

Diffusion MRI of myelin vacuolization in NF1

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Section: Neuroradiology

Imaging Technique: MR

Imaging Technique: MR-Functional imaging

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Case Type: Clinical Cases

Authors: R.N.Sener

Patient: 6 years, female

Clinical History:

Cafe-au-lait spots, and axillary freckling

Imaging Findings:

The patient was a member of a family with known neurofibromatosis type 1. She had multiple cafe-au-lait spots, and axillary freckling. CT and MRI studies were performed several times. On the recent follow-up by MRI, a diffusion imaging protocol was added to the conventional T1 and T2-weighted sequences.

Discussion:

Neurofibromatosis type 1 (NF1) is primarily a disease of nerves and astrocytes, and its intracranial manifestations mainly include optic gliomas, astrocytomas, hamartomas, and neurofibromas. Its incidence is approximately 1:2000-3000. On the other hand, NF2 is a disease of coverings of the central nervous system, and its intracranial manifestations mainly include schwannomas and meningiomas. Its incidence is approximately 1:50000. In patients with neurofibromatosis, myelin vacuolization represents a characteristic histopathological change in which layers of myelin separate from each other as they spiral around the axon. These occur at the regions where myelin is dysplastic, and they are commonly seen in the cerebellar white matter, pons, midbrain, callosal splenium, and internal capsule. On MRI areas of myelin vacuolization characteristically appear as multiple nodular lesions which are hyperintense on T2-weighted images. They usually are not seen on T1-weighted images, and there is no contrast-enhancement after intravenous administration of contrast medium, and they do not cause vasogenic edema, hence no mass effect. These features distinguish regions of myelin vacuolization from a developing astrocytoma.

Differential Diagnosis List: NF1

Final Diagnosis: NF1

References:

Barkovich AJ. Pediatric neuroimaging. Lippincott Williams & Wilkins, Philadelphia (2000).
Aoki S, Barkovich AJ, Nishimura K, Kjos BO, Machida T, Cogen P, Edwards M, Norman D. Neurofibromatosis types

1 and 2: cranial MR findings.

Radiology. 1989 Aug;172(2):527-34. (PMID: [2501822](#))

DiPaolo DP, Zimmerman RA, Rorke LB, Zackai EH, Bilaniuk LT, Yachnis AT. Neurofibromatosis type 1: pathologic substrate of high-signal-intensity foci in the brain.

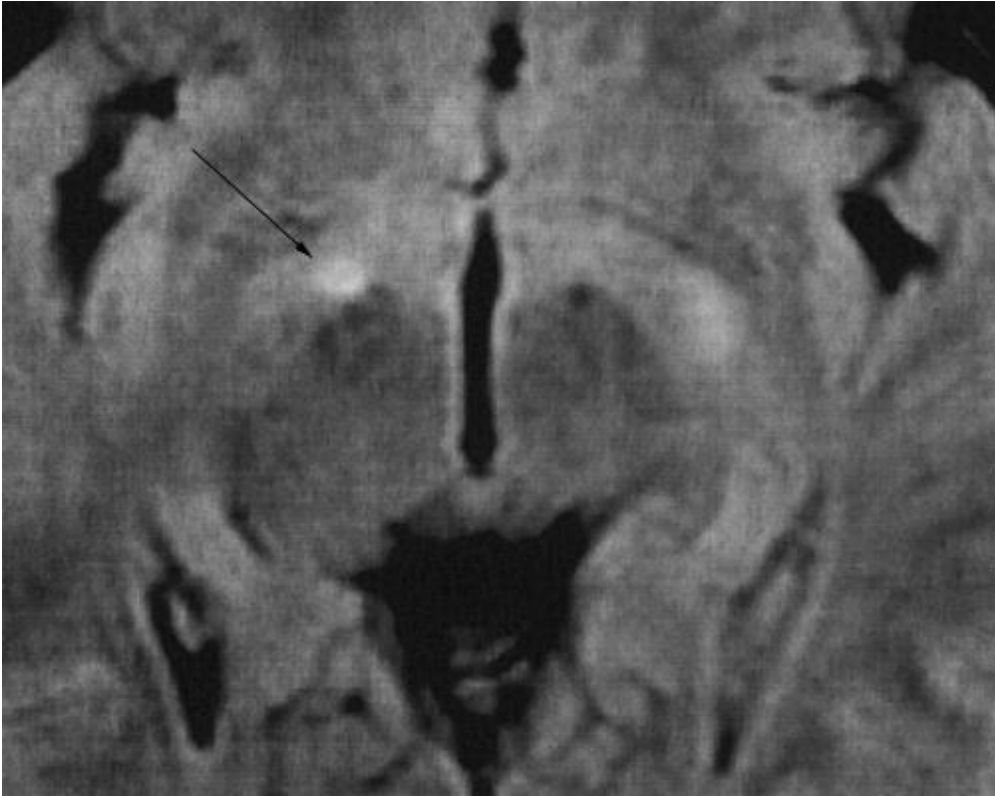
Radiology. 1995 Jun;195(3):721-4. (PMID: [7754001](#))

Sevick RJ, Barkovich AJ, Edwards MS, Koch T, Berg B, Lempert T. Evolution of white matter lesions in neurofibromatosis type 1: MR findings.

AJR Am J Roentgenol. 1992 Jul;159(1):171-5. (PMID: [1609692](#))

Figure 1

a



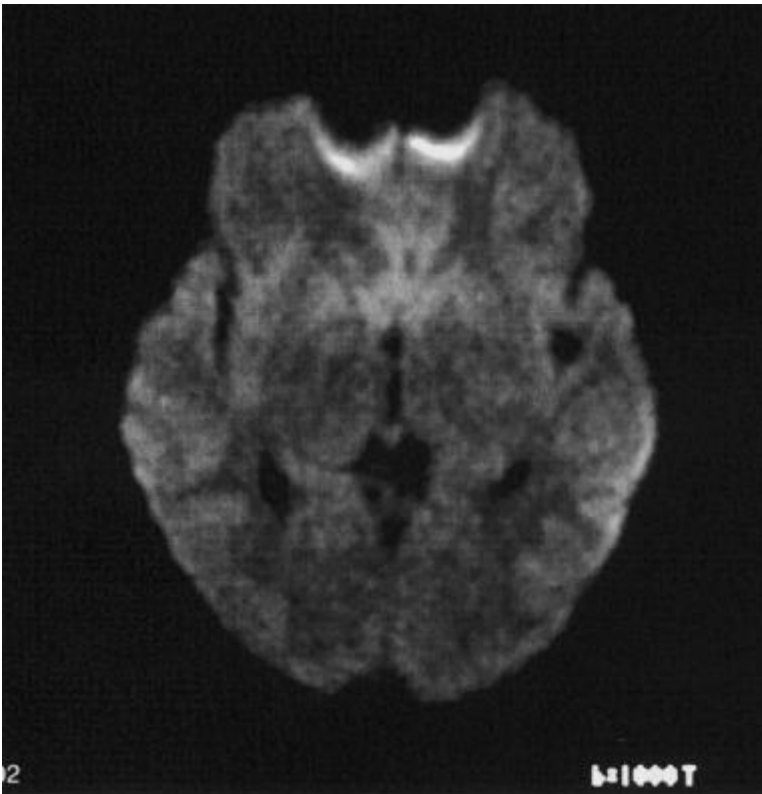
Description: T2-weighted image reveals bilateral high-signal nodules in the globus pallidus, consistent with hamartomas. **Origin:**

b



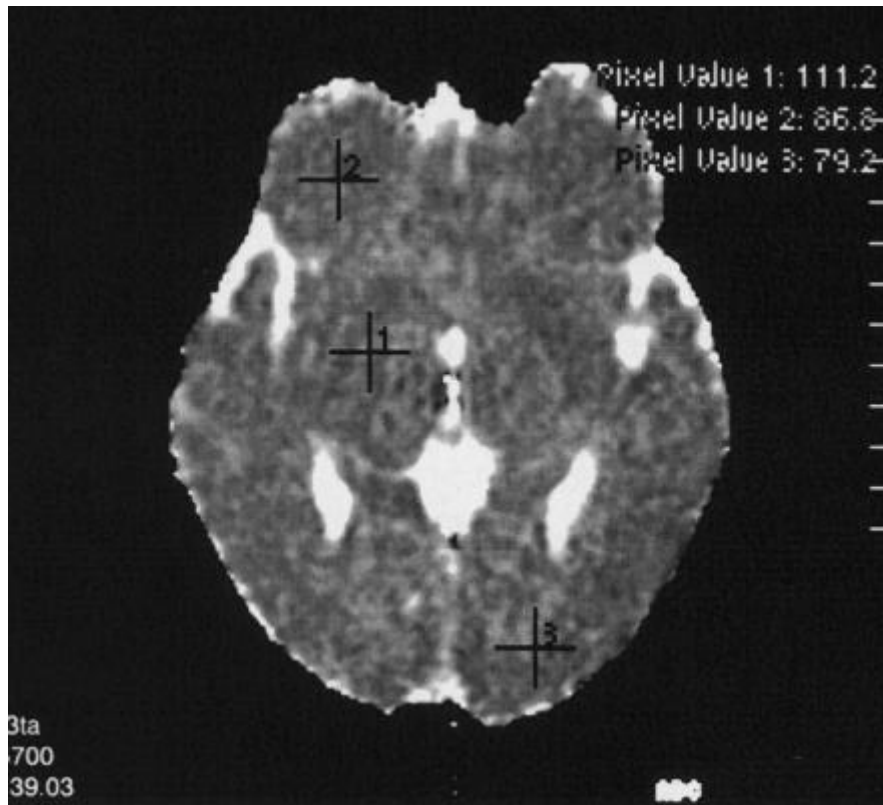
Description: FLAIR image reveals high-signal hamartomas in the globus pallidus. That on the right side is well defined (arrow). This is a common finding in NF1. **Origin:**

c



Description: Heavily diffusion-weighted (b=1000 sec/mm²) image is negative for the hamartomas. **Origin:**

d



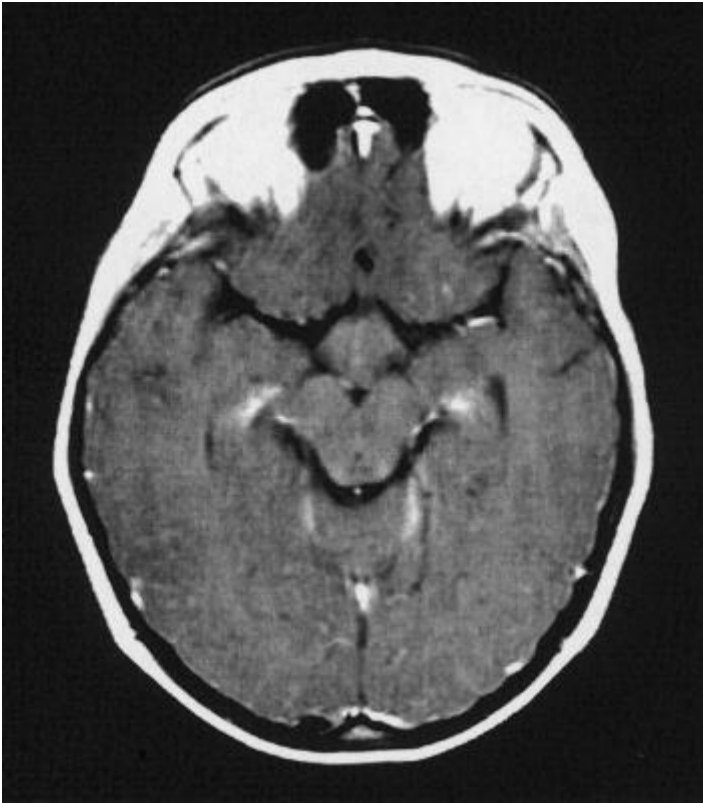
Description: ADC map from an echo-planar diffusion imaging sequence reveals a higher ADC value: $1.11 \times 10^{-3} \text{ mm}^2/\text{sec}$ in the right-sided hamartoma, compared to normal white matter values: 0.86 and $0.79 \times 10^{-3} \text{ mm}^2/\text{sec}$. **Origin:**

e



Description: Left parasagittal T1-weighted image with paramagnetic contrast medium reveals an unenhancing, thickened left optic nerve, consistent with low-grade glioma (arrow). Note that the nodule-like structure anterosuperior to the optic nerve is the normal gyrus paraterminalis, which should not be taken as a mass lesion. **Origin:**

f



Description: Transverse image with contrast medium reveals thickened chiasm due to bilateral optic gliomas. **Origin:**

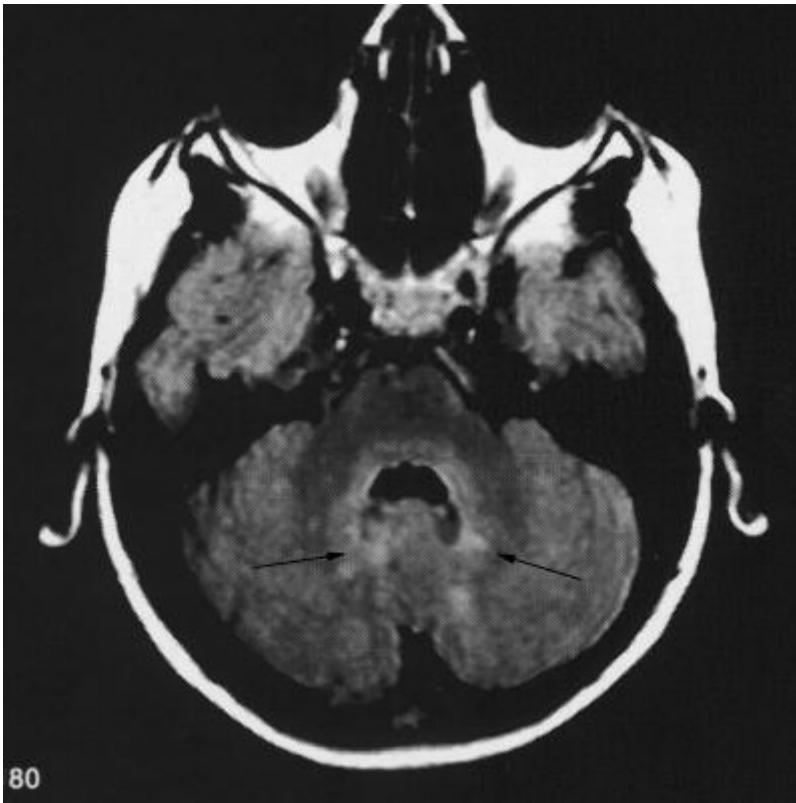
Figure 2

a



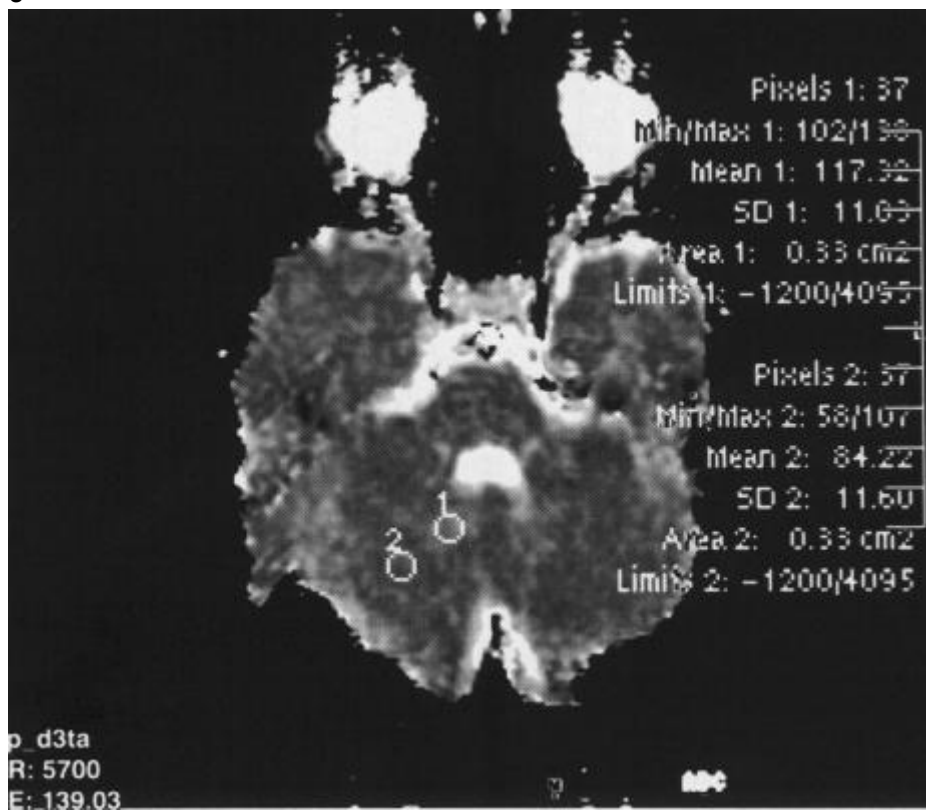
Description: T2-weighted image reveals high-signal globular foci in the cerebellum in the vicinity of the fourth ventricle. **Origin:**

b



Description: FLAIR image reveals the globular foci (arrows), representing myelin vacuolization, to better advantage. **Origin:**

c



Description: ADC map from an echo-planar diffusion imaging sequence reveals a higher ADC value: $1.17 \times 10^{-3} \text{ mm}^2/\text{sec}$ in the region affected with myelin vacuolization, compared to that of normal cerebellar parenchyma: $0.84 \times 10^{-3} \text{ mm}^2/\text{sec}$. **Origin:**