VASCULAR TUMORS OF CHILDHOOD

By

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Table 69-1 International Society for the Study of Vascular Anomalies (ISSVA) Classification of Vascular Anomalies

Vascular tumors	Infantile hemangioma Hemangioendotheliomas Angiosarcoma Miscellaneous
Vascular malformations	Slow-flow: Capillary malformation (CM) Lymphatic malformation (LM) Venous malformation (VM) Fast-flow: Arterial malformation (AM) Combined

Vascular tumors of childhood

- Usually benign and consist of four major types: infantile hemangioma, congenital hemangioma, kaposiform hemangioendothelioma, and pyogenic granuloma.

- They are manifested during infancy or childhood.

- May involve any location.

- **Local complications:**
Bleeding, destruction of tissue, obstruction, and pain.

- **Systemic sequelae:**
Thrombocytopenia, congestive heart failure, and death.

Vascular tumors must be differentiated from vascular malformations of childhood:

Tumor	Malformation
Not present at birth	Present at birth
Have proliferating endothelium	Quiescent endothelium
Exhibit postnatal growth	Slowly expand with growth

Table 70-1 Common Types of Childhood Vascular Tumors

Correct Biologic Term	Incorrect Descriptive Name
Infantile hemangioma	Capillary hemangioma Strawberry hemangioma Cavernous hemangioma
Congenital hemangioma	Infantile hemangioma
Kaposiform hemangioendothelioma	Hemangioma
Pyogenic granuloma	Hemangioma Lobular capillary hemangioma

INFANTILE HEMANGIOMA

Clinical Features:

- Is most common neoplasm of infancy.
- It affects about 4% to 5% of white infants.
- It is more frequent in premature infants and girls (4 : 1).
- Most single (80%) and involve head & neck (60%), trunk (25%), or extremity (15%).
- Visible at birth as a small pale spot, telangiectatic stain, or ecchymotic area.
- The median age at appearance is 2 weeks after birth.

Proliferating phase:

• During the first 9 months of life, the lesion grows rapidly, faster than the growth of the child.

Growth plateaus

•By 10 to 12 months of age, the tumor increases in size at the same rate of growth as the child.

In superficial dermis, it appears red.

In deep dermis, overlying skin may be bluish normal.

Involuting phase:

Begins at 1 year of age and is characterized by shrinkage of the tumor.

Involution is completed in most children by 3.5 years of age.

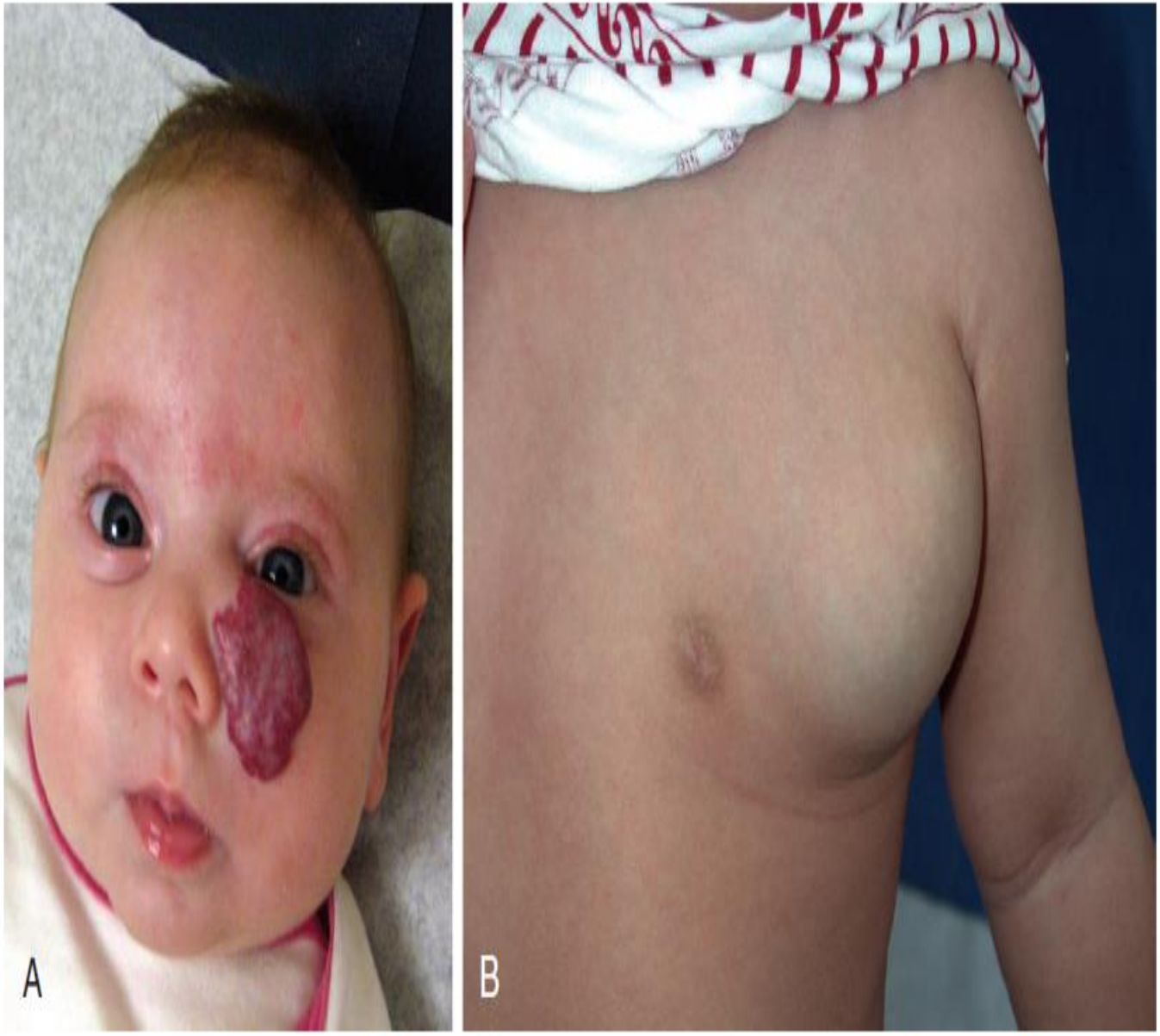


Figure 70-1 Proliferating infantile hemangioma. **A,** A 2-month-old girl with a superficial tumor. **B,** A 9-month-old girl with a deep lesion of the chest.

50 % of patients have residually damaged skin, fibrofatty tissue, telangiectasias, discoloration, scarring, or redundant skin

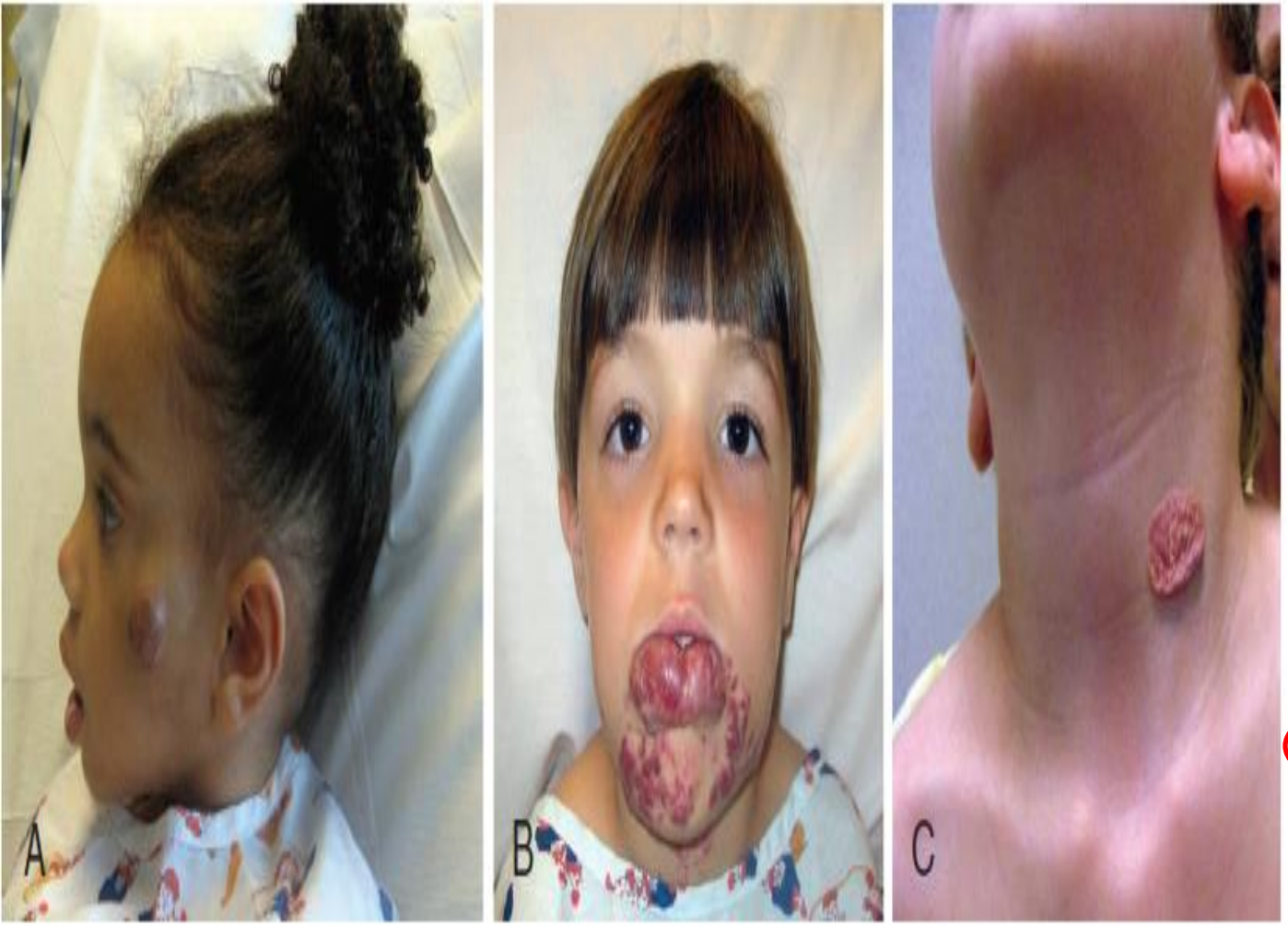


Figure 70-2 Involuting infantile hemangioma. **A,** A 3-year-old girl with a residual fibrofatty lesion of the cheek. **B,** A 4.5-year-old girl with fibrofatty residuum and telangiectasias of the lower lip. **C,** A 4-year-old boy with redundant skin of the neck.

Multiple Hemangiomas:

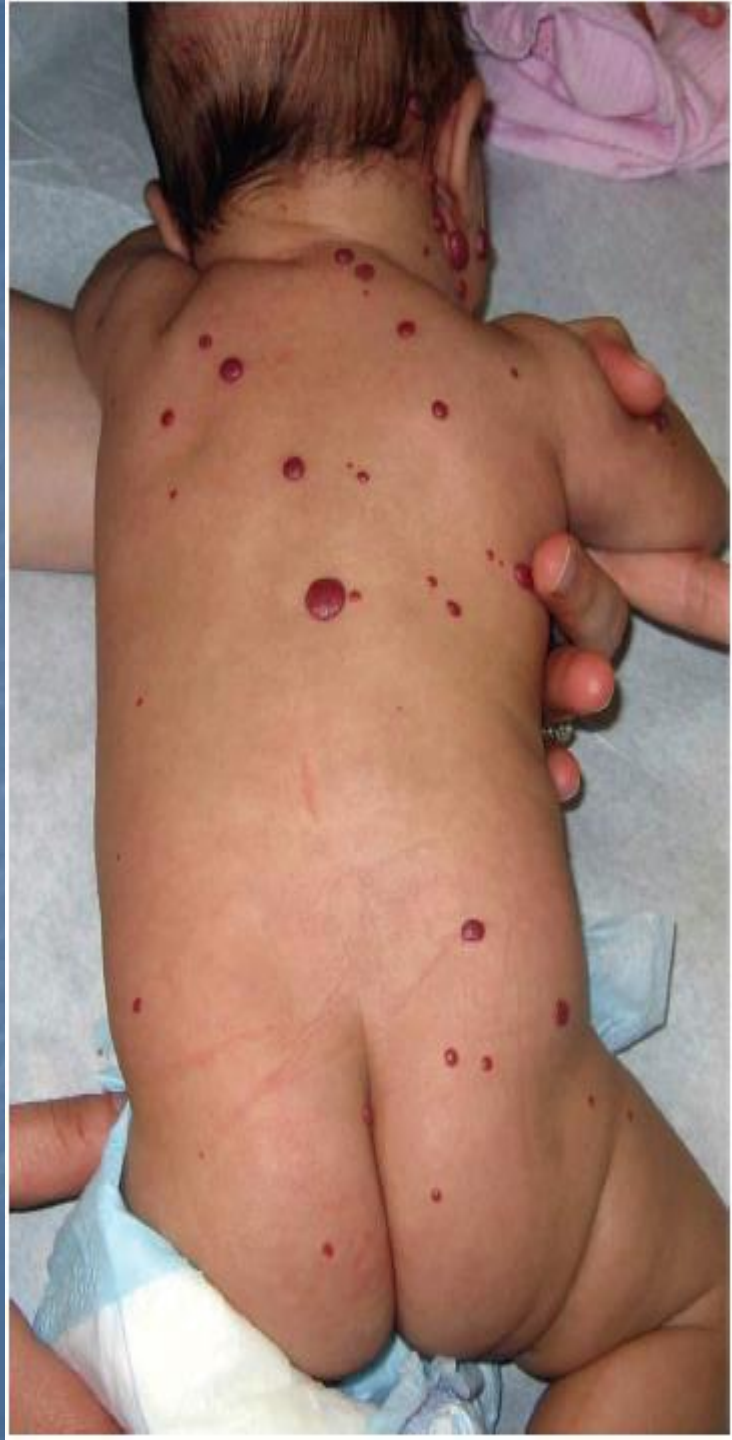
On occasion, children have **multiple cutaneous hemangiomas** (hemangiomatosis).

The lesions usually are **less than 5 mm** in diameter and are **domelike.**

Individuals with **≥ 5** have a **16% chance of having visceral lesions** that are almost always located in the **liver**.

Patients are **screened with ultrasonography** to rule out hepatic hemangiomas.

The **brain, gut, and lung are rarely involved.**



Hepatic Hemangiomas

- Liver is most common site of extracutaneous hemangioma.
- Hepatic hemangiomas may be focal, multifocal, or diffuse.
- They are asymptomatic and discovered incidentally.

- A focal lesion is a rapidly involuting congenital hemangioma and not associated with cutaneous hemangiomas

- Multifocal hepatic hemangiomas are infantile hemangiomas and may be associated with multiple cutaneous lesions.

- Patients with diffuse hepatic hemangioma must have thyroid-stimulating hormone monitoring.
- Massive intravenous thyroid hormone replacement may be necessary to prevent irreversible mental retardation until the hemangioma regresses.

Lumbosacral Location:

- An infant with a large midline infantile hemangioma involving the lumbosacral area has an approximately 1 in 3 chance of having an underlying spinal anomaly (e.g., tethered cord, lipoma, intraspinal hemangioma).

- MRI is performed between 3 and 6 months of age to rule out an occult spinal dysraphism.

PHACES Association

- PHACES association (posterior fossa brain malformations, hemangioma, arterial anomalies, coarctation of the aorta and cardiac defects, eye abnormalities, sternal clefting/ supraumbilical raphe)

Diagnosis

History and Physical Examination:

Not present at birth, although a light stain may be noted in 50% of infants.

At age of **2 weeks**, the lesion will **grow rapidly** larger.

By **12 months** of age, will **begin to involute**, becoming **gray, soft, and smaller.**

A **superficial** appears **red**, whereas **deeper** lesions may not be noted until later as a **blue** mass visualized through the skin.

Imaging

• Less than 10% of infantile hemangiomas require imaging for a definitive diagnosis to be obtained.

US is the first-line study and shows a well-circumscribed hypervascular mass.

Low-resistance arterial waveforms are present with increased venous drainage.

By MRI, during the proliferative phase, shows a parenchymal mass with dilated vessels .

An involuting phase has increased lobularity and adipose tissue & reduced number of vessels.

Histopathology:

Less than 1% of infantile hemangiomas require histopathologic evaluation for diagnosis.

Biopsy is indicated if **malignant disease** is suspected or if the **diagnosis remains unclear** after imaging.

A **proliferating** lesion shows **tightly packed capillaries with plump endothelial cells** and **minimal stroma.**

During **involution**, **reduced capillarie**, enlargement of channels, and **increased stroma**.

Infantile Hemangioma expresses an erythrocyte-type glucose transporter (**GLUT1**) that can be used to **differentiate the Tumor from other vascular tumors and malformations.**

Management

Observation:

Observation is the mainstay of management because 90% of infantile hemangiomas are small, are localized, and do not involve aesthetically or functionally important areas.

Parents should be reassured.

Wound Care:

During the proliferative phase, approximately 16% of lesions will have skin ulceration.

Tumors can be covered with petroleum gauze barrier.

Ulcerated are treated with soap and water irrigations twice/d.

Small areas are covered with topical antibiotic.

Topical Pharmacotherapy:

Has minimal efficacy, if in the deep dermis and subcutis. Topical corticosteroid (e.g., clobetasol) may be effective for small, superficial lesions, but hypopigmentation and skin atrophy can occur.

Intralesional Corticosteroid

is indicated for small, well-localized infantile hemangiomas that obstruct vision or the nasal airway or are at risk for damaging an aesthetically important area (eyelid, lip, nose).

Triamcinolone (Kenacort) stabilizes growth in 95% of cases, and 75% will decrease in size.

Injections are administered at 6-week intervals during the proliferative phase.

Risks include subcutaneous fat atrophy and ulceration. Blindness has been reported due to embolic occlusion of the retinal artery.

Systemic Pharmacotherapy

Indicated for a problem hemangioma that is too large to be treated with a local injection.

Two primary drug options: prednisolone and propranolol.

Prednisolone (3 mg/kg) is given once in the morning for 1 m. Dose is then tapered every 2 to 4 weeks until it is discontinued between 10 and 12 months of age.

S/E:
20% develop a cushingoid appearance that resolves when the drug is discontinued.

12% have a ↓ in their gain in height but return to their pretreatment growth curve by 24 months of age.

Propranolol:

(2 mg/kg/day) is divided into two or three daily doses.
Then tapered until it is discontinued around 12 months of age.

Risks (<5%) :

-Bronchospasm -Bradycardia -Hypotension
- Hypoglycemia -seizures, and hyperkalemia.

Contraindications :

- Asthma or reactive airway disease
- Glucose abnormalities -Congenital heart disease
- Hypotension -Bradycardia

- **Second-line drugs for treatment of infantile hemangioma include interferon and vincristine.**

- **They are rarely used because tumors will respond to corticosteroid or propranolol.**

- **Interferon is no longer recommended in children younger than 12 months because it can cause neurologic problems.**

- **Vincristine may be considered in the unlikely event that a child has failed to respond to or has a CI to prednisolone and propranolol.**

Embolic Therapy:

Large infantile hemangiomas, most commonly multifocal or diffuse hepatic lesions, can cause high-output CHF.

Embolization indicated for initial control of HF while the therapeutic effects of systemic drug therapy are pending.

Embolization has little or no role in hepatic hemangioma without heart failure.

Drug therapy should be continued after embolization until the child is approximately 12 months of age.

Laser Therapy:

Almost all hemangiomas are beyond the reach of the laser.

The laser penetrates only 0.75 to 1.2 mm into the dermis and thus affects the superficial portion of the tumor only.

Although lightening of the color may occur, the mass is not affected & increased risk of skin atrophy and hypopigmentation

The thermal injury delivered by the laser to ischemic dermis increases the risk of ulceration, pain, bleeding, and scarring.

The pulsed dye laser is indicated, during involuted phase to treat residual telangiectasias.

The carbon dioxide laser is indicated, during proliferative phase to treat a subglottic hemangioma.

Operative Management:

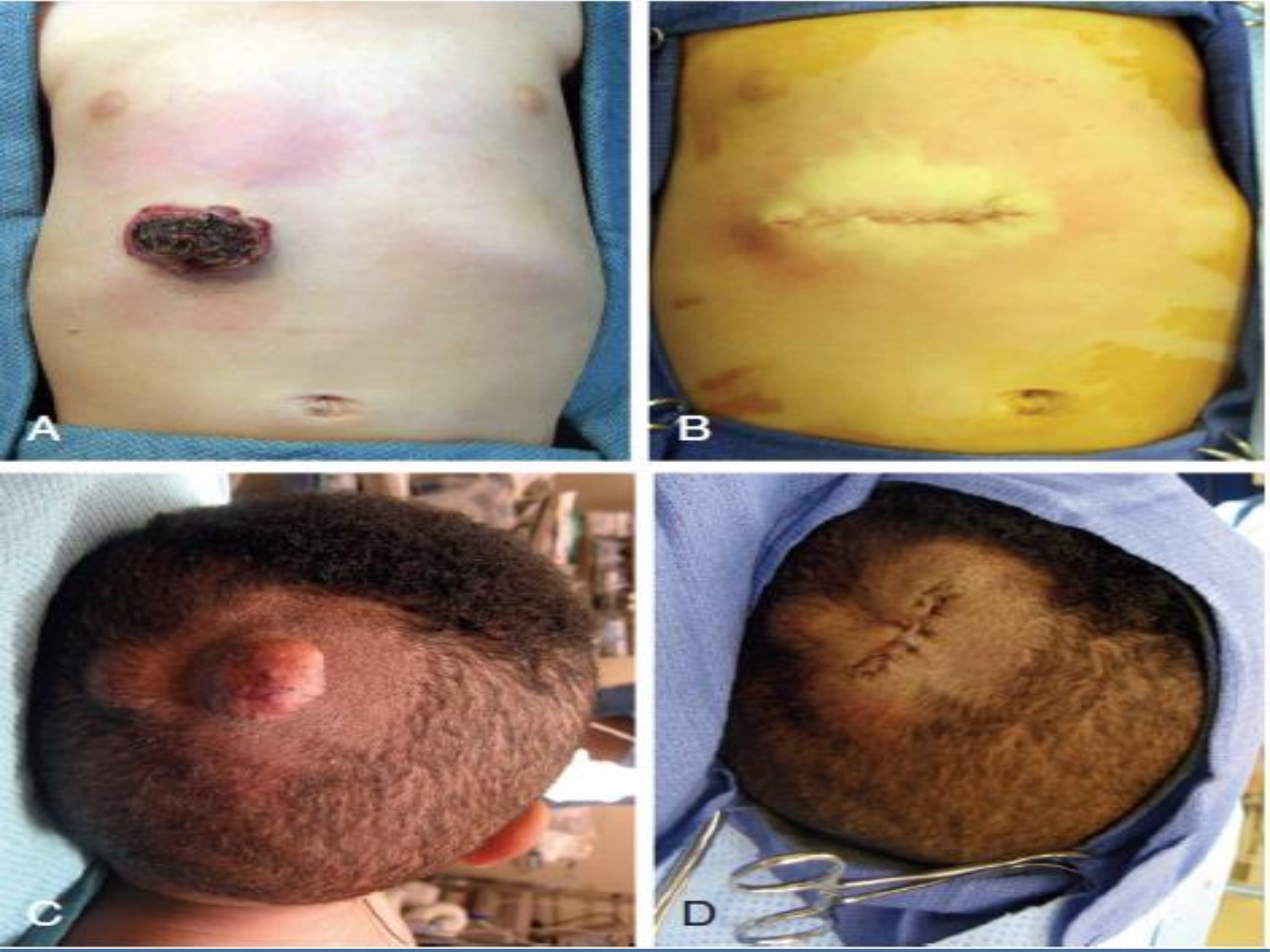
The tumor is highly vascular thus the patient is at greater risk for blood loss, iatrogenic injury.

Anesthetic morbidity is also higher during infancy compared with children older than 12 months.

A well-localized or pedunculated lesion that is ulcerated or bleeding may be excised

Operative intervention during the involuted phase (after 3 years of age) is safer

Because the lesion has been permitted to shrink, the area to be excised and reconstructed is smaller and thus the outcome is superior.


A
B
C
D

CONGENITAL HEMANGIOMA

Clinical Features:

- **Full grown at birth** and no postnatal growth.

It is:

- **Red** with coarse **telangiectasias**
- **Central pallor** with **peripheral pale halo**
- **Solitary** with an average diameter of 5 cm.
- More **common in extremities**
- **Equal sex** distribution.
- Two types : **rapidly involuting and noninvoluting**

Rapidly involuting : RICH

Involutes quickly after birth;
50% have completed regression by 7 months of age.
The remaining lesions are fully involuted by 14 months.

Affects head or neck (42%), limbs (52%), or trunk (6%).

After involution, does not leave adipose component.
Often, results in atrophic skin and deficient SC fat.

Noninvoluting : NICH

Does not involute after birth.

Affects head or neck (43%), limbs (38%),or trunk (19%).

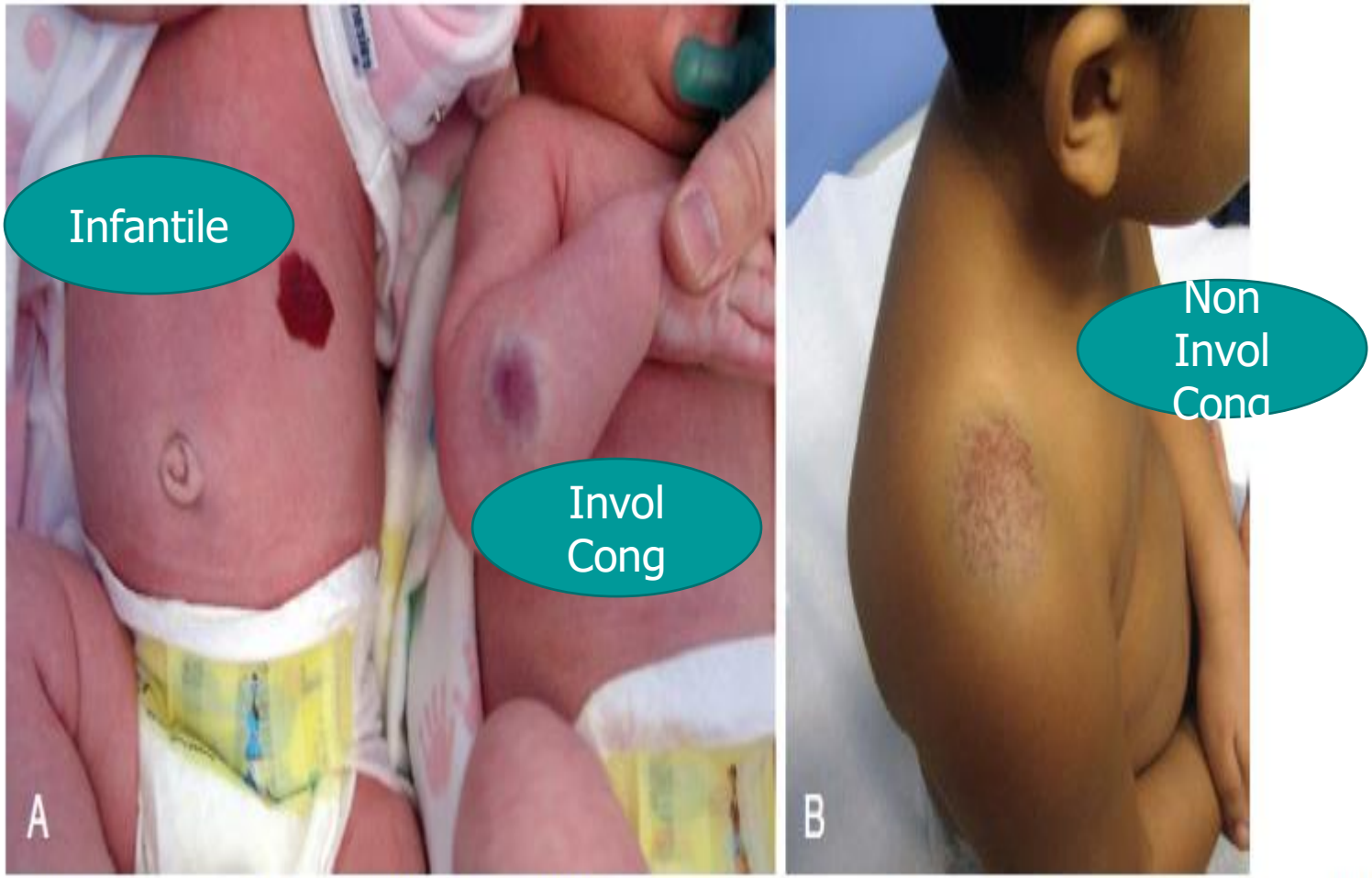


Figure 70-6 Congenital hemangioma. **A,** The 6-week-old twins illustrate the difference between an infantile hemangioma and a rapidly involuting congenital hemangioma. The twin on the left has an abdominal infantile hemangioma. The lesion, which was noted to be a dime-sized, light red area at 2 weeks of age, has grown significantly larger. The twin on the right was born with a rapidly involuting congenital hemangioma of the upper extremity that was bright red at birth. Postnatally, it has decreased in size and faded in color. **B,** A 6-year-old boy with a noninvoluting congenital hemangioma of the shoulder. The lesion was present at birth and has remained unchanged.

Management:

- Most are managed by observation.
- Drug treatment does not accelerate its involution.
- If atrophic tissue remains, reconstruction with grafts (dermal, fat, acellular dermis) or resection before 4 years of age.
- Rarely, a large lesion may cause high-output HF and thus require excision or embolization.
- Its color may be lightened with pulsed dye laser treatment or sclerotherapy.

KAPOSIFORM HEMANGIOENDOTHELIOMA

Clinical Features

- Is a rare vascular neoplasm 1/100000 that is locally invasive but does not metastatic.

- Multifocal cases are rarely reported and should be confirmed by multiple-site biopsy when suspected.

- 60% are manifested in the neonatal period and 93% are manifested in infancy .

- Has an equal sex distribution, is solitary, and affects the head or neck (40%), trunk (30%), or extremity (30%).

- It is reddish purple with an ill-defined border, often first mistaken for ecchymosis, and is often larger (>5 cm).

Hirsutism in involved skin, probably secondary to cutaneous edema, is common and transient.

The natural history includes expansion in infancy and early childhood and then partial regression.

When biopsy is performed, spindled lymphatic endothelial cells, and fibrosis is common and may cause pain and contractures.

More than 50% of affected children exhibit Kasabach-Merritt phenomenon, a profound thrombocytopenia (<25,000/mm3) that has a mortality of 12% to 24%.

KMP shows Consumptive coagulopathy with ↓ fibrinogen, ↑ PT & PPT , ↑FDP and D-dimers is present in severe cases.

• Large tumors, those that present during infancy, and visceral or muscle involvement increase the risk of KMP.

• kaposiform hemangioendothelioma has overlapping histopathologic and clinical features with tufted angioma, including Kasabach-Merritt phenomenon.

• Although kaposiform hemangioendothelioma is considered a pediatric tumor, adult-onset and posttraumatic cases have been described.

• In adults, the mean age at diagnosis is 43 years (range, 22-64 years), the majority are male(80%), and distribution is similar to that in children.

• Lesions presenting in adulthood are smaller than those affecting children; two thirds are less than 2 cm (average, 4.5 cm; range, 0.6-15 cm).

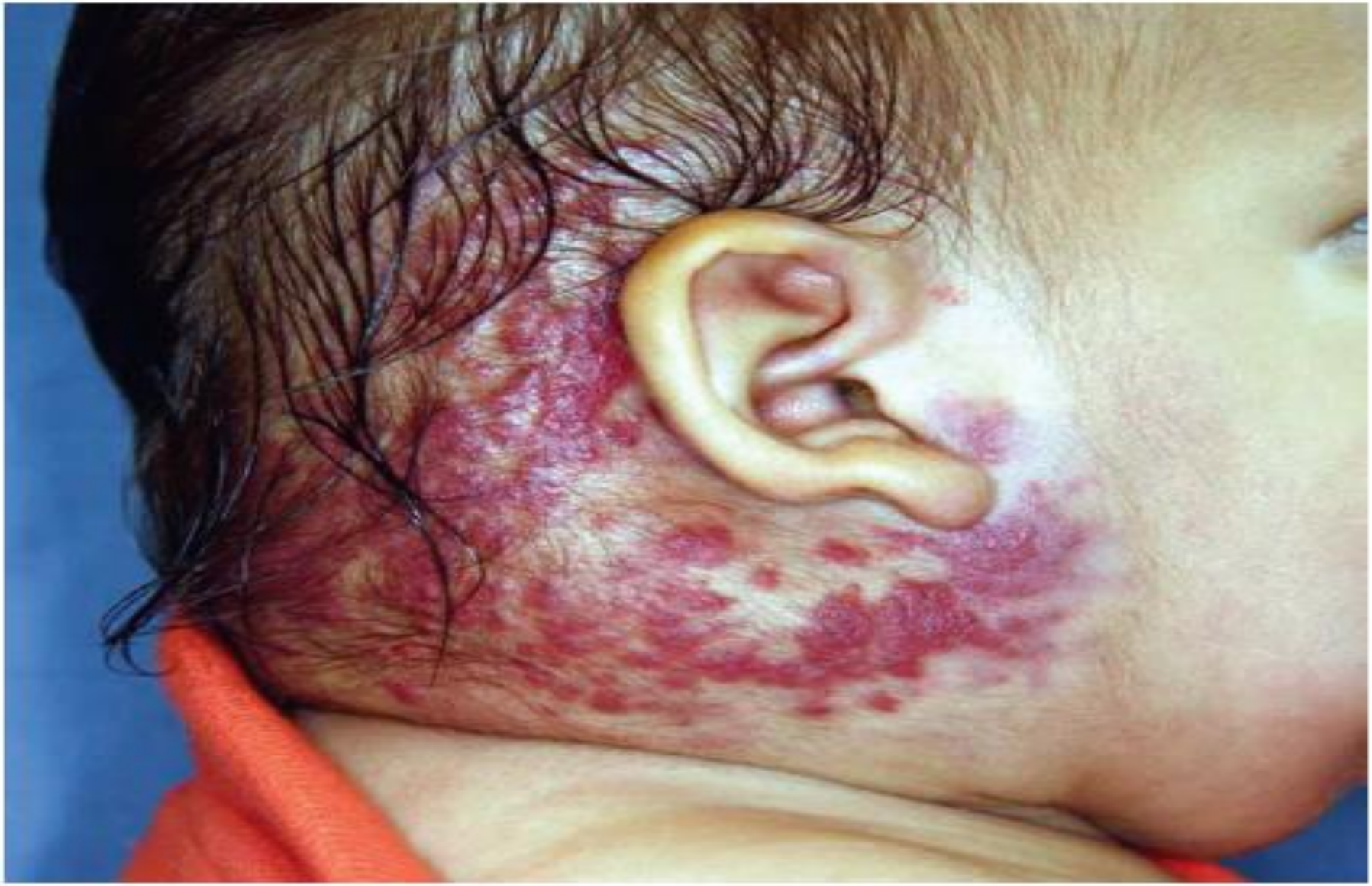


Figure 70-7 A 1-year-old boy with kaposiform hemangioendothelioma complicated by Kasabach-Merritt phenomenon. His lesion partially regressed and his platelet count improved after treatment with vincristine.

Management:

Treatment depends on size of lesion and presence of KMP.

Excellent responses to medical therapy.

Surgical resection is rarely indicated as are large, infiltrative, and involve multiple tissues, preventing complete excision.

Patients with unresectable tumors or KMP require systemic treatment to prevent life-threatening complications.

Patients without KMP are treated to minimize fibrosis and thus chronic pain, stiffness, and contractures.

Responds best to vincristine (~90%), followed by interferon (~50%), and corticosteroid (~10%).

- Standard initial therapy is corticosteroid 2 mg/kg/day and vincristine 0.05 mg/ kg intravenously weekly.

- Corticosteroids are tapered once KMP begins to improve, often within 4 to 6 weeks.

- Length of vincristine therapy is about 6 months & continues until KMP is resolved .

- Sirolimus (Rapamycin) led to improvement in refractory case with KMP, and this has been repeated in several cases.

- Sirolimus offers potential advantages of oral administration and dose adjustment to serum trough levels to minimize side effects.

- Interferon is rarely used because of concern of spastic diplegia when it is used in infancy.

- KMP is a destructive thrombocytopenia; platelet transfusions cause a transient improvement in platelet counts and active bleeding but result in significant swelling and pain because the platelets are trapped in the lesion.

- Platelet transfusion should be avoided unless there is active bleeding or a surgical procedure is indicated.

- Anticoagulation (e.g., heparin) should not be given because it significantly increases bleeding risk and thrombocytopenia and may stimulate growth of the tumor.

- Improvement in KMP is an accepted biomarker of treatment response, and serial evaluations of cutaneous lesions .

PYOGENIC GRANULOMA

Clinical Features:

- Has been called lobular capillary hemangioma.
- It is a solitary, red papule that grows rapidly, forming a stalk.
- It is small, with an average diameter of 6.5 mm; 75% of lesions are less than 1 cm.
- The male-to-female ratio is 2 : 1.
- Complicated by bleeding (65%) and ulceration (35%).
- The mean age at onset is 6-7 years; only 12% develop during the first year of life.
- The majority affect the skin (88.2%) and mucous membranes in (11.8%).

- They are distributed on head or neck (62%), trunk (19%), upper extremity (13%), or lower extremity (5%).

- Within the head and neck, affected sites include the cheek (30%), oral cavity (15%), scalp (10%), forehead , eyelid and lips (9%).

- 25 % of patients have a history of preexistent trauma or an underlying cutaneous condition (including capillary malformation, dermatologic disorder, viral infection, and insect bite).

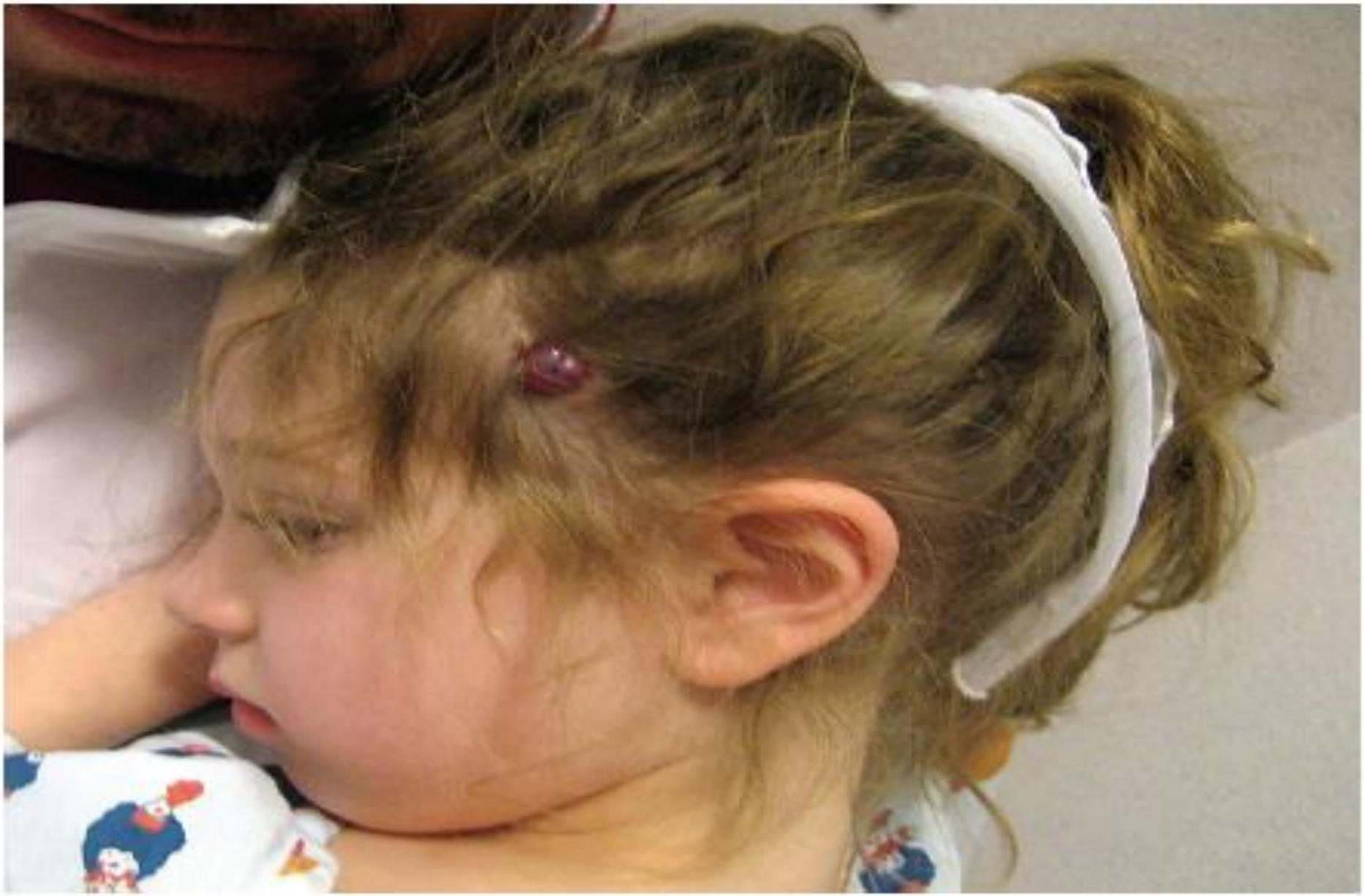


Figure 70-8 A 5-year-old girl with a 6-month history of pyogenic granuloma of the scalp. The lesion was complicated by bleeding and was excised.

Management

- May become temporarily smaller after bleeding and crusting, which is followed by regrowth.

- Treatment methods include : curettage, shave excision, laser therapy, or excision.

- Because the lesion can involve the reticular dermis, it may extend beyond reach of pulsed dye laser, electrocauterization, or shave excision.

- These modalities have a recurrence rate as high as 43%.

- Definitive treatment requires fullthickness skin excision, which has an approximately 100% cure rate.

Thank you for listening..

Any questions?