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Diffuse neonatal hemangiomatosis with extensive involvement of the brain and cervical spinal cord

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Abstract *Background.* Diffuse neonatal hemangiomatosis (DNH) is a rare disorder first recognized at birth or during the neonatal period. DNH is characterized by numerous cutaneous and visceral hemangiomas involving three or more organ systems.
Materials and methods. Although the skin and liver are most frequently affected, we present a case of DNH demonstrating an unusual predilection for the central nervous system (CNS).
Results and conclusion. We report the imaging findings in a patient with this disorder, paying particular attention to the features seen on cranial sonography and spinal MR imaging.

Introduction

Diffuse neonatal hemangiomatosis (DNH) is a rare and often fatal condition that manifests at birth or during the neonatal period and is characterized by numerous cutaneous and visceral hemangiomas. This case report describes the findings of DNH in a male neonate with multiple cutaneous hemangiomas, subsequently found to have extensive involvement of the brain and cervical spinal cord. To a lesser extent, the liver and heart were also involved.

Case report

A male infant weighing 3830 g was born by cesarean section to a 36-year-old, gravida 2, para 1 mother. The infant's Apgar scores were 8 and 9. Immediately after delivery, signs of respiratory distress were noted, and the patient was transferred to the neonatal intensive care unit for ventilation therapy, as well as for further evaluation of cutaneous hemangiomas that were disseminated over the entire body. Initial chest radiographs suggested wet-lung disease, and the infant was weaned to room air within 36 h after birth.

Imaging studies were obtained for evaluation of possible visceral hemangiomas. Cranial sonography performed on day 3 of life showed numerous hyperechoic lesions of varying size throughout the cerebral hemispheres (Fig. 1). Color-flow sonography did not reveal any appreciable flow within these lesions. Cranial computed tomography (CT), also performed on day 3, showed multiple well-circumscribed, round, hyperdense hemorrhagic lesions throughout the brain parenchyma. Some of these lesions showed enhancement and many were surrounded by extensive edema (Fig. 2). Hydrocephalus was not seen at this time. Abdominal CT performed on the same day revealed a number of small enhancing lesions, consistent with hemangiomas, within the subcutaneous tissues and muscles of the abdominal wall. Abdominal CT also revealed a 2.0-cm enhancing lesion within the left lobe of the liver, seen with sonography as a heterogeneous, predominately hyperechoic lesion. MR imaging of the brain, performed on day 9 of life, showed numerous intra-axial lesions, with involvement of the cerebral hemispheres, cerebellum, and brainstem. These lesions had markedly increased signal intensity on T1-weighted images, mixed signal intensity on T2-weighted images, and variable enhancement with gadolinium. Many of the lesions were surrounded by extensive edema, and blood-fluid or sedimentation levels were present in several lesions,

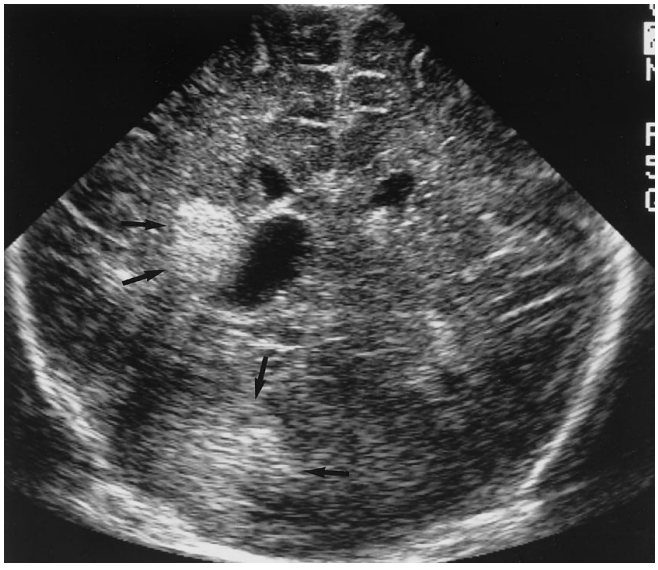

Fig. 1 Cranial sonogram in the coronal plane reveals large echogenic lesions, consistent with hemangiomas, within the right parietal and occipital lobes *(arrows)*. Similar lesions were seen throughout the cerebral hemispheres bilaterally

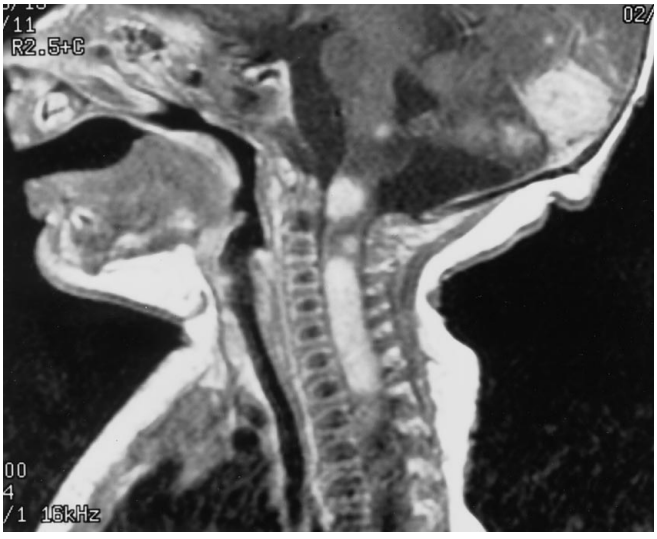

Fig. 3 Gadolinium-enhanced sagittal T1-weighted image (TR/TE, 500/14) demonstrates expansion and enhancement of the cervical spinal cord, as well as enhancing lesions in the right occipital lobe and brainstem

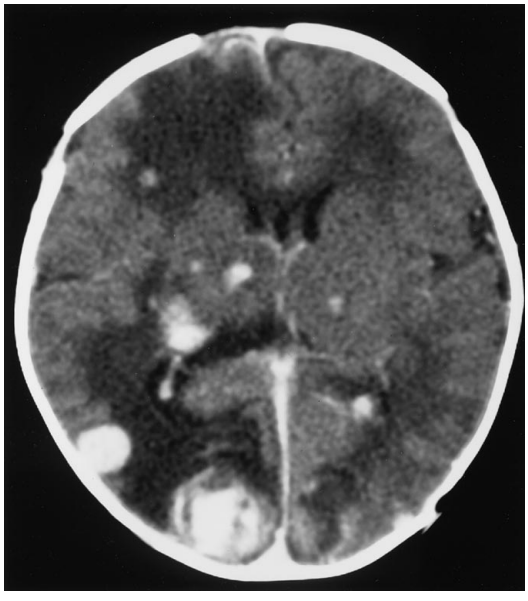

Fig. 2 Enhanced axial CT scan of the brain demonstrates multiple hyperdense hemorrhagic lesions consistent with hemangiomas, as well as extensive edema within the right frontal and right parietooccipital lobes

presumably resulting from layering of hemoglobin breakdown products. This examination also revealed several areas of encephalomalacia, suggesting prior infarctions, as well as interval development of a moderate degree of hydrocephalus. Also noted was extensive involvement of the entire cervical spinal cord, which showed diffuse enlargement, intermediate signal intensity on T1-weighted images, increased signal intensity on T2-weighted images, and marked diffuse enhancement with gadolinium (Fig. 3). An echocardiogram performed on day 9 revealed numerous small nodules, probably hemangiomas, within the outflow tracts of the left and right ventricles. Seventy cutaneous hemangiomas over the entire body were eventually treated with pulsed dye laser, and several lesions within the oral cavity were excised and proven to be capillary hemangiomas.

During the following week, the patient's neurological status worsened. He failed to demonstrate head and neck control, and there was no evidence of spontaneous movement of the upper extremities. Follow-up MR imaging of the brain on day 13 of life showed increasing hydrocephalus, but the numerous intracranial and spinal lesions were otherwise stable. Increasing blood flow within the hepatic hemangioma was also seen at that time with color flow sonography. Direct laryngoscopy revealed no evidence of hypopharyngeal, laryngeal, or tracheal lesions. The infant had been treated with prednisolone, 4 mg/kg per day, throughout his hospital stay, and had no signs of congestive heart failure or coagulopathy. The neurological status on day 17 of life showed continual worsening, and treatment plans at that time centered on possible interferon therapy after rapid weaning of the patient from prednisolone.

Discussion

DNH is a rare, often fatal, disorder first recognized at birth or during the neonatal period. DNH is characterized by the widespread presence of cutaneous and visceral hemangiomas involving three or more organ systems [1]. The hemangiomas are usually of the capillary type and show no evidence of malignancy. The cutaneous hemangiomas are generalized, have a diameter of 0.5–1.5 cm, and range from 50 to 100 in number. Viscer-

al hemangiomas can affect various organ systems, and involvement of the liver (64–100 %) is second only to that of the skin in frequency [2]. Other commonly involved organs are the lungs, the intestines, and the CNS. The mouth, tongue, spleen, kidneys, mesentery, heart, and genitourinary tract are involved less often.

Approximately 60 % of infants with DNH die during the first few months of life as a result of high-output cardiac failure, visceral hemorrhage, CNS involvement, or a combination of these [1]. Other potential complications of this disorder include respiratory insufficiency, ocular abnormalities, and consumptive coagulopathy, resulting in thrombocytopenia and fatal hemorrhage. Because DNH is potentially lethal if untreated, the goal of therapy is to accelerate the involution of the hemangiomas, usually by means of corticosteroid therapy. When the visceral lesions are unresponsive to steroid therapy, other therapeutic options include surgical resection, radiation therapy, laser therapy [3], and the use of angiogenesis inhibitors such as interferon [4]. Finally, hepatic lesions that are unresponsive to medical therapy may be treated with hepatic arterial embolization [3].

The clinical, histological, and imaging features of DNH should be compared to those of the more common solitary juvenile hemangiomas. In this latter group, the hemangiomas are usually discovered within the first several weeks of life, grow rapidly, and then frequently regress over a variable period of time later in childhood. Histologically, these benign vascular lesions typically show endothelial proliferation during the period of rapid growth, followed by a slower phase of involution with fibrofatty replacement [5]. The imaging appearance of a hemangioma therefore depends on the histologic phase of the lesion. During the proliferative phase, the typical hemangioma usually presents as a soft-tissue mass, hypointense or isointense to muscle on T1-weighted spin-echo MR imaging, moderately hyperintense on T2-weighted images, and uniformly enhancing with gadolinium. Flow voids are typically seen associated with these lesions on spin-echo images, and gradient imaging shows high-flow or high-signal vessels. During this proliferative phase, the soft-tissue mass is typically hypodense or isodense with muscle on CT. During the phase of involution, the imaging appearance on CT and MR will reflect the increasing and variable fatty composition of the lesion [6].

DNH, on the other hand, is generally associated with a high mortality during infancy, as previously noted. Histologically, the hemangiomas are composed of dilated thin-walled channels lined by a single layer of flattened endothelial cells [7]. In regard to imaging findings, this report has demonstrated that the hemangiomas in DNH, particularly the intracranial lesions, are hyperdense on CT and hyperintense on T1-weighted MR images, likely due to the presence of hemorrhage. In addition, flow voids were not seen in association with these lesions on spin-echo MR imaging. These findings are consistent with those reported by other investigators [1, 8].

This case is atypical because of the preponderance of brain and spinal cord involvement. The findings on MR imaging of the brain, including abundant hemorrhagic lesions, associated edema, and sedimentation levels within some of the lesions, were similar to those reported by Poirier et al. [8]. However, this may be the first report demonstrating the cranial sonographic findings of DNH and the MR imaging appearance of cervical spinal cord involvement in this disorder. Although CT and MR imaging are superior to ultrasound in demonstrating these intracranial lesions, cranial sonography nonetheless depicted many of these hemangiomas effectively and would likely be valuable in demonstrating the progression of hydrocephalus on follow-up scans. In conclusion, we have described a patient with DNH characterized by numerous hemangiomas involving the cutaneous and subcutaneous tissues, brain and spinal cord, and to a lesser extent, the liver and heart.

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