PEDIATRIC DERMATOLOGY

NEONATAL DERMATOSES : AN OVERVIEW

Jayakar Thomas

Benign dermatoses in newborns must be distinguished from more serious disorders with cutaneous manifestations. Erythema toxicum neonatorum, transient neonatal pustular melanosis, sucking blister, miliaria and Mongolian spots are among the many benign skin conditions that can occur in newborns. Recognition of these dermatoses allows the physician to proceed appropriately, reassure the parents and initiate the further evaluation or treatment as necessary. To avoid adverse sequelae, special attention must be given to more persistent conditions and those with the potential for complications or malignant transformation. Consultation with a pediatric dermatologist, a plastic surgeon or a neurosurgeon may be necessary.

Dermatoses in newborns can cause a great deal of anxiety for parents. While the vast majority of these skin conditions are benign and transient, each case needs to be evaluated carefully to rule out the possibility of a serious disorder with cutaneous manifestations. Once a dermatosis is identified, the physician can reassure the parents and proceed with treatment, if necessary.

Erythema toxicum neonatorum

Erythema toxicum neonatorum affects 30 to 70 percent of all infants. It is less common in premature infants than in full- term infants.[2] In most cases, the eruption is initially noted 24 hours to two weeks after birth.

Characteristically, the lesions are yellow or white papulovesicles, 1 to 2mm in diameter, that develop primarily on the trunk, arms and legs. Typically, each lesion is surrounded by an irregularly shaped erythematous area. As a result, the eruption has been described as having a "flea-bitten" appearance.

Immaturity of the pilosebaceous follicles plays a role in the development of erythema toxicum neonatorum. The lesions are not present on the palms and soles, because these areas contain no pilosebaceous follicles.

Cultures of the lesions are negative for bacteria, fungi and viruses. Wright's staining of the vesicle contents reveals eosinophils. Eosinophilia has been described in some cases. Erythema toxicum neonatorum resolves spontaneously without treatment.

Transient neonatal pustular melanosis

Transient neonatal pustular melanosis occurs in 0.2 percent of white infants and 4.4 percent of black infants.[2,3] The lesions are usually present at birth.

In this dermatosis, superficial pustules or pigmented macules develop anywhere on the body but are especially likely to appear on the chin, neck, upper chest and lower back. The pustules rupture easily, and a collarette

Address correspondence to :

Dr. Jayakar Thomas, Dermatologist, Institute of Child Health and Government Hospital for Children, Chennai 600 008. India.

of white scale may be present around the pigmented macule. When only the pigmented macules are present, the pustular phase may have occurred in utero.

Cultures are negative, but the lesions are found to contain polymorphonuclear leukocytes. The macules usually resolve spontaneously within three months.

Suction blister

Suction pressure can produce a blister on any part of the body that an infant's mouth can reach.[4,5] The blister is a clear bullous lesion with no surrounding erythema. The lesion can be present at birth as bulla, an erosion or a crusted lesion. If the affected arm or leg is moved to the infant's mouth, the infant will usually begin sucking.

Cultures of the lesion are negative for bacteria, fungi and viruses. If the blister ruptures, a topical antibiotic ointment must be applied to prevent secondary bacterial infection.

Miliaria

Miliaria rubra and miliaria crystallina most often occur in the intertriginous areas, on the scalp, face and neck, or in any occluded area, such as on the back of a very ill neonate who cannot be turned.

Miliaria rubra is characterized by vesicular, pustular or papular lesions, occasionally with some associated erythema. The lesions are distributed in a follicular-type pattern. In miliaria crystallina, the lesions resemble drops of water on the skin. When ruptured, the lesions drain clear fluid (i.e., eccrine duct contents). The crystallina lesions are delicate, roofed by a thin layer of stratum corneum and rupture easily.

On histopathologic examination, miliaria rubra lesions display spongiosis and vesicle formation in the eccrine ducts. Histopathologic examination of miliaria crystallina lesions shows intracorneal or subcorneal vesicles. Immaturity of the eccrine ducts plays a role in the development of the lesions in both miliaria rubra and miliaria crystallina.

Miliaria resolves following elimination of environmental and physical factors that cause sweat gland occlusion or local temperature increase. No specific therapy is indicated.

Mongolian spots

Mongolian spots are flat, bluish black macules or patches caused by the arrest of melanocytic migration in the dermis of the embryo. These lesions are present in 96 percent of black infants, 90 percent of Native American infants, 81 to 90 percent of Asian infants, 46 to 70 percent of Hispanic infants and 10 percent of white infants.[6] The lesions are most commonly located in the lumbosacral area, but they can also occur on the legs, back, flank and shoulders.

The macules and patches of Mongolian spots were once thought to disappear some time between the ages of seven and 13 years. However, recent evidence indicates that the lesions may be more persistent.[1,4]

Mongolian spots require no treatment. The lesions are not at risk of malignant transformation.

Subcutaneous fat necrosis

Subcutaneous fat necrosis of the new-born is characterized by erythematous nodules or plaques on the cheeks, buttocks, posterior trunk, arms or legs. These characteristically painless lesions can appear in infants up to one month old. The occurrence of this dermatosis in the newborn period is thought to be related to the susceptibility of fat to become necrotic in the presence of trauma, asphyxia, hypothermia or a combination of these factors.[7]

Histopathologic examination of the nodules or

plaques reveals fat necrosis and foreign body giant cells. In most cases, the lesions resolve spontaneously. If a lesion becomes fluctuant, pressure can be relieved by aspiration with a sterile needle.

Hypercalcemia has been reported in association with subcutaneous fat necrosis of the newborn.[8]

Milia

Milia are superficial epidermal inclusion cysts. They occur in approximately 40 percent of newborns.[4] These whitish papules are 1 to 2mm in diameter and are often noted on the forehead, cheeks and nose. Similar lesions in the oral cavity are known as Epstein's pearls. Milia usually resolve spontaneously.

Acne neonatorum

The lesions of acne neonatorum develop on the face. Inflammatory papules, open and closed comedones or less frequently, nodules may be present. Androgenic hormones stimulate the sebaceous glands[9] and the acne tends to improve as the hormone levels decrease.

If treatment is necessary, over-the-counter acne soaps (which have a low pH) can be used. Prescription medications such as a 5 percent benzoyl peroxide wash or a 2.5 percent benzoyl peroxide gel can be used sparingly if further treatment is needed.

An association between acne neonatorum and adolescent acne has been noted but remains unclear.

Smooth muscle hamartoma

A smooth muscle hamartoma is composed of arrector pili muscles.[10] Clinically, the lesion exhibits hyperpigmentation and hypertrichosis overlying a firm, nodular base. On occasion, a lesion may "ripple" when it is stroked.

It may be difficult to distinguish between a hamartoma and a congenital melanocytic nevus. However, on microscopic examination of material obtained by punch biopsy, bundles of smooth muscle fibers are seen in the hamartoma.

Smooth muscle hamartomas are not at increased risk of malignant transformation.

Nevus sebaceus of Jadassohn

Nevus sebaceus of Jadassohn occurs in approximately 0.3 percent of newborns.[11] This dermatosis is characterized by the presence at birth of a yellowish hairless plaque on the scalp or face.

Widespread lesions may appear as grouped papules with a linear or swirled pattern. The lesions typically flatten during childhood and become elevated and/or papular with puberty. Histopathologic examination shows sebaceous glands and incompletely differentiated hair structures.

Since nevus sebaceus undergoes malignant transformation to cutaneous carcinoma in 10 to 15 percent of cases, surgical removal is recommended, usually at puberty. If the lesions are widespread or if surgical removal would be difficult, close observation is recommended. In the patient with epidermal nevus syndrome, various systemic abnormalities may also be present.[12]

Aplasia cutis congenita

Approximately 0.03 percent of newborns are afflicted with aplasia cutis congenita, or a congenital absence of skin.[4] The lesion is present at birth and may be ulcerated, bullous or atrophic in appearance. It is solitary in 70 percent of cases. The most common location is the scalp, near the vertex, although these lesions may occur anywhere on the body.

Histopathologic examination reveals an absence of epidermis and dermal appendages. In the newborn period, it is important to avoid trauma and secondary infection.

Smaller lesions typically heal as an atrophic scar and are easily covered by scalp hair. Larger lesions may require surgical intervention.

The lesions of aplasia cutis congenita clinically are somewhat heterogenous. A specific subtype associated with an embryologic defect has recently been described.[13] This finding may explain the varying modes of inheritance reported previously.

Nevus simplex

" Nevus simplex" is the term applied to a flat, pink, vascular lesion found in the glabellar area, on the upper eyelids or at the nape of the neck in 40 percent of newborns. The lesion is composed of dilated dermal capillaries and represents persistent foetal vessels. The erythema associated with nevus simplex may become more prominent when an infant cries.

Hemangioma

Hemangioma occurs in 1.1 to 2.6 percent of all infants and appears to be more common in premature infants. The lesion often is not present at birth. In retrospect, however, a blanched area may have been present.

The hemangioma is an erythematous lobulated lesion that usually develops within the first four weeks after birth. The lesion generally has a proliferative phase that lasts until the child is between nine and 12 months of age. At this point, the lesion begins to involute. In some cases, the hemangioma ulcerates due to its rapid growth and the consequent thinning of the epidermis.

Spontaneous resolution of hemangiomas occurs by the age of five years in 50 percent of cases, by the age of seven years in 70 percent of cases and by age of nine years in 90 percent of cases.[14] If complications are not present, careful observation of the lesion is sufficient.

If the hemangioma compromises a vital structure, is associated with platelet trapping (i.e. Kasabach-Merritt syndrome) or is persistently ulcerated, intervention is necessary. Lesions that cause closure of the eye and prevent light from entering can result in loss of vision in a short period of time. In such cases, urgent evaluation by an ophthalmologist is recommended.

Corticosteroids have been used both orally and intralesionally in the treatment of hemangiomas. Prednisolone, 1 to 2mg per kg. is administered once daily until the lesion stops growing or diminishes in size, or until the ulceration begins to heal. The prednisolone is tapered as rapidly as possible to avoid side effects. When Kasabach-Merritt syndrome is diagnosed, treatment is directed toward restoring the platelet count to normal.

Biosynthetic dressing can be placed over an ulceration. These dressings, which are changed every 24 hours can be used alone or in combination with systemic corticosteroids. Interferon alfa has been used to treat hemangiomas that do not respond to corticosteroids.[15] Pulsed-dye laser therapy may be helpful in eliminating some ulcerations, but it has a limited effect on the deeper components of hemangiomas.[16]

Compression garments or Coban tape can be used, especially with hemangiomas located on arms or legs. In some cases, these measures can promote the early involution of lesions.

Surgical excision or embolization (e.g.,for Kasabach-Merritt syndrome) can be considered if the hemangioma is localized and other treatments have been unsuccessful.

Lumbosacral hemangiomas may be associated with a tethered cord.[17] Disseminated neonatal hemangiomatosis has been associated with visceral hemangiomas and multiple cutaneous hemangiomas.[18]

Nevus flammeus

Nevus flammeus, often referred to as the port-wine stain, is a vascular malformation composed of mature

ectatic capillaries. This lesion occurs in approximately 0.3 percent of newborns.[18]

The flat, reddish blue nevus flammeus is present at birth. During childhood, the lesion lightens only minimally due to skin thickening and changes in pigment. In or following adolescence, the lesion may begin to darken and develop varicosities, nodules or pyogenic granulomas.

Nevus flammeus is associated with Klippel-Trenaunay-Weber syndrome, Sturge-Weber syndrome and ipsilateral glaucoma. Klippel-Trenaunay-Weber syndrome is characterized by the overgrowth of soft tissue, bone and all involved structures in an area of the body (especially an arm or leg) in which a nevus flammeus is present. In particular, discrepancies in leg length can occur, resulting in gait abnormalities and/or scoliosis.

Sturge-Weber syndrome is estimated to occur in 5 to 8 percent of children with nevus flammeus involving the upper eyelid or temple areas. If the lesion is completely below the palpebral fissure, there is no risk for Sturge-Weber syndrome.

Glaucoma is one of the classic features of the Sturge-Weber triad (i.e., nevus flammeus, seizures and glaucoma). When a nevus flammeus is located around the eye, glaucoma can occur with or without a diagnosis of Sturge-Weber syndrome. Therefore, infants with such lesions should be evaluated by an ophthalmologist.

Pulsed-dye laser therapy can be used to lighten a nevus flammeus lesion, to reduce the risk of Klippel-Trenaunay-Weber syndrome and to minimize the deformities that can occur with progression of the lesion.[19] Compression garments can help to minimize overgrowth, especially when the lesion is confined to an extremity.

References

1. Hurwitz S. Clinical Pediatric Dermatology : A Textbook of Skin Disorders of Childhood and Adolescence. 2nd ed. Philadelphia: Saunders, 1993;7:15.

2. Laude TA. Approach to dermatologic disorders in black children. Semin Dermatol 1995; 14:15-20.

3. Simon MW. Transient neonatal pustular melanosis : an uncommon rash (Letter). Am Fam physician 1995;51:1401.

4. Weston WL, Lane AT. Color Textbook of Pediatric Dermatology. St. Louis : Mosby: 1991;233-235.

5. Murphy WF, Langely AL. Common bullous lesions presumably self-inflicted occurring in utero in the newborn infant. Pediatrics 1963; 32:1099-1101.

6. Jacobs AH. Walton RG. The incidence of birth marks in the neonate. Pediatrics 1976;58:218-222.

7. Silverman AK, Michels EH, Rasmussen JE. Subcutaneous fat necrosis in infant, occurring after hypothermic cardiac surgery. Case report and analysis of etiologic factors. J Am Acad Dermatol 1986; 15:331-336.

8. Sharata H. Postellon DC. Hashimato K. Subcutaneous fat necrosis, hypercalcemia and prostaglandin E. Pediatr Dermatol 1995; 12:43-47.

9. Duke EM. Infantile acne associated with transient increases in plasma concentrations of luteinising hormone, follicle-stimulating hormone, and testosterone. Br Med J (Clin Res Ed) 1981;282:1275-1276.

10. Grau-Massanes M. Raimer S, Colome-Grimmer M, et al. Congenital smooth muscle hamartoma presenting as a linear atrophic plaque: case report and review of the literature. Pediatr Dermatol 1996;13:222-225.

11. Karvonen SL. Vaajalahti P. Marenk M. Birthmarks in 4346 Finnish newborns. Acta Derm Venereol 1992; 72:55-57.

12. Rogers M. Epidermal nevus syndrome; a review of 233 cases. Pediatr Dermatol 1992; 9:342-344.

13. Drolet D, Prendiville J. Golden J, et al. 'Membraneous aplasia cutis' with hair collars. Congenital absence of skin or neuroectodermal defect? Arch Dermatol 1995; 131:1427-1431.

14. Esterly NB. Cutaneous hemangiomas, vascular stains and malformations, and associated syndromes. Curr Probl Dermatol 1995; 7:65-108.

15. Ezekowitz RA, Mulliken JB, Folkman J. Interferon alfa 2a therapy for life-threatening hemangiomas of infancy. N Engl J Med 1992; 326:1456-1463.

16. Geronemus RG. Pulsed dye laser treatment of vascular lesions in children. J Dermatol Surg Oncol 1993; 19:303-310.

17. Goldberg NS, Hebert AA, Esterly NB. Sacral hemangiomas and multiple congenital abnormalities. Arch Dermatol 1986; 122:684-687.

18. Mulliken JB, Young AE. Vascular birthmarks: hemangiomas and malformations. Philadelphia; Saunders. 1988; 170:181.

19. Van Gemert MJ. Welch AJ, Pickering JW, Wavelengths for laser treatment of port wine stains and telangiectasia. Lasers Surg Med 1995;16:147-155.