Management of Congenital Vascular Lesions of the Head and Neck

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Congenital vascular lesions are often misdiagnosed and, for the most part, left untreated. The absence of a uniformly accepted classification of these lesions and confusion over their natural history are partly responsible. A new classification of these lesions recognizes two distinct groups of lesions, hemangiomas and vascular malformations.

Introduction

Until recently, the treatment of congenital vascular lesions lacked a protocol. This stemmed, in part, from the absence of a uniformly accepted classification and the lack of a clear understanding of the natural history of these lesions. Further compounding the problem was the widely held notion that, given time, most such lesions would spontaneously resolve. Congenital vascular lesions were therefore misdiagnosed and, for the most part, left untreated. The plight of these unfortunate patients led them from physician to physician in search of help.

The renaissance began with a (biologic) classification first proposed by Mulliken and Glowacki. They dispensed with the old, often confusing terminology and replaced it with a clear, concise set of terms and definitions. Mulliken and Glowacki recognized two entirely distinct groups of congenital vascular lesions: hemangiomas and vascular malformations. They defined hemangiomas as lesions that were usually not present at birth, proliferated during the first year of life, and then involuted over the ensuing years. The term "vascular malformations" was reserved for lesions that were always present (although not necessarily apparent) at birth, never proliferated, and never involuted.

Hemangiomas

Most hemangiomas first appear soon after birth, but as many as 30% may be apparent at birth. They characteristically proliferate during the first year of life and then involute sometime during childhood.

Proliferative Phases

Although the rate and timing of proliferation within this period is extremely variable, two periods of rapid growth are frequently seen. The first and most common of these proliferative phases takes place during the neonatal period and early infancy and the second, between 4 and 6 months of age. Histologic features of proliferation include tubules of plump, proliferating endothelial cells with frequent mitoses and an abundance of mast cells. Little or no intervascular stroma is apparent, and the lesion appears cellular (Figure 1).

Hemangiomas may originate in the superficial papillary and/or reticular dermis. These lesions were previously known as capillary hemangiomas, but have been renamed cutaneous hemangiomas (Figure 2). Lesions that arise from deeper subcutaneous tissues, previously known as a cavernous hemangiomas, are now termed subcutaneous hemangiomas (Figure 3). Hemangiomas that stem from either the superficial or deeper tissues but extend across both planes are referred to as compound hemangiomas.

Involution Phase

Proliferation invariably slows by the end of the first year of life, and a long period of involution follows. During involution, the once active proliferating endothelial cells become less active and progressively flatten until late in involution, when they are not dissimilar from cells seen in normal vascular channels (Figure 4). The involution process is usually accompanied by an increase in intervascular fibro-fatty tissue, which eventually replaces most of the hemangioma. Thus, the lesion changes from a predominantly cellular to a predominantly vascular tumor and finally to a fibro-fatty mass of tissue with a few ectatic vascular channels.
Rate and Patterns of Involution--The rate of involution is extremely variable. To date, no known characteristics appear to influence either the rate or completeness of involution. Waner and Suen have identified two distinct patterns of involution (Figure 5). In the first group, lesions involute rapidly, and the involution process is complete by between 5 and 6 years of age. This group is more likely to have a satisfactory outcome and are thus less likely to require corrective surgery. Finn et al reported that only 40% of such patients required corrective surgery [3]. In the second group, lesions involute at an extremely slow rate; this may take up to 12 years and is less likely to be complete. Approximately 80% of patients with this type of lesion ultimately require corrective surgery.

Diagnosis

The diagnosis of a hemangioma is made, for the most part, on clinical grounds. The classic feature, namely, proliferation during the first year of life, sets this lesion apart from all other vascular lesions. A superficial or cutaneous lesion is a red, bosselated mass, whereas a deeper lesion presents as a bluish mass. The mass is much firmer during proliferation and softer and more easily compressible during involution. Parents often notice the mass expanding when the child cries or the lesion is in a dependent position. Subcutaneous hemangiomas are often more difficult to diagnose. When the diagnosis is in doubt, an MRI is useful in confirming the presence of a hemangioma. Hemangiomas are high-flow, solid-tissue lesions of intermediate intensity on T1-weighted presaturation images.

Treatment

Current doctrine dictates that hemangiomas should, for the most part, be left untreated (a policy known as benign neglect). However, contrary to widely held belief, a cosmetically unacceptable result is often left at the end of involution [3-5]. If this is so, then the consequences of benign neglect are unjustified. The psychosocial trauma of a readily visible hemangioma, the disappointment of an unacceptable cosmetic result at the end of involution, and the further trauma of corrective surgery all support the need for an alternative approach. Treatment should be aimed at early intervention and/or complete resolution by the age of 3 to 4 years.

Early Proliferation

Laser photocoagulation of superficial proliferating hemangiomas results in complete destruction of the hemangioma with little or no scarring (Figure 6) [4-6]. Using a 5-mm spot size and overlapping individual treatment areas by about 30%, the entire lesion should be treated with a flashlamp pumped dye laser at between 6.5 and 8 J/cm² until a purplish-gray discoloration is achieved. Considerably more overlap may be needed to achieve this. Garden et al recommended administering treatment in four weekly cycles until a complete response is noted [5].

Late Proliferation

Most established hemangiomas or deeper lesions have acquired a thickness and/or depth beyond the reach of yellow light and thus cannot be treated effectively with this modality. For lesions that have reached this stage [1], a decision should be made as to whether active intervention is warranted, or alternatively, whether benign neglect is preferable. Benign neglect, in the form of parental reassurance and careful follow-up, is often appropriate during this phase. Exceptions include an obvious disfiguring facial hemangioma, a massive lesion of the head and neck, or any complication. In these cases, a strategy aimed at complete ablation of the lesion, or if this is not possible, diminution of the ultimate lesion size by the end of involution, will suffice.

Prednisone--A course of oral prednisone (2 to 4 mg/kg) given over a 4-week period and tapered over a further 2 weeks is recommended. Response to prednisone is usually rapid and quite obvious within a week after therapy is begun. Should no response be noted, the drug dose should be tapered and an alternative strategy sought. Prednisone is only effective during proliferation and should not be used during any other phase. In 60% of patients, the hemangioma will respond to prednisone. Half of responsive lesions shrink and do not recur, whereas the other half reappear after the course of steroids has been completed. For recurrent lesions, a second course of prednisone, after an interval of 6 weeks, or alternatively, a maintenance dose of ½ to 1 mg/kg given daily or on alternate days should be considered and should be weighed against the potential risks and benefits. At least one group of investigators recommend that prednisone be continued despite potential side effects [7]. The list of these side effects is considerable. Fortunately, relatively few manifest over a short 6-week regimen. Of these, the most
frequent include temporary growth retardation, transient fluid retention, central obesity, and appetite stimulation. Infants receiving steroids also are at increased risk of infection as a consequence of depressed T-cell function.

Azzolini et al first made a case for using intralesional steroids in 1970 [8]. They contended that intralesional administration would reduce or even eliminate systemic side effects of steroid therapy. However, many investigators now believe that intralesional steroids are probably absorbed and then act systemically. Thus, there is no clear advantage to intralesional injection and perhaps even a reduced local effect.

Interferon Alpha 2a (Roferon-A) has also been shown to exhibit antiangiogenic activity. Ezekowitz et al reported promising results with this agent in treating life-threatening lesions, as well as lesions that showed signs of obstructing the visual axis [9]. The recommended dose is 3 million U/m² of body surface area given daily as an SC injection. Unfortunately, long periods of treatment are necessary (up to 13 months in cases of high-output cardiac failure).

**Complications**

Although complications of hemangiomas are infrequent, they should be anticipated and treated as they appear (Table 2).

**Ulceration**, perhaps the most frequent complication, is due to rapid proliferation within the papillary dermis, which eventually splits the overlying skin. Since the hemangioma grows at a far more rapid rate than the basal epidermal cells, ulceration may persist for several months if left untreated. Treatment with a flashlamp pumped dye laser has been shown to be extremely beneficial in temporarily retarding superficial proliferation of the hemangioma, which enables epidermal closure to take place. Usually only one or two treatments within a 4-week period are necessary to accomplish this.

**High-output cardiac failure** is a rare, but often fatal, complication of hemangiomas. It is usually seen during rapid proliferation and may be associated with a single large hemangioma, multiple cutaneous lesions, a visceral lesion, and/or a hepatic lesion. Cardiac failure should be treated in the usual manner; however, in refractory cases, prednisone, interferon, or surgical resection may be helpful, especially when a large solitary hemangioma has been incriminated.

**Airway obstruction** can be treated conservatively with a tracheostomy and extubation once the hemangioma has involuted. On the other hand, Healy et al reported a high rate of success treating subglottic hemangiomas with a carbon dioxide laser [10]. As a result, it is recommended as the treatment of choice. However, since subglottic stenosis is possible after carbon dioxide laser ablation, this modality should be used with caution. Yellow light lasers should be more effective, but experience with these lasers is limited.

**Visual axis obstruction** (deprivation amblyopia) is probably the most common cause of blindness in first world countries. Prednisone, interferon, and, if necessary, surgical resection should be undertaken to ensure unimpeded vision.

**Kasabach-Merritt syndrome**--Of more concern, however, is the phenomenon known as the Kasabach-Merritt syndrome. This syndrome is characterized by profound thrombocytopenia resulting from platelet sequestration and destruction within the hemangioma, as well as a consumptive coagulopathy. If left untreated, Kasabach-Merritt syndrome results in a generalized bleeding disorder and is potentially fatal.

The syndrome usually is clinically evident in the first few weeks of life. Edema and ecchymosis surrounding a rapidly proliferating hemangioma are early signs (Figure 7). The edema spreads rapidly to involve a large surface area and should heighten one's clinical suspicion for this syndrome. A complete blood count and coagulation profile are diagnostic. Profound thrombocytopenia, with or without an elevation in fibrin degradation products, is frequently seen.

The early aggressive management with prednisone, 4 mg/kg body weight, and interferon alpha-2a (3 million U/m² of body surface area given daily as a subcutaneous injection) is essential. Fresh frozen plasma and platelet concentrates should be infused as needed until the complication is brought under control. Once the platelet count recovers and remains within the normal range, the prednisone dose can be tapered, but the child should remain on subcutaneous injections of interferon until at least 8 months of age. Early withdrawal from this regimen may result in a recurrence of this potentially lethal complication [9].

**Involution**

Once the phase of proliferation has ended, the patient is best treated expectantly. Follow-up examinations should be done every 6 months to monitor the progress of involution.

**Rapid Involution**--Rapid progression of involution warrants a "wait and see" policy; ie, one should wait until this process is complete before deciding whether or not any treatment is necessary. As
mentioned previously, 60% of patients whose lesions involute rapidly experience complete involution and do not require any further treatment. In the remainder, residual telangiectasia can be readily photocoagulated with a laser, and any contour deformity resulting from residual fibro-fatty tissue can be surgically reduced.

**Slow Involution**—If the process of involution proceeds at a much slower rate, a residual defect that requires surgical correction is likely in at least 80% of cases. It is therefore prudent to intervene when the child is 3 to 4 years of age, before he or she enters school.

Laser photocoagulation is valuable for the treatment of superficial lesions, whereas surgical resection is essential for removing deeper lesions. A compound or combined lesion should be treated first with a laser and subsequently excised (Figure 8), as initial treatment with a laser enables the surgeon to preserve more skin and even raise a skin flap over the deeper component [2].

### Vascular Malformations

Vascular malformations are true developmental anomalies and, as such, are always present at birth [1]. However, in contradistinction to hemangiomas, vascular malformations never proliferate or involute. Instead, slow steady growth, commensurate with the growth of the child, is the norm. Rapid expansion is seen with infection, after trauma, and around puberty, but this occurs through a process of hypertrophy rather than hyperplasia. Existing vascular channels become more ectatic. Vascular malformations have been further subclassified according to their vascular component. The subcategories recognized thus far are listed in Table 3. Since the features and management of this diverse group of lesions are dependent on the vascular content, they will be discussed separately.

#### Arteriovenous Malformations

An arteriovenous malformation is an acquired malformation that consists of a nidus, an afferent blood supply, and venous drainage. Waner and Suen recently studied the histologic features of the nidus and found that it consists of a dilated capillary bed (January 1995). Presumably, an absence of precapillary sphincters results in arteriovenous shunting across this capillary bed, and over time, expansion of the lesion by recruitment of adjacent capillary beds occurs, along with hypertrophy of the feeding arteries and dilatation of draining veins. Therefore, early in the process, the nidus is quite distinct and can be easily excised (Figure 9). At a later stage, however, it is far more difficult to separate the nidus from its blood supply and venous drainage.

**Treatment**—Of all vascular malformations, arteriovenous lesions are the most difficult to manage. Surgical resection remains the only way to completely eradicate an arteriovenous malformation. The timing of surgery is crucial. Resection should be undertaken soon after the diagnosis has been made since progressive expansion of the lesion will make surgery far more difficult. The nidus must be completely removed during resection, as any residuum will invariably result in a recurrence. The margins of resection are less distinct in an older patient. Consequently, when removing an arteriovenous malformation in such a patient, an aggressive approach should be taken. Embolization as a single modality does not cure an arteriovenous malformation. This modality should be considered only as a preoperative adjunct to surgery to reduce intraoperative blood loss. Even if preoperative embolization is done, however, meticulous attention to hemostasis during surgery is also necessary. Surgical ligation of the afferent arteries should be discouraged since the lesion will invariably parasitize a blood supply through a collateral source and can become even more difficult to embolize and/or resect.

#### Venous Malformations

Unlike arteriovenous malformations, venous malformations tend to be more diffuse and are composed of a collection of ectatic, malformed, dilated veins. The initial cause is believed to be a relative or complete absence of an autonomic venous innervation with a loss in venomotor tone. This, in turn, results in progressive venous ectasia and expansion of the lesion. Venous malformations usually are readily visible and most often are easily diagnosed clinically. **Treatment** is aimed at complete ablation/excision of the lesion. A superficial component, especially one involving the mucosa, can be readily treated with a copper vapor laser or a neodymium/yttrium-aluminum-garnet (Nd:YAG) laser in the noncontact mode [11]. Deeper components should be surgically excised. Meticulous attention to hemostasis is essential and will enable preservation of important structures. As with arteriovenous malformations, any residuum left at the end of treatment of venous malformations will invariably result in a recurrence. Treatment, therefore, should be aimed at complete ablation whenever possible. Unfortunately, in the presence of an extensive lesion, this may
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be difficult and sometimes impossible. At least two groups are exploring the possibility of interstitial laser photocoagulation [12,13]. Placement of a quartz fiber coupled to an Nd:YAG laser, under MRI guidance, enables one to deliver sufficient energy to coagulate a predictable amount of the venous malformation. This therapy can be repeated until the entire lesion has been ablated.

**Lymphatic Malformations**

Lymphatic malformations may be either cystic or diffuse. Cystic lesions commonly occur in the neck and are referred to as cystic hygromas, whereas diffuse lesions more often are found on the face and upper aerodigestive tract. Both patterns appear to be equally common and may coexist. **Treatment**—Lymphatic malformations should be excised as completely as possible. Excision may have to be repeated several times before a satisfactory result is obtained. It is possible to preserve important structures while excising as much of the malformation as possible. As is the case with other vascular malformations, any residuum will recur, although it may take several years before a recurrent lesion is noticeable.

**Capillary and Venular Malformations**

These two subclassifications include all the cutaneous vascular birthmarks and are thus considered together. **Capillary malformations** are quite distinct from venular malformations in that they are usually midline and nonprogressive. These lesions typically are found on the forehead, upper eyelids, and nasal alae, as well as the central portion of the upper lip. The nape of the neck is another common site. Capillary malformations disappear spontaneously in about 40% of cases. Those that persist are readily amenable to treatment. Waner et al reported a complete response to photocoagulation with a flashlamp pumped dye laser in over 90% of cases [14]. **Venular malformations**, or port-wine stains, are made up of ectatic postcapillary venules in the reticular and papillary dermis. Waner et al postulated that the presence of a "sick dermatone" is central to the pathogenesis of these lesions [14]. A developmental relative or absolute deficiency of an autonomic nerve supply to the cutaneous venous plexus results in progressive dilatation of this plexus, and hence, the port-wine stain. It is important to realize that port-wine stains are not static, but rather, are steadily progressive lesions. In time, almost all will thicken and eventually cobblestone (Figure 10). These lesions can be quite disfiguring and should therefore be treated when first diagnosed. Most pediatric and early port-wine stains are best treated with a flashlamp pumped dye laser, whereas more advanced and cobblestoned lesions are treated initially with the copper vapor laser. Somewhere between 15% and 20% of lesions will respond completely to laser therapy, and the vast majority will lighten significantly [14,15].

**Mixed Malformations**

The most common mixed malformations are venous lymphatic malformations. Occasionally, a mixed venous venular malformation or a venous malformation overlying an arteriovenous malformation may be seen. **Treatment** of mixed lesions should be determined by the vascular component(s) present. For example, a deep mixed venous lymphatic malformation should be excised; after the deeper lesion has been removed, however, the venular component of a venous malformation can be treated with a laser. The same treatment approach can be used for a venular malformation that overlies an arteriovenous malformation.

**Summary**

The new classification of congenital vascular lesions of the head and neck recognizes distinct groups of lesions: hemangiomas and vascular malformations. Knowledge of this classification is essential to the proper diagnosis and management of these lesions. The time-honored policy of calling all vascular lesions hemangiomas and treating them conservatively should finally be abandoned; instead, each type of lesion should be treated individually.

**References:**


Links:
[1] http://www.diagnosticimaging.com/review-article