

# Disappearing Lesion in Linear Nevus Sebaceous Syndrome

J Zimmer, M Edwards-Brown, D Sokol

## Citation

J Zimmer, M Edwards-Brown, D Sokol. *Disappearing Lesion in Linear Nevus Sebaceous Syndrome*. The Internet Journal of Neurology. 2004 Volume 5 Number 1.

## Abstract

Disappearing lesions, or unidentified bright objects (UBOs), have been demonstrated on MRI of patients with neurofibromatosis type I (NF1). Clinically UBOs have been associated with learning disorders. We report the phenomena of UBO in a girl with linear nevus sebaceous syndrome, suggesting that UBOs may not be specific to NF1.

## INTRODUCTION

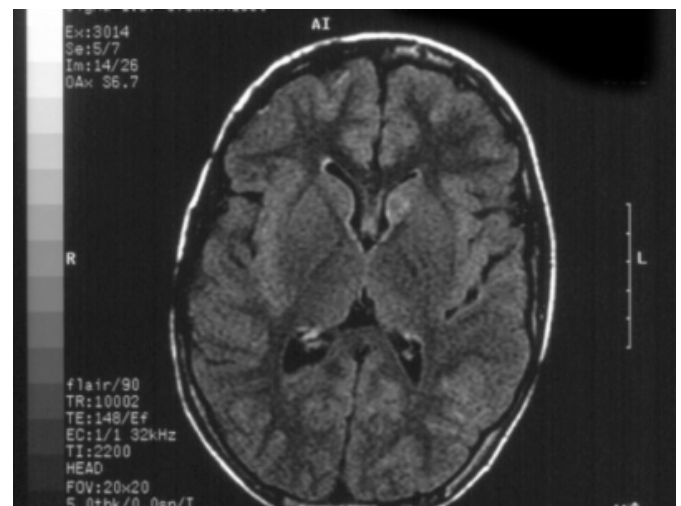
Linear nevus sebaceous syndrome consists of the triad of linear nevus sebaceous, seizures, and mental retardation. Nevi are typically near the midline of the face but may also be on the extremities or torso. Nevi do not cross midline. Since up to 15% of lesions may undergo malignant transformation in the patient's second to third decade of life they are surgically removed. Patients often have ocular findings such as hemianopsia, cortical blindness, and colobomas of the iris, choroid, optic nerve, and retina. Neuroradiologic findings include porencephaly, ipsilateral megacephaly and ventriculomegaly, and hamartomas ipsilateral to the skin lesions. <sup>1</sup>

## CASE REPORT

A five-year-old female with linear nevus sebaceous syndrome presented with behavior suspicious for seizures. She experienced crying spells occasionally associated with left arm weakness and incontinence of stool. She would recover to her normal baseline within two hours after onset. MRI showed several hyperdense lesions on flair images, most prominent in the left caudate nucleus (Figure 1a) and adjacent to the frontal horn of the right lateral ventricle. She was placed on valproic acid and has remained seizure-free. Follow-up MRI approximately one year later showed complete resolution of the left caudate lesion (Figure 1b) and decreased intensity of the lesion near the right lateral ventricle.

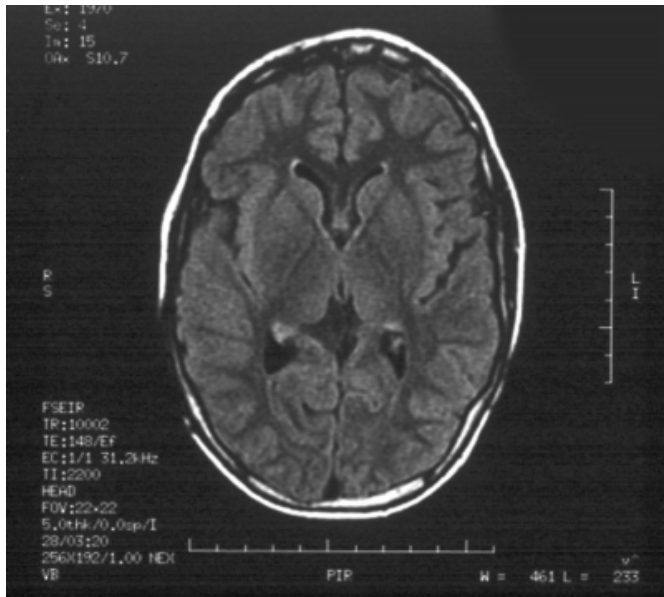
## Figure 1

Figure 1a: Axial FLAIR MRI showing an unidentified bright object in the head of the left caudate nucleus (arrow).



**Figure 2**

Figure 1b: Axial FLAIR MRI performed 14 months after scan in Figure 1a showing resolution of the unidentified bright object.



Disappearing lesions, or unidentified bright objects (UBOs), have been demonstrated on MRI of patients with neurofibromatosis type I (NF1).<sup>2</sup> These lesions may represent abnormal glial cell proliferation and increased fluid within the myelin.<sup>3</sup> Clinically UBOs have been associated with learning disorders<sup>4</sup> and with the incidence of

other features of NF1 such as the existence of optic gliomas, neurofibromas, and Lisch nodules.<sup>5</sup> To our knowledge, this is the first report demonstrating disappearing lesions in a patient with linear nevus sebaceous syndrome. Thus, the phenomena of UBOs may not be specific to NF1.

**CORRESPONDENCE TO**

Deborah K. Sokol, PhD, MD Pediatric Neurology-RI 1757  
Riley Hospital for Children Indiana University School of  
Medicine 702 Barnhill Drive Indianapolis, IN 46202  
317-278-2219 FAX: 317-274-3622 dksokol@iupui.edu

**References**

1. Sokol DK, Dunn DW, Treadwell PA. Evaluation and treatment of patients with neurocutaneous syndromes. In Batjer HH, Loftus CM (Eds): Textbook of Neurological Surgery. Philadelphia, Lippincott, Williams, and Wilkins 2003:346-358.
2. Raininko R, Thelin L, Eeg-Olofsson O. Non-neoplastic brain abnormalities on MRI in children and adolescents with neurofibromatosis type 1. *Neuropediatrics* 2001;32:225-230.
3. DiPaolo DP, Zimmerman RA, Rorke LB, et al. Neurofibromatosis type 1: Pathologic substrate of high-signal-intensity foci in the brain. *Radiology* 1995;195:721-724.
4. North K, Riccardi V, Samango-Sprouse C, et al. Cognitive function and academic performance in neurofibromatosis type-1: Consensus statement from the NF1 Cognitive Disorders Task Force. *Neurology* 1997;48:1121-1127.
5. Szudek J, Friedman JM. Unidentified bright objects associated with features of neurofibromatosis 1. *Pediatric Neurology* 2002;27(2):123-127.

**Author Information**

**Jennifer Zimmer, M.D.**

Department of Pediatrics, Riley Hospital for Children, Indiana University School of Medicine

**Mary Edwards-Brown, M.D.**

Neuroradiology, Department of Radiology, Riley Hospital for Children, Indiana University School of Medicine

**Deborah K. Sokol, Ph.D., M.D.**

Pediatric Neurology, Department of Neurology, Riley Hospital for Children, Indiana University School of Medicine