

Leukoencephalopathy, cerebral calcifications, and cysts

Entity that can mimic a neoplasm

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Leukoencephalopathy with intracranial calcifications and cysts (LCC) is a rare and recently described entity characterized radiologically by white matter abnormalities, calcifications, cysts and enhancing nodules¹. LCC is a disorder of unknown etiology and its neurological manifestations include cognitive decline, seizure, obstructive hydrocephalus, progressive cerebellar, besides extrapyramidal and pyramidal signs with juvenile onset². We report a case of LCC with remarkable radiological and clinical features.

CASE

A 30-year-old man presenting frontooccipital headache for the last 7 days, worsening in the morning, associated with progressive anorexia and vomiting. Eight years ago, he had an acute neurological event characterized by paresis and aphasia of expression. His past medical history demonstrated moderate cognitive impairment since childhood. Family history, were unremarkable and no consanguinity was known. Neurological examination revealed left paresis grade 4+ with positive Babinski sign besides ataxia. Brain MRI (1.5T) revealed an expansive solid / cystic interhemispheric lesion measuring 55×25 mm with important mass effect. T2 im-

aging showed remarkable hemosiderin deposition in its walls. MR perfusion demonstrated high capillary density within the walls. Multiple enhancing nodular lesions were observed involving supra and infratentorial brain parenchyma. Some of these nodules had hemorrhagic component on T2 images. Broad and symmetric calcifications in periventricular white matter, basal ganglia, brainstem and dentate nuclei were observed.

Patient underwent surgery for relieving intracranial hypertension and biopsy. Pathological examination was inconclusive due to the large amount of clotted blood within the material. No neoplastic cells were observed. Patient was discharged in good clinical condition, without signs of intracranial hypertension, remaining hemiparesis and aphasia.

DISCUSSION

This is the first report of MR perfusion findings in a case of LCC. The enhancing walls presented a striking high capillary density that could mislead for an erroneously diagnosis of neoplastic lesion. In fact, this finding may be explained for one of the theories of cerebral microangiopathy as the etiology of this entity. This theory could explain the hemorrhagic content of the lesions.

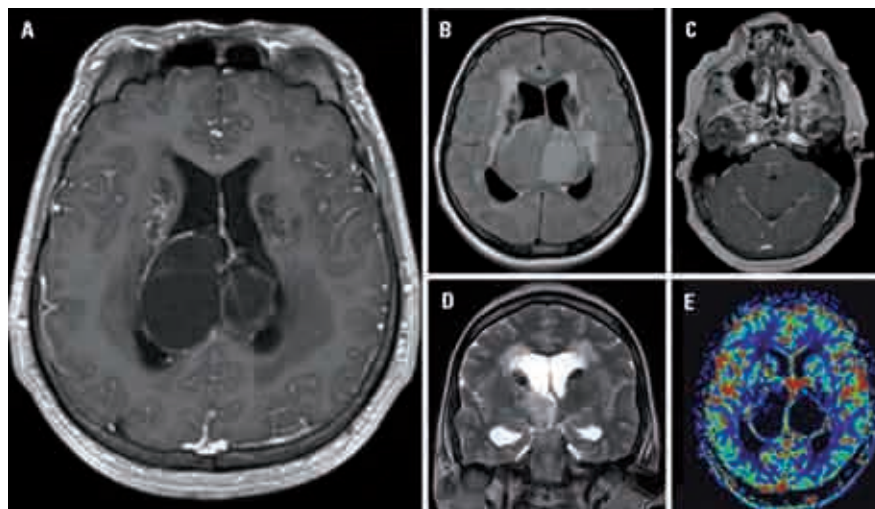


Figure. [A] Axial post contrast T1 weighted MR image shows cystic lesions with impregnating the walls. [B] Axial flair weighted MR image shows leukoencephalopathy cysts and areas with low signal in basal ganglia corresponding to calcifications. [C] On Coronal T2 weighted MR image, calcifications in the basal ganglia with low signal and hemorrhagic lesion protruding to the 3rd ventricle. [D] Nodular lesion with calcification and prominent vessels surrounding the pons on post-contrast T1 weighted MR. [E] MR perfusion demonstrated high capillary density within the walls.

Furthermore high vascular lesions may present high perfusion as well as brain cancer.

The symmetrical calcifications and the diffuse enhancing nodular lesions are other important findings that suggest a diffuse cerebral condition. Actually the nodular enhancing lesions constitute pre-cystic lesions as same cases show in the literature. Actually the leucodystrophy described in this entity is rather related to vasogenic edema and gliosis surrounding the cysts and the nodules¹⁻⁵.

LCC is a very rare disease and less than 30 cases reported since the original description by Labrune and colleagues, all of them with characteristically infantile or juvenile onset¹⁻³. Sener et al. were the first to suggest the existence of an adult form of this disease, based on the observation of a slow clinical progression in a patient with onset in late adolescence⁴. Wargon et al. report a 30-year-old woman with a lacunar infarct as the first manifestation of LCC⁵. In our patient, the diagnosis of LCC was based on the highly suggestive neuroradiological findings which were almost identical to previous report. Our biopsy showed no neoplastic cells, but clotted blood within the lesion.

LCC is an entity can present with cystic expansive lesions with mass effect, hemorrhagic content and high vascular density that can be misunderstood as neoplastic lesions. Symmetrical brain calcifications and dif-

fuse enhancing hemorrhagic nodules are the other crucial findings. These abnormalities points out that this entity should rather be described as a vascular entity in the books than as a primary leucodystrophy.

REFERENCES

1. Labrune P, Lacroix C, Goutieres F, et al. Extensive brain calcifications, leukodystrophy, and formation of parenchymal cysts: a new progressive disorder due to diffuse cerebral microangiopathy. *Neurology* 1996;46: 1297-1301.
2. Kaffengerger T, Valko PO, von Meyenburg J, et al. A case of late onset leukoencephalopathy with cerebral calcifications and cysts in 59-year-old woman. *Eur J Neurol* 2009;16:278-281.
3. Marelli C, Savoriardo M, Fini N, et al. Late presentation of leukoencephalopathy with calcification and cysts: report of two cases. *J Neurol Neurosurg Psychiatry* 2008;79:1303-1304.
4. Sener U, Zorlu Y, Men S, Bayol U, Zanapalioglu U. Leukoencephalopathy, cerebral calcifications and cysts. *Amer J Neuroradiol* 2006;27:200-203.
5. Wargon I, Lacour MC, Adams D, Denier C. A small deep infarct revealing leukoencephalopathy, calcifications and cysts in an adult patient. *J Neurol Neurosurg Psychiatry* 2008;79:224-225.

LEUCOENFALOPATIA, CALCIFICAÇÕES CEREBRAIS E CISTOS: ENTIDADE QUE PODE SIMULAR UMA NEOPLASIA

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Pial arteriovenous fistula in the posterior fossa

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Intracranial pial arteriovenous fistulas (AVFs) are rare congenital abnormality that can cause severe morbidity and mortality, particularly in neonates¹. AVFs are rare cerebrovascular lesions of the brain that were considered to be distinct from other arteriovenous malformations (AVMs) by Lasjaunias, in 1986². Intracranial pial AVFs have a single or multiple arterial connections to a single venous channel. They differ from brain AVMs in that they lack a true nidus. They differ from dural AVFs in that they derive their arterial supply from pial or cortical arteries and are not located within the dura mater³. They may be localized anywhere in the brain but display a preference for the supratentorial regions³.

Pial AVFs can be acquired traumatically or iatrogenically or may be congenital. Their natural evolution is unfavorable, conservative management of pial AVFs has been associated with mortality in 63% of patients⁴.

CASE

A 6-month old boy present macrocephaly, asymptomatic, without apparent motor injury, did not show any skin lesion and no signs of heart failure. A transfontanele ultrasound Doppler showed evidence of large arteriovenous shunt (AVS) at posterior fossa with patent flow (Fig 1A and 1B). Additional investigation with computed tomography (Fig 1C) and magnetic resonance