Birthmarks: I. Vascular Nevi
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Birthmarks: I. Vascular Nevi

Alvin H. Jacobs, MD

Birthmarks are nevoid anomalies resulting from faulty embryogenesis. In other words, they are hamartomas or benign tumor-like malformations of an organ composed of inappropriate admixtures of its histologic components. These new growths of congenital origin may be visible at birth or may not actually be apparent until some time after birth.

This review of birthmarks (nevi) will be presented in two parts: vascular nevi (I) and melanocytic and epidermal nevi (II) to appear in a future issue of Pediatrics in Review.

FLAT VASCULAR NEVI

Salmon Patch

The commonest of the vascular birthmarks is the salmon patch (Fig 1) (nevus simplex, erythema nuchae, angel's kiss, stork bite, etc). Pratt,1 in a study of 1,096 neonates, found 43.5% had salmon patches. More recently Jacobs and Walton2 examined 1,058 newborn infants and found 40.3% of them to have salmon patches. These dull pink birthmarks which are present at birth, occur most frequently on the nape of the neck, mid-forehead, glabella, and upper eyelids. Many infants have salmon patches in more than one of these locations. Of the total number of infants with salmon patches in the Jacobs and Walton series, 81% had the lesions at the nape, 45% on the eyelids, and 33% on the glabella.

Salmon patches are composed of ectatic dermal capillaries that represent the persistence of fetal circulatory patterns in the skin. No treatment is indicated since they usually fade away during the first year of life, although those at the nape may take longer to disappear.3 About 5% of those at the nape remain permanently.

Port-Wine Nevi

The port-wine stain is often referred to as nevus flammeus. However, this term is also used in reference to the salmon patch. Since these two types of lesions have a different significance and different prognosis, it is preferable not to use the inclusive term. The port-wine stain is also present at birth, but shows no tendency to resolve. This type of lesion is usually primarily unilateral; in general, it respects the midline, although in some cases a portion of the lesion does extend to the other side (Fig 2). The color is usually pink in infancy but tends to develop a dull erythrocyanotic hue or purple tone with advancing age. Initially the lesions are entirely macular with a smooth surface. With increasing age, especially after the fourth decade, the surface may become irregular, thickened, and nodular. This type of nevus may appear on any area of the body although these nevi seem to be somewhat more common on the face and neck. The frequency of port-wine stains is 0.3%.

Microscopically, the port-wine stain is composed of dilated mature capillaries in the dermis. There may be associated connective tissue hypertrophy when deeper vessels are involved.

No uniformly acceptable therapy exists for the cosmetically disfiguring port-wine stain. Excision, tattooing, radiation, cryosurgery, and dermabrasion have been employed without convincing success. Only the very smallest of lesions can be excised successfully. Tattooing with insoluble light colored pigments usually produces a mottled appearance. X-irradiation should be mentioned only to condemn its use in any benign, functionally uncompromising condition affecting patients in the pediatric age group. Camouflaging...

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cosmetics with opaque bases are still the best treatment available for port-wine nevi on exposed areas. Recently much publicity has been given to treatment of port-wine nevi with the argon laser. Apfelbert and associates have treated many vascular lesions with the argon laser and are pleased with the results, although they admit that the lesions are not completely eliminated and that scarring often occurs. Goldman, a pioneer in the medical uses of the laser, states, "For practical reasons, at present, only relatively small port-wine marks can be treated effectively." He also states, "It is recommended for the present that treatment of port-wine marks by argon lasers be restricted to investigation in medical centers...."

One syndrome that involves facial port-wine stains deserves special mention. This is encephalotrigeminal angiomatosis or Sturge-Weber syndrome in which there is an association of the port-wine stain with contralateral focal seizures and often with congenital glaucoma. The seizures have been found to be secondary to ipsilateral leptomeningeal angiomatosis.

An important feature of the port-wine stain in the Sturge-Weber syndrome is that it diffusely involves the skin area innervated by the first branch of the trigeminal nerve (Figs 3a and 3b). This characteristic of the nevus was documented in a monograph on Sturge-Weber syndrome in which 257 cases of the syndrome were reviewed and no instance of CNS involvement in which the nevus was entirely below the palpebral fissure was found. The explanation of this association can be understood by knowing the embryology of the vasculature of the brain. The superficial portion of the primordial vascular plexus develops into the vascular supply of the upper face, scalp, meninges, and choroid.

Aside from the facial nevus, the first evidence of the presence of the Sturge-Weber syndrome is the occurrence of focal seizures. Skull x-rays taken in infancy rarely yield ev-
idence of the meningeal involvement, since calcification may take years to develop. Recently, early diagnosis has been established by computerized tomography.

RAISED VASCULAR NEVI

Involuting hemangiomas are elevated above the skin surface. Their nomenclature tends to be confused. The superficial capillary type is usually referred to as a strawberry nus. The cavernous hemangioma is basically the same pathologic process, but it is deeper and is made up of larger vessels. Most lesions seen in infants are composed of various degrees of both types.

Capillary and cavernous hemangiomas are quite common, occurring in about 10% of all babies under 1 year of age.7 They may be single or multiple and vary considerably in ultimate size. Any area of the body may be involved. There is no sexual or genetic predilection. The lesions are not usually present at birth but become apparent during the first 3 or 4 weeks of age. Occasionally they may be seen in the newborn infant as faint telangiectasias surrounded by a pale halo (Fig 4). These become solid red in color and elevated during the subsequent weeks. The lesions when not thus apparent at birth begin as an erythematous macule that rapidly enlarges and becomes elevated with a bright red, leather-like surface. Hemangiomatous elements may remain histologically superficial or penetrate into the subcutaneous tissues, in which case considerable anatomical distortion may result. The surface epithelium is usually thin and delicate, but may be of normal thickness if the hemangioma is exclusively composed of deep elements. Ulceration with secondary infection may occur following minor trauma. However, significant hemorrhage is uncommon. The rapid growth and associated cutaneous changes described above may erroneously suggest malignancy. After a variable period of months, the surface shows a mottled bluish-white appearance indicative of thrombosis of vascular channels with resulting diffuse scarring. This is the hallmark of spontaneous involution which is usually complete after the fifth or sixth year.7 If the lesion has not been excessively elevated and deforming no evident scar remains (Figs 5a and 5b). In the larger hemangiomas with considerable cavernous element, a relatively loose, atrophic pale scar is the only residual clinical defect unless extensive ulceration and infection have occurred during the course (Figs 6a and 6b). In the latter case, scarring may be more significant, but no restorative procedures should be undertaken until complete and unequivocal resolution has occurred. In most cases of involuted extensive hemangiomas, excision of the redundant scar is all that is required (Figs 7a and 7b).

No specific therapy is indicated for capillary and/or cavernous hemangiomas unless a vital function is disturbed by its presence (ie, sucking, breathing, vision, etc). In these cases systemic corticosteroid therapy will usually result in rapid diminution in size of the lesion. It is usually necessary to continue this therapy for four to six weeks. In some cases enlargement of the lesion requires a second course of therapy.10

Occasionally a large cavernous hemangioma is complicated by the sequestration of platelets and other clotting factors with resulting thrombocytopenia (Kasabach-Merritt syndrome). Systemic corticosteroid therapy and platelet transfusion are usually necessary.
Fig 7. Top, Large lesion that was not treated. The eye did open at times each day; vision, therefore, was not threatened. Bottom, Same lesion three years later; there is a remarkable degree of spontaneous resolution.

REFERENCES


SUMMARY

Features of:
—Salmon patch—no treatment necessary
—Port-wine stain—cosmetics best therapy when on upper face associated with Sturge-Weber syndrome
—Raised vascular nevi—no therapy necessary unless interfering with vital functions

Fig 6. Above, Large cavernous hemangioma in a 9-month-old infant. Right, Same hemangioma six years later; with no treatment, only atrophic skin remains. At this stage relatively simple plastic procedures are indicated.
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