Late onset of leucoencephalopathy with cerebral calcifications and cysts

Début tardif d’une leucoencéphalopathie calcifiée et kystique

We report on the case of a 69-year-old Caucasian woman who was hospitalized for confusion, ataxia, gait unsteadiness, repetitive falls and left homonymous hemianopsia that had been progressing over several months. Her past medical history was noteworthy because of a concussion 14 years earlier, at which time, computed tomography (CT) and magnetic resonance imaging (MRI) showed cerebral calcifications and leukoencephalopathy. The patient had been asymptomatic and lost to follow-up until her retirement. Her family history revealed stroke in first-degree relatives.

Neurological examination on admission revealed a right parieto-occipital syndrome, left facial paresis and left hyperreflexia. Cerebral CT and MRI showed calcifications in the thalamus, basal ganglia, deep white matter, brain stem and dentate nucleus (Fig. 1). The associated cystic lesions were most prominent in the right medial temporal region. T2-weighted and FLAIR signals revealed extensive white-matter hyperintensities (Fig. 2). A cerebral biopsy, which included a cyst, was performed to exclude a metastatic lesion. Histopathological examination showed organizing hemorrhagic tissue with gliosis, although no angiomatous or hyalinized blood vessels, myelin loss, calcifications or Rosenthal fibers were identified. Oncological, biochemical, ophthalmological and hematological workups were all normal. Serological tests for cytomegalovirus, toxoplasmosis, Echinococcus and cysticercosis were also negative.

Leukoencephalopathy with calcifications and cysts (LCC) is an extremely rare disease. It is primarily described in childhood and adolescence as presenting with neurological impairment of relatively rapid progression and intracerebral hemorrhage that frequently leads to death [1]. Systemic involvement is commonly seen, and includes hematological and ophthalmological abnormalities [1,2]. Only six cases of adult-onset LCC have been reported in the literature so far [2–4] and, in most of these cases as in ours, there is no

Figure 1   Brain CT scan shows asymmetrical calcifications in the thalamus (a), dentate nucleus (b), basal ganglia and deep white matter (not illustrated here).
Figure 2  Axial T2-weighted MRI shows cysts with calcifications in the dentate nucleus and thalamus (a and b, arrows), associated with large areas of white-matter hyperintensities (c, arrows) and small calcifications in the deep white matter (c, arrowheads).

mention of early neurological impairment during infancy or any systemic features. Thus, we can hypothesize that the adult form presents as a much more indolent and benign disease.

CT and MRI both offer clues to the diagnosis by demonstrating the unique pathognomonic triad that includes cysts, leukoencephalopathy and asymmetrical calcifications [5]. On histology, the key feature of LCC is a proliferative small-vessel angiopathy with Rosenthal fibers [1]. In our case, only non-specific hemorrhagic changes were seen, as the cyst was damaged during the sampling procedure.

To our knowledge, our patient is the oldest of all cases reported so far to present with the onset of LCC. Furthermore, this patient had no neurological dysfunction and was professionally active until her retirement. Thus, this case supports the possibility of a long-lasting asymptomatic form of LCC. As evidenced by the CT and MRI scans of our patient taken 14 year earlier, the disease may remain latent for years. The cysts developed slowly, over decades, and the diagnosis was made at the time of intracystic hemorrhage, a highly unpredictable event. This may explain the wide interindividual variability among patients with LCC. Diagnostic imaging and follow-up are essential, while brain biopsy can be avoided as the neuroimaging triad is highly characteristic.

Conflict of interest statement

The authors report no conflicts of interest.

References


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Primary dural lymphoma with vault involvement mimicking meningioma

Lymphome dural primitif avec envahissement de la voute simulant un méningiome

A 25-year-old immunocompetent man presented with frontal headaches, vomiting and left hemiparesis. Magnetic