Chapter 20: Vascular Anomalies of the Head and Neck

Trevor J. I. McGill, John B. Mulliken

The intricate vascular anatomy of the head and neck probably predisposes the region to vascular anomalies. This very same anatomy certainly complicates the management of cervicofacial vascular anomalies.

Our understanding of vascular anomalies has been hampered by confusing nosology. This confusion has been responsible for improper diagnosis, illogical treatment, and misdirected research efforts. Furthermore, interdisciplinary communication has been limited in the field of vascular anomalies as each medical specialty has its own nosologic jumble. A classification is useful only if it allows accurate diagnosis, predicts prognosis, guides treatment, and stimulates research into pathogenesis.

This chapter is based on a biologic classification of vascular anomalies introduced in 1982 by Mulliken and Glowacki and divides the differential diagnosis of cervicofacial vascular anomalies into either *hemangioma* or *malformation.* Furthermore, vascular malformations are subdivided into slow-flow or fast-flow lesions. The management of these various types of vascular lesions is also described, based on current biologic and rheologic knowledge.

Classification

The clinical presentation of vascular anomalies is confusing because all lesions appear in the color spectrum of blue, pink, and red. Standard nosology of vascular anomalies offers an array of overlapping descriptive and histopathologic terms. Unfortunately, the same word has been used to describe entirely disparate vascular lesions. For example, "hemangioma" has been a generic term for various vascular lesions with distinctive natural histories and differing etiologies. The most common tumor of infancy is known as a "strawberry", "capillary", "cellular", or "juvenile" hemangioma. This lesion typically undergoes rapid growth during infancy, followed by slow, invariable involution. Port-wine stain, a lesion that never regresses, has also been classified as a "capillary hemangioma" (Popkin, 1977; Thomson, 1979). The term "cavernous hemangioma" has been used to describe lesions that involute, never involute, or rarely involute (Lampe and Latourette, 1956; Simpson, 1959). The hyphenated appellation "capillary-cavernous hemangioma" has further compounded the issue. This confusing nomenclature has been largely responsible for incorrect diagnosis and illogical treatment of cutaneous vascular anomalies.

There is no useful classification without properly defined terms. The Greek noun suffix *-oma* denotes a swelling or tumor. In modern usage, however, a tumor is characterized by cellular hyperplasia. The semantic refinement is crucial to a precise nosology for vascular anomalies. The biologic classification proposed by Mulliken and Glowacki in 1982 defines the cellular features of vascular anomalies and correlates them with clinical behavior. Correlative studies of natural history and surgical specimens, using histopathologic techniques, demonstrate that there are two major categories of vascular anomalies in infancy and childhood: (1) *hemangioma,* a lesion that grows rapidly in neonatal life, is characterized by endothelial proliferation, and invariably undergoes slow regression; and (2) *vascular*

malformation, a lesion that is present at birth, is characterized by a normal rate of endothelial cell turnover, and grows commensurately with the child (Fig. 20-1). Malformations are structural anomalies, errors of vascular morphogenesis. It is clinically useful to separate the vascular malformations into "slow-flow" (either capillary, venous, lymphatic, or combined forms) or "fast-flow" (arteriovenous fistulas and arteriovenous malformation) categories.

Characteristics that distinguish hemangioma from vascular malformation

Clinical

Hemangioma is usually not seen at birth. Approximately a third, however, will present in the nursery as a reddish macule or telangiectasia. This lesion occurs more commonly in females, with a ratio of 3:1 (Mulliken and Glowacki, 1982). This tumor is also more frequent in whites than in blacks. Clinically, hemangioma is characterized by a rapid postnatal growth (the proliferative phase) for the first 8 to 12 months, followed by a slow regression over 5 to 8 years (involutive phase) (Figs. 20-2 to 20-4).

Vascular malformations, by definition, are present at birth. Many, but not all, are clearly seen in the nursery. Most lymphatic and many venous anomalies appear during infancy. Arteriovenous anomalies more often manifest later in childhood. A vascular malformation grows proportionately with the child, although the lesion may expand secondary to trauma, infection, hormonal changes, or embolic or surgical intervention.

Cellular

Hemangioma in its proliferating phase is composed of rapidly dividing endothelial cells forming syncytial masses with and without lumens (Fig. 20-5). Autoradiograms demonstrate this endothelial hyperplasia with incorporation of tritiated thymidine into replicating endothelial DNA (Mulliken and Glowacki, 1982). Electron microscope examination of proliferating hemangioma reveals multilamination of the basal lamina beneath the epithelium. Mast cells are significantly increased in proliferating hemangiomas and return to normal late in the involuting phase (Glowacki and Mulliken, 1982). The role of the mast cell in angioneogenesis is poorly understood. During the involutive phase, endothelial cell activity diminishes and the cellular parenchyma is replaced by fibrofatty tissue (Dethlefsen et al, 1986). At this stage, a surgical specimen may have a "cavernous" histologic appearance, which may be confused with a venous malformation.

In contrast, histologic evaluation of vascular malformations shows no evidence of cellular proliferation, but rather a progressive dilatation of vessels of abnormal mural structure. Vascular malformations are lined by flat, quiescent endothelium, lying on a thin single laminar basement membrane. Mast cell counts are normal throughout the evolution of vascular malformations (Glowacki and Mulliken, 1982).

Hematologic

Large or extensive hemangiomas may cause platelet trapping, the Kasabach-Merritt syndrome (Kasabach and Merritt, 1940). Neonates with this syndrome present with purpura and life-threatening bleeding into the pharynx, GI tract, and/or brain. Although Kasabach-

Merritt thrombocytopenic coagulopathy occurs in only 1% of infants with hemangiomas, it is associated with death in 30% to 40% of those affected despite therapy (El-Dessouky et al, 1988).

In contrast, those with arterial or lymphatic malformations usually have normal bleeding studies. Large or extensive venous malformations, however, may be associated with localized or disseminated intravascular coagulopathy (DIC).

Radiographic

Imaging studies show that a hemangioma is a well organized mass arranged in a lobular configuration. Magnetic resonance imaging (MRI) may reveal fast flow in a proliferative-phase hemangioma; this finding may be confused with an arteriovenous malformation. Vascular malformations consist entirely of vessels of a different caliber without intervening parenchyma (Meyer et al, 1991). Capillary, lymphatic, venous, and combined channel lesions have slow-flow characteristics that are apparent using ultrasonography, MRI, or computed tomography (CT).

Skeletal

Hemangiomas only rarely cause bone or cartilaginous hypertrophy. They may obstruct the cartilaginous ear canal or distort the nasal bony-cartilaginous pyramid. Slow-flow vascular anomalies, specifically lymphatic, venous, or lymphatovenous types, can cause significant hypertrophy and distortion of the craniofacial skeleton. Fast-flow anomalies typically cause interosseous destruction (Boyd et al, 1984).

Summary

This simplified classification of vascular anomalies of childhood as hemangioma or malformation can be called "biologic" because it is based on cellular and clinical behavior. It is a practical system; one that does not necessitate diagnostic studies such as angiography or a biopsy. An accurate history and clinical examination permits distinguishing between a hemangioma and vascular malformation in most patients (Finn et al, 1983). Once a diagnosis is made, appropriate therapy can be planned.

Hemangioma

Diagnosis and natural history

Hemangioma is the most common tumor of infancy. There is an equal incidence in premature (1500 to 2500 g) and full-term white infants of 10% to 12% by 1 year of age (Holmdahl, 1955). There is, however, a 23% incidence of hemangiomas in those premature children weighing below 1000 g (Amir et al, 1986). A majority of hemangiomas appear during the first 6 weeks of life. The first sign of a hemangioma is a macular patch, blanched spot, or a localized area of telangiectasia surrounded by a halo (Hidano and Nakajiuma, 1972). Rarely, a fully grown hemangioma is present at birth. Hemangiomas undergo rapid proliferation in the early months of life as a localized tumor in a single area or simultaneously proliferate in several sites throughout the body. Eighty percent of hemangiomas occur as an

isolated lesion, whereas 20% are multiple hemangiomas (Margileth and Museles, 1965). The head-and-neck region of the body is the most commonly involved, followed by the trunk and extremities (Finn et al, 1983). It is rare for an infant with a visceral hemangioma not to have multiple cutaneous lesions. Cervicofacial lesions may be associated with laryngeal and tracheal lesions (Healy et al, 1980, 1984).

When hemangioma begins or extends into the superficial dermis, the cellular proliferation causes the skin to become raised with a vivid bright red color. Most lesions will remain well cricumscribed, measuring 0.5 to 5.0 centimeters in diameter, whereas some spread in a geographic fashion. Some hemangiomas proliferate in the lower dermis or subcutaneous tissue with little involvement of the superficial or capillary dermis. These lesions may be slightly raised. The overlying skin is smooth with a bluish hue: enlarged, radially draining veins are often a clue. In the past, these deeper lesions were erroneously called "cavernous" hemangiomas (Fig. 20-6). When a hemangioma involved both deep and superficial skin layers, it was called a "mixed", or "capillary-cavernous", hemangioma. In fact, histologic examination of hemangiomas with these morphologic appearances show the proliferating endothelial cell pattern is remarkably consistent throughout the depth of the tumor (Mulliken and Glowacki, 1982). The microscopic terms "capillary" and "cavernous" are therefore confusing and should not be used in clinical practice. It is more accurate to refer to a bright red lesion as a *superficial* hemangioma, to designate a lesion with normal overlying skin as a *deep* hemangioma, or to describe a combination thereof.

Hemangioma is the most common tumor of the parotid gland in infancy. This usually presents during the second or third month of life. Later in its period of rapid proliferation, there may or may not be invasion of the skin. If there is cutaneous involvement, the diagnosis is obvious. Deep-seated hemangiomas may be associated with a blue tinge on the overlying skin, but sometimes it may be difficult to distinguish such a lesion from a lymphatic malformation.

In 95% of cases, a hemangioma can be distinguished from a vascular malformation based on clinical history and physical examination (Mulliken and Young, 1988). The hemangioma feels firm and rubbery and is difficult to compress compared with the readily compressible venous malformation. In certain instances in the head-and-neck region, it may be difficult to differentiate a deep hemangioma from a lymphatic or venous malformation, particularly in the preauricular or cervical area.

Radiologic imaging

Notwithstanding the accuracy of clinical findings in head and neck vascular anomalies, there are times when imaging technology is needed for a precise diagnosis. The following techniques should be considered in this order: ultrasonography (with Doppler flow study), MRI, occasionally CT, and if necessary, angiography. Ultrasonography is highly operator-dependent; however, in experienced hands, it will differentiate slow-flow malformations (specifically venous or lymphatic anomalies) from hemangioma. MRI is more expensive but provides more information and is highly sensitive and specific (Meyer et al, 1991). MRI portrays the extent of involvement within tissue planes and demonstrates flow characteristics. Hemangioma, in either the proliferative or involutive phase, can be differentiated from arteriovenous malformation. Furthermore, slow-flow anomalies can be subcategorized as

capillary, lymphatic, venous, or combined forms. The most difficult lesions to distinguish with MRI are lymphatic, venous, and lymphatovenous malformations. These, however, ca usually be discriminated by administration of intravenous gadolinium and repetition of the T1-weighted sequence (Meyer et al, 1991). CT with contrast enhancement may not distinguish slow-flow from fast-flow anomalies unless dynamic scanning is done. There may still be a place for CT evaluation of intraosseous vascular lesions. Angiography still has a well-deserved place in the diagnosis of vascular anomalies. It is now usually part of treatment of an arteriovenous malformation by super-selective embolization or is performed before surgical extirpation and reconstruction. Venous angiography is needed for sclerotherapy of venous malformations.

Complications

Obstruction

Visual. Obstruction of the visual axis by a hemangioma can result in deprivation amblyopia and failure to develop binocular vision. Even a small hemangioma of the upper eyelid may cause distortion of the growing cornea, producing refractive error leading to astigmatic amblyopia (Robb, 1977). Any infant with a hemangioma in the upper periorbital region should have a prompt ophthalmologic examination. Lower eyelid or cheek hemangiomas rarely cause cornea distortion.

Subglottic hemangioma. Subglottic hemangioma is a potentially life-threatening lesion that usually presents after the first 6 weeks of life (Brodsky et al, 1983; Ferguson and Flake, 1961; Healy et al, 1980, 1984). More than half of these infants have an associated cervicofacial cutaneous hemangioma (Ferguson and Flake, 1961).

Clinically, these patients, otherwise healthy, present with the onset of biphasic stridor. The child is usually 2 to 3 months old. With increasing size of the hemangioma, there is reduction in the subglottic airway and the insidious onset of respiratory distress. On other occasions, the child may first appear with a protracted episode of laryngotracheal bronchitis (Calcaterra, 1968). Conversely, the child may be diagnosed with failure to thrive because of continued respiratory distress.

A lateral radiograph of the neck or a fluoroscopic study of the upper airway will show a smooth, usually posteriorly based round swelling in the immediate subglottic space (Fig. 20-7). Diagnosis requires a direct laryngoscopy, which demonstrates the hemangioma as a smooth, easily compressible mass in the subglottic space (McGill, 1990). The subglottic hemangioma may occupy 20% to 80% of the subglottic space (Fig. 20-8). On rare occasions, it may extend circumferentially around the subglottic space. Biopsy of this lesion is not necessary to make the diagnosis. Further evaluation of the distal airway is important to rule out other hemangiomas.

Ulceration and bleeding

Local complications during the proliferative phase in infancy, such as ulceration and bleeding, may necessitate active treatment.

When a hemangioma penetrates the epidermal basement membrane, ulceration or bleeding may occur. Bleeding is often sudden, punctate, and frightening. The parents should be taught how to compress the area with a clean pad, applying pressure for 10 minutes. Repeated bleeding is rare. If it occurs, a mattress suture may be indicated. Localized bleeding is usually not a manifestation of platelet-trapping coagulopathy (Kasabach-Merritt syndrome).

Ulceration is particularly common in hemangiomas of the lips. Secondary infection invariably accompanies ulceration. Superficial ulceration usually responds to daily cleansing and application of topical antibiotic ointment. Deeper ulceration may require dressings - these lesions often take several weeks to heal. Recurrent ulceration after healing is rare. Extensive and/or refractory ulceration may be an indication for pharmacologic therapy.

Alarming complications

More worrisome are hemangiomas in the head/neck region that cause obstruction of vital structures, distortion, or life-threatening coagulopathy. These are all indications for pharmacologic therapy.

Congestive heart failure. Congestive heart failure, a life-threatening complication, is typically seen with multiple cutaneous hemangiomas and with hemangiomatous proliferation within the viscera, typically the liver. High-output congestive heart failure can also occur with large cervicofacial hemangioma. Despite multimodality treatment, the overall mortality rate is reported to be as high as 54% (Berman and Lim, 1978).

Obstruction and/or distortion. The proliferating hemangioma in the head and/or neck is likely to impinge on or deform a critical anatomic structure. Distortion of the growing cornea by a periorbital hemangioma is an obvious example. A hemangioma within the nasal tip may obstruct the vestibular passages. This can occur during the first 3 months of life when the infant is an obligatory nose breather. Usually, however, the obstruction is unilateral and the narrowing occurs slowly so that the infant adapts and learns to breathe orally.

More life-endangering and insidious is hemangiomatous proliferation in the larynx (Brodsky et al, 1983; Healy et al, 1984). A preauricular hemangioma may cause obstruction of the external auditory canal. Curiously, there is often bilateral preauricular involvement. This results in a temporary mild-to-moderate conductive hearing loss.

A large facial hemangioma may grow to distort the features. Such a hemangioma, in effect, acts as a tissue expander, destroying the normal collagenous-elastin framework of the dermis. If this is allowed to continue, the skin may fail to contract with regression. The result will be loose, wrinkled skin and a fibrofatty tumor residuum.

Platelet-trapping coagulopathy (Kasabach-Merritt syndrome). Kasabach-Merritt syndrome, the hematologic complication of hemangioma, was first documented in 1940 by Kasabach and Merritt. It occurs early in the postnatal rapid-growth phase. Characteristically, the involved skin is deep red-purple, tense, and shiny. In large lesions, there may be central area of softness, suggesting intralesional bleeding. Petechiae and ecchymoses are seen overlying and adjacent to the hemangioma. Hematologic evaluation reveals a profound thrombocytopenia (2.000 to 40.000/mm^3). Early on, the fibrinogen level is slightly low. In

time, fibrinogen falls to trace levels, whereas prothrombin time (PT) and thromboplastin time (PTT) become dangerously prolonged. There is a risk of acute hemorrhage in the GI tract or the pleural, peritoneal, pulmonic, or central nervous systems. There is also the hazard of rapid expansion of the hemangioma secondary to intralesional bleeding, causing compression of a vital structure.

Treatment

Most hemangiomas in the head-and-neck region grow as small tumors and invariably regress, leaving inconsequential skin changes. Clinical studies confirm that complete resolution of hemangiomas occurs in over 50% of children by age 5 years and in over 70% by the age of 7 years, with continued improvement in the remaining children until ages 10 to 12 (Bowers et al, 1960; Simpson, 1959). Typically, the skin after involution exhibits mild atrophy, or it may have a wrinkled quality, or a few telangiectatic vessels. The skin may be slightly more pale than normal skin. In summary, for the small hemangioma in an inconspicuous location, nothing should be done - "primum non nocere" (Fig. 20-9).

Laser excision for subglottic hemangioma

Treatment of all infants with a subglottic hemangioma is unnecessary, especially in those with a minimal lesion occupying anything less than 20& of the subglottic airway. The treatment of choice is watchful waiting until involution occurs. However, an adequate airway has to be established with the onset of respiratory distress. Formerly, a tracheotomy was left in place for approximately 2 years until there was sufficient involution of the hemangioma. Systemic steroid therapy is a pharmacologic option, as discussed earlier (Cohen and Wang, 1972; Hawkins et al, 1984). Sixty percent of subglottic hemangiomas respond to steroids. The usual dose is prednisone, 2 to 3 mg/kg/day. A lower dosage or an alternate-day treatment of steroids may be continued for 4 to 6 weeks or longer.

The carbon dioxide laser is the therapeutic option in infants who fail to respond to corticosteroids. With general anesthesia, the subglottic hemangioma is exposed with an appropriate laryngoscope. An operating microscope coupled to a carbon dioxide laser is used to visualize the lesion. Jet ventilation with a Venturi apparatus is used to maintain oxygen while the infant is paralyzed. Carbon dioxide laser beam is used in the intermittent mode at a setting of 0.05 seconds at 20 watts. The laser is very useful for treating eccentrically placed subglottic hemangiomas; often such a lesion can be removed in one operative procedure (Healy et al, 1980, 1984; Sie, in press). The rare circumferential subglottic hemangiomas should be removed in stages in an effort to prevent subglottic stenosis, or tracheostomy may become necessary.

Pharmacologic therapy for alarming complications

Corticosteroid. Since first reported in 1967, high-dose corticosteroid remains the premier pharmacologic agent for control of endangering hemangiomas (Zarem and Edgerton, 1967). The decision to proceed with drug therapy should be made early. There is empirical evidence that the young proliferating hemangioma is far more responsive to corticosteroid therapy than is a lesion in an older infant. The usual dosage is prednisone, 2 to 3 mg/kg/day, given orally (Edgerton, 1976). Intravenous corticosteroid may be used in an infant with

respiratory complications. There is no evidence, however, that the response is more likely or profound with intravenous than with oral administration.

A responsive hemangioma exhibits signs of accelerated regression within several hours to days after beginning corticosteroid. The signs are usually obvious; that is, lightening of color, softening, and diminished growth. If there is no evidence of accelerated regression at 2 to 3 mg/kg/day prednisone for 7 days, then the hemangioma must be termed unresponsive. Such a lesion will not respond to a higher dose, and the drug should be discontinued. If the lesion does respond, the dosage can be lowered slowly over several weeks, or the patient can be switched to alternate-day therapy. The duration and dosage of corticosteroid therapy depends on the tumor's location and maturity. Rebound growth may occur in a proliferative-phase hemangioma at a low steroid level. To minimize regrowth, the corticosteroid must be continued until the hemangioma is well into the involuting phase, usually until the infant is 8 to 10 months old.

Intralesional corticosteroid should be considered for small protuberant hemangiomas in the face, particularly for upper eyelid and nasal tip lesions. The dosage is based on the size of the lesion and the infant's weight. The dosage is no more than 40 mg triamcinolone acetate and 6 mg betamethasone at each injection (Kushner, 1985; Sloan et al, 1989). Usually several injections (1 to 5) are necessary, spaced 4 to 6 weeks apart, to control the hemangioma.

The response rate is 30% to 60% with either systemic or intralesional corticosteroid administration (Bartoshesky et al, 1978; Enjolras et al, 1990; Kushner, 1985; Sloan et al, 1989). The mortality rate for infants with platelet-consumption coagulopathy is 30% to 40% despite the use of corticosteroids (El-Dessouky et al, 1988).

Interferon-alpha-2a. The antiangiogenic properties of interferon (IFN)-alpha-2a were discovered fortuitously as the recombinant drug was used for acquired immunodeficiency syndrome (AIDS). White et al (1989) observed remarkable regression of "pulmonary angiomatosis" in a 7-year-old boy after IFN-alpha-2a therapy. Orchard et al (1989) and White et al (1991) reported favorable response to this drug in a small number of infants with Kasabach-Merritt syndrome. In our institution a study of IFN-alpha-2a therapy for endangering hemangiomas is in progress (Eczekowitz et al, in press). All infants entered into the study have serious lesions that failed to respond to systemic corticosteroids. Most of our 20 patients have shown resolution of their hemangioma after an average of 11 months of treatment. There have been, to date, two deaths on therapy. This preliminary study suggests that IFN-alpha-2a should be used in infants with life-endangering hemangiomas that do not respond promptly to corticosteroids and that this is the agent of choice for Kasabach-Merritt syndrome. No serious side-effects have been seen in the short-term use of this drug.

Surgical therapy. Surgical excision during childhood is usually indicated for removal of the fibrofatty residuum or skin laxity that remains after complete regression of hemangioma. There are, however, indications for earlier operative intervention. If a hemangioma is causing visual problems and is unresponsive to corticosteroid therapy, subtotal excision may relieve pressure on the developing cornea. Another example is early excision of an obstructing subglottic hemangioma using CO_2 laser as discussed earlier. There are instances when the hemangioma is pedunculated and ulcerated. These lesions should be removed rather than waiting for involution to occur.

As the child with hemangioma reaches age 2.5 to 3 years, he or she may evidence psychosocial problems. This is the age when a child first manifests a defined body or facial image. Usually the child with facial hemangioma is accepted by nursery school and kindergarten classmates. Problems are more likely to develop during the first grade, when the child is exposed to older classmates. This is a time for considering subtotal or possibly total excision of an involuting-phase hemangioma. Excision for psychologic indications must be carefully discussed, and often a psychologist can help with the decision. Excision is also a consideration when it is obvious that skin removal will be necessary in the future, either because of color, quantity, or contour, notwithstanding the final result of involution. Hemangioma of the lip and the nasal tip are psychologically sensitive foci. Subtotal or contour excision may be of benefit during early childhood (Fig. 20-10). As a general rule, subtotal or staged excision is the best approach. Caution should be taken for fear of causing late-contour deficiency or deformity. Residual fibrofatty tissue and skin can always be removed, often with local anesthesia, when the child is older.

Vascular Malformations

Vascular malformations usually grow commensurately with the child. Each channel type, however, may change because of different pathophysiologic influences. Venous anomalies often expand because of hormonal changes such as puberty, pregnancy, antiovulants, or secondary to trauma. Lymphatic malformations typically enlarge with infection or intralesional bleeding. Arteriovenous malformations may enlarge in association with trauma, puberty, other hormonal changes, or after incomplete surgical excision. It is essential not to confuse these nonproliferative enlargements of vascular malformations with the proliferating phase of hemangioma.

Capillary malformation

Histology

The skin or mucosa involved with a port-wine stain contains abnormally dilatated capillaries or venule-sized vessels in the superficial dermis (Noe et al, 1980). The anomaly is present at birth and changes slowly with growth to a purple color in adulthood. Furthermore, the lesion may become raised and nodular with progressive vascular ectasia (Fig. 20-11).

Clinical features

Patients with port-wine stain within the V1 area alone or extending into the maxillary and mandibular region are at serious risk for having choroidal and intracranial vascular anomalies; that is, the Sturge-Weber syndrome (Enjolras et al, 1985). Those children with dermal staining of V2 and/or V3 general areas alone not at increased risk. The capillary malformation in Sturge-Weber syndrome may involve the entire face and neck or trunk and distal extremities. The soft tissue is often hypertrophic, and frequently skeletal overgrowth occurs in the maxilla, particularly the alveolar region. Skeletal overgrowth may not be obvious at birth, but it is progressive throughout adolescence.

Cerebral angiography of patients with Sturge-Weber syndrome displays capillary,

venous, and arteriovenous anomalies of the leptomeninges (Poser and Taveras, 1957). The anomalous circulation is responsible for the progressive degeneration and atrophy of the cerebral hemispheres (Lichtenstein, 1954). Early diagnosis of intracranial anomalies can be made with single-photon-emission CT (SPECT), positron-emission tomography (PET) or MRI. In time, serpentine calcifications may be seen in the temporal and occipital and occipital lobes. A 45% incidence of associated glaucoma occurs if the port-wine stain involves both the ophthalmic and maxillary divisions of the trigeminal nerve. The diagnosis is based on the presence of elevated intraoperative pressure, increased corneal diameter, and typical defects in the visual fields.

Treatment

Laser therapy. Laser therapy is now the established treatment for port-wine stains in both children and adults. Thermal thrombosis occurs within the port-wine stain, with resultant lightening of the port-wine stain and flattening of its surface. The efficiency of argon laser therapy can be predicted on the basis of age, color of the port-wine stain, and a preliminary biopsy (Noe et al, 1980). Argon laser is used for adult patients.

The flashlamp pulsed tunable dye laser is preferable in treating young children, patients with light-colored port-wine stains, and those whose skin is very sensitive to heat (Tan et al, 1986, 1989) (Fig. 20-10, B-C).

Surgical excision. In selective cases, it may be possible to surgically excise the port-wine stain and obtain primary closure by skin advancement, split-thickness, or full-thickness skin. There are serious potential problems after excision and grafting, including scarred hypertrophy at the junction of the graft and normal skin and unpredictable pigmentation within the skin graft itself. Tissue expansion techniques may also be considered in removing a port-wine stain. Contour excision of hypertrophied lip is often necessary.

Venous malformation

In the past, venous anomalies were erroneously termed "varicose" or "cavernous" hemangiomas or "lymphangiohemangiomas". They are not true hemangiomas, but rather developmental anomalies of veins. Venous malformation (VM) usually occurs in pure form but may be combined as capillary (CVM) or lymphatic (LVM) anomalies.

Histology

Histologic examination shows dilatated or ectatic vascular channels lined by normal endothelium. Thrombosis is common and is associated with dystrophic calcification, clinically and radiologically manifested as phlebitis.

Clinical features

Various presentations of VM can occur, ranging from an isolated skin varicosity or localized spongy mass to complex lesions infiltrating various tissue planes (Fig. 20-12). The overlying skin may be normal, or it may exhibit a bluish tinge caused by involvement of the dermis. The combined venous-lymphatic lesions often exhibit dermal lymphatic vesicles

overlying the deep venous anomaly.

Venous anomalies are common in the skin and subcutaneous tissue of the head-and-neck region, particularly so in the lips and cheeks. They also may be formed in skeletal muscle; intramasseteric lesions are the most common (Welsh and Hengerer, 1980). The clinical characteristic is a soft, compressible nonpulsatile mass with rapid refilling. Expansion will occur on compression of the jugular vein or Valsalva maneuver, or with the head in a dependent position. These lesions grow proportionately with the child and tend to expand following puberty, trauma, or attempted subtotal excision. Sluggish flow and stasis lead to phlebothrombosis, which presents clinically as recurrent pain and tenderness. Characteristic phleboliths can be palpated and seen on radiographic examination. Venous anomalies may also occur within the craniofacial skeleton and are most common in the mandible, less common in the maxilla, and rare in the nasal and cranial bones and zygomas. Mandibular venous anomalies may present with increased mobility of the teeth, expansion of the buccal cortex, or spontaneous bleeding (Kaban and Mulliken, 1986). The radiographic appearance of an intraosseous VM is pathognomonic: plain films demonstrate a localized hypolucency with a honey combed, or "soap bubble", appearance (Sherman and Wilner, 1961). Profile or tangential films show spicules of bone radiating in a sun-burst pattern.

Treatment

Most VMs do not require any specific treatment apart from reassurance and an explanation of the natural history of the lesion.

Sclerotherapy. Direct injection of a sclerosing agent into the center of the soft-tissue VM is an accepted mode of treatment. The sclerosing agent, 95% ethanol and sodium tetradecyl sulfate (1% or 2%) or cryoprecipitate, is injected into the epicenter of the venous anomaly during occlusion of the arterial inflow and venous outflow. A skilled interventional radiologist is needed.

Surgery. Surgical resection is indicated for large or symptomatic venous anomalies. Often it is advisable to initially shrink the VM with sclerotherapy. Under most circumstances, total surgical extirpation is impossible, and a subtotal resection is indicated to reduce bulk and improve contour and function or relieve pain. Lesions of the jaw, nasal bones, and zygoma are managed by curettage and packing with a hemostatic agent.

Lymphatic malformation

Perhaps in no other category of vascular anomalies has nomenclature played such a perverse role as in lymphatic malformation (LM). Most authors agree that these lesions are abnormalities of lymphatic development. Therefore the term *lymphatic malformation* seems most appropriate. However, a persistent notion exists that these anomalies have the potential for neoplastic growth and recurrence after excision. This misconception is fostered by the old terms "cystic hygroma" and "lymphangioma". In a strict semantic sense, the suffix *-oma* denotes a potential for growth by cellular mitosis. There is, however, little evidence that untouched lymphatic anomalies show increased cellular turnover. But once transsected, LMs do, indeed, have a decided tendency to proliferate into the surgical scar.

LMs are uncommon congenital lesions that may occur throughout the body, although the head-and-neck region is the most common site. Most LMs are seen at birth. There is equal frequency in both sexes and in all races (Gross, 1953). Symptoms are related to the anatomic location of the malformation and the extent of involvement.

Histology

The classical LM consists of multiple dilatated lymphatic channels lined by a single layer of flattened endothelium. The vessel walls are of variable thickness and are fibro-muscular, with both striated-muscle and smooth-muscle components. Collections of lymphocytes are common throughout the contained connective tissue. Hemorrhage within the cystic spaces is common, indicating recent trauma or spontaneous bleeding, or the lesion may be a combined capillary-lymphatic malformation (CLM) or lymphatovenous malformation (LVM) (Mulliken and Glowacki, 1982; Mulliken and Young, 1988).

Clinical features

LM presents in various diverse forms. Clusters of watery vesicles are common on the buccal mucosa, tongue, and conjunctiva. The vesicles often are red or black, the result of microscopic bleeding. There may be a generalized infiltration of the tongue, a common cause of macroglossia. LM is also a common basis for macrocheilia, macrotia, macromala, and macrodontia (Mulliken and Young, 1988). The cystic variety, formerly called a "cystic hygroma", occurs in the anterior and posterior triangles of the neck. This lesion consists of large, thick-walled cysts, with less infiltration of surrounding tissue. The more severe forms of LM malformation extensively infiltrate throughout the head and neck, producing extensive deformities. This variety commonly involves the oral cavity, oropharynx, and preepiglottic space. Isolated supraglottic malformations are rare and usually occur and are associated with these extensively infiltrative lesions.

LMs are associated with hypertrophy of bone as well as of soft tissue. This is frequently seen as progressive distortion of the mandibular body, causing prognathism. LMs grow commensurately with the patient; however, spontaneous, sudden, and rapid increase in size associated with infection or intralesional bleeding may occur. Spontaneous decompression or shrinkage of LM is extremely uncommon but may occur in cystic lesions of the lower neck (Mulliken and Young, 1988).

LMs can be divided into two treatment categories based on anatomic distribution, computerized imaging, and histologic findings (Friedman et al, in press).

Type 1 LMs are located below the level of the myohyoid muscle and involve the anterior and posterior cervical triangles. CT reveals ringlike margin enhancement with sharp demarcation of the cystic areas, which appear to be well circumscribed and discreet (Fig. 20-13). Histologically, type I malformations have large cystic structures without infiltration of surrounding soft tissue. These malformations correlate most closely with the lesions previously called "cystic hygromas" (Fig. 20-14).

Type II LMs are found above the level of the myohyoid muscle and involve the oral cavity, lip, and tongue. CT scans of type II lesions reveal iso-dense masses that are poorly

defined and show obscured muscle and fatty planes. These lesions do not appear well circumscribed or discreet, and the ringlike enhancement seen in the type I lesions is notably absent. Histologic examination of type II lesions reveals smaller lymphatic channels with infiltration of the surrounding tissue.

Treatment

Therapeutic modalities such as repeated incision and drainage, aspiration, radiotherapy, and injection of sclerosing agents should be avoided. Surgical resection is the treatment of choice for control of this vascular anomaly. Appropriate surgical planning should be based on an understanding of the two forms of LMs and realistic expectations for each patient.

The physical examination of the patient and the preoperative imaging results are extremely important, bearing in mind the clinical significance of the typical infiltrative disease above the myohyoid muscle. Photographic documentation is important because the care of many of these patients spans several years. It is helpful to the surgeon and the family to recall the initial condition as well as interval results.

Surgical excision should be undertaken recalling that LMs are benign; therefore, complete and total resection of the lesion is not necessary and sometimes not possible. Aggressive resection is not warranted, nor is an excessively conservative approach to be condoned. The long-term psychologic and social ramifications of these malformations should not be underestimated.

Cold knife dissection is frequently the modality of choice. Type I lesions are ideally removed in one procedure because repeated excisions are complicated by fibrosis and anatomic distortion. As many of the type I lesions are resected fairly easily, surgical intervention can be safely carried out within the first 9 to 12 months of life.

Type II lesions are the most difficult to manage because no distinct tissue planes between the malformation and the normal structures exist. As these lesions are not curable, the timing of intervention is less critical. Repeated procedures are necessary, and complete removal is almost impossible. Resection of LM before the age of 5 or 6 years is recommended. In planning such a procedure, restrictions should be set for the extent of dissection, duration, and acceptable blood loss.

Meticulous dissection is necessary because anatomic structures frequently are not in their normal position. Magnification and the use of the nerve stimulator are necessary. The disruption of the abnormal lymphatic channels frequently leads to prolonged wound drainage. Suction drains should remain in position for an adequate period to avoid reaccumulation of lymphatic drainage under the skin flaps.

The laser has a role in the management of these lesions. The use of the laser is reserved for disease that is not readily resectable by sharp dissection. Lesions involving the oral cavity and tongue are particularly amenable to CO_2 laser resection, as is involvement of the supraglottic region. The CO_2 laser may be helpful in localized lesions in the lip and buccal area. Nd-YAG has been relatively successful in controlling the vesicular lesions that are commonly found in the tongue.

Special considerations

Airway obstruction. Airway obstruction caused by a cervical lymphatic anomaly and requiring intervention occurs primarily within the first 12 months of life. The lesion is usually an infiltrative LM involving the tongue, floor of the mouth, and preepiglottic space. A sudden increase in size demands immediate attention. Attempts at aspiration are unsuccessful. With progressive respiratory distress, evidenced by tachypnea and increasing chest retractions, orotracheal intubation becomes necessary. This procedure should be performed under general anesthesia, and it provides an opportunity for detailed examination of the oropharynx, hypopharynx, and supraglottic larynx. If extensive involvement of the supraglottic airway exists, tracheotomy is indicated.

Hemorrhage. Hemorrhage into a cystic LM in the anterior triangle is sometimes associated with the sudden onset of acute respiratory distress. The airway should be secured with nasotracheal intubation, and intravenous antibiotics should be started. If there is a discreet mass, it may be removed and cause significant improvement in the airway.

Sepsis

A rapid increase in the size of the LM within the floor of the mouth or the tongue is usually secondary to cellulitis. There are usually systemic signs of infection. These clinical situations necessitate admission to the hospital, monitoring of the airway, and often prolonged intravenous antibiotics. If there is recurrent cellulitis, prophylactic antibiotics may be needed. Dental hygiene should be stressed in these children.

Arteriovenous malformation

The high-flow vascular anomalies in the head and neck are arteriovenous malformation (AVM) or, less commonly, arteriovenous fistulae (AVF). These fast-flow lesions are relatively uncommon as compared with slow-flow vascular anomalies. Most reports describe an experience with less than 15 cases (Malan and Azzolini, 1968). In contrast, AVMs are 20-fold more common in the intracranial vasculature than in branches of the external carotid (Olivecrona and Ladenheim, 1957).

Histology

Histopathologic examination using serial sectioning technique may demonstrate the AV shunts. The dysmorphic arteries are thick-walled and of irregular caliber. Under high-power magnification, the arteries exhibit fragmentation of the internal elastic lamina and highly disorganized smooth muscle in the media. Secondary changes occur within the veins; it is, progressive reactive hypertrophy, intimal thickening, and sclerosis caused by increased blood flow occur.

Clinical findings

AVMs in the head and neck are rarely symptomatic in the neonatal period. Instead, they manifest during late childhood, adolescence, or early adulthood. Many lesions have either a warm erythematous blush or a true port-wine stain in the overlying skin. Distressing

symptoms of AVM or AVF in children are throbbing pain, buzzing, or pulsatile tinnitus, which may prevent sleeping (Coleman, 1973; Malan and Azzolini, 1968). In time, the involved skin has an elevated temperature, and a thrill may be felt. AV shunting is confirmed by the presence of a bruit on auscultation or Doppler examination. These anomalies remain stable for years, only to expand following minimal trauma, infection, or hormonal changes. Shunting of blood diminishes nutritive flow, which may result in skin necrosis, ulceration, and bleeding. Slow destruction of facial bones may occur; the patient seeks treatment for swelling, pain, or sudden hemorrhage.

Treatment

If an AVM or AVF is asymptomatic, no treatment is necessary. But with complications, such as pain, ulcerations, bleeding, or heart failure, therapy is necessary. There *is no place for proximal ligation* of the feeding arterial system (Fig. 20-15).

MRI and angiography are essential for evaluating symptomatic malformations. MRI best portrays the extent of involvement within tissue planes and demonstrates the flow characteristics. Angiography is usually reserved until treatment planning is complete and is used for superselective embolization before surgical extirpation. Superselective embolization may have a role in palliation (ie, for control of pain, tinnitus, or hemorrhage), or it may be used as primary therapy for surgically inaccessible AVM or AVF.

The only therapy that carries any hope for long-term success is total resection of the tissue that is involved with the AV anomaly. Leaving behind residual and dormant anomalous channels only invite further collateral formation, shunting, and expansion (Mulliken and Young, 1988). Preoperative superselective embolization will not diminish the extent of the resection. It will, however, minimize intraoperative bleeding. Preoperative embolization of proximal feeding vessels only complicates the problems. Embolization must be of the nidus, or epicenter, of the AVM. Often a two-team approach (for resection and reconstruction) is applicable to surgical management of these lesions. The critical decision is how far must the resection go to include all the pathologic vasculature. Reconstruction often necessitates closure and soft-tissue replacement using microvascular tissue transfer. Given proper indications and with careful planning, extensive resection is curative and justified.

In the past, these fast-flow vascular anomalies were considered insoluble problems. Accepting these patients for treatment is a challenging commitment.