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Multifocal intracranial MR abnormalities in neurofibromatosis.

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Multifocal Intracranial MR Abnormalities in Neurofibromatosis

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MR imaging of four children with neurofibromatosis demonstrated areas of increased T2 signal involving widespread multifocal regions of basal ganglia and brainstem. Such abnormalities have not been previously reported. No corresponding CT abnormalities were present. These regions may represent hamartomatous or low-grade gliomatous change, and appear to be characteristic of neurofibromatosis in children.

The association between CNS neoplasms, especially optic gliomas of childhood, and neurofibromatosis is well known, with the reported frequency of CNS tumors as high as 19% [1].

Four children with neurofibromatosis, three with optic glioma and one with an orbital plexiform neurofibroma, were examined with MR imaging. In addition to the expected abnormalities involving the orbit and optic chiasm, areas of abnormally increased intensity on T2-weighted images were seen to involve the basal ganglia, posterior visual pathways, and brainstem of all patients. The ability to recognize and characterize these widespread abnormalities is of potential diagnostic and therapeutic importance.

Subjects and Methods

All MR examinations were performed on a 1.0-T Siemens Magnetom system. T1-weighted (TR = 500, TE = 20) and T2-weighted (TR = 2500, TE = 30 and 60) spin-echo (SE) pulse sequences in the sagittal and axial or coronal planes were obtained in all patients. All CT examinations were performed on a GE 9800 scanner.

Patients were selected for scanning on the basis of clinical indications noted in the Results section. These subjects represent neurofibromatosis patients scanned so far at our institution.

Results

Case 1

A 7-year-old boy was followed since age 6 months for neurofibromatosis originally diagnosed because of multiple cafe-au-lait spots and a neurofibroma involving the right side of the face. Subsequent development included large neurofibromas involving the facial bones on the right side resulting in asymmetric bony growth with complicating ptosis and ectropion. CT demonstrated orbital asymmetry with dysplasia of the right sphenoid bone and probable bilateral optic nerve enlargement. MR confirmed bilateral optic nerve enlargement, thought to be compatible with optic nerve gliomas. T2-weighted MR images additionally demonstrated increased signal involving the optic tracts and radiations bilaterally as well as areas of involvement in the globus pallidus and cerebellar white matter (Figs. 1 and 2).

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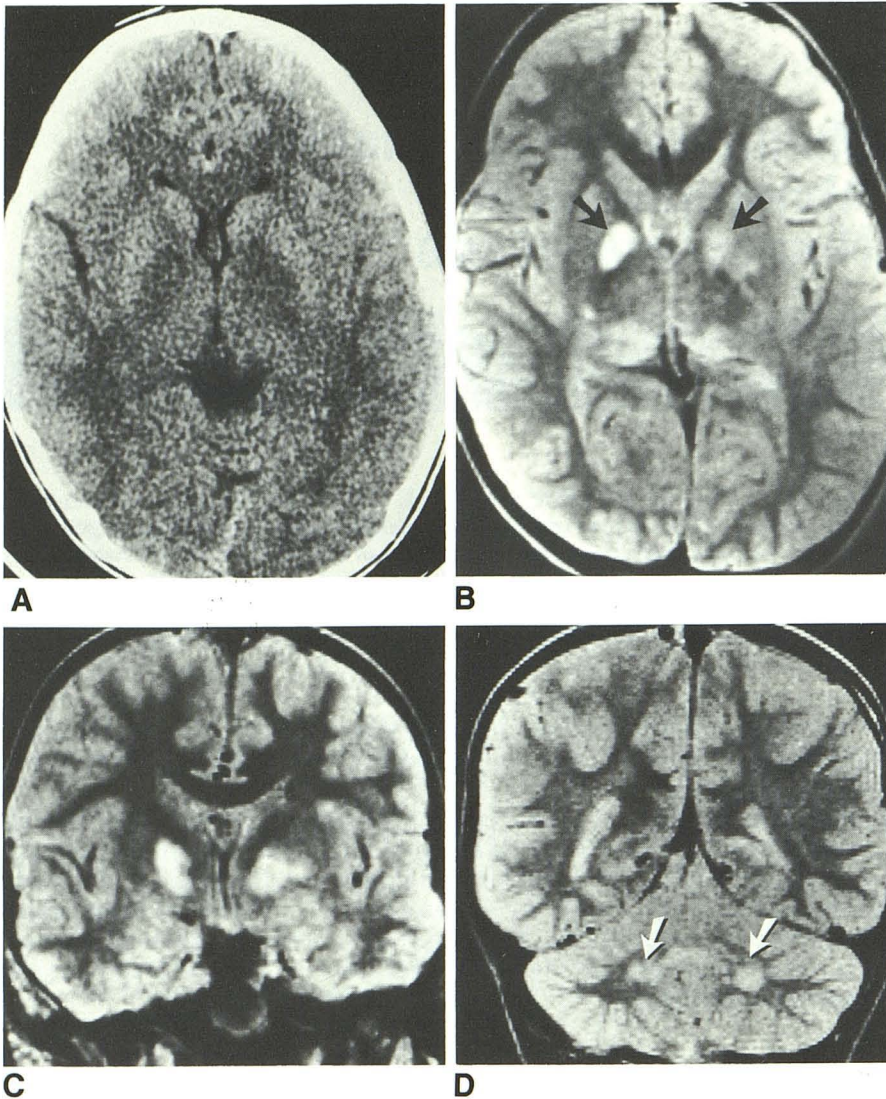


Fig. 1.—Case 1.
A, CT scan shows no abnormality of basal ganglia.
B, T2-weighted MR image (2500/60) shows increased signal involving globus pallidus and internal capsule bilaterally (arrows).
C, T2-weighted coronal MR image (2500/60).
D, T2-weighted coronal MR image (2500/60) shows increased signal from cerebellar white matter (arrows).

Case 2

A 5-year-old girl was found to have decreased visual acuity on the right side on routine screening examination. An afferent pupillary defect was noted and she was referred for neurologic evaluation. Work-up revealed multiple cafe-au-lait spots with visual acuity on the right of 20/4900 and on the left of 20/25. CT demonstrated right optic nerve enlargement extending through the optic canal. MR confirmed an optic nerve glioma extending to the chiasm. In addition, there was evidence of increased T2 signal emanating from the left thalamus (Fig. 2).

Case 3

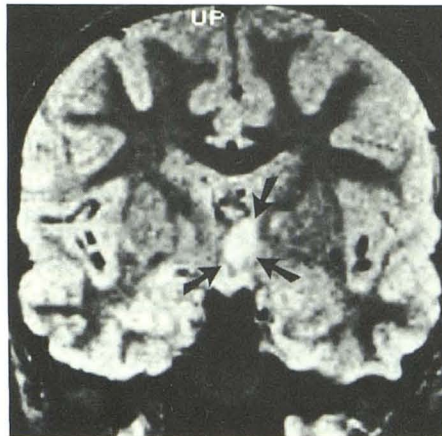
A 7-year-old boy with a known positive family history of neurofibromatosis was noted to have "difficulty tracking" by a school nurse. Cafe-au-lait spots were present and the patient was referred for neuroophthalmic evaluation. Visual acuity was 20/20 on the right and 20/60 on the left with a definite left afferent pupillary defect. MR revealed irregular

thickening of the left optic nerve with an increased signal on T2-weighted images, extending back to involve the chiasm and both optic tracts. In addition, high T2 signal was noted from the globus pallidus bilaterally (Fig. 3).

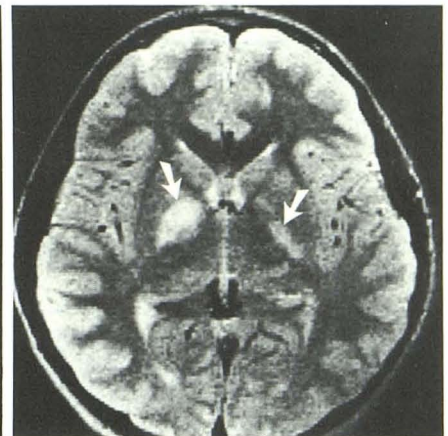
Case 4

A 7-year-old girl who was noted to have neurofibromatosis shortly after birth presented with a large plexiform neurofibroma involving the right face and orbit. She had progressive hypertrophy of the right side of the face and was treated with multiple craniofacial and plastic surgical procedures. CT revealed evidence of soft-tissue asymmetry involving the right side of the face and multiple bony changes involving the right orbit and sphenoid region (related to congenital anomaly and postsurgical changes). MR demonstrated no evidence of optic nerve glioma, but an area of increased T2 signal in the left globus pallidus and genu of the internal capsule was present (Fig. 4).

Fig. 2.—Case 2. T2-weighted coronal MR image (2500/60) shows increased signal from left thalamus (arrows).



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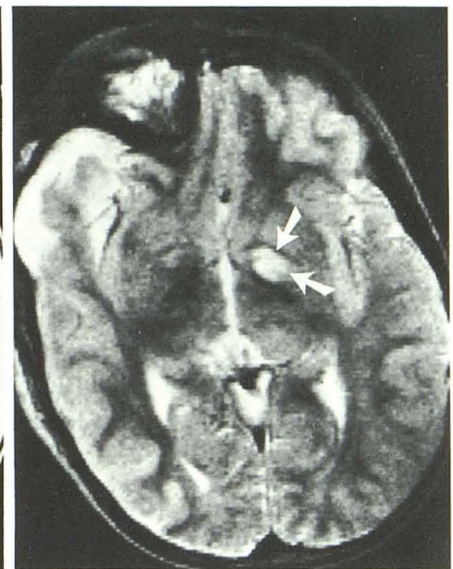


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Fig. 3.—Case 3. T2-weighted MR image (2500/60) shows areas of increased signal involving globus pallidus bilaterally.



A



B

Fig. 4.—Case 4.

A, Enhanced CT scan shows compromise of right anterior fossa secondary to plexiform neurofibroma.

B, T2-weighted MR image (2500/60) reveals increased signal intensity of left internal capsule (arrows). Increased signal in right temporal region represents postsurgical change.

Discussion

Neurofibromatosis is the most common of the phakomatoses and has an autosomal dominant inheritance. The most useful approach to the poorly understood pathogenesis of this disorder is to consider the origin of many of its cellular components in the transient embryonic neural crest. Neural crest cells migrate widely to form neuronal, glial, and other tissues whose disordered growth in neurofibromatosis results in a spectrum of abnormalities ranging from hamartomas to highly malignant neoplasms [2]. Multifocality is a common characteristic of pathologic change in neurofibromatosis.

Optic glioma, the most common CNS tumor occurring in the first decade in patients with neurofibromatosis, was present in three of our patients [3]. Differences in extent and initial sites of involvement by optic glioma between patients with and without neurofibromatosis have been described, with

multicentric glioma of the visual pathways found only in those patients with neurofibromatosis [4].

Multiple meningiomas are more likely to occur in association with neurofibromatosis, as is diffuse meningiomatosis with cortical invasions [4, 5]. Support for the occurrence of widespread multicentric disease of the CNS in neurofibromatosis thus accrues from both the presumed pathogenesis of the condition as well as the documentation of actual occurrence. This propensity for multifocal disease seems to be further supported by our findings in these patients.

MR in four children with neurofibromatosis, three with associated optic glioma, demonstrated multicentric abnormalities characterized by increased T2 signal involving multiple brain structures in each case. The globus pallidus was the most commonly involved structure, with abnormalities in three of the four cases. In two cases the globus pallidus was involved bilaterally. Other regions of diencephalic involvement

TABLE 1: MR Abnormalities in Neurofibromatosis

Case	Gender	Age (years)	Optic Glioma	Abnormality	Comment
1	M	7	Yes	Increased T2 signal from optic tracts bilaterally, globus pallidus bilaterally, and cerebellar white matter	
2	F	5	Yes	Increased T2 signal from left thalamus	
3	M	7	Yes	Increased T2 signal from optic chiasm and tracts, and bilateral globus pallidus	
4	F	7	No	Increased T2 signal from left globus pallidus and genu of internal capsule	Plexiform neurofibroma of right face

included the thalamus and internal capsule. The patient with the most widespread involvement (case 1) also had abnormalities of the cerebellar white matter. Only in this patient did abnormal signal intensity involve the posterior optic pathways. No involvement of the occipital lobe was seen in any patient. In no case were the regions of abnormal T2 signal intensity contiguous with regions of known tumor. No mass effect was associated with any of the regions of increased T2 signal. CT was performed in three of the patients, and in no case did it demonstrate abnormalities in the above locations. No focal neurologic abnormalities were present in any patient aside from the visual symptoms noted previously.

While posterior extension of glioma is an etiologic consideration for some lesions in three of the cases, the absence of CT abnormalities, the absence of continuity between the T2 abnormalities and the main tumor mass, and the lack of mass effect are features not in keeping with previous examples of such extension. A report [6] of seven cases of posterior visual pathway involvement by optic gliomas demonstrated characteristic CT abnormalities in all patients. In the single patient studied by MR, abnormally increased T2 signal was seen in continuity with the main tumor bulk with associated mass effect, findings at variance with those in our cases. Furthermore, the involvement of basal ganglia and cerebellar structures as well as other brainstem structures without concomitant neurologic symptoms somewhat mitigates against, but does not exclude, tumor etiology. These findings are instead more suggestive of widespread abnormalities such as hamartoma or low-grade gliomatosis, which may characterize central neurofibromatosis, perhaps predisposing to neoplastic transformation. Indeed, although considered rare, several reports exist of diffuse gliomatosis in neurofibromatosis characterized by abnormal hypertrophic glia, giant astrocytes, and multinucleate cells. The described distribution of abnormalities included the basal ganglia, pulvinar, other thalamic nuclei, and midbrain similar to the distribution of T2 abnormalities in our patients [7]. While the question of the premalignant nature of such lesions remains open, an increased number of nonoptic gliomas also occurs in neurofibromatosis, also in similar

distribution, including involvement of the third ventricular region, brainstem, and cerebellum [8].

The observed T2 abnormalities in our patients affect regions outside the visual pathways, where abnormality has not been considered characteristic of neurofibromatosis. Such lesions may well contribute to the specific learning disabilities, attention-deficit disorder, and speech impairment that are the most common neurologic complications of neurofibromatosis, or to the intellectual retardation or seizure disorders that eventually occur in about 5% of these patients [9].

Recognition of such lesions in children with neurofibromatosis is the first step toward their proper management. While present evidence suggests that they do not represent tumor and, therefore, do not warrant surgical or radiation therapy, further study is imperative to determine any potential for neoplastic transformation and to delineate their functional significance.

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