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Spontaneous Regression of a Tectal Mass in Neurofibromatosis 1

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Summary: MR images showed an enhancing, enlarging mass in the tectum of the midbrain in a child with neurofibromatosis type 1. The mass was presumed to be a tectal glioma, which initially enlarged then regressed in size over a 3-year period and ceased to enhance. Although a tissue diagnosis was not available, we believe the temporal evolution of this lesion is strong presumptive evidence of a hamartoma. This case argues for the conservative management of patients with neurofibromatosis type I when possible.

There are many CNS lesions that occur in neurofibromatosis (NF). In NF type 1, two of the most common lesions are gliomas and hamartomas. Differentiating between these two lesions can be difficult, because gliomas and hamartomas share many imaging features. When hamartomas enlarge, enhance, or appear in atypical locations, differentiating them from gliomas is problematic. We report a case of a child with NF type 1 and a large, enhancing tectal mass that spontaneously regressed over a 3-year follow-up period.

Case Report

The patient initially presented at the age of 3 weeks with a left-sided periauricular soft-tissue mass, which subsequently enlarged and was resected when the patient was 8 years old. Pathologically, the mass proved to be a plexiform neurofibroma. The patient remained well but reported visual blurring at the age of 13. Clinical examination at that time showed a normal range of eye movements with no defects in gaze. Funduscopic examination revealed papilledema. A CT study showed obstructive hydrocephalus with dilatation of the lateral and third ventricles. A shunt was placed in the left lateral ventricle and an MR study was obtained 3 days later. The MR examination showed multiple intracerebral lesions, two of which enhanced: one in the globus pallidus and the other in the tectum (Fig 1A–C). About 6 months later, the lesion in the tectum enlarged slightly (Fig 1D–F) while the other lesions, including the one in the globus pallidus, remained static. A decision was made to reexamine the patient in 6 months rather than to immediately biopsy this tectal lesion.

At the 6-month follow-up study, at age 14, the lesions had decreased in size and were less enhanced (not illustrated). At age 16, when the patient was treated for shunt malfunction, an MR study revealed resolution of enhancement and a marked decrease in the size of the tectal lesion (Fig 1G–I). The globus pallidus lesion was minimally decreased in size and also enhanced less. No treatment, except shunting, had been instituted at any time.

Discussion

The most common lesions in the CNS in NF 1 are optic gliomas, astrocytomas, and hamartomas. Astrocytomas may occur anywhere in the brain; hamartomas are found most often in the globus pallidus, cerebellum, and brain stem (1, 2). Hamartomas in the tectum have not been reported previously. Most hamartomas appear as focal areas of hyperintensity on T2-weighted images and on T1-weighted images they may be hypointense, isointense, or hyperintense (1, 2). Although there are reports of enhancing hamartomas (3), enhancement is a distinctly unusual feature and thought to be indicative of a neoplastic lesion. Hamartomas are much more common in children than in adults. With respect to temporal changes, hamartomas usually regress as a child matures, but up to 10% show growth on serial examinations. Growth is unusual after age 10 years (3, 4).

Histologically, hamartomas are composed of atypical glial infiltrates, foci of microcalcifications, perivascular gliosis, and spongy peripheral changes (2). Recently, Dipaolo et al (5) investigated these masses with pathologic/radiologic correlation and described spongiform myelinopathy or vacuolar changes in the myelin. The importance of these lesions lies in their differentiation from low-grade astrocytomas. Often, biopsy is not a viable option, as these masses may lie in eloquent areas.

The patient cited in this report had a number of intracerebral lesions that were of high signal intensity on T2-weighted images. The smaller lesions showed neither mass effect nor edema and were presumed to be hamartomas. The lesion in the right globus pallidus, although displaying enlargement and enhancement, was also thought to be a hamartoma owing to its typical location (1, 2). In contradistinction, the mass in the tectum was unlike any previously reported

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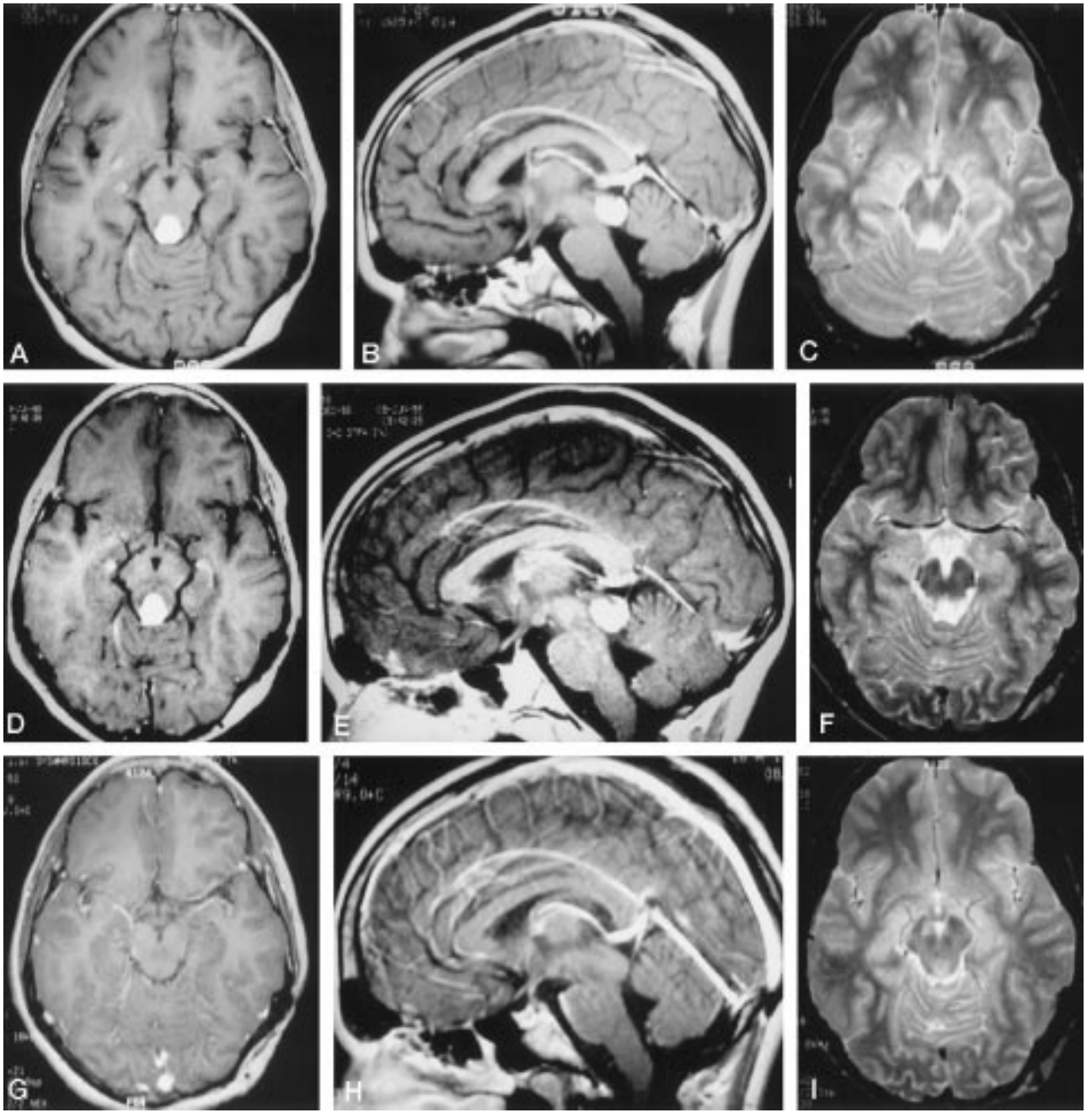

FIG 1. Child with neurofibromatosis type 1 and a large, enhancing tectal mass that spontaneously regressed over a 3-year follow-up period.

A, Contrast-enhanced axial T1-weighted MR image (550/11/2 [TR/TE/excitations]) at age 13 shows a homogeneously enhancing mass, 1.2 × 1.2-cm (width × anteroposterior dimension), in the tectum.

B, Contrast-enhanced sagittal T1-weighted image (566/11/2) at age 13 shows the mass narrowing the aqueduct of Sylvius. The superoinferior dimension is 1.2 cm. A ventriculostomy had been placed previously to decompress the resultant hydrocephalus.

C, Unenhanced axial T2-weighted image (2500/90/2) also at age 13 shows the mass to have uniformly increased signal intensity.

D, Contrast-enhanced axial T1-weighted image (570/15/2) at age 13½ shows the enhancing tectal mass, which now measures 1.4 × 1.6 cm in maximum dimension.

E, Contrast-enhanced sagittal T1-weighted image (425/10/2) at same time shows the mass has slightly increased in size, now measuring 1.3 cm in the superoinferior plane.

F, Unenhanced axial T2-weighted image (2500/90/2) also at age 13½ shows the high-signal tectal lesion.

G, Contrast-enhanced axial T1-weighted image (417/25/2) obtained at age 16½ shows a bulky appearance to the mass, which now measures approximately 0.8 × 0.9 cm. The mass no longer enhances.

H, Contrast-enhanced sagittal T1-weighted image (633/24/2) also at age 16½ confirms the lack of enhancement of the mass, which still measures 0.8 cm in the superoinferior dimension.

I, Unenhanced axial T2-weighted image (2500/90/2) at same time no longer shows the previously identified hyperintensity of the mass.

hamartoma, and these atypical features prompted a presumptive diagnosis of astrocytoma. We observed, however, a decrease in size and enhancement of the mass over time, a fact that was presumed to exclude the possibility of an astrocytoma and that supported the diagnosis of hamartoma.

Conclusion

Conservative management of patients with NF 1 whenever possible is emphasized by the spontaneous regression of a tectal mass observed in this case.

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