AJNR

Bilateral arachnoid cysts of the temporal fossa in four children with glutaric aciduria type I.

J K Hald, P H Nakstad, O H Skjeldal and P Strømme

AJNR Am J Neuroradiol 1991, 12 (3) 407-409
http://www.ajnr.org/content/12/3/407

Bilateral Arachnoid Cysts of the Temporal Fossa in Four Children with Glutaric Aciduria Type I

John K. Hald[1]
Per H. Nakstad[1]
Ola H. Skjeldal[2]
Petter Strømme[2]

Glutaric aciduria type I is an uncommon inborn error of metabolism. It is a serious disease, often with a fatal outcome. This study reports the presence of bilateral temporal fluid collections, probably bilateral arachnoid cysts, in association with glutaric aciduria type I. The CT and, when available, MR studies from five patients with this disorder were reviewed. Four of the patients had findings consistent with bilateral arachnoid cysts of the temporal fossa. This is a rare occurrence, with only 11 such cases reported in the literature.

The observed association between temporal fluid collections and glutaric aciduria type I suggests that patients with bilateral arachnoid cysts should be investigated for this metabolic disorder.

***AJNR* 12:407–409, May/June 1991**

Neuroradiologic investigations have demonstrated retarded myelination and possible cortical dysplasia in the temporal regions in patients with glutaric aciduria type I [1]. Ventricular enlargement, loss of caudate nuclei, and frontotemporal atrophy have also been reported [2, 3]. This article reports the findings of bilateral temporal fluid collections, consistent with arachnoid cysts, in children with glutaric aciduria type I.

Materials and Methods

The study population consists of five children (from three families) known to have glutaric aciduria type I (Table 1). The CT examinations of the patients were retrospectively reviewed, and two of the children were also examined with MR imaging. CT was performed with either a Somatom DRH (Siemens, Erlangen, Germany), or a GE 9800 (General Electric, Milwaukee, WI), except for one patient, who was examined in 1982 on a Delta Scan 50 FS (Ohio Nuclear, Solon, OH). MR was performed on a 1.5-T Magnetom (Siemens, Erlangen, Germany). Sagittal T1-weighted images, 600/15/2 (TR/TE/excitations), and double echo T2-weighted axial images, 3100/15, 90/1, were obtained.

Results

One patient (case 4), an 11-year-old boy, had diffusely reduced attenuation of white matter in both cerebral hemispheres demonstrated by CT. The other patients had normal attenuation. All the children had slight ventricular enlargement, but no evidence of loss of the caudate nuclei. Except for slight ventricular enlargement, one girl (case 5) had normal CT and MR scans. The other four children had bilateral fluid collections of varying size in the middle cranial fossa at the tip of the temporal lobes (Figs. 1–4). The fluid collections varied in size from 2 to 4 cm in largest diameter, and were symmetrical in both size and shape except in one patient

Received July 2, 1990; revision requested September 12, 1990; revision received October 16, 1990; accepted October 26, 1990.

Presented in part at the annual meeting of the American Society of Neuroradiology, Los Angeles, March 1990.

[1] Department of Radiology, Section of Neuroradiology, Rikshospitalet, The National Hospital, University of Oslo, 0027 Oslo 1, Norway. Address reprint requests to J. K. Hald.

[2] Department of Pediatrics, Rikshospitalet, 0027 Oslo 1, Norway.

0195–6108/91/1203–0407

TABLE 1: Data on Five Children with Glutaric Aciduria

Case No.	Age at Diagnosis	Sex	Neurologic Deficits	Type of Examination
1*	2 yr	M	Dead	CT
2*	2 da	M	Slight	CT and MR
3*	13 mo	F	Moderate	CT
4*	11 yr	M	Severe	CT
5	8 mo	F	Severe	CT and MR

* Indicates presence of bilateral temporal fluid collections.

(case 2), in whom the right fluid collection was slightly larger than the left.

Two patients (cases 1 and 3) had bilateral frontoparietal subdural hematomas that were shunted (case 1) and evacuated by trephination (case 3), respectively. Subsequently, they developed hydrocephalus and both were shunted to their lateral ventricles. The hematomas showed no communication with the temporal fluid collections, and there was no apparent change in the size and shape of the temporal fluid collections when comparing the CT scans of one of the patients (case 3) from before the hematoma evacuation with the scans obtained after the surgical procedure. We do not have any scans of the other patient with hematoma (case 1) from before the shunting procedures, but the two available CT examinations obtained after shunting demonstrate reduction in the size of the hematomas and unchanged temporal fluid collections.

Discussion

Glutaric aciduria type I is an inborn error of metabolism caused by a deficiency of glutaryl CoA dehydrogenase, which catalyzes one step in the catabolism of lysine, hydroxylysine, and tryptophan [1, 4, 5]. The disease has an autosomal recessive mode of inheritance, and is characterized by a progressive choreoathetosis, hyperkinesia, and spasticity. Mental retardation has been reported, and by the second half of the first decade the motor disorder may be so severe that the patient is completely helpless [2, 3].

Arachnoid cysts may be congenital, posttraumatic, or postinflammatory. Congenital or primary arachnoid cysts are benign developmental cavities situated within the arachnoid membrane; they are lined by collagen and cells of the arachnoid mater, and contain clear CSF-like fluid [6]. They usually occur as single lesions [7], and a review of the literature has revealed only 11 cases of bilateral arachnoid cysts, two of the patients being siblings [8–12].

We do not have any surgical or autopsy proof that the fluid collections we report represent arachnoid cysts, and with noninvasive techniques only, we cannot eliminate the possibility of atrophy as the cause. Previous publications on the subject suggest frontotemporal lobe atrophy [1, 4, 5], and this might be correct for at least one of the previously reported cases [1]. We do, however, consider the published CT findings of six other patients with glutaric aciduria type I to be consistent with arachnoid cysts [4, 5].

The MR examination of case 2 (Fig. 2) does not demonstrate undulation of the temporal lobe margins adjacent to the cysts, a finding that could have indicated temporal lobe hypogenesis [13]. However, the contour of extracerebral arachnoid cysts may correspond to irregularities of the brain [14]; well-defined, rounded cysts pushing back the brain tissue are less common [14]. Arachnoid cysts of the temporal region are associated with subdural hemorrhage [7], a complication that was present in two of the patients (cases 1 and 3) in our study. However, the subdural hematomas were at some distance from the temporal regions, and the size and shape of the temporal fluid collections were not affected by the surgical procedures. This is indicative of arachnoid cysts rather than atrophy or dysplasia.

CT and MR are comparable in detecting arachnoid cysts,

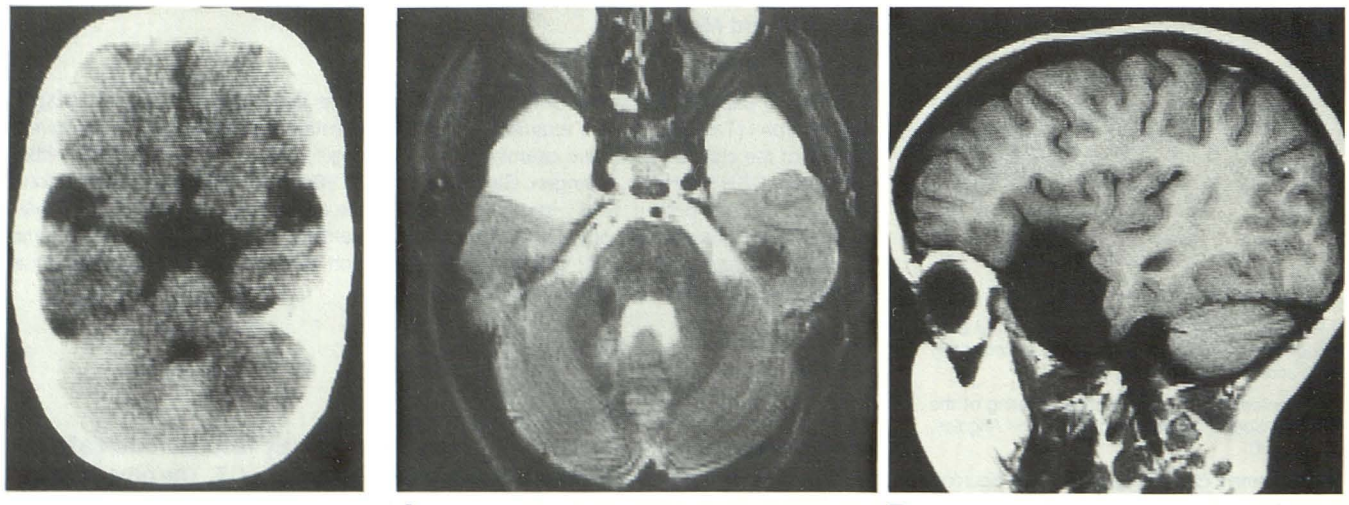
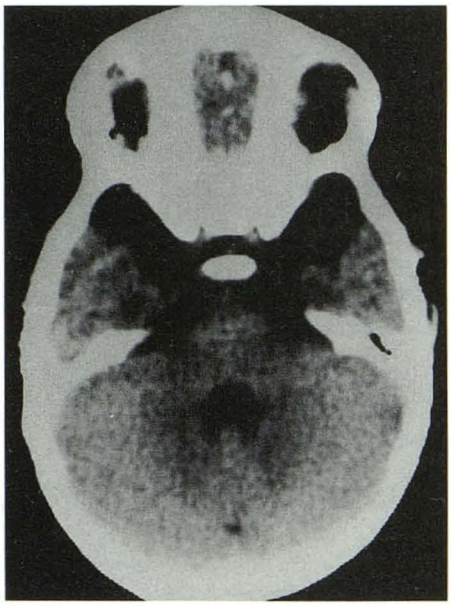


Fig. 1.—Case 1. Axial CT scan of 2-year-old boy shows fluid collections of the temporal regions, consistent with arachnoid cysts.

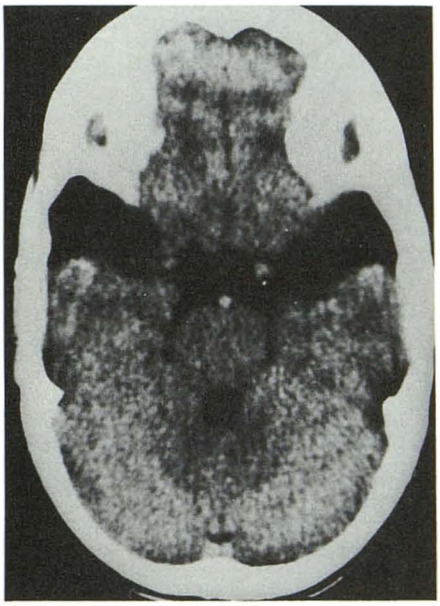
Fig. 2.—Case 2. Five-year-old brother of patient in case 1.
A and *B*, Axial (3100/90) (*A*) and sagittal (600/15) (*B*) MR images show fluid collections of the temporal regions, consistent with arachnoid cysts.

Fig. 3.—Case 3. Axial CT scan of 13-month-old girl shows fluid collections consistent with bilateral arachnoid cysts.



3

Fig. 4.—Case 4. Axial CT scan of 11-year-old brother of patient in case 3 shows same findings of fluid collections consistent with bilateral arachnoid cysts.



4

but MR is superior in defining cysts in several planes and in tissue discrimination, thereby differentiating arachnoid cysts from other lesions that are hypodense on CT [15]. The association between severe neurologic deficits and bilateral arachnoid cysts of the temporal fossa is well documented [4, 11, 12], but not always the case [8]. It has been suggested that the neurologic deficits can be due to primary dysgenesis of the operculum [8], and it has been shown that middle cranial fossa cysts can be present together with temporal lobe hypogenesis [13]. In this study all the patients had some degree of neurologic deficits, but not necessarily associated with dysgenesis of the operculum, as demonstrated by the normal MR appearance of the temporal lobes in case 5, a patient who was severely handicapped.

We cannot explain the association between the bilateral temporal fluid collections and glutaric aciduria type I. The possibility exists that this metabolic disorder might be concealed among children confirmed to have developmental retardation, cerebral palsy, or acute encephalopathy due to unknown causes. Patients with bilateral arachnoid cysts should be investigated for this enzyme deficiency.

REFERENCES

1. Valk J, van der Knaap MS. *Magnetic resonance of myelin, myelination and myelin disorders.* Berlin: Springer Verlag, **1989**:141–147
2. Amir N, El-Peleg ON, Shalev RS, Christensen E. Glutaric aciduria type I: enzymatic and neuroradiologic investigations of two kindreds. *J Pediatr* **1989**;114:983–989
3. Amir N, El-Peleg ON, Shalev RS, Christensen E. Glutaric aciduria type I: clinical heterogeneity and neuroradiological features. *Neurology* **1987**; 37:1654–1657
4. Goodman SI, Markey SP, Moe PG, Miles BS, Teng CC. Glutaric aciduria: a "new" disorder of amino acid metabolism. *Biochem Med* **1975**; 12:12–21
5. Goodman SI, Kolhoff JG. Glutaric aciduria: inherited deficiency of glutaryl-Co A dehydrogenase activity. *Biochem Med* **1975**;13:138–140
6. Rengachary SS, Watanabe I, Brackett CE. Pathogenesis of intracranial arachnoid cysts. *Surg Neurol* **1978**;9:139–144
7. Naidich TP, McLone DG, Radowski MA. Intracranial arachnoid cysts. *Pediatr Neurosci* **1985–86**;12:112–122
8. Nagaya T, Doi T, Katsumata T, Kuwayama N. Bilateral middle cranial fossa arachnoid cysts. *Pediatr Neurosci* **1987**;13:184–187
9. Shirane R, Tanaka T, Andoh A, Suzuki J. Bilateral symmetrical middle cranial fossa arachnoid cysts in a neonate. *Surg Neurol* **1986**;26:395–398
10. Abhyankar SC, Bichile LS, Chabra A. Bilateral cerebral arachnoid cysts with mental retardation. *Indian J Pediatr* **1985**;52:103–105
11. Rossitch E, Oakes WJ. Klüver-Bucy syndrome in a child with bilateral arachnoid cysts: report of a case. *Neurosurgery* **1989**;24:110–112
12. Handa J, Okamoto K, Sato M. Arachnoid cyst of the middle cranial fossa: report of bilateral cysts in siblings. *Surg Neurol* **1981**;16:127–130
13. Robertson SJ, Wolpert SM, Runge VM. MR imaging of middle cranial fossa arachnoid cysts: temporal lobe agenesis syndrome revisited. *AJNR* **1989**;10:1007–1010
14. Giudicelli G, Hassoun J, Choux M, Tonon C. Les kystes "arachnoidiens" supra-tentoriels. *J Neuroradiol* **1982**;9:179–201
15. Gandy SE, Heier LA. Clinical and magnetic resonance features of primary intracranial arachnoid cysts. *Ann Neurol* **1987**;21:342–348

The reader's attention is directed to the commentary on this article, which appears on pages 413–416.