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Diffuse Neonatal Hemangiomatosis: A Case Report

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Diffuse neonatal hemangiomatosis (DNH) is a rare condition characterized by the presence of numerous cutaneous and visceral hemangiomas that become manifest at birth or within the neonatal period. The cutaneous lesions are generalized, vary from 0.5 to 1.5 cm in diameter, and often range from 50 to 500 in number. Visceral lesions are most commonly found in the liver, CNS, intestine, and lungs. Skeletal involvement also has been reported. Approximately 60% of infants with DNH die during the first few months of life due to high-output cardiac failure, hemorrhage, or CNS involvement [1]. Steroid therapy is thought to accelerate involution of these lesions and has raised prospects for long-term survival [2].

This report describes the usefulness of MR imaging in demonstrating the numerous intracranial hemorrhages seen in DNH. MR imaging was much more sensitive than CT in demonstrating these lesions and also was more specific in showing the characteristic sedimentation levels seen with subacute hemorrhages. Other imaging examinations confirmed the presence of additional hemangiomas in the long bones and liver. Despite a stormy course over the first 2 weeks of life, this infant responded successfully to steroid therapy and is doing well at 16 months of age.

Case Report

A 7 lb 9 oz full-term infant was noted to have a cephalohematoma and multiple cutaneous hemangiomas (greater than 30) at birth. The infant's medical history was unremarkable except for a 9-year-old sibling who had a strawberry hemangioma of the cheek treated with cryotherapy early in childhood. At 5 days of age the infant had bright red blood per rectum. The child was hospitalized and a barium enema, technetium-99 red blood cell scan, and endoscopy were performed and failed to show the source of bleeding. The patient's hematocrit on admission was 22.2 and the platelet count was 137,000. CT scan of the head without IV contrast administration demonstrated seven well-defined, smooth, round and ovoid hemorrhagic lesions measuring 0.5 to 2.0 cm in diameter throughout the brain (Fig. 1). Nearly all of these hemorrhages had a heterogeneous density and associated surrounding edema. Only one lesion demonstrated a well-defined sedimentation level. An abdominal CT scan without IV contrast administration demonstrated multiple well-circumscribed, round, low-attenuation lesions within the liver. The smaller lesions became isodense to normal liver parenchyma after IV contrast administration. No lesions were identified in the kidneys or spleen. A skeletal survey demonstrated multiple lytic lesions in the metaphyseal region of the long bones. Plain films of the skull were negative. A chest radiograph demonstrated slight prominence of the vascular markings.

The infant was treated with transfusions of packed red blood cells on several occasions. On the sixth day of life the infant had decreased neurologic function and was treated with steroids to reduce cerebral edema. The following day the patient became hemodynamically unstable and was intubated for approximately 48 hr. During this time the patient's platelet count fell to 17,000, requiring platelet transfusion. After a stormy 48-hr course, the infant stabilized and was ultimately discharged at 9 days of age. The infant was placed on oral prednisone (1 mg/kg/day) at the time of discharge. An MR brain scan performed at 5 weeks of age demonstrated multiple hemorrhagic lesions throughout the brain, most of which had a cystic appearance with sedimentation levels (Fig. 2). The number of hemorrhagic lesions demonstrated on the MR scan was considerably greater than had been identified on the earlier CT scan. These lesions were seen throughout both cerebral hemispheres, predominately in the subcortical white matter as well as in the brainstem and cerebellum. There was a mild degree of hydrocephalus secondary to compression of the aqueduct by one of these lesions in the tectum. On T2-weighted images those lesions with a sedimentation level demonstrated a hypointense dependent layer and a hyperintense supernatant, probably representing intracellular deoxyhemoglobin and free methemoglobin, respectively. Those lesions that did not demonstrate sedimentation levels contained either central or peripheral hypointense areas, consistent with hemosiderin. Many of the hemorrhagic lesions had surrounding edema.

The patient was seen at 1½ months for an outpatient follow-up visit. The stool was negative for occult blood at that time. Because of the hydrocephalus evident on the MR scan, enlarging head size, and slight fullness of the fontanelle, a ventriculoperitoneal shunt was recommended; however, the patient's parents refused any further medical treatment. The infant has been followed by an outside pediatrician and, as of age 16 months, has normal growth and development for age with no residual cutaneous hemangiomas evident, and no physical findings to suggest the presence of hydrocephalus.

Discussion

The MR appearance of DNH involving the CNS has not yet been described. The hemangiomas, which are of the capillary type, show the same tendency for rapid growth and subse-

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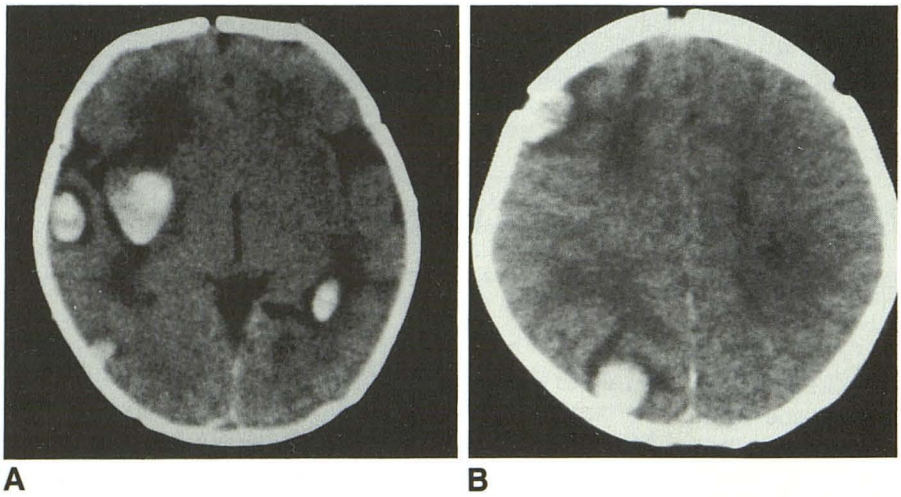

Fig. 1.—*A* and *B*, CT brain scans without IV contrast medium at age 1 week show multiple intracranial hemorrhages with surrounding edema.

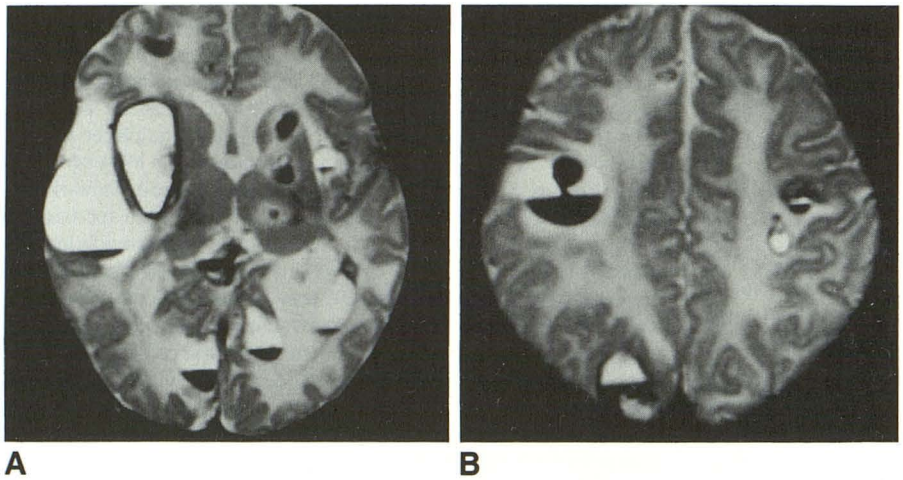

Fig. 2.—*A* and *B*, MR images (3000/140/1) at age 5 weeks show a significantly greater number of intracranial hemorrhages than seen on CT scans 1 month earlier. The patient's clinical condition had improved markedly at this time. Multiple fluid-fluid levels of hemoglobin breakdown products are seen in most of these lesions.

quent involution as do solitary juvenile hemangiomas [1]. Involution of juvenile cutaneous hemangiomas occurs between ages 6 and 12 months in 65% of cases. Involvement of the brain often has resulted in hydrocephalus due to compression of the aqueduct or fourth ventricle by these lesions in the midbrain, pons, or cerebellum, similar to our case [3–5]. Only two case reports have described the use of CT to evaluate the CNS lesions with this disease [1, 5]. In one case these lesions appeared diffusely throughout the brain as numerous hyperdense foci that enhanced with administration of IV contrast material. Neither of the previously reported cases with CT head scans demonstrated sedimentation levels in the hemorrhagic lesions. In our case, only one of the hemorrhages demonstrated a typical sedimentation level, whereas the MR examination 4 weeks later showed sedimentation levels in most of the hemorrhages. The CT images underestimated the number of hemangiomas present in the brain on the subsequent MR examination at age 5 weeks. Although IV contrast material was not administered for the MR or CT scans in this case, it most likely would have caused enhancement of additional hemangiomas, as has been reported with CT [1].

MR is able to demonstrate the presence of subacute blood products [6, 7]. In those hematomas with a sedimentation level, the supernatant represents free methemoglobin and the dependent layer represents intracellular deoxyhemoglobin, both of which have specific appearances on T1- and T2-weighted MR imaging sequences [7]. Abrahams et al. [8] have found MR to be superior to CT in detecting intracranial fluid levels. Sedimentation levels have been described in CT imaging of liver hemangiomas [9]. It is unusual to see sedimentation levels and persistent edema in MR images of intracerebral hematomas at age 5 weeks, as was seen in our case. This presumably indicates recurrent bleeding from these hemangiomas, which accounts for the presence of layers of intracellular deoxyhemoglobin and extracellular methemoglobin. Delayed temporal evolution of hemoglobin breakdown products and persistent edema have been described in hemorrhagic malignant intracranial neoplasms [10]. Atlas et al. [10] reported a case of a hemorrhagic metastasis with a persistent sedimentation level such as ours 16 days after the hemorrhage. They also noted an absent, diminished, or irregular rim of presumed hemosiderin with these hemorrhages, as well as the presence of focal abnormal tissue not corre-

sponding to hemorrhage (i.e., presumed tumor). We did not note any abnormal tissue other than hemorrhagic products in this case of DNH. The literature does not describe any characteristic CT or MR appearance of uncomplicated capillary hemangiomas except for the contrast enhancement pattern seen with hepatic lesions, as in this case (i.e., lesions became isodense with normal liver). Possibly, this enhancement feature will be useful in differentiating these lesions in the future with use of contrast-enhanced MR imaging.

Some of the lesions in this case resemble occult vascular malformations, as described by Gomori et al. [11], with areas of hematoma in different stages of evolution surrounded by a hemosiderin ring. According to Atlas et al., 16–33% of these cavernous angiomas may be multiple, and they are most often located in the subcortical white matter. They may be associated with edema but usually are not discovered in infancy. Other differential diagnostic considerations include hemorrhages secondary to other types of bleeding diathesis, hemorrhagic metastatic disease, and Osler-Weber-Rendu disease. The latter may be present at birth, but is hereditary, whereas DNH is not a hereditary disease.

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