

Pediatric Facial Plastic and Reconstructive Surgery

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Chapter 22: Dermatologic Diseases in Pediatric Facial Plastic and Reconstructive Surgery

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Dermatologists and facial plastic and reconstructive surgeons enjoy close collaboration in the evaluation and treatment of patients with cutaneous disorders of the head and neck. Each specialty can benefit from the scientific and therapeutic expertise of the other. Although many of the diseases mentioned in this chapter are covered elsewhere in this book, discussion is repeated here because a different perspective is often valuable. Particular emphasis will be devoted to melanocytic and vascular disorders.

Melanocytic Lesions

Disorders of melanocytes cause abnormalities of pigmentation resulting in skin discolorations of tan, brown, bluish-gray, blue, and black. Mixtures and gradations of these colors may be seen, as well as the lack of melanocytic pigment altogether, as in albinism and vitiligo. The vast majority of pigmentary abnormalities seen in children are benign. Malignant melanoma in children is distinctly rare. We will discuss congenital and acquired benign melanocytic tumors, focusing on the precursor lesions of melanoma that the facial plastic surgeon will most likely treat.

Mongolian Spot

A mongolian spot is a flat, grayish-blue discoloration with either sharp or ill-defined borders. Histologically it is caused by the presence of melanocytes in the mid- to deep dermis. Lesions are usually seen over the spine or occasionally on the shoulders or neck. Mongolian spots are a normal variant in Asians (90% to 99% of infants), blacks (96% of infants), and in more darkly pigmented races such as Hispanics (46% of infants). Most lesions involute by age 5 years. Histologically, they are similar to the nevus of Ota and nevus of Ito.

Nevus of Ota and Nevus of Ito

The nevus of Ota or nevus of Ito is a flat, hyperpigmented lesion that may be bluish-purple to bluish-gray to bluish-brown in color. Discoloration within the area of skin may be diffuse or concentrated in small macules against a background of fainter hyperpigmentation. The spots are typically larger than Mongolian spots and they do persist. Sixty percent are present at birth, and 40% develop in adolescence. The nevus of Ota is located within the dermatomal distribution of the ophthalmic and/or maxillary branch of the trigeminal nerve. Involvement of mucosae is common, including the hard palate, pharynx, and nasal mucosa as well as the auditory canal. In addition, ocular structures are pigmented in around 50% of cases. The nevus of Ito is usually unilateral and in the rough dermatomal distribution of either the lateral brachial or posterior supraclavicular cutaneous nerve branches; thus, the supraclavicular, deltoid, or scapular regions are most often affected.

Camouflaging makeup is the only effective therapy for Mongolian spot or nevus of Ota/Ito, although the Q-switched ruby laser shows promise. Since the melanocytes are deep in the dermis, wounding procedures such as dermabrasion or chemical peels are ineffective. Likewise, the yellow dye pigmented lesion laser will not reach the necessary depth. Surgical intervention is not required unless melanoma supervenes; fortunately, melanoma is only slightly more common in patients with either the nevus of Ota or nevus of Ito than in unaffected subjects.

Melanocytic Nevi

Melanocytic nevi (nevocellular nevi; moles) are benign growths composed of nests of melanocytes. Whether they are hamartomas or true neoplasms is controversial. Melanocytes are derived from the neuroectoderm, and the migration of melanoblasts from the skin during development may help explain the different presentations of congenital melanocytic nevi.

Melanocytic nevi may be either congenital or acquired. They are congenital in only 1% of Caucasians. However, most Caucasians will gradually acquire melanocytic nevi, with a peak number of lesions around age 15 in males and in the third decade in females. Following a peak of about 15 to 50 lesions, nevi undergo gradual involution. By the ninth decade, the average Caucasian has only four nevi.

Acquired Melanocytic Nevi

Acquired melanocytic nevi undergo a typical life cycle. Lesions start out flat, gradually become raised, then involute completely. There are color changes and histologic appearances corresponding to each of these stages. Flat, dark brown lesions are junctional nevi, in which nevus cell nests are confined to the lowest portions of the epidermis at the dermo-epidermal (DE) junction. Lesions that are medium-brown to flesh-colored and are dome-shaped have nevus cell nests both at the DE junction and in the upper dermis. These are termed compound nevi. Soft, rounded, tan to flesh-colored lesions are known as intradermal nevi, since nevus cell nests are confined to the dermis alone.

Prediction of the histology of the lesion will help determine the optimal mode of excision. If one is suspicious of malignant melanoma, it is desirable to remove the entire lesion, down to subcutaneous tissue, with a 2-mm margin. If the specimen is found to be a melanoma, a wider excision is required. When an entirely banal-appearing elevated nevus is removed for cosmetic reasons, it is axiomatic that cosmesis be maximized. Often "shave" or tangential excisions give the best results. It is easy to see that a junctional nevus is most likely, and an intradermal nevus least likely, to be completely removed by tangential excision. However, even in the case of intradermal nevi, cosmesis is often best served by a tangential excision, with the understanding that reexcision may be required. The lesion is most easily removed after distending it with local anesthetic, making the surrounding skin taut by stretching with the contralateral hand, and then applying either a razor blade (the double-edged variety is sharpest) or a No 15 blade with sweeping motions in a slightly concave arc.

Congenital Melanocytic Nevi

By definition, congenital melanocytic nevi (CMN) are present at birth or shortly thereafter; all other nevi are acquired. The CMN is typically medium to dark brown in color. CMNs are typically raised, papillated, verrucous, and often hairy. They are often large; almost any nevus larger than 1.5 cm in diameter is congenital, whereas almost every nevus smaller than 7 mm is acquired.

Some histologic features may be suggestive of CMN, but none of them is definitive. Histologic features suggestive of congenital nevi are the following:

1. nevus cells present in the lower two-thirds of the reticular dermis in almost all cases, and into the subcutaneous layers in more than half of cases;
2. nevus cells disposed between collagen bundles or in "Indian" files;
3. nevus cells commonly involving appendages, nerves, and vessels in the lower two-thirds of the reticular dermis or subcutaneous tissue.

In addition, there may be a fibroblastic stromal response in the dermis, giving CMN a thickened, pachydermatous texture. Horn cysts of the epidermis may be seen, especially in verrucous CMN.

There are two principal considerations in the management of CMN: the increased incidence of melanoma in giant CMN and the cosmetic morbidity of CMN of any size. The larger the lesion, the greater the potential for malignancy. There is no doubt that there is an increased incidence of malignant melanoma in giant CMN. Data are less conclusive for smaller lesions.

The definition of a "giant" CMN in the literature varies. Definitions include lesions greater than 20 cm in the largest dimension, lesions involving the major portion of an anatomic area, lesions larger than 144 square inches, or lesions that cannot be excised and their defect closed primarily in a single setting. Kopf's criteria seem the most practical: giant CMN are 20 or greater, medium CMN are greater than 1.5 cm but less than 20 cm in diameter, and small CMN are 1.5 cm or smaller.

Giant CMN are sometimes termed "garment" or "bathing trunk" nevi because they may encase an anatomic unit such as the trunk or an extremity. Cosmetic morbidity is considerable. Giant CMN are typically hairy (hence, the term "giant hairy nevus"), irregular in texture and shape, and varied in coloration, ranging from tan to brownish-black. They occur in less than 1 in 20,000 births.

Patients with giant CMN have an increased lifetime risk for development of malignant melanoma, variously estimated to be between 2% and 20%. Most series find an incidence of malignant transformation around 8% to 9%.

It is important to note that the majority of melanomas associated with giant CMN appear before the age of 5 years. This fact is especially impressive since melanoma is least

common in the first decade of life. Only 2% of all cases of melanoma occur in childhood.

Another risk of giant CMN is leptomeningeal melanocytosis, in which nevus cells involve the brain, spinal cord, or meninges. It is seen in large CMN involving the head or midline of the back. Proliferation of cells or supervening melanoma may develop and result in a mass effect causing hydrocephalus, nerve damage, or seizures. Prognosis is poor in these patients.

To prevent melanoma and improve cosmesis, complete surgical excision of giant CMN is advised. Excision should be performed as soon as practical, taking into account the patient's general health and risk for complications of general anesthesia. Excision should extend down to and include muscle fascia, since nevus cells may infiltrate to this level. Dermabrasion, cauterization, laser vaporization, cryotherapy, or any other superficial modality does not ablate the majority of nevus cells and should not be used. Nevi may extend too widely or deeply (eg, to periosteum or bone) to make complete excision possible. In these patients the most abnormal areas should be excised, with close follow-up of the CMN that remains. The dermatologist can help identify the most abnormal portions of the nevus.

Data are less conclusive regarding the lifetime risk for developing melanoma in small and medium CMN. Prospective studies have not been done. Retrospective studies using historical data (ie, whether the patient has developed melanoma in a precursor nevus thought to have been present at birth) have shown that patients with small CMN have a 21-fold increase in melanoma incidence. Correspondingly, patients who develop melanoma in nevi with histologic features typical of CMN were found to have a three- to tenfold increase in melanoma. However, retrospective data are not always reliable, especially when relying on historical data. In addition, since congenital and acquired melanocytic nevi may appear similar histologically, lesions cannot be definitively identified as congenital on histologic grounds.

Regardless of the relative risk, melanoma may develop in small or medium CMN; there are, however, important differences from melanoma seen in giant CMN. Melanoma in small or medium CMN usually develops after age 12 and begins with radial growth typical of superficial spreading melanoma that is more easily detected. Melanoma in giant CMN, on the other hand, most commonly develops before 5 years of age and, because of predominantly deep histologic components, is often nodular or metastatic before 5 years of age and, because of predominantly deep histologic components, is often nodular or metastatic before it is detected. In practical terms, this means that the decision whether to excise small or medium CMN can often be delayed until adolescence. Melanoma in these lesions usually will not present before adolescence, and even if it does occur, it can usually be detected in early, more curable stages of growth.

Factors that may favor the excision of small or medium CMN include

1. lesions whose removal would result in minimal cosmetic deformity;
2. lesions that are difficult to follow clinically because of anatomic location, eg, scalp (patient self-examination is difficult);
3. lesions in patients with poor reliability either in self-examination or in regular

physician examinations;

4. lesions whose continued presence is psychologically troubling to the patient.

Spitz Nevus

Another variety of acquired melanocytic nevus is Spitz nevus. Another synonym is spindle and epithelioid cell nevus; outmoded terminology includes the term *benign juvenile melanoma*. The latter term arose because the Spitz nevus may be histologically confused with melanoma. However, these lesions are benign, usually arising in childhood, typically on the extremities or face. Growth is rapid; many lesions arise over weeks or months. Lesions typically have a reddish hue and may be pink to brownish-red to bluish-black with a shiny, sometimes slightly scaly surface. They are firm in texture and are usually entirely banal in appearance.

Histologically, most lesions are compound melanocytic nevi with vascular ectasia (hence the reddish color). The nevus cell nuclei are pleomorphic-appearing and may be confused with malignant melanoma. For this reason, it is usually prudent to have biopsy material reviewed by an experienced dermatopathologist. The lesions should be completely excised with 1-2 mm margins since recurrent benign Spitz nevi may be easily confused with melanoma clinically or histologically.

Blue Nevus

Blue nevi are deep blue to bluish-black (steel blue) nodules. On the nose, lesions may appear grayish-brown. They are most commonly seen on the dorsa of the hands, feet, forearms, shins, and face. The common blue nevus is a type of benign intradermal melanocytic nevus. Onset is early in life, but lesions grow slowly and rarely measure over 10 mm in diameter. Most are less than 7 mm in size. The melanocytes in these lesions are dendritic and deep, causing the lesion's bluish appearance (Tyndall effect). Because of the depth of the cells, tangential excision will not extirpate the lesions; full-depth excision of epidermis and dermis is required for complete removal.

Recurrent Melanocytic Nevi

Melanocytic nevi that are incompletely removed, especially after tangential excision, may result in scars with irregular pigmentation and may clinically resemble superficial spreading melanoma. There may be histological confusion with melanoma as well, but the lesions are benign.

Vascular Growths and Malformations

A variety of vascular lesions may be seen in infants and children. Although they all have the common feature of vascularity, their appearance, natural history, histopathology, and treatment differ widely. Some patients may benefit from early surgery, whereas in the majority of cases optimal cosmesis and function are best achieved by allowing natural involution.

Vascular lesions can be classified according to their clinical appearance. Raised lesions usually involute with time, whereas flat lesions tend to persist.

Flat Vascular Growths

Salmon patches and port-wine stains are flat lesions that are true vascular malformations. The term *nevus flammeus* should not be used because it has been applied to both lesions and is, therefore, ambiguous.

Salmon Patches

Salmon patches are very common; they are seen on the nape of the neck ("stork bites") in 50% of infants. They may also be seen on the glabella ("angel's kiss") or upper eyelids. Lesions are pink or light red in color, and are accentuated by physical exertion or crying. Eyelid lesions generally fade away by age 3 to 6 months and glabellar lesions by age 5 to 6 years, but many nape lesions have a greater tendency to persist and are seen in 25% of adults. The vast majority of lesions do not require treatment, but persistent, cosmetically disturbing lesions may be removed using yellow dye vascular lasers (see discussion under Port-Wine Stains). Histologically, lesions are composed of ectatic capillaries thought to represent persistent fetal circulatory patterns. This is perhaps due to localized dysfunction of nerves of the vascular plexus, such as loss of sympathetic neurons. The etiology is not genetic; discordance among monozygotic twins has been reported.

Port-Wine Stains

Port-wine stains (PWS) are flat, congenital, vascular malformations named for their red to reddish-purple color. They may cause a pink discoloration of the skin in early infancy, but eventually assume their characteristic color, which is darker than that seen in salmon patches. A PWS is composed of dilated mature capillaries with no tendency to involute. Unlike juvenile hemangiomas, there is no cellular proliferation and no tendency to grow out of proportion to normal somatic growth. PWSs are usually unilateral, most commonly located on the face or extremities, but any body area may be involved. When the ophthalmic dermatome of the trigeminal nerve is involved, either alone or in conjunction with other areas, the patient should be evaluated for the presence of the Sturge-Wever syndrome, in which there is also a vascular malformation of the ipsilateral meninges and cerebral cortex that may result in seizures, mental retardation, hemiplegia, or glaucoma.

Port-wine stains have a natural history that may favor early intervention. Lesions tend to darken with time, and, although they are initially flat, may eventually develop raised cavernous portions that are easily traumatized. The skin may become thickened, nodular, and irregular. Early intervention may be indicated to forestall cosmetic and hemorrhagic complications.

The treatment of choice for PWS is a yellow light laser. These lasers produce light at 577 or 585 nm, the wavelengths of local peaks of the light absorption curve of oxyhemoglobin. Equally important, these wavelengths are relatively less absorbed by other components of the skin such as melanin or water. Thus, the light energy from the laser is selectively absorbed by hemoglobin, causing coagulation and sludging of red blood cells,

leading to occlusion and fibrosis of the abnormal vessels. Some of these lasers pulse the laser output to bursts shorter than the thermal relaxation time of hemoglobin, thus minimizing the spread of heat beyond the vessel walls, reducing the risk of damaging and scarring surrounding tissues. Some yellow dye lasers lack pulsing. Studies have shown scarring rates less than that seen with the use of the 514-nm argon laser, in which light output is neither as selective for hemoglobin nor as short in duration as the pulsed dye laser. As with any laser modality, scarring can be minimized by using the lowest effective power settings; this is determined by performing test treatments at different power fluences. Different body sites respond to different power fluences. Experience with the argon laser has shown a greater tendency for scarring of the lips and neck than other areas of the head and neck.

With the argon laser, significant scarring occurred in 20% to 25% of patients, with the highest rates seen in infants and young children. For this reason, treatment was often deferred as late as possible, even though the PWS continued to worsen in severity. The pulsed dye laser does not scar as much as the argon laser, and thus can be used in infancy and early childhood before cavernous lesions and psychosocial trauma develop. The yellow dye laser does not work as well with cavernous lesions as it does with flat ones. The copper vapor laser, which also produces yellow light, is a newer modality that shows promise for treatment of the cavernous portions of port-wine stains.

Raised Vascular Growths

Juvenile Hemangiomas

Juvenile hemangioma is a term that includes the capillary hemangioma, the cavernous hemangioma, and the mixed capillary-cavernous hemangioma. Other synonyms include strawberry hemangioma, strawberry nevus, immature capillary hemangioma, and cellular hemangioma. The capillary type of juvenile hemangioma typically has a strawberry-red color and raised surface. Cavernous lesions are deep, bluish-hued nodules with normal overlying skin. An individual lesion may have a mixture of superficial ("capillary") or deep ("cavernous") components. Juvenile hemangiomas are seen in 0.8% to 2.5% of all newborns and in 10% to 12% of Caucasian infants by age 1 year. Most lesions appear by age 2 months and are most prevalent on the head and neck. Thirty percent of patients have multiple lesions. If the lesion is not obvious at birth, the first manifestation may be a pale, circumscribed area of skin with telangiectasias with or without subjacent swelling. Early in life the lesions undergo a proliferative phase that may be alarming in rate. It is essential to inform the patient's parents of the nature of the lesion and its natural history. The most important datum is that at least 95% of these lesions undergo complete resolution. Superficial lesions typically enlarge for 6 to 8 months, achieving the characteristic "strawberry" appearance, whereas deeper, cavernous components may grow for up to 12 months. Fading of color is the first sign of involution, occurring at 12 months of age in the superficial lesions. Flattening occurs by the end of the 2nd year. Involution is seen in 50% to 65% of patients by age 5 years, in 70% to 80% by age 7 years, and in 90% to 95% by age 9 years. Fewer than 2% of lesions have required further treatment.

Untreated, most patients with hemangiomas have no complications. Minor sequelae of cutaneous hemangiomas include erosion, ulceration, bleeding, and secondary bacterial infection. Areas subject to friction, trauma, or poor hygiene (such as the diaper area) are more

susceptible to ulceration or necrosis, which is seen in 15% to 20% of lesions. Raw, oozing surfaces may be treated with compresses of aluminum acetate solution and mupirocin ointment.

More serious sequelae include ocular sequelae, compression of an important anatomic structure, coagulopathy (Kasabach-Merritt syndrome), high-output cardiac failure, and intracranial extension. The exact rate of serious complications in untreated patients is unknown, but is probably close to 5% to 10%, although some studies quote rates as high as 80%. If a periorbital hemangioma blocks vision, amblyopia leading to irreversible visual impairment ranging up to cortical blindness may result, even after a few weeks of visual occlusion; cooperation with a pediatric ophthalmologist is imperative. Periorbital hemangiomas may also extend to the retrobulbar area and compress the optic nerve, causing atrophy. Impingement of the globe may cause astigmatism. Strabismus, eyelid ptosis, and proptosis are other ocular sequelae. Infantile subglottic hemangioma causes narrowing of the airway and inspiratory stridor; without intervention, mortality rates approach 45%. Systemic steroids or CO₂ laser ablation cures around 80% of patients. Lesions involving the lower lip may interfere with feeding, have a higher degree of scarring, and tend not to completely resolve. The Kasabach-Merritt syndrome is the sudden, painful, rapid enlargement of a cavernous hemangioma associated with coagulopathy, thought to be due to platelet trapping and local intravascular coagulation. Cardiac failure may result from large arteriovenous shunts present in some very large hemangiomas; patients in which this is a concern should be seen in cooperation with a pediatrician. Some facial hemangiomas have been associated with intracranial extension with arteriovenous malformations, hypoplasia of cerebral vessels, ventricular dilation, and hemicerebral atrophy. Diagnostic imaging of the head may help delineate the extent of the hemangioma. Disfigurement, although not a functional complication per se, may occur to a degree that merits classification as a serious complication.

The decision for early surgical intervention is often difficult. Intervention occurs most commonly with complicated large hemangiomas. High-dosage systemic steroids have an established benefit in reducing the size or rate of growth of a complicated hemangioma if it is treated during the proliferative phase. The latest literature should be consulted regarding dosage, as recommendations have varied. Repeat treatment is to be avoided or delayed as long as possible so that risks for corticosteroid side effects such as growth retardation are minimized. Intralesional steroid injections (triamcinolone acetonide 40 mg/mL) have also been a mainstay therapy but their use may be accompanied by rapid necrosis or hemorrhage of the lesion. Treatment of periorbital hemangiomas may be associated with inadvertent embolization of retinal vessels by steroid particles, causing blindness that may be permanent; consultation with a pediatric ophthalmologist may be helpful. Interferon-alpha-2a has been used as adjuvant therapy in severe cases with massive lesions, consumptive coagulopathy, or visceral involvement. Finally, intravascular embolization therapy conducted by an interventional radiologist has proven useful in reducing the size of large, cavernous lesions.

Although most hemangiomas resolve spontaneously, and only 6% of them are considered unsightly, some physicians have recommended a greater role for surgical intervention, specifically laser therapy. There are anecdotal reports that yellow dye or argon laser treatment of hemangiomas within the first weeks of life may arrest or slow the growth of a hemangioma while it is still small. If this proves true, not only will the usual morbidities of hemangiomas be reduced, but also the psychosocial morbidity that all hemangiomas, even

small ones, evoked will also be reduced. The yellow dye laser has been helpful especially in healing ulcerated juvenile hemangiomas without adding to the expected amount of scarring. Long-term studies using the yellow dye laser are in progress. The argon laser may be more useful for lesions with a greater thickness, but it also has a greater risk for scarring. There are also anecdotal reports of the use of contact neodymium:yttrium-aluminum-garnet (Nd:YAG) lasers, but this modality may be associated with short-term necrosis and bleeding, as well as a more unsightly, scarred appearance in the long run.

Children with juvenile hemangiomas may benefit from reconstructive facial plastic surgery late in the evolution of the disease. Lesions that remain unchanged for 3 years are unlikely to undergo further involution. Many patients will have loose, atrophic, redundant tissue at the site of an involuted lesion. This tissue may also be scarred if ulceration or infection have occurred. Reconstructive surgery often proves beneficial to these children, especially if surgery is completed before school age. Half of these children will achieve maximal involution by school age; a dermatologic can help determine the degree of involution. In general, the larger the lesion, the more delayed will be the resolution and the more likely it will never completely resolve.

Spider Angiomas

Spider angiomas (nevus araneus) are small arteriovenous fistulae named for their characteristic clinical appearance. Lesions have a central red papule from which several fine telangiectatic vessels radiate, similar to the legs of a spider. Spider angiomas are seen in 30% of children by age 4 years and in 40% by age 8 years. Only 10% to 15% of adults have spider angiomas.

Although most spider angiomas appear in normal children and adults, they may be associated with high estrogen states, such as pregnancy, hepatic insufficiency, and exogenous estrogen use, especially when large numbers of lesions are found on the trunk.

In children, the most common sites of involvement are the face, upper trunk, and upper extremities. Although most lesions resolve, some patients or their parents may seek treatment for cosmetic reasons. Lesions may be treated with gentle electrodesiccation of the central feeding artery; a small depressed scar may result. The pulsed yellow dye vascular laser will also ablate the lesion, almost always without scarring. The argon or CO₂ lasers are also effective, but scarring has been observed accompanying their use.

Pyogenic Granuloma

Pyogenic granulomas are benign vascular proliferations similar to granulation tissue. Although the name, a relic of past beliefs, suggests a bacterial etiology, these lesions are most likely an exaggerated and perpetuated healing response to injury, although the injury is usually not remembered and may have been trivial. Most arise in acral skin such as the distal extremities, face, or oral mucous membranes. Lesions are usually dull red, dome-shaped, or pedunculated nodules that may have a moist and eroded or crusted surface. Bleeding may be the presenting symptom; they may be quite friable. Satellite lesions may be seen, but are not malignant.

Treatment is simply ablative; scalpel saucerization excision or scissors snip excision with 1-mm margins is usually sufficient. Alternatively, many lesions can be shelled out with a dermal curette, leaving an epidermis-lined concave space that will heal without scarring; only the remaining central stalk with its feeding vessel need be fulgurated. All tissue removed should be submitted for pathologic examination since amelanotic melanoma or certain benign entities may resemble pyogenic granuloma. Lesions may recur or persist and are simply retreated. Satellite lesions usually resolve within 6 weeks of treatment of the primary lesion.

Lymphangiomas

Lymphangiomas are congenitally lymph vessel hamartomas that usually appear at birth or within the first 2 years of life. Like juvenile hemangiomas, lesions range from a circumscribed, more superficial form, corresponding to the strawberry hemangioma, to a deep, cavernous form.

Lymphangioma circumscriptum is the more superficial form, consisting of multiple gelatinous papules and plaques that tend to be grouped. The appearance is likened to frog spawn. There are often interconnections with the venous system, so some lesions may be hemorrhagic.

Cavernous lymphangiomas are skin-covered nodules with a rubbery consistency. Lesions located in the head and neck region are termed cystic hygromas. Like their hemangioma counterpart, cavernous lymphangiomas may become quite large and disfiguring and may have a rapid growth phase in early infancy.

Treatment of either form of lymphangioma is unsatisfactory. Lesions tend to recur after excision, even with wide margins and repair with myocutaneous flaps. Laser ablation is no more effective than cold steel surgery. Small lesions are most likely to be cured. Supportive therapy includes the use of compressive garments and prophylactic antibiotics to prevent cellulitis and scarring lymphangitis and chronic lymphedema.

Genetic Diseases

Nevoid Basal Cell Carcinoma Syndrome

The nevoid basal cell carcinoma syndrome (NBCCS) is an autosomal dominant disorder of high penetrance and variable expressivity with numerous cutaneous and extracutaneous findings. Although the primary defect is not known, patients appear to have a defect in DNA repair, especially DNA damaged by irradiation.

The key cutaneous feature of the syndrome is the development of numerous (often hundreds) basal cell carcinomas, most predominantly on the head, neck, thorax, and upper extremities. The carcinomas usually begin to develop in adolescence, but have been reported in young children, where they may resemble skin tags or melanocytic nevi. The lesions are usually translucent and pink or pigmented, but any clinical or histologic pattern of basal cell carcinoma may be seen. Left untreated, lesions may become invasive or metastasize. The incidence of basal cell carcinoma is increased in relation to cumulative sunlight or x-ray irradiation. So-called pitting or pinpoint papules may be seen on the hands and feet, especially

the palms and soles, and are considered by some to be a forme fruste expression of basal cell carcinomas; these lesions, however, never become clinically aggressive. Other cutaneous manifestations include epidermal cysts, milia, comedons, and lipomas.

Extracutaneous features are equally characteristic of the syndrome. Seventy percent of patients have keratinizing odontogenic cysts of the maxilla or mandible, some of which develop into ameloblastoma. About 70% to 75% of patients have skeletal abnormalities, which may include bifid or splayed ribs, scoliosis, spina bifida, and bridging of the sella turcica. Shortening of the fourth metacarpal results in dimpling of the overlying skin, known as Albright's sign. By adulthood, craniofacial changes are apparent, consisting of sunken eyes, frontal and temporal bossing, and a broadened nasal root with true or pseudohypertelorism. Central nervous system abnormalities, include calcification of the falx cerebri or falx cerebelli, agenesis of the corpus callosum, and medulloblastoma. Following surgery and radiotherapy of the latter, there is an increased incidence of basal cell carcinomas of skin overlying the radiation ports. There have also been case reports of ovarian fibrosarcoma following radiotherapy to the entire neuraxis for medulloblastoma. Many other abnormalities in these and other organ systems have been reported.

As expressivity of the manifestations of the syndrome is variable, treatment must be tailored to the individual. Patients should protect themselves scrupulously from sunlight and avoid x-ray irradiation if possible. Long-term and short-term goals should be kept in mind when planning excision of multiple lesions. A triage approach to tumor management may prove helpful. The most biologically aggressive lesions should be removed first. Because patients are likely to develop scores of lesions during their lifetime, as much normal surrounding tissue as possible should be preserved. Because of the likelihood of the presence of scars from previous treatments as well as the presence of new basal carcinomas in overlying skin, it may be difficult to determine whether a lesion has recurred in an area reconstructed by flaps or grafts. Curettage and electrodesiccation followed by second intention healing is often the most practical modality of treatment for optimal reduction of long-term morbidity and mortality. Microscopically controlled excision (Mohs surgery) should be considered for treatment of recurrent lesions because of tissue sparing and high cure rates. A more recently described therapy is the use of systemic retinoids to prevent formation of new lesions while the drug is being used. Once the drug is discontinued, however, new carcinomas form as frequently as before. Side effects complicate long-term use of the current generation of retinoids. Photodynamic therapy using lasers following the systemic administration of photosensitizing agents is another promising experimental modality.

Xeroderma Pigmentosum

Xeroderma pigmentosum is a group of autosomal recessive genodermatoses in which children manifest an increased number of skin carcinomas of the head and neck, including basal cell carcinomas, squamous cell carcinomas, and malignant melanomas. All of the varieties of xeroderma pigmentosum, which number at least eight, have defective capacity for repair of ultraviolet light-induced DNA damage. In addition, there is a variant of xeroderma pigmentosum in which DNA repair is normal but S-phase DNA synthesis is impaired following ultraviolet irradiation (defective postreplication repair). Like the skin, the ocular epithelium has similar hypersensitivity to ultraviolet irradiation. Some forms of xeroderma pigmentosum have varying neurologic abnormalities.

Xeroderma pigmentosum manifests in some populations more than others, but is found worldwide. In European races, the incidence is 1 per 250,000 births, and in Japan the incidence is 1 per 40,000 births.

Onset is congenital; photophobia in infancy is usually the first manifestation. Infants exposed to ultraviolet light, either from natural or, to a lesser extent, artificial sources, will begin to develop dose-related conjunctivitis and sunburn-like erythema and scaling of the skin as evidence of ultraviolet-induced damage. If the acute dose of light is strong enough, blistering may be seen. As damage progresses, freckling and poikiloderma (hyperpigmentation, hypopigmentation, telangiectasia, atrophy, and scarring) of the skin will form and the ocular conjunctiva will become xerotic, hyperpigmented, and scarred. Symblepharon and blepharitis are also seen. As damage accumulates, the skin forms actinic keratoses. Frank carcinomas subsequently develop, usually by the age of 6 years. Basal cell carcinomas, squamous cell carcinomas, and malignant melanomas, in order of decreasing frequency, are commonly seen. Other skin tumors include keratoacanthomas, fibrous tumors, and sarcomas. The conjunctivae and corneas become clouded, hypervascularized, and scarred, and develop malignancies. Vision is correspondingly impaired. Some patients, such as those with the De Sanctis-Cacchione syndrome, also have progressively worsening neurologic abnormalities including mental retardation, cerebellar ataxia, and seizures. Some patients have developmental abnormalities such as microcephaly and dwarfism.

In the past, death usually occurred by the third decade. Patients are now living longer due to better protection from ultraviolet light and better medical care. Strict avoidance of both natural and artificial sources of ultraviolet light have forestalled the progressive damage to eyes and skin. Patients with neurologic abnormalities, however, face progression of neurologic disease despite strict protection from ultraviolet irradiation.

Patients should be monitored closely for the development of cutaneous malignancy, with prompt and judicious biopsy performed on suspicious lesions to allow the earliest possible treatment of cancer. Skillful use of cryosurgery or topical 5-fluorouracil on premalignant lesions such as actinic keratoses or lentigo maligna will help forestall the inevitable onset of malignancy. Use of systemic retinoids such as isotretinoin will also slow the formation of carcinomas, but the effect does not last after the patients discontinue the drug, and long-term side effects may be expected.

Tuberous Sclerosis

Tuberous sclerosis is an autosomal-dominant disease with a wide variety of cutaneous and internal findings. The classic triad is epilepsy, mental retardation, and "adenoma sebaceum", which is a misnomer, since these lesions are actually cutaneous angiofibromas. Angiofibromas are seen in 65% to 95% of patients, almost always over the cheeks, nasolabial folds, and chin; more rarely, they are seen elsewhere on the head. Angiofibromas are shiny, translucent papules usually millimeters in size, with a yellowish-pink to brownish-red hue. Patients typically have dozens of lesions. Some patients have fibrous plaques of the face that lack a significant vascular component.

Patients may have other cutaneous findings besides angiofibromas. The most common are "ash-leaf" macules, which are oval-shaped hypopigmented macules seen on the trunk;

"shagreen" patches, which are slightly raised, slightly textures, leather-like plaques seen on the trunk; periungual fibromas, which are firm, budlike growths around the nail plates, sometimes termed Koenen's tumors. Visceral involvement includes potato-shaped (tuberous) or flat, sclerotic plaques of the cerebral cortex, cardiac rhabdomyomas, renal hamartomas, retinal hamartomas, and a variety of other ocular tumors.

There are many methods for management of angiofibromas of the face. Lesions may be removed by CO₂ laser vaporization, tangential excision, dermabrasion, electrodesiccation, or cryosurgical ablation. Further surgery may be required at intervals, as new lesions continue to form.

Miscellaneous Cutaneous Tumors

Epidermal Cysts

The most common superficial tumor seen in childhood is the epidermal cyst (epithelial cyst; epidermoid cyst). The term *sebaceous cyst* is a misnomer, since the epithelial lining of the cyst arises from epidermis identical to the infundibulum of the hair follicle, and not sebaceous glands. The cysts are dermal or subcutaneous, and are filled with laminated keratin normally shed from the surface of the skin. Most epidermal cysts arise spontaneously, but rarely may arise traumatically if epidermis inverts into the skin; this variant is termed an epidermal inclusion cyst.

Clinically, epidermal cysts are firm, spherical nodules that vary in size from millimeters to centimeters; most are less than 2 cm in diameter. In children, the most common location is the lateral eyebrow or elsewhere in the periorbital area. Cysts may be superinfected, in which case inflammation with redness and tenderness is seen. Multiple cysts in children should raise the suspicion for Gardner's syndrome, an autosomal dominant disorder in which premalignant intestinal polyposis, muscular desmoid tumors, and jaw osteomas may also be seen.

Epidermal cysts require no therapy as such, as they are harmless. They occasionally regress spontaneously, but may also be subject to repeated infection with cyclical enlargement and drainage. Removal of the entire cyst wall is curative. Cysts may be excised en bloc, but removal through the punctum of the cyst or a small slit incision is often possible and reduces the size of the scar. Using this method, a 2- to 4-mm incision is made over the cyst; if the punctum is large, a tiny 2- to 4-mm ellipse incision is made to include the punctum. The cyst contents are then decompressed, if necessary, and the cyst wall is everted through the opening. Applying subjacent pressure, scooping out the cyst wall with a small curette, and teasing out the cyst wall using steady traction with forceps are helpful. Removing the cyst wall in this fashion is akin to everting a balloon through its own neck. If the cyst has been subject to repeated infection, adherent scarring around the cyst wall may make this conservative approach impossible. Regardless of the method used, rupture of the cyst during removal has no effect on recurrence. It is retention of any portion of the cyst wall, including its neck or punctum, that causes recurrence.

Dermoid Cysts

Dermoid cysts are round intradermal or subcutaneous nodules most commonly seen on the face or scalp. They are thought to result from the invagination of ectoderm at embryonic fusion planes. Thus, they represent developmental anomalies. Unlike epidermal cysts, which are formed of epidermis alone, dermoid cysts contain epidermis, hair follicle structures, sebaceous glands, and apocrine glands, and thus may form keratin, hair, sebum, and apocrine secretions. The lateral eyebrow is a site of predilection. Lesions located over the midline, such as the nose, glabella, and skull, as well as over skull sutures, should be approached with caution, as they may have sinus tracts that may extend deeply. A typical presentation of such a cyst is a nodule over the nose or glabella that has hairs protruding from a punctum. Some dermoid cysts even connect with the meninges. Magnetic resonance or computed tomography scans should be considered in the preoperative evaluation of midline cysts with neurosurgical consultation as necessary.

Epidermal Nevi

Epidermal nevi are hamartomas that may form from any of the components of skin and its adnexal structures, including keratinocytes, hair follicle cells, sebaceous glands, or sweat glands. Combinations of these cutaneous structures with a variety of reactive patterns such as inflammation, hyperkeratosis, or papillomatosis result in a variety of histologic and clinical patterns. Thus, epidermal nevi may clinically or histologically resemble many other proliferative disorders of the skin including warts, acanthosis nigricans, Darier's disease, ichthyoses, or psoriasis.

For the most part, epidermal nevi are not genetic in etiology and appear sporadically. Although the lesions are composed of epithelium, there appears to be a component of interaction with other tissues of ectodermal as well as mesodermal origin, since patients may also have other developmental abnormalities; permanent destruction of lesions seems to require ablation of underlying papillary (superficial) dermis.

The nosology of epidermal nevi is inconsistent; most lesions are described by clinical features, and there is overlap. The nevus sebaceus (of Jadassohn) will be discussed separately.

The *verrucous epidermal nevus* is warty, with color ranging from pink to gray to brown. Lesions often have a dirty appearance due to hyperpigmentation and hyperkeratosis. Age of onset is usually at birth or in the first decade, but may range up to the third decade.

Nevus unius lateris is a widespread, generally linear, epidermal nevus that usually appears on the trunk or extremities. The configuration is usually longitudinal on the limbs and transverse on the trunk; Blaschko's lines refer to the pattern these lesions usually follow. Lesions are usually unilateral, stopping at the midsagittal plane. Bilateral distribution is sometimes called ichthyosis hystrix, but this term may be confused with varieties of ichthyosis and should be avoided.

Inflammatory linear verrucous epidermal nevus (ILVEN) refers to a pruritic, inflammatory lesion with a generally later onset. It is usually seen on the extremities in girls.

The *epidermal nevus syndrome* is a severe condition in which widespread epidermal nevi are seen along with other developmental defects of varying severity. Craniofacial or other skeletal defects involving the spine and extremities occur in two-thirds of patients. Abnormalities of the central nervous system, including seizures and mental retardation, are seen in half of patients. Ocular abnormalities, including extension of the nevus onto the eyelids, ocular dermolioid tumors, and colobomas of the lid, iris, or retina, are seen in one-third of patients. Besides ocular involvement, other challenges for the facial plastic surgeon include widespread involvement of the ear or other facial structures.

Other dermatologic conditions seen in patients with the epidermal nevus syndrome include vascular lesions (eg, nevus flammeus or juvenile hemangiomas), giant congenital melanocytic nevi, and keratoacanthomas. Some patients developed carcinomas in the epidermal nevi, especially those with the nevus sebaceus form of the syndrome.

Treatment, if any, for epidermal nevi depends on the symptomatology, and extent and distribution of the lesions. Topical therapy with tretinoin, aquaglycolic acids, or 5-fluorouracil may be sufficient for some patients, although it is not curative. Simple excision and closure may be practical for smaller lesions. Widespread plaques may be treated with deep tangential excision, electrosurgery, cryosurgery, dermabrasion, or CO₂ laser vaporization; all of these modalities require healing by second intention.

Nevus Sebaceus

The nevus sebaceus (organoid nevus; nevus sebaceus of Jadassohn) is a hairless, raised, yellowish to yellowish-brown plaque most commonly seen on the scalp or face. It is generally present from birth, and grows at the same relative rate as the child. The lesion is comprised of abnormally developed apocrine and sebaceous glands with papillomatosis of the superficial dermis and epidermis. Under hormonal influence at puberty, the glandular elements will proliferate, and the lesion assumes a more thickened, verrucous and papillomatous texture. In one-third of patients, benign or malignant neoplasia will develop. The most common tumor is syringocystadenoma papilliferum, a benign tumor of apocrine origin, seen in 12% to 20% of lesions. A wide variety of other benign adnexal tumors may be seen.

Basal cell carcinoma (BCC), seen in 6% to 16% of lesions, is the most common malignant growth in nevus sebaceus. Fortunately, BCC in this setting is slow-growing and is usually seen before the fourth decade of life. Other malignancies are also reported, some of which have grown aggressively.

Neoplasia in nevus sebaceus is unusual before puberty, so surgical excision can usually be delayed until then unless concerns regarding cosmesis supervene. Since the lesion is benign, narrow margins of excision may be used, with wider excision or microscopically controlled excision (Mohs surgery) being used only if pathologic evaluation discloses malignancy.

Adnexal Tumors

The epidermal adnexae include the hair follicles, sebaceous glands, apocrine sweat glands, and eccrine sweat glands. The first three structures are continuous with the hair

follicle, whereas the eccrine sweat gland empties directly to the skin surface. Some benign neoplasms derived from these structures are discussed elsewhere, eg, the epidermal nevus and nevus sebaceus. Adnexal tumors are more common in adults, but three types, besides epidermal nevi and nevus sebaceus, are more prevalent than other in children or adolescents: pilomatricomas, trichoepitheliomas, and syringomas.

Pilomatricomas

Pilomatricomas (calcifying epithelioma of Malherbe) are hard, cutaneous nodules with normal overlying skin. Lesions are usually solitary and arise on the face or limbs. Onset may range from birth, but infancy or early childhood is the usual time of onset. Rarely, lesions are associated in a syndrome with myotonic dystrophy. Lesions measure 2 to 5 mm in size, and arise from cells of hair matrix. Calcification may occur. Lesions may be removed by excision; they usually do not recur.

Trichoepitheliomas

Trichoepitheliomas, when multiple, are the result of an autosomal dominant trait. They may also be found in a nonhereditary form as single lesions. Lesions are round, usually measure up to a few millimeters in diameter, are flesh-colored to reddish brown, and are primarily found in the midface around the nose or elsewhere on the head and neck. Onset is usually in adolescence. Histologically, they are comprised of horn cysts and islands of basaloid cells, probably of hair follicle origin. Solitary lesions may be confused clinically or histopathologically with basal cell carcinoma. When the lesions are multiple, with new lesions arising continually, cosmetically acceptable treatment is difficult. Tangential excision, dermabrasion, or CO₂ laser vaporization may be applied, similarly to the treatment of angiofibromas of tuberous sclerosis.

Syringomas

Syringomas are derived from sweat gland epithelium. Clinically, they present as small, flat, waxy or translucent papules around the eyes, usually on the eyelids or in an infraorbital or zygomatic distribution. Lesions may also appear on the upper trunk or vulva; there is a female preponderance. Papules are skin-colored with hues of yellow or brown. Onset is usually around adolescence. Since the nests of eccrine epithelium deeply infiltrate the dermis and are not well circumscribed, ablation of the lesions short of full dermal excision is difficult. Lesions are usually too numerous to excise. Sometimes patients are satisfied with tangential or saucerization excision of a few of the larger lesions, with the expectation that new and recurrent lesions will arise and that cosmesis will not be perfect. Laser surgery, including the CO₂ laser, is also used, as well as electrosurgical ablation.

Juvenile Xanthogranulomas

Juvenile xanthogranulomas are soft, orange to yellowish-brown nodules appearing on the skin and other tissues, including internal organs and the iris. They usually arise before 1 year of age, are found on the scalp, face, and neck, and occasionally elsewhere, and are often multiple. Although histologically the lesions are comprised of lipid-filled granuloma cells, there is no relation to abnormalities of lipid metabolism. The process is idiopathic and benign,

and lesions usually resolve within 1 year, occasionally requiring several years for complete involution. No treatment is necessary for skin lesions. Biopsy of a single lesion for diagnosis and referral for ophthalmologic evaluation may be indicated. Lesions may be confused with Spitz nevi.