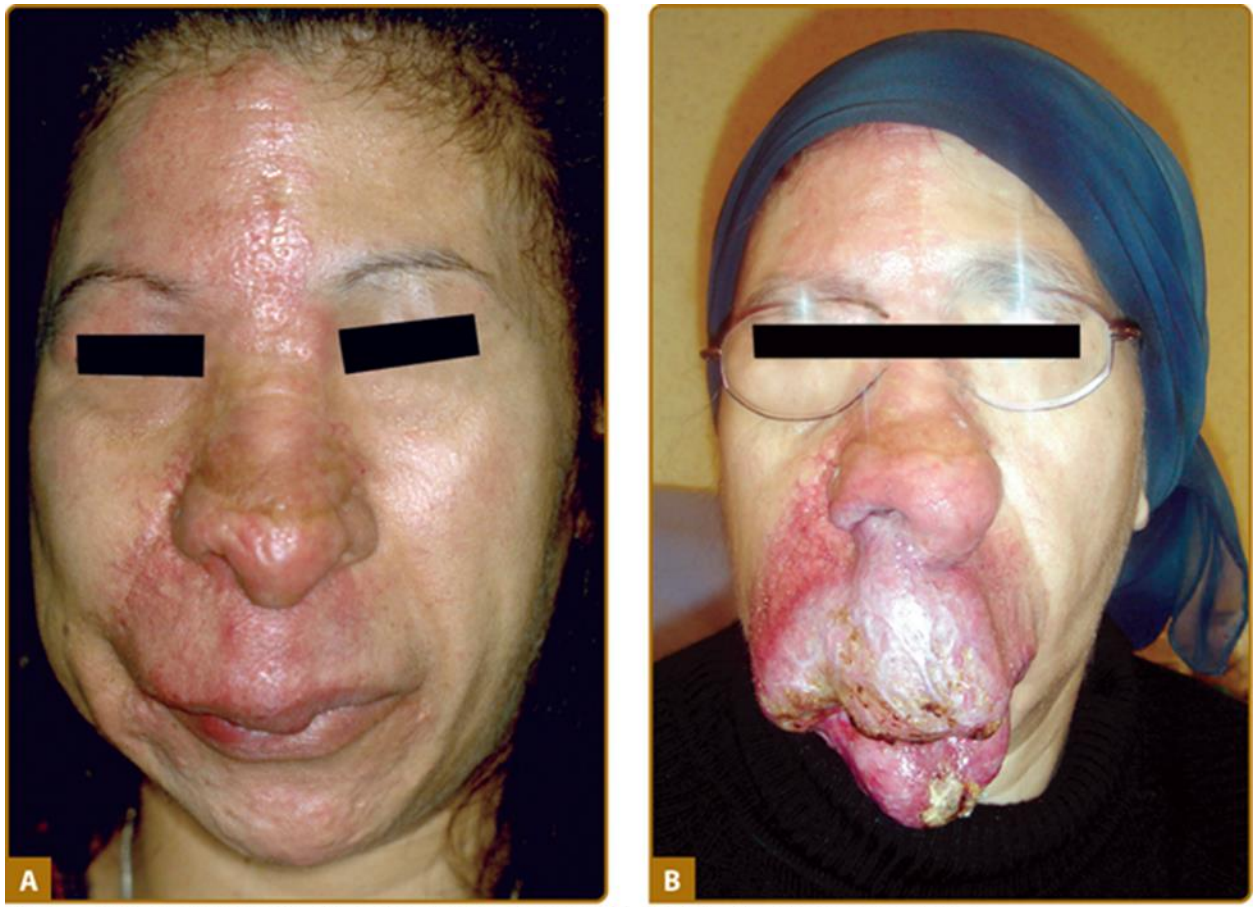
- During childhood, AVM is often mistaken for a CM or a hemangioma, as, at birth, AVM usually manifests as a faint, ill-defined, macular red stain. However palpation reveals its fast-flow component as it is warm.
- About one third of AVMs are present at birth, another one-third appear during childhood or at puberty, and the rest in adulthood. Puberty and trauma usually trigger the growth of an AVM.
- AVMs are staged using the Schobinger classification AVMs manifest as cutaneous, red-to-purple warm masses with a thrill, a bruit, or a pulsation of increased amplitude. They worsen with time evolving from stage I to III or even IV, never regre

STAGE 1	Local mass, blush or red stain Doppler ultrasound: increased vascularization
STAGE 2	+ Prominent draining veins, pulsation, thrill Doppler ultrasound: arteriovenous shunting
STAGE 3	+ Dystrophic changes
STAGE 4	+ Congestive heart failure



Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

Small atypical capillary malformations of capillary malformation-arteriovenous malformation (**A–D**). Note the pale halo (**D**).

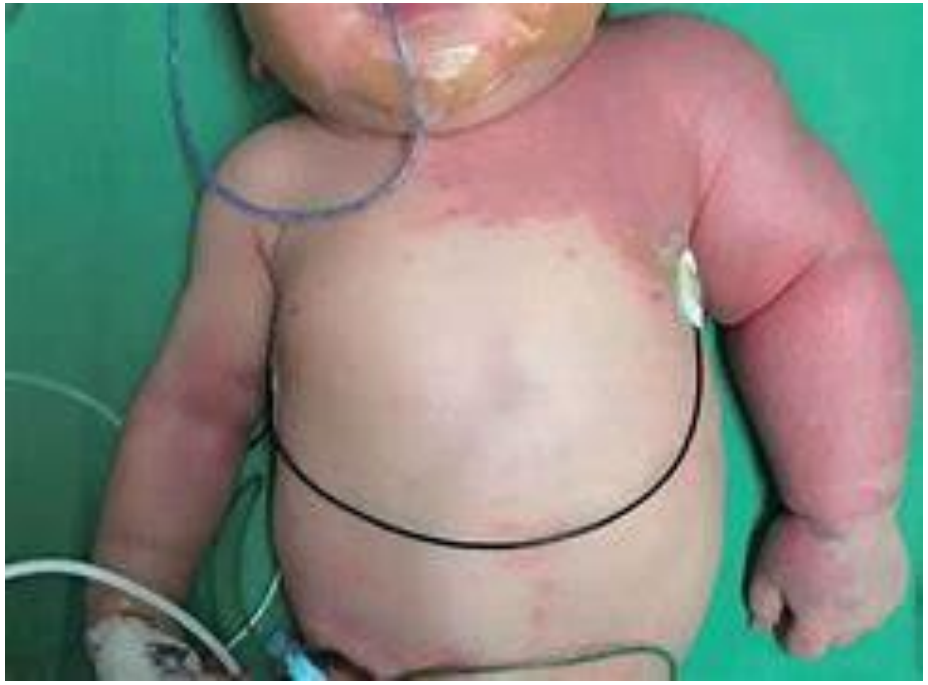


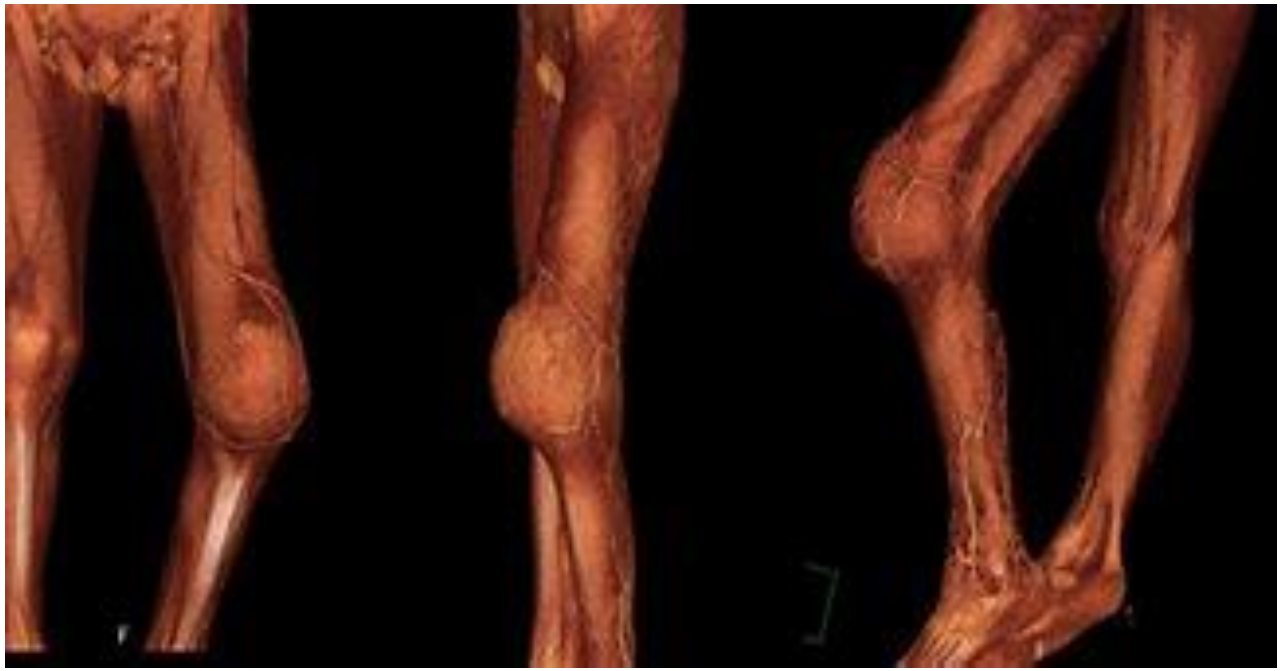
Source: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K: *Fitzpatrick's Dermatology in General Medicine, 8th Edition*: www.accessmedicine.com

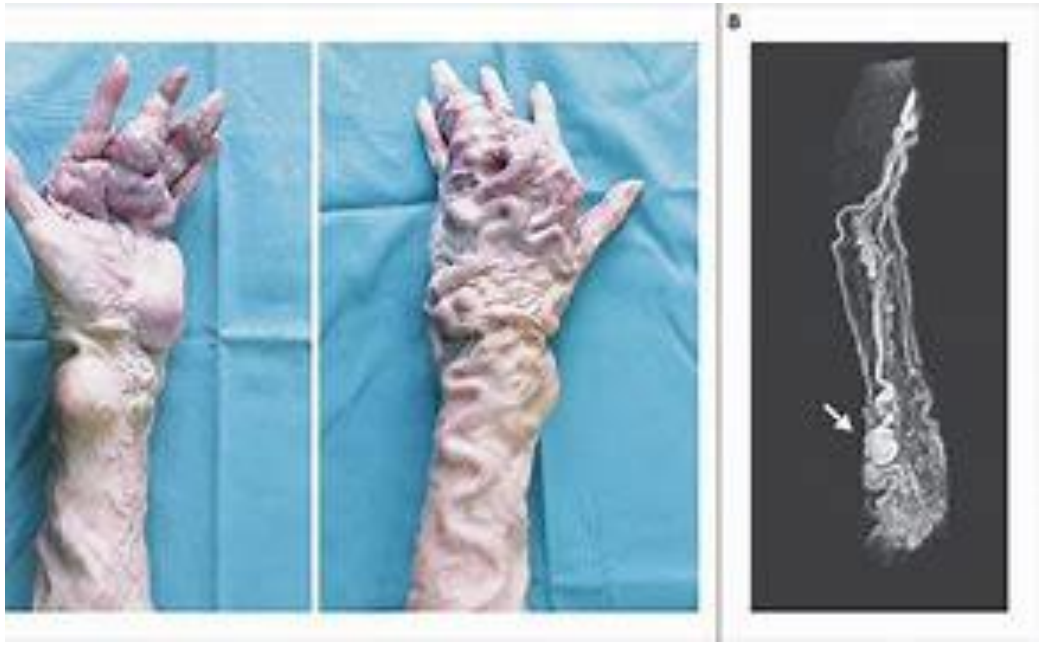
Stage 2 arteriovenous malformation (AVM) (**A**) evolving into stage 3 AVM (**B**).

Treatment

- This fast-flow vascular malformation is the most complex and difficult vascular anomaly to treat. AVM should be followed by periodic evaluation.
- In a limb AVM or multiple AVFs, elastic stockings can stabilize the lesion and protect the skin. The goal of cure of an AVM is based on obliteration (by embolization) and complete removal of the nidus or the epicenter. Early intervention for "quiescent AVM" (Schobinger Stage I) is debatable but should be considered only if complete resection is possible.
- In contrast to AVF, superselective arterial embolization alone is only palliative and is done only in unresectable complicated AVM. The embolized particles need to reach the epicenter of the lesion to avoid refilling of the nidus through new collaterals. Direct puncture of the nidus is another possibility in patients with previous arterial ligation or embolization.
- Surgical resection is usually done only after embolization, to minimize perioperative bleeding. AVM needs to be widely excised. Reconstruction often necessitates microsurgical free flap transfer.
- At least 5-year follow-up by annual Doppler ultrasound and/or MRI is mandatory after any treatment. For patients with Parkes Weber syndrome, the treatment should be as conservative as possible (e.g., elastic stockings).
- For patients with Hereditary Hemorrhagic Telangiectasia (HHT), multiple treatments have been tried to reduce bleeding, for example, topical application of anti-inflammatory drugs, laser, or surgery. Due to the extensiveness of the lesions, they all give limited free symptoms-intervals. Thalidomide has recently been successfully used to reduce the frequency and duration of nosebleeds in patients with HHT.

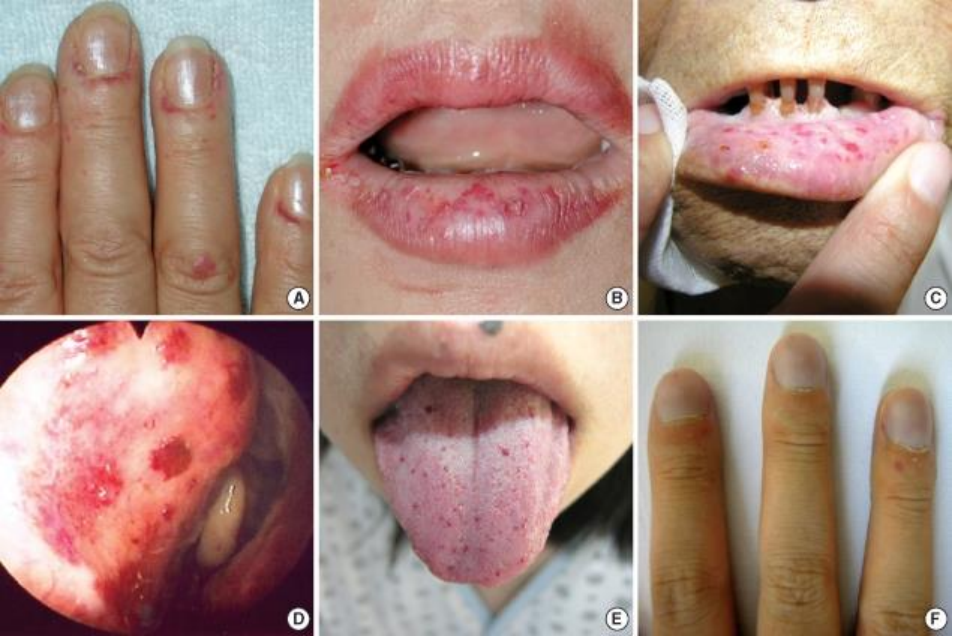


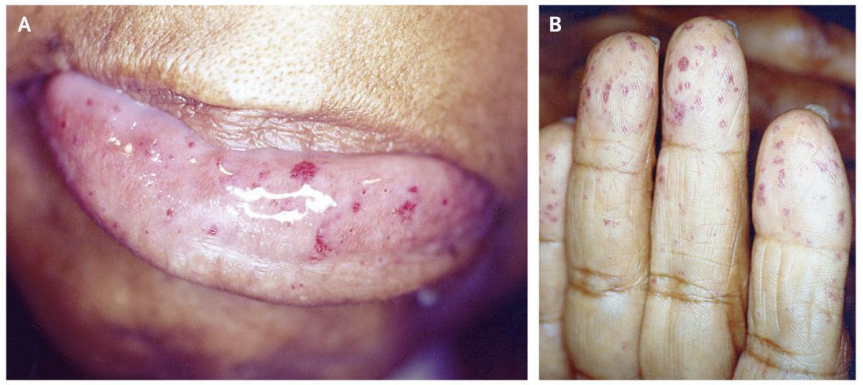


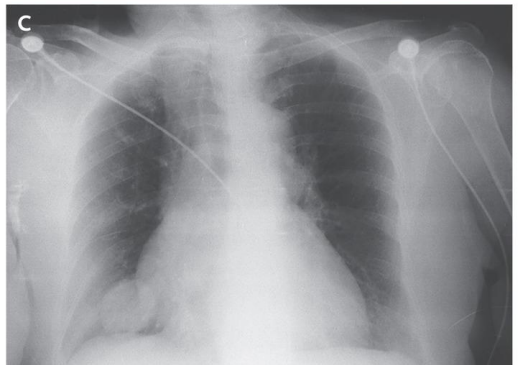


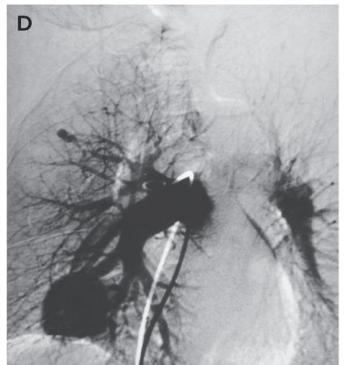
Parkes Weber Syndrome

This condition is characterized by a large, congenital, cutaneous, vascular stain on an extremity in association with soft tissue and skeletal hypertrophy of the affected limb, and with underlying multiple arteriovenous microfistulas









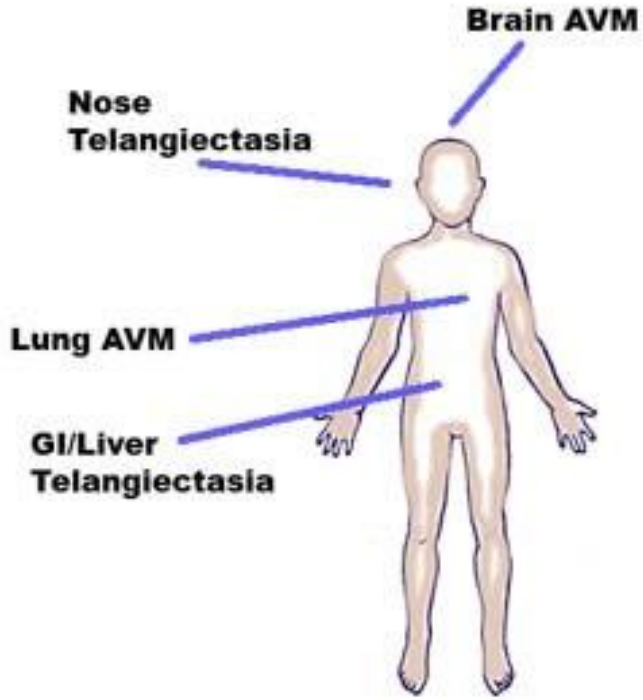
Hereditary Hemorrhagic Telangiectasia

Preventable

- Complications of Arterio-Venous Malformations (AVMs)
- Brain: hemorrhage, death
- Lung: stroke, brain abscess, hemorrhage, death

Preventable

- Complications of Small Malformed Blood Vessels (Telangiectases)
- GI Tract and Nose: Transfusion Dependant Bleeding
- Liver: liver failure, Heart failure



Hereditary hemorrhagic telangiectasia (HHT) is characterized by the presence of multiple arteriovenous malformations (AVMs) that **lack intervening capillaries** and result in direct connections between arteries and veins.

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