European Journal of Radiology Extra 79 (2011) e85–e88

Contents lists available at ScienceDirect

European Journal of Radiology Extra

journal homepage: intl.elsevierhealth.com/journals/ejrex

Leukoencephalopathy with extensive brain calcifications and parenchymal cysts in a child

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ARTICLE INFO

Article history:
Received 23 May 2010
Received in revised form
21 September 2010
Accepted 6 April 2011

Keywords:
Leukoencephalopathy
Brain calcifications
Cysts
Magnetic resonance imaging

ABSTRACT

Leukoencephalopathy with cerebral calcifications and cysts (LCC) is a recently described and extremely rare entity of unknown origin characterized radiologically by white matter abnormalities, calcifications and cysts. The onset of this disorder occurs from early infancy to adolescence and there is no specific clinical feature suggestive of this disorder. We report a new case of this rare entity and emphasize CT and MRI findings that permit to assess diagnosis.

Contents

1. Introduction

The association of brain calcifications, leukoencephalopathy and cysts is a very uncommon disorder which was recently described by Labrune et al. in 1996 [1,2]. The clinical presentation of this entity is not specific but neuroradiological findings are grouped in a very suggestive triad of signs [1–3] which are parenchymal cysts, calcifications in the basal and cerebellar grey nuclei and white matter diffuse signal abnormalities on MRI [2]. The development of parenchymal cysts can lead to compressive complications and surgery is sometimes indicated [4]. We report the case of a 5-year-old girl with the uncommon syndrome of leukoencephalopathy, brain calcifications, and cysts (LCC) revealed by tetraparesis and seizure.

2. Case report

A 5-year-old girl was admitted to the emergency unit with a generalized tonic clonic seizure. Her parents report a one month history of vomiting, headache, progressive spasticity and disorientation. After stabilization of the seizure, neurological examination revealed a spastic tetraparesis and ophthalmologic examination showed a bilateral papillar edema without other abnormalities.

A computed tomography of the brain was performed and showed numerous calcifications in the basal ganglia (Figs. 1 and 2). CT also showed marked hypodensity in the periventricular white matter (Fig. 1) and intraparenchymal bilateral cystic lesions. Some of the cysts present a peripheral enhancement after intravenous injection of an iodine contrast agent. MRI of the brain gave similar results and showed diffuse high signal intensity in the periventricular white matter on T2 weighted images, mainly in the anterior areas, relatively sparing the U fibers and corpus callosum with extensive calcifications in the basal ganglia and thalamus (Figs. 3–8). There were also parenchymal cysts; a voluminous right

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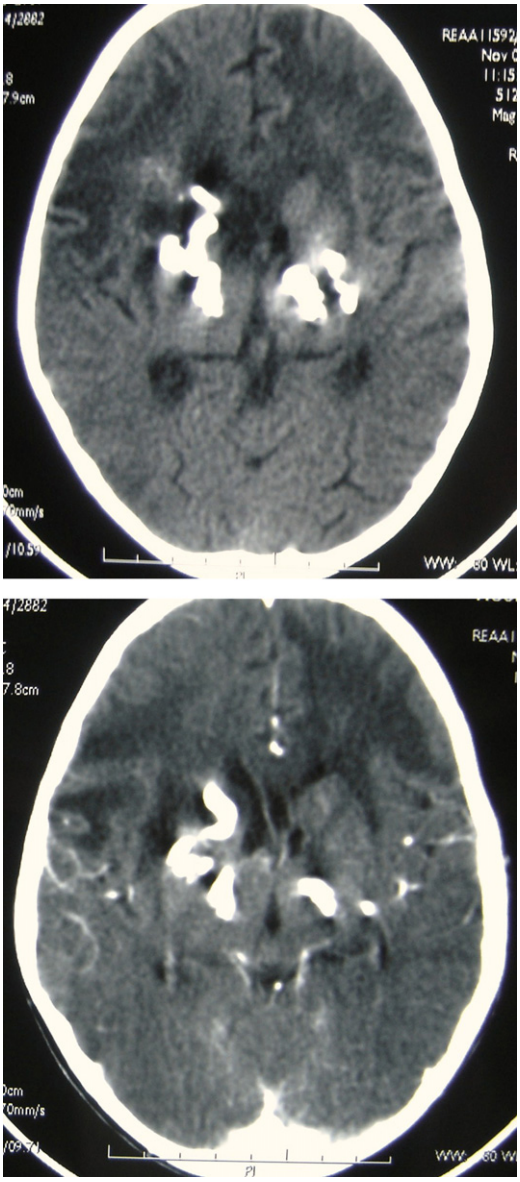
doi:10.1016/j.ejrex.2011.04.009



Figs. 1 and 2. Brain CT scan before (1) and after (2) intravenous administration of a contrast agent showing bilateral extensive calcifications of the basal ganglia and marked hypodensity in the anterior periventricular white matter.

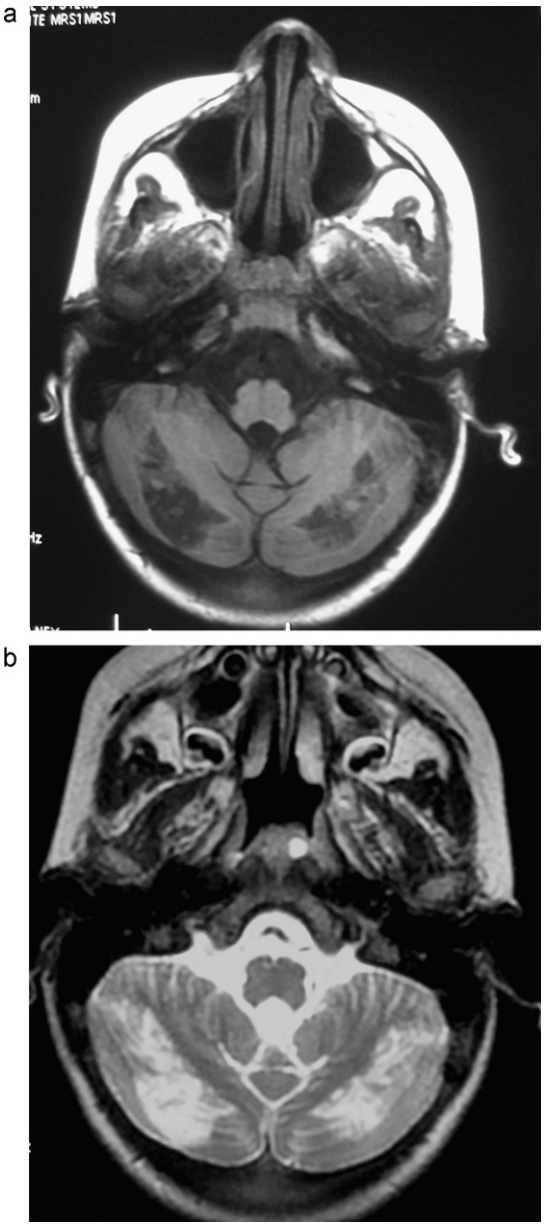


Fig. 3. (a) and (b) MRI, T1 and T2 weighted images: bilateral signal abnormalities in the cerebellar hemispheres.

frontal cyst was responsible of a compression of the frontal horn of the right lateral ventricle (Figs. 4–8). After intravenous injection of Gadolinium, there was an enhancement of the cysts wall and enhancement along the perforating vessels of the white matter (Fig. 6a and b).

Complete blood count, sedimentation rate, liver and renal function tests, serum thyroid and parathyroid hormones, calcium, phosphate, alkaline phosphatase, and lactate levels were within normal limits. Serological tests for cytomegalovirus, toxoplasma gondii, cysticercosis, hydatic cyst, and HIV 1 and 2 were all negative. No serologic confirmatory evidence of parasitic brain infection was found. Regarding the relevant literature, LCC was considered.

Parents of the patient refuse any treatment and the patient was discharged against medical advice.

3. Discussion

Leukoencephalopathy with intracranial calcifications and cysts (LCC) is a new and very uncommon entity that was first described in 1996 in three unrelated children [1] and reported in less than 10 cases so far [4].

The clinical presentation of this brain disorder includes progressive extrapyramidal and pyramidal signs, slowed cognitive performances and seizures [1,2]. This disease is most exclusively observed in early infancy and adolescence but adult onset cases have been reported [2,5,6]. Seizures can reveal LCC but to the best of our knowledge, this is the first report in which LCC was also revealed by a tetraparesis.

Histopathological examination in LCC reveals "angiomatous like rearrangements of the microvessels" with exuberant proliferation of abnormal small vessels associated with Rosenthal fibers, gliosis and calcifications [1,2,4]. Cerebral angiomatous rearrangement of microvessels is the primary pathological feature of LCC while perivascular foci of calcifications, hyaline deposits and formation of Rosenthal fibers appear compatible with secondary changes [1]. The development of parenchymal cysts can lead to compressive complications [2,4].