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Lacunarlike Skull in Neurofibromatosis

Neurofibromatosis is an autosomal dominant disorder resulting from mesodermal and ectodermal dysplasia and neoplasia [1]. Calvarial involvement by lytic lesions is relatively uncommon compared with the rest of the skeleton [2]. When the skull is affected, radiolucent defects usually are seen in the occipital bone and frequently are found bordering the left lambdoid suture [2]. We report a case of neurofibromatosis in which multiple calvarial defects occurred diffusely throughout the skull, giving it a lacunarlike appearance. We think this is the first time such a diffuse pattern of skull defects has been described in neurofibromatosis.

Case Report

A 13-year-old girl was brought to the emergency department because she had collapsed at home. Physical examination showed lethargy, hyperreflexia, and papilledema. The patient had cutaneous stigmata of neurofibromatosis, including multiple café-au-lait spots on the extremities, axillary freckles, and fleshy nodules (fibroma molluscum) on the chest and left ankle. No scalp nodules were present. CT scan showed a large left frontal nonenhancing cystic mass, hydrocephalus, and multiple calvarial defects involving both inner and outer tables (Figs. 1A and 1B). A left frontal burr hole was established, and an Ommaya reservoir was inserted. Skull radiographs showed multiple radiolucent defects and an appearance similar to lückenschädel or lacunar skull (Fig. 1C). (When she was 9 months old, the patient had had a linear occipital skull fracture, but no other bony abnormalities were seen on radiographs obtained then.) MR showed that the large cystic mass contained a smaller round solid component located at the base of the left anterior fossa (Fig. 1D). This was resected and was found to be a pilocytic cystic astrocytoma. Follow-up CT 6 months later showed no recurrence of tumor, decompressed ventricles, and unchanged multiple calvarial defects.

Discussion

In neurofibromatosis, radiolucent skull lesions are thought to be the result of either mesodermal dysplasia or erosions by neurofibromas [2-4]. Although these lesions usually are found along the left lambdoid suture and in the parietal bone, other sites of involvement in the skull have been described [2-6]. In our case, the bony involve-

ment was quite extensive, as it included the occipital, parietal, and frontal bones with defects found within both the inner and the outer tables. Skull radiographs showed multiple radiolucent defects resembling those of a lacunar skull. Normally, lacunar skull disappears by the time a baby is 6 months old and nearly always is seen with developmental abnormalities such as myelocele, encephalocele, or meningocele [7, 8]. In our patient, the skull was normal at 9 months except for the fracture, and she had none of the conditions associated with cranialacunia. Moreover, lacunar skull results from depressions in the inner table, whereas in our case, both tables clearly were involved [8]. Similarly, digital impression associated with hydrocephalus affects the inner table only and cannot account for the defects in the outer table seen in our case. Therefore, this lacunarlike appearance is probably the result of a mesodermal dysplasia of the skull associated with neurofibromatosis.

Lacunarlike skull joins the long list of radiologic findings associated with neurofibromatosis. Recognition of this entity will prevent misdiagnosis of conditions with similar appearances, such as lückenschädel and metastatic disease.

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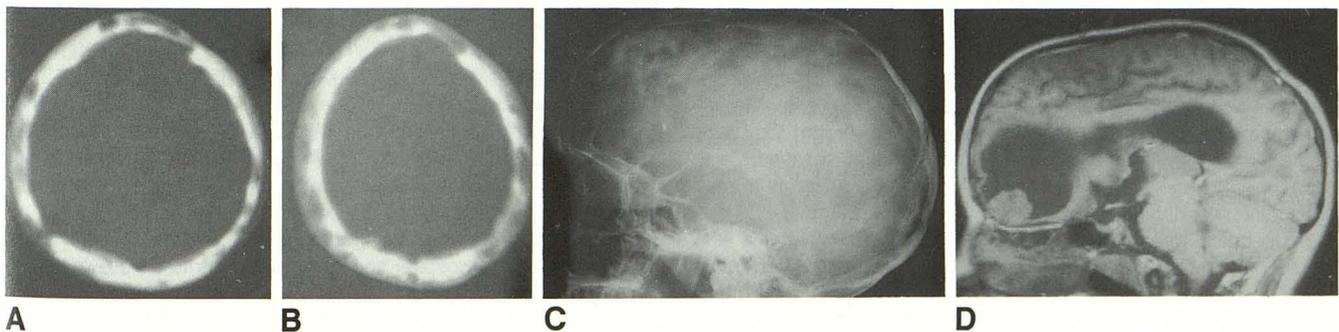


Fig. 1.—Lacunarlike skull in neurofibromatosis.

A and B, Bone-window CT scans show multiple radiolucent defects within calvarium.

C, Lateral radiograph shows lacunarlike appearance of skull.

D, Sagittal T1-weighted MR image shows solid and cystic components of pilocytic cystic astrocytoma in anterior fossa.