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Letters

Hemangiomatous Calvarial Trabecular Patterns in Newborns

Many pediatric conditions can present clinically with cranial dysmorphism and cutaneous hemangiomas [1]. Early distinction among these conditions is important for counseling parents and medical planning. Because of variable and often overlapping clinical features, the radiologist is often the first to suggest the precise diagnosis of a baby with cutaneous hemangiomas.

Gyriform calcifications on skull radiographs are highly specific for the Sturge-Weber-Dimitri (SWD) or the combined Sturge-Weber-Dimitri–Klippel-Trenaunay-Weber (SWD–KTW) syndromes [2, 3]. However, these calcifications are rarely present at birth and may not be obvious for months. Our case illustrates a lesser known feature of SWD or combined SWD–KTW syndromes which may be present on the newborn skull film—a sunburst trabecular pattern of the type seen with direct hemangiomatous involvement of bone [4, 5].

A 3,350 g, term male was delivered by cesarean section because of cephalopelvic disproportion. Physical examination after delivery demonstrated macrocephaly, a normal body length and weight, a normal ophthalmologic examination, and extensive port wine hemangiomas involving the head, face, neck, upper chest, and extremities.

Skull radiographs at age 6 days revealed a "spoke wheel" radiating trabecular pattern involving the frontal and parietal regions bilaterally (fig. 1). Unenhanced computed tomography (CT) demonstrated thickening of the calvarium. Because of bilateral paroxysmal sharp waves in the frontal region at electroencephalography, the baby was placed on phenobarbital before discharge from the nursery.

At age 2 months, the baby developed jacksonian seizures. Physical examination continued to reveal macrocephaly, but a weight and length at the 50th percentile. The cutaneous hemangiomas remained unchanged. Skull radiographs continued to reveal a hemangiomatous trabecular pattern.

Follow-up CT examination at age 9 months demonstrated cerebral atrophy and extensive, predominantly left-sided gyriform calcification. Despite the presence of cerebral atrophy, the skull remained macrocephalic. Plain skull films at age 9 months continued to show a hemangiomatous pattern, but failed to demonstrate the gyriform calcifications seen on CT.

Of the newborn conditions presenting with cranial dysmorphism and cutaneous hemangiomas [1], calvarial angiomatosis has been reported only in SWD or combined SWD–KTW syndromes. The radiographic appearance of the skull in these babies has been variable [4, 5]. As in our case, the skull may have obvious thickening, striations, and diploic accentuation. On the other hand, a histologically proven angiomatous involvement of the calvarium may radiographically produce only a subtle increase in bone porosity. Any of these radiographic alterations may be generalized or localized.

Our case demonstrates that a striking hemangiomatous calvarial pattern may present at birth long before the gyriform calcifications are apparent by CT or plain film. This radiographic finding therefore facilitates early distinction of SWD or SWD–KTW syndromes from the other conditions that can be associated with cutaneous hemangiomas and cranial dysmorphism [1]. A hemangiomatous calvarial trabecular pattern also has added significance prognostically because of the growth disturbance potentially produced by direct hemangiomatous involvement of bone.

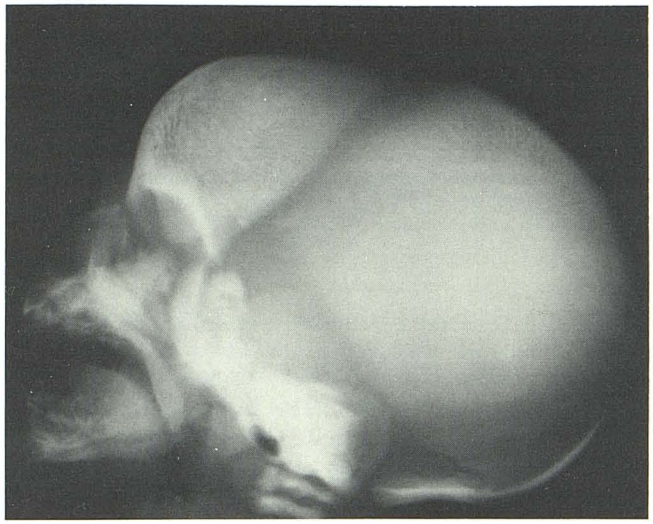

Fig. 1.—Age 6 days. Frontal and parietal bones have "sunburst" trabecular pattern, and there are prominent striations in the mandible. These changes were bilateral on the posteroanterior film. (Increased skull density is normal in a newborn).

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