Benign Neonatal Hemagiomatosis with Unusually Persistent Lesions and Conjunctival Hemangioma

Bang Soon Kim, M.D, Hyang Joon Park, M.D., Jin Nam Park, M.D.

Department of Dermatology, Sangkye Paik Foundation Hospital, Inje University College of Medicine, Seoul, Korea

Most neonatal cutaneous hemangiomatosis are complicated by visceral involvement and are associated with a high mortality rate in the first month of life. Some neonates with multiple cutaneous hemangiomas, however, may follow a benign course of spontaneous resolution without visceral involvement. Such cases are called benign neonatal hemangiomatosis (BNH). BNH is characterized by a lack of mucosal or symptomatic visceral involvement, rapid spontaneous regression of cutaneous hemangimas, and an excellent prognosis.

We report herein a case of BNH which is atypical compared to previously reported BNH cases. Our patient had a more prolonged course than those of usual cases with BNH and showed a conjunctival involvement.(Ann Dermatol 6:(2) 244-248, 1994)

Key Words: Benign neonatal hemangiomatosis

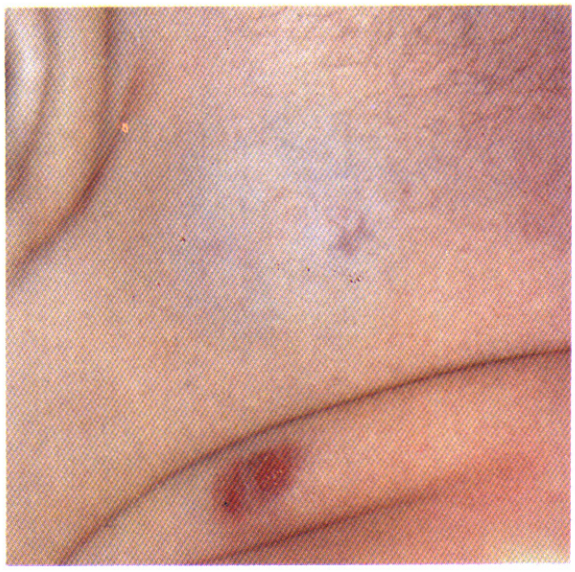
Cutaneous hemangiomas are present at birth or appear shortly thereafter in approximately 10% of all neonates[1]. While most often the lesions are solitary and regress spontaneously[2], patients occasionally may have multiple involvement of skin and viscera[3,4]. Most of the reported cases of neonatal cutaneous hemangiomatosis are complicated by visceral involvement and are associated with a high mortality rate in the first month of life. The presence of multiple cutaneous lesions with life-threatening multisystem involvement has been termed diffuse or disseminated neonatal hemangiomatosis(DNH)[4]. Some neonates with multiple cutaneous hemangiomas, however, may follow a benign course of spontaneous resolution without visceral involvement. Such cases are called benign neonatal eruptive hemangiomatosis or benign neonatal hemangiomatosis(BNH)[5].

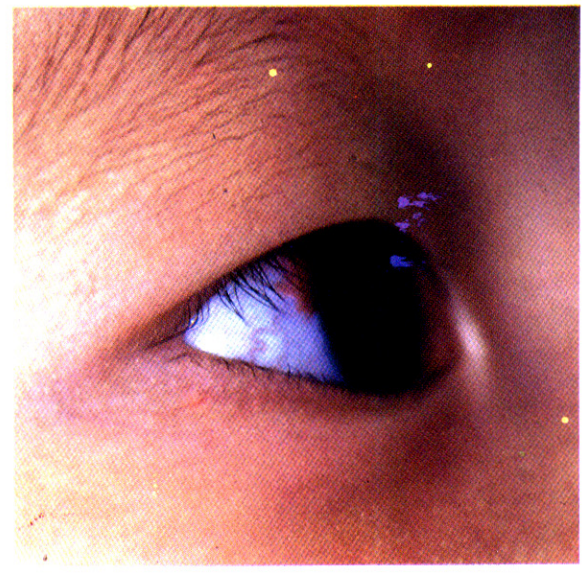
Case reports of BNH are rare[5-10] and only one case has been reported in Korean dermatologic literature[7]. We report a patient who somewhat differs from those previously described. This child showed a more prolonged course than that of cases with typical BNH and had a conjunctival hemangioma.

Received November 29, 1993.
Accepted for publication March 28, 1994.
Reprint request to: Bang Soon Kim, M.D., Department of Dermatology, Sangkye Paik Foundation Hospital, Inje University College of Medicine, Seoul, Korea

REPORT OF A CASE

A female infant was born at full term, weighing 3.65kg. At birth red spots were noted on her left thumb and right conjunctiva. The cutaneous lesions continued to grow in size and new lesions arose. At the age of 3 months, fourteen hemangiomas were scattered over the trunk, extremities, face, nape, and right conjunctiva. The cutaneous lesions were red to purple papulonodules ranging in size from 2mm to 2cm(Fig. 1A). Physical examination did not reveal an abnormal finding except for the cutaneous lesions. Ophthalmologic examination revealed a right conjunctival hemangioma (Fig. 1B) without any other involvement. The results of the following laboratory tests were normal or negative; complete blood count, platelet count, urinalysis, and liver function test. A roentgenogram of the chest showed findings of bronchiolitis in both lungs. Ultrasonographic ex-





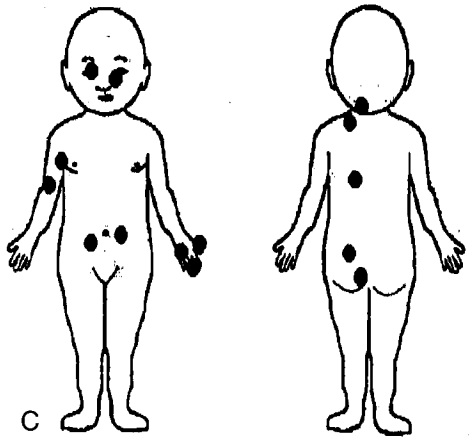


Fig. 1. Hemangiomas on the nape(A) and right conjunctiva(B) and the schematic figures of cutaneous lesions(C).

amination did not reveal any visceral abnormalities. Other studies such as occult blood test, UGI series, liver-spleen scan or brain CT were not performed. A skin biopsy taken from a lesion on the back showed an increased number of aggregated vascular spaces lined by plump endothelial cells and containing erythrocytes within the reticular dermis; a histopathologic pattern compatible with capillary hemangioma(Fig. 2).

By the age of 9 months, the cutaneous hemangiomas had gradually increased in size. At the age of 10 months, one hemanioma on the periumbilical area had resolved but the others persisted without change of size and new hemangioma arose on the left axilla. The child, now 16 month old, has 14 hemangiomas and the size of them has not changed since 10 month old. Anticipating spontaneous regression of lesions, we are closely observing the status of the lesions.

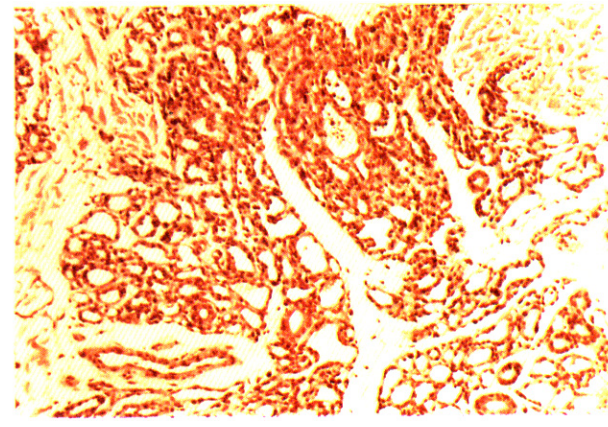


Fig. 2. An increased number of aggregated vascular spaces within the dermis(H&E, × 40).

DISCUSSION

Patients with multiple, diffuse hemangiomas most likely represent a spectrum of disease ranging from only cutaneous involvement(i.e., BNH) to cutaneous and visceral involvement (i.e., DNH)[6]. The distinction is artificial since one cannot exclude the presence of systemic lesions without intensive investigation, which most clinicians would regard as unnecessary. Clinically the cuta-

Table 1. Clinical findings in patients with BNH

Sex	Age at onset	No. of lesions	Size of lesions	Other findings	Treatment	Course	Reference
F	At brith	Approximately 30 at 3wks	1-5 mm	congestive heart failure, hepatomegaly	None	Size and Number increased until 3 wks; spontaneous regression with complete resolution at 10 mo	5
F	After birth	Unknown	0.3-1.2 mm	None	Oral prednisone	Lesions began to involute after 2 mo of oral steroids	6
F	At birth	Numerous	Pinhead -little finger tip size	Hepatomegaly, hemangioma in buccal mucosa	None	Size & number increased until 2 mo spontaneous complete regression at 10 mo	7
F	10 days premature	Multiple	2-3 mm	Small bowel atresia	None	Lesions continued to appear and grow at 18 mo	8
M	At birth	Numerous	2-7 mm	Significant hemorrhage from hemangioma	Electorcautery oral prednisone surgical excision transfusion	Prolonged and rampant proliferation with significant hemorrhage	9
F	At birth	Unkown	Unkown	PDA, ASD with pulmonary hypertension	None	Number increased over the 1st few wks complete resolution at 12 mo	10
F	6 months	Unkown	Unkown	Meningocele Arnold-Chiari malformation	None	No follow up	10
M	20 days	Unkown	Unkown	Hyaline membrane disease, hypothyroidism	None	At 16 mo, hemangiomas were involuting; at 28 mo only one hemangioma persisted	10
F	At birth	14	2-20 mm	Hemangioma in conjunctiva	None	At 16 mo, the size of hemangiomas has not changed since 10 mo	our case

neous features cannot be distinguished from those seen in cases where the hemangiomas have been restricted to the skin. The clinical characteristics of BNH are multiple, small, bright red to purple capillary hemangiomas that range in size from that of a 2 mm to 2 cm in diameter [11]. The hemangiomas are usually present at birth or appear during the first week of life. They rapidly increase in size and number until approximately 3 weeks of age, at which time they begin to spontaneously regress.

Initially, BNH was defined by Stern et al[5] as multiple cutaneous hemangiomas with a lack of symptomatic visceral involvement, rapid spontaneous regression, and an excellent prognosis. In addition to their original case, Stern et at[5] identified 6 cases in the literature as BNH. According to them, all 6 cases had multiple cutaneous hemangiomas with spontaneous regression. Although

case reports of BNH are rare[5-10], this disorder is probably more common than is suggested by the literature. To our knowledge, 8 cases have been reported under the name of BNH since the first report by Stern et al. Clinical characteristics of the 8 cases identified as BNH are outlined in Table 1[5-10]. One case was not followed up[10]. In 4 cases the hemangiomas were almost completely resolved within 12 months[5-7,10]. Such a course is similar to that of typical BNH. The other 3 cases, however, showed somewhat different courses[8-10]. The course of one case was remarkable for the prolonged and rampant proliferation of cutaneous hemangiomas with significant hemorrhage. It was unresponsive to systemic corticosteroids and necessitated surgical intervention[9]. The lesions in another case were present at 16 months of age[10] The other case had continued todevelop new cutaneous hemangiomas through 18 months[8]. Virtually nearly 100% of the usual solitary strawberry hemangiomas undergo spontaneous regression, which is complete or incomplete in about 95%[2,12,13]. About 30% of strawberry hemangiomas will have resolved by the fourth birthday, about 50% by the fifth, and 75% by the seventh. It is said that the likelihood of early resolution is not affected by the size of lesion, its site, or by the number of lesions present[13]. However, the small, multiple hemangiomas involved spontaeously, generally more repidly than would be anticipated in other usual hemangiomas; in such cases the skin lesions may disappear by the age of 12 months[12]. In our patient, cutaneous hemangiomas had gradually increased in size by the age of 9 months. Even though one hemangioma resolved after then, new one arose and the others have persisted without involution until the age of 16 months, and such a course is atypical compared with previously reported patients with BNH. It is possible that each of the lesions in our patient will follow a course similar to that expected of most solitary strawberry hemangioma and therefore, she will have a more prolonged course than those of cases with typical BNH.

Six of the 8 cases identified as BNH in the literature manifested signs of congestive heart failure, hepatomegaly, small bowel atresia, patent ductus arteriosus, atrial septal defect, pulmonary hypertension, meningocele, Arnold-Chiari malformation, hyaline menbrane disease or hypothyroidism(Table 1). It is possible that congestive heart failure and hepatomegaly was caused by hepatic hemangioma, but in that case[5], she recovered rapidly from an episode of congestive heart failure without persistent arteriovenous shunts. Because of the rapid resolution of hepatomegaly after treatment for heart failure, radioactive scanning and/or arteriography was not performed. It seems that the other conditions had no direct relation to hemanginma. Because the incidenu of other associated conditions had no direct realtion to hemangioma.

Because the incidence of other associated conditions was high, however, we think that clinicians have to pay attention not only to the possibility of visceral involvement of cutaneous hemangomas but also that of occurrence of other associating conditions which are not directly related to hemangioma.

Unlike DNH, it is said that BNH do not involve mucous membranes[11]. However, Ha et al[7] reported a BNH case which had buccal mucosal hemangiomas, and the conjunctival hemangioma occurred early in life in our case. Eyes are involved in about one third of patients with DNH and the most common ocular manifestation is iris hemangiomata, followed by involvement of the conjunctiva, chorioretinal vasculature, and eyelids [14]. Considering diffuse hemangiomas as a spectrum of disease ranging from BNH to DNH, it is not surprising for BNH to involve oral or ocular mucous membranes.

In patients with DNH, the organs most affected were the skin, liver, central nervous system(CNS), intestine, and lungs[14]. It is very important to differentiate BNH from DNH. They both may have similar skin lesions, but patients with DNH may die of secondary conditions such as gastrointestinal tract hemorrhage, congestive heart failure associated with an arteriovenous shunt, consumption coagulopathy (Kassabach-Merritt syndrome), hydrocephalus or seizures[3,4]. On the contrary, those with BNH have a good prognosis, and unless they show signs of cardiac failure, interference with vision, or bleeding, they usually do not require specific treatment. Although differences in opinion exist regarding appropriate workup, complete evaluation of a patient with onsef of multiple hemangiomas shortly after birth should attempt to determine the extent of such involvement. Held et al[8] recommended full physical examination including

ophthalmologic examination, and laboratory investigations including chest radiograph, computerized tomography of the brain, gastrointestinal series or equivalent radiologic examination, liver-spleen scan and abdominal ultrasound, liver function tests and urinalysis, and careful review of hematologic and hemodynamic indices.

REFERENCES

1. Jacobs AH, Walton RG: The incidence of birthmarks in the neonate. Pediatrics 58: 218-222, 1976.
2. Bowers RE, Graham EA, Tomlinson KM: The natural history of the strawberry nevus. Arch Dermatol 82: 667-680, 1960.
3. Burman D, Mansell PWA, Warin RP: Miliary haemangiomata in the newborn. Arch Dis Child 42: 193-197, 1967.
4. Holden KR, Alexander F: Diffuse neonatal hemangiomatosis. Pediatrics 46: 411-421, 1970.
5. Stern JK, Wolf JE, Jarratt M: Benign neonatal hemangiomatosis. J Am Acad Dermatol 4: 442-445, 1981.
6. Ronan SG, Solomon LM: Benign neonatal eruptive hemangiomatosis in identical twins. Ped Dermatol 1: 318-321, 1984.
7. Ha BS, Won YH, Chun YK, Kim YP: A case of benign neonatal eruptive hemangimatosis. Kor J Dermatol 26: 960-966, 1988.
8. Held JL, Haber RS, Silvers DN, Grossman ME: Benign neonatal hemangiomatosis: Review and description of a patient with unusually persistent lesions. Ped Dermatol 7:63-66, 1990.
9. Rothe MJ, Rowes D, Grant-Kels JM: Benign neonatal hemangiomatosis with aggressive growth of cutaneous lesions. Ped Dermatol 8:140-146, 1991
10. Smith DD, Cowen P: Benign neonatal hemangiomatosis. Int J Dermatol 31: 336-338, 1992.
11. Messaritakis J, Anagnostakis D, Feingold M: Benign neonatal hemangiomatosis. Am J Dis Child 140:447-448, 1986.
12. Atherton DJ: Angiomatous naevi. In Champion RH, Burton JL, Ebling FJC(eds): Textbook of dermatology. Blackwell Scientific Pub, Oxford, 1992, pp469-482.
13. Nakayama H: Clinical and histological studies of the calssification and the natural course of the strawberry mark. J Dermatol 8:277-291, 1981.
14. Goltz LE, Rudikof J, O'Meara P: Diffuse neonatal hemangiomatosis. Ped Dermatol 3: 145-152, 1986.