Leukoencephalopathy with calcifications and cysts

Herein, we report a case of LCC. A 16 yr female presenting with headache vominting, seizures, loss of balance since 20 days.

MRI brain with contrast reveals, multifocal dense asymmetric areas of blooming in bilateral capsuloganglionic regions, thalami, right cerebellum, part of left centrum semiovale associated with extensive diffuse hyperintensities involving the bilateral frontoparietal, temporal subcortical white matter, centrum semiovale and an approximately 2.7x 2.5 x 2.5 cm round cystic lesion in the right cerebellum with thin enhancing walls, eccentric wall calcification and perilesional vasogenic edema.

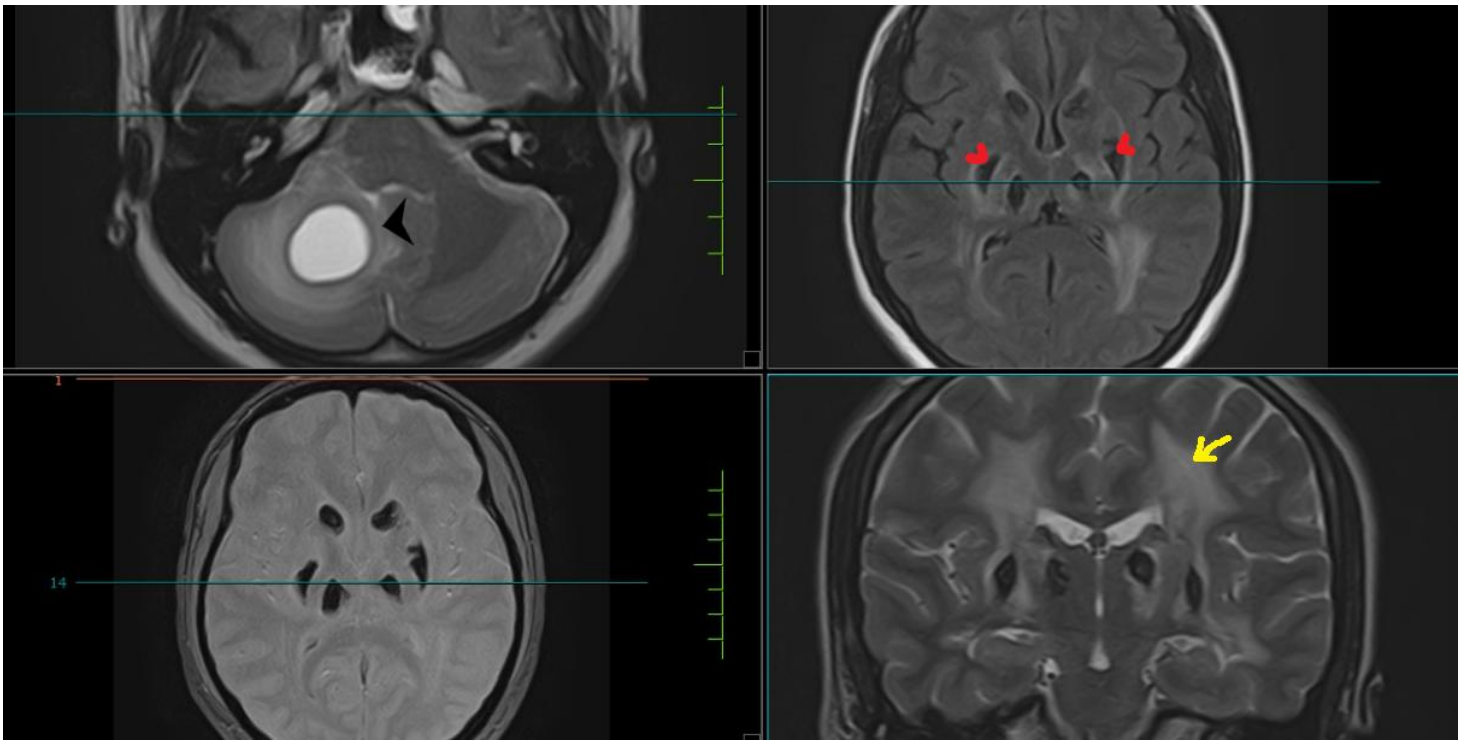


Fig: Dense asymmetric calcifications (red arrowheads) in bilateral capsuloganglionic regions, thalami, right cerebellum, part of left centrum semiovale . Extensive diffuse hyperintensities involving the bilateral frontoparietal, temporal subcortical white matter, centrum semiovale (yellow arrow) A rounded cystic lesion in the right cerebellum with thin enhancing walls, eccentric wall calcification and perilesional vasogenic edema (black arrowhead)

Leukoencephalopathy with calcifications and cysts

Introduction:

Leukoencephalopathy with calcifications and cysts (LCC) is a rare entity that is radiologically characterized by edematous leuoencephalopathy, cerebral calcifications, and formation of parenchymal cysts.[1–3] , also known as Labrune syndrome.[4]

Epidemiology

A rare entity, a small number reported in the literature [2,5]. Clinical onset can occur early infancy to late adulthood with female predeliction.

Clinical presentation

Headache, epileptic seizure, ataxia, cognitive dysfunction, papilledema, dystonia, and visual disturbance.

Etiopathology

Associated with mutation in SNORD118 gene

Pathophysiology : diffuse cerebral microangiopathy with early micro cystic disease followed by large cystic whitematter degeneration [1].

Histological features Renshaw cell proliferation, Rosenthal fibers and glial proliferation , encountered including demyelination, necrosis

CT

Multifocal progressive calcifications, involving basal ganglia and cerebellum and sub cortical white matter, with diffuse low attenuation involving the white matter associated with expansile cystic areas.[5].

MRI

MRI is the modality of choice for whitemmater disease and cysts [3,5].

Cysts predominantly involving corona radiate, centrum semiovale, capsuloganglionic regions , cerebellum which are hypointense on T1, hyperintense on T2 without restriction of diffusion. Post contrast scan may show rim enhancement of the cysts.

Leukoencephalopathy : diffuse T2 hyperintensity involving the whitematter sparing subcortical U fibers. [3]

- Calcification seen as blooming on GRE /SWI sequences in basal ganglia, cerebellum.

MR spectroscopy of the cysts may demonstrate a lactate peak and no evidence of the normal metabolites of the brain .