

## Professor Yamada's warning.....March 1, 2014

I would like to **stress** the following two points when choosing the cases.

#1 The case **should be educational** and it **does not have to be a very rare case**. Rather, a common disease could be more appealing to the audience.

#2 It would be nice if one can **reach the correct conclusion in theoretical fashion**. The case wouldn't attract the audience if it is just a "gamble" to get the correct diagnosis.



## Professor Yamada's warning.....March 1, 2014

I would like to **stress** the following two points when choosing the cases.

#1 The case **should be educational** and it **should be** **practical**. In other words, a common disease could be more appealing to the audience.

#2 It would be nice if one can **reach the correct conclusion in theoretical fashion**. The case wouldn't attract the audience if it is just a "gamble" to get the correct diagnosis.



**I am afraid, this case against all rules : )**

## Chief Complaint:

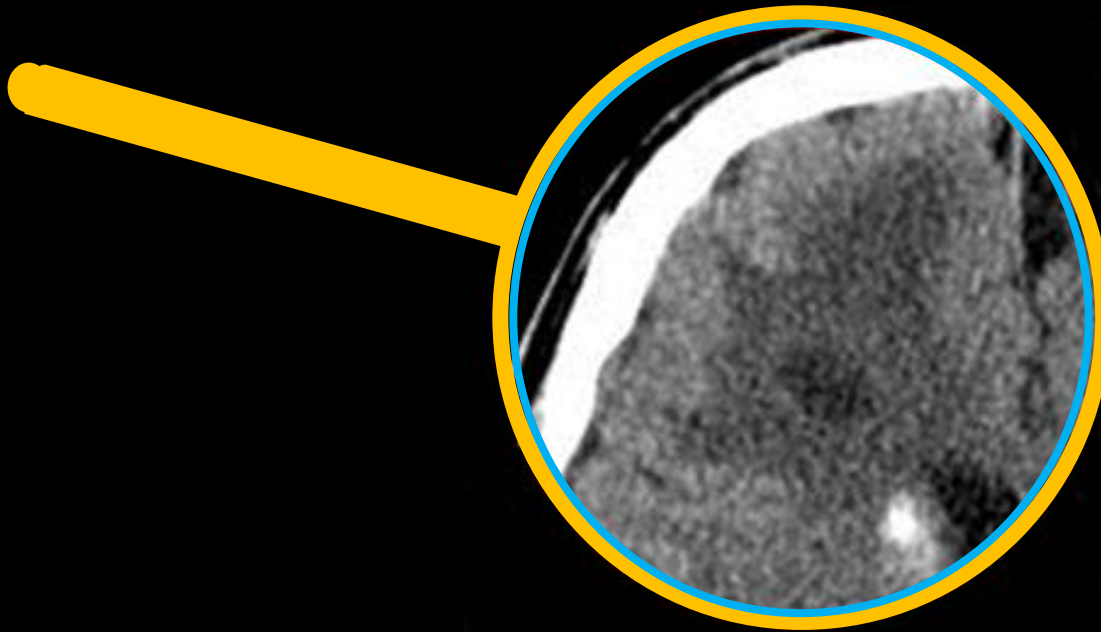
44-year-old man with headache and unstable gait for 2 weeks. Prior history of brain surgery on 2009 and 2012 for cerebellar and frontal lobe masses, respectively.



07/13/2009



07/13/2009



**White matter lesions**  
**Cyst?**  
**Calcification**

Google

white matter lesions cyst calcification

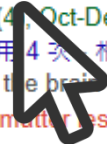


### Leukoencephalopathy with intracranial calcifications and ...

[www.ncbi.nlm.nih.gov](http://www.ncbi.nlm.nih.gov) > ... > v.13(4); Oct-Dec 2010 ▾ 翻譯這個網頁

由 K Ummer 著作 - 2010 - 被引用 4 次 - 相關文章

Magnetic resonance imaging of the brain showed multiple bilateral cerebral **cystic lesions** along with diffuse **white matter lesions** in the cerebral and cerebellar ...



### A case of leukoencephalopathy, cerebral calcifications and ...

[www.ncbi.nlm.nih.gov](http://www.ncbi.nlm.nih.gov) > ... > v.14(4); Oct-Dec 2011 ▾ 翻譯這個網頁

由 A Gulati 著作 - 2011 - 被引用 4 次 - 相關文章

A supratentorial **cystic lesion** with blood fluid level was seen in left occipitotemporal region. MRI examination revealed diffuse symmetric **white matter** ...



### Full Text - American Journal of Neuroradiology

[www.ajnr.org/content/27/1/200.full](http://www.ajnr.org/content/27/1/200.full) ▾ 翻譯這個網頁

由 U Sener 著作 - 2006 - 被引用 21 次 - 相關文章

The supratentorial **lesions** were predominantly located in the subcortical **white matter**. Some of the **calcified lesions** were associated with **cystic** formations (Fig ...



### [PDF] Leukoencephalopathy, cerebral calcifications, and cysts

[www.scielo.br/pdf/anp/v69n4/a27v69n4.pdf](http://www.scielo.br/pdf/anp/v69n4/a27v69n4.pdf) ▾ 翻譯這個網頁

由 MM Bertotti 著作 - 2011 - 被引用 1 次 - 相關文章

characterized radiologically by **white matter** abnormal- ... an expansive solid / **cystic** interhemispheric **lesion** mea- ... ular **lesion** with **calcification** and prom-



### [PDF] The Rare Association of Leukoencephalopathy ... - SciELO

[www.scielo.br/pdf/jecn/v16n3/v16n3a06](http://www.scielo.br/pdf/jecn/v16n3/v16n3a06) ▾ 翻譯這個網頁

由 GR Isolana 著作 - 2010 - 被引用 2 次 - 相關文章

cerebral **calcifications**, and **cysts** (LCC) is a very rare entity described in 1996.5 We present a new case of LCC ... initial CT showed numerous **foci** of **calcifications** scattered through the ... lobes **white matter** whereas the cortical gray matter was.



You can't miss it!

Ann Indian Acad Neurol. 2010 Oct-Dec; 13(4): 299–301.

PMCID: PMC3021936

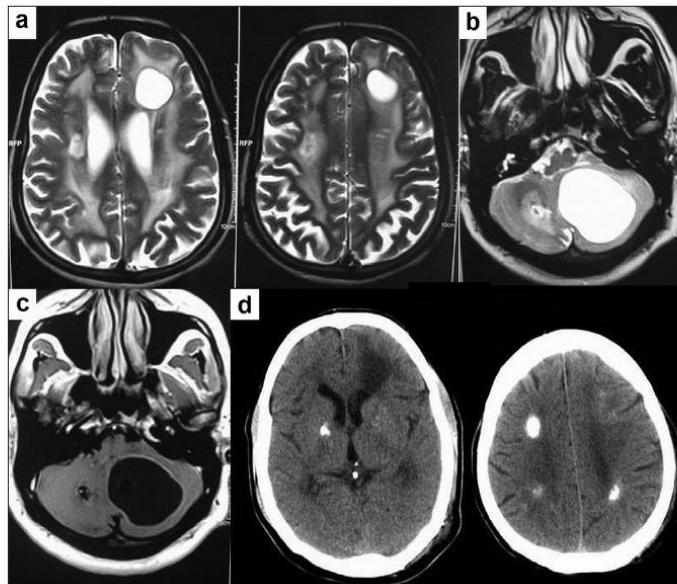
doi: [10.4103/0972-2327.74198](https://doi.org/10.4103/0972-2327.74198)

## Leukoencephalopathy with intracranial calcifications and cysts in an adult: Case report and review of literature

[K. Ummer](#), [K. A. Salam](#), [Mohan L. Noone](#), [V. G. Pradeep Kumar](#), [Neena Mampilly](#),<sup>1</sup> and [S. Sivakumar](#)

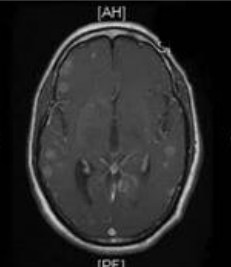
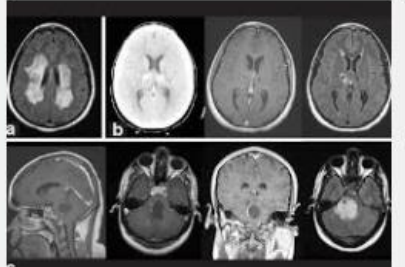
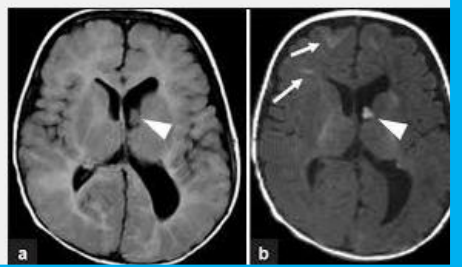
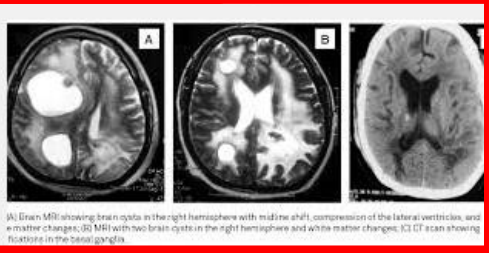
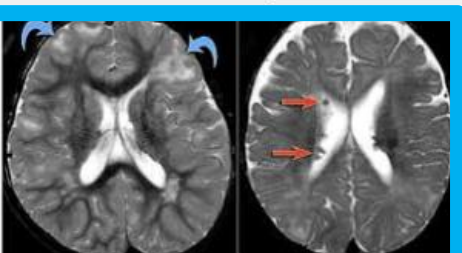
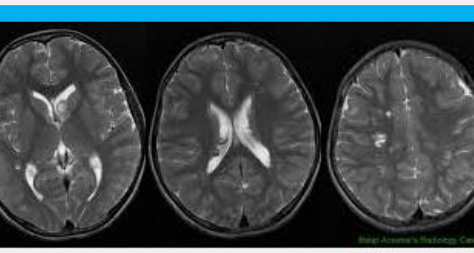
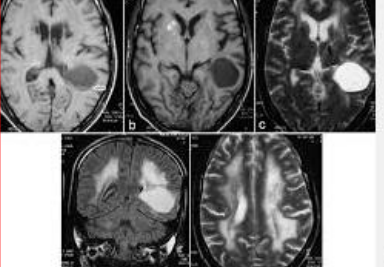
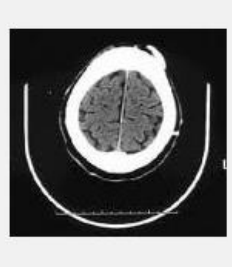
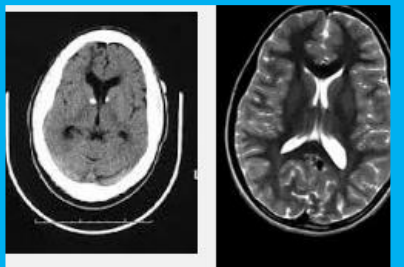
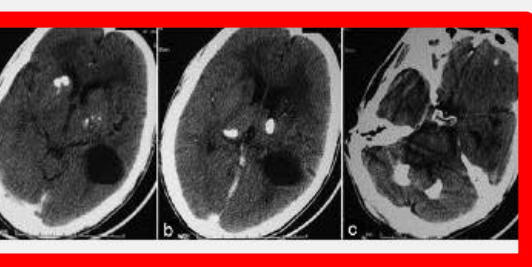
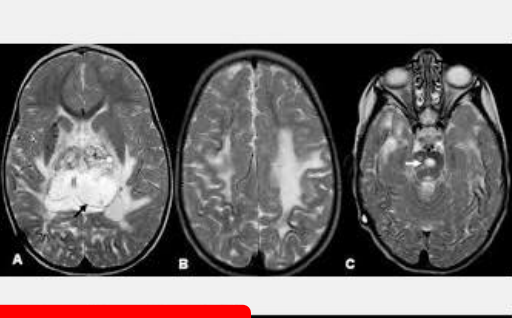
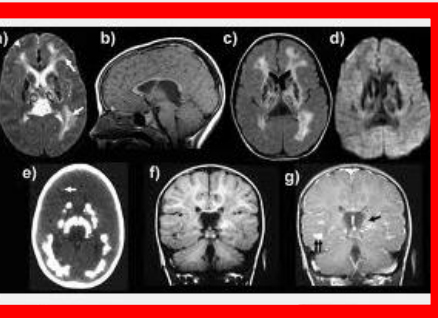
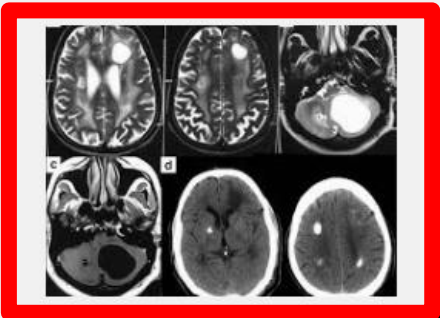
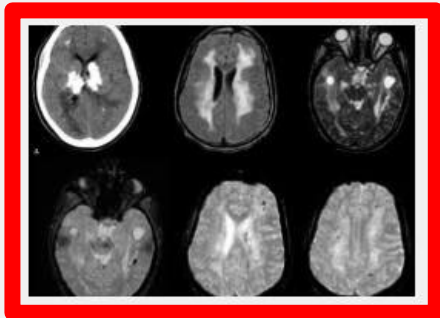
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Figure 1



**This is only  
the first click!**

**The diagnosis  
is embarrassingly  
quick!**



**Standard gears!**

**Have you ever been Classic recently?**



# Working lists

1. Nature history of LCC
2. Pathological findings
3. Differential diagnoses



## Natural history of LCC

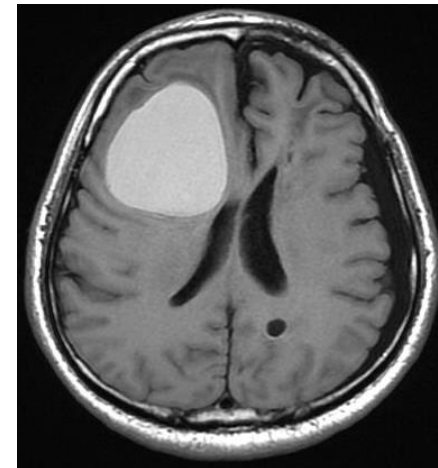
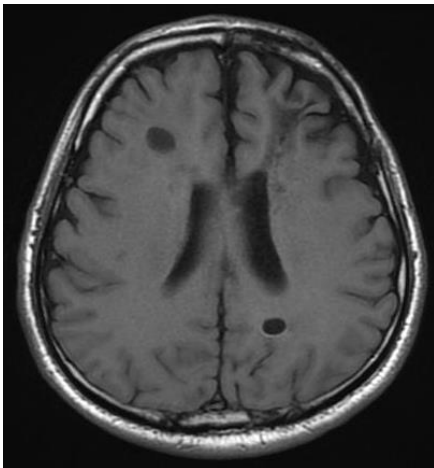
1. Early (**childhood**) onset; adult type
2. Progressive **calcifications** in the BG, Cerebellar gray nuclei, central white matters
3. **White matter** dystrophy
4. Development of **parenchymal cysts** in the cerebellum and cerebrum that may require surgical decompression -- **localized necrosis and progressive osmotic fluid accumulation.**

# The Cyst

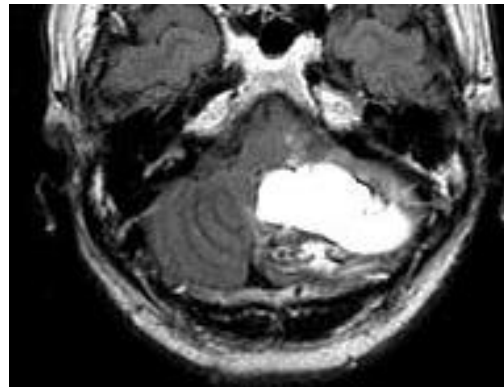
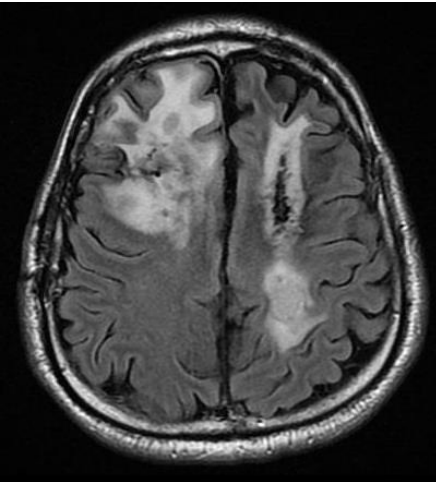
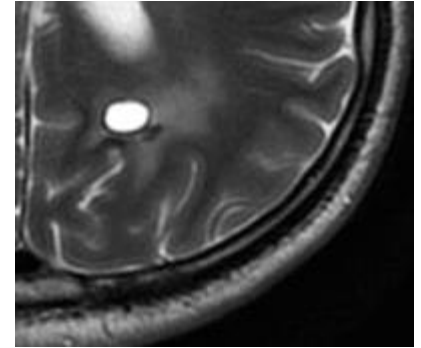
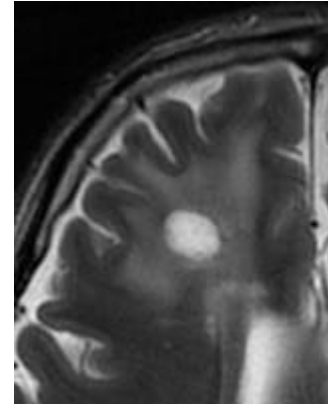
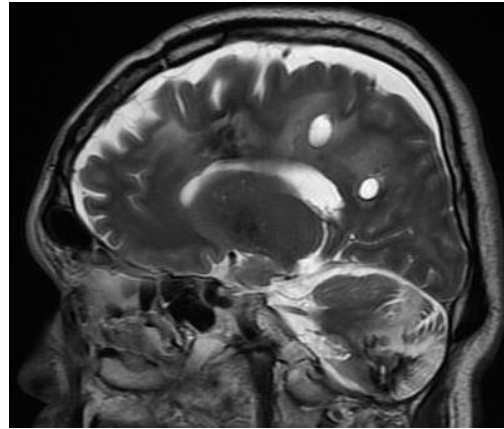
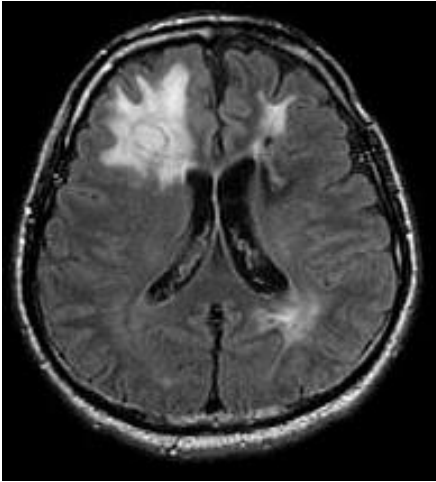
July  
2009



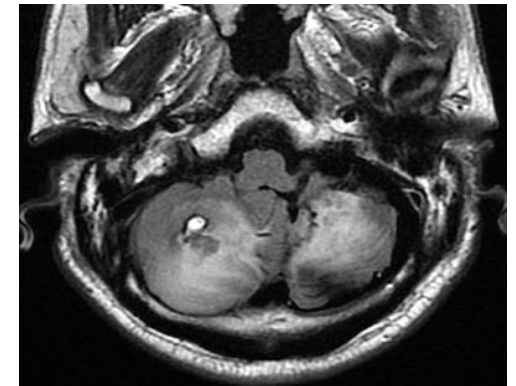
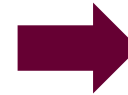
Dec.  
2012



# The cysts and leukodystrophy

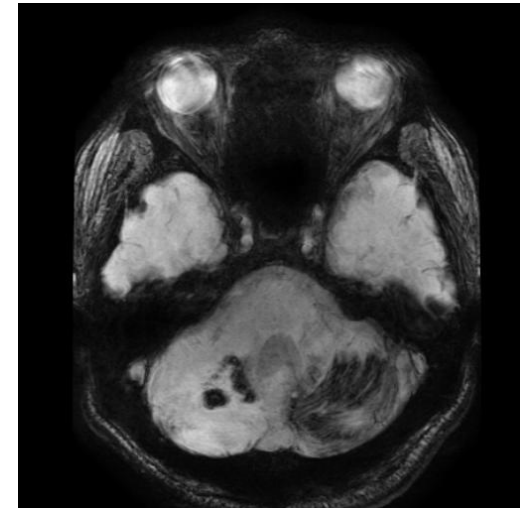
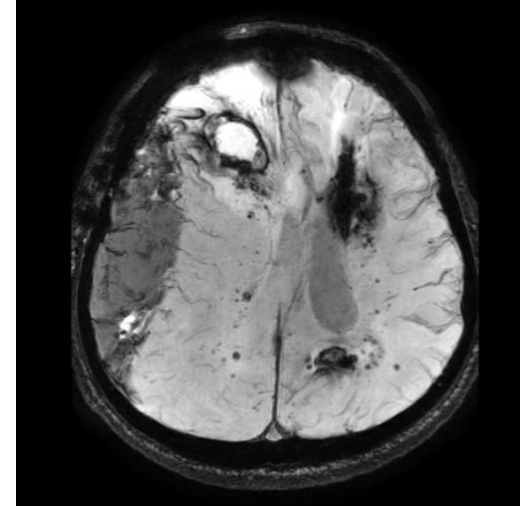
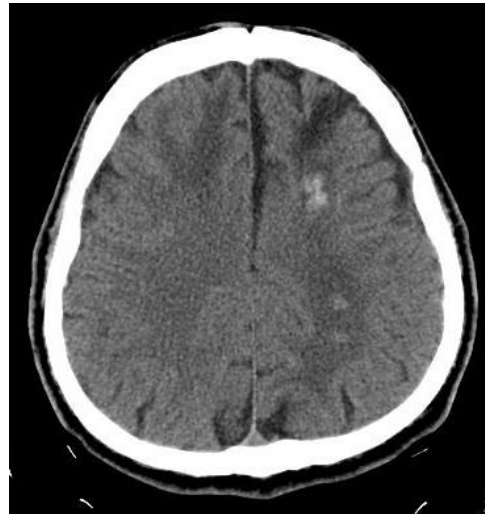
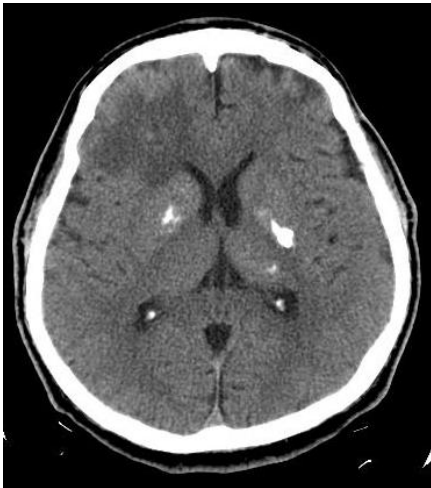


July 2009



April 2014

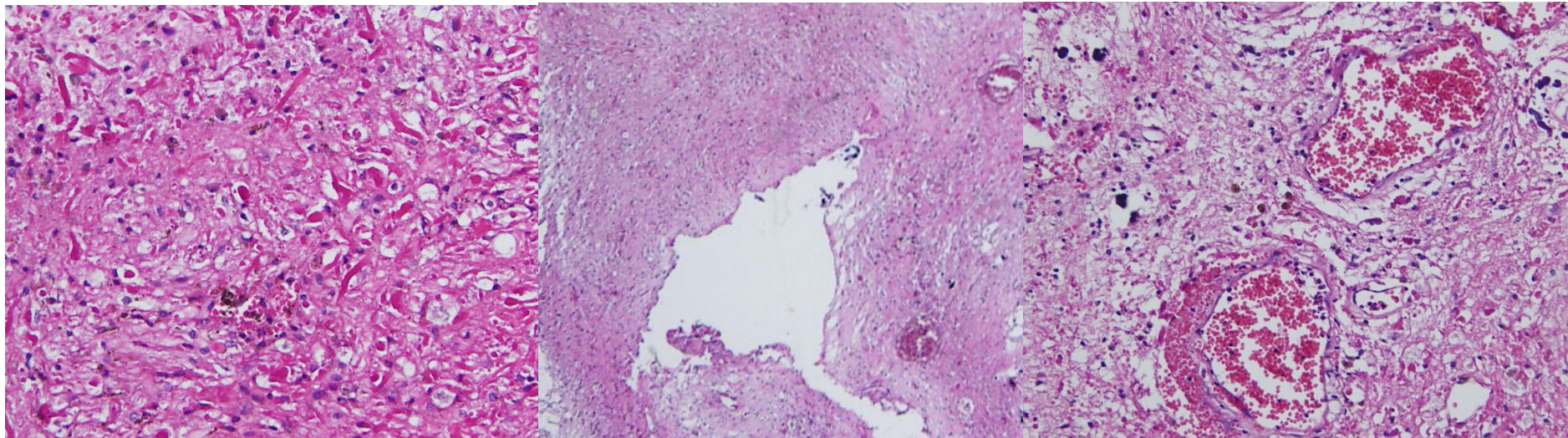
# Calcifications and hemosiderin



# Histopathology of LCC

- Calcifications; hemosiderin
- Cysts
- Angiomatous lesions
- Rosenthal fibers; astrogliosis

Labrune 1996  
Wang 2013



**Rosenthal fibers**  
**Hemosiderin**

**Brain cysts**

**Angiopathy**  
**Calcifications**

# Differential diagnoses

1. Toxoplasmosis
2. Neurocysticercosis
3. CMV infection
4. Tuberosus sclerosis complex
5. Inherited metabolic disease
6.
  - a. Leukodystrophy, calcification, and cysts
  - b. Coats plus syndrome (CTC1 gene mutation)
  - c. Cerebroretinal microangiopathy with calcifications and cysts – mixed LCC and Coats plus syndrome.

**Table I.** Clinical Overlapping Features of Patient and Syndromes.

	Patient	ND Bilateral involvement	Coats Unilateral involvement	Coats plus Bilateral involvement	FEVR Bilateral involvement	LCC Bilateral involvement	CRMCC Bilateral involvement
<b>Ophthalmologic findings</b>							
Exudative retinopathy	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Retinal telangiectasias	Yes	Yes	Yes	Yes	Yes	No	Yes
<b>Brain MRI</b>							
Cerebral calcifications	Yes	No	No	Yes	No	Yes	Yes
Leukoencephalopathy	Yes	No	No	Yes	No	Yes	Yes
Intracranial cysts	Yes	No	No	Yes	No	Yes	Yes
Cerebellar atrophy/ hypoplasia	No	One case	No	No	No	No	Sometimes
<b>Neurologic symptoms</b>							
Developmental delay	Yes	Yes	No	Yes	No	Yes	Yes
Pyramidal/cerebellar signs	Yes	One case	No	Yes	No	Yes	Yes
Mental retardation	Yes	Yes	No	Yes	No	Yes	Yes
Seizures	Yes	Yes	No	Yes	No	Yes	Yes
Sensorineural hearing loss	No	Yes	No	Yes	No	Yes	Yes
<b>Other abnormalities</b>							
IUGR and growth retardation	Yes <sup>a</sup>	Sometimes	Sometimes	Yes	No	Yes	Yes
Sparse hair	No	No	No	Yes	No	No	Yes
Nail dystrophy	No	No	No	Yes	No	No	Yes
Skeletal defects	No	No	No	Yes	No	No	Yes
Gastrointestinal bleeding	No	No	No	Yes	No	No	Yes
Liver failure	No	No	No	Yes	No	No	Yes
Hematologic abnormalities	Yes	No	No	Yes	No	No	Yes

Google

kyoto



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# Thank you for kind attention

