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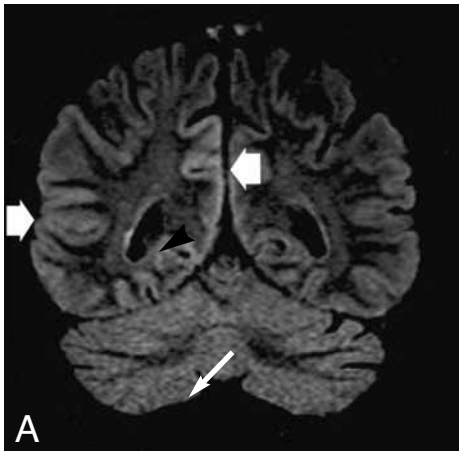
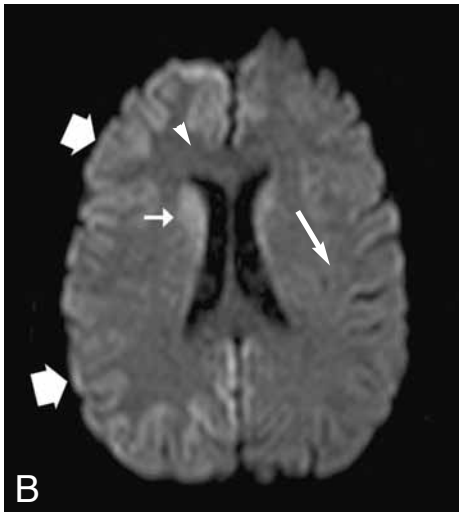
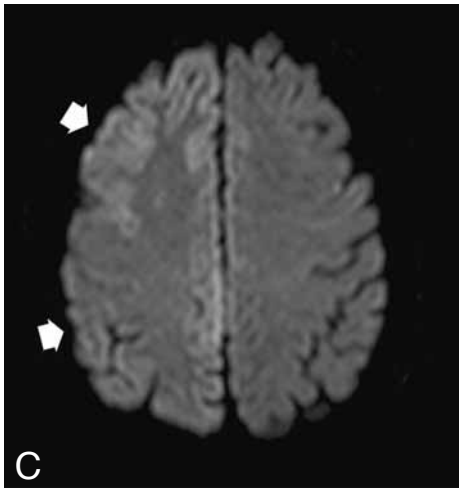
IMAGES IN CLINICAL RADIOLOGY







Fast progressive memory loss in a 63-year-old man

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A 63-year-old man presented to the neurology department with fast progressive memory loss especially short term memory. For 2 weeks he had experienced loss of orientation, judgment difficulties, and concentration problems. A CT scan of the brain was normal.

MRI showed normal findings on T1 and T2 weighted images.. There was no contrast enhancement. On diffusion weighted images hyperintense signal was seen at the cortex of the right hemisphere and in the basal ganglia. On PET regional hypometabolism of glucose correlated with sites of increased intensity on diffusion weighted images. An electroencephalogram showed periodic synchronous bi- or triphasic sharp wave complexes. Laboratory findings showed a positive CSF 14-3-3 immuno-assay. Given these findings the diagnosis of sporadic Creutzfeldt-Jacob disease was suggested. Dementia progressed rapidly and the patient subsequently developped myoclonic jerks. The patient died of pneumonia one month later. Brain biopsy confirmed the diagnosis of Creutzfeldt Jacob.

Comment

Creutzfeldt-Jacob disease is a rapidly progressive fatal form of dementia caused by a prion. It is potentially transmittable. Sporadic CJD occurs most commonly in the elderly, Spontaneous CJD occurs throughout the world, and there is no male or female predilection. CJD can be inherited (familial form), sporadic (sCJD), or acquired (nvCJD). The familial form of CJD is related to mutations in the PRPN gene on chromosome 20. Acquired CJD is related to an infection from prion containing materials such as surgical instruments, cadaveric dura mater grafts, EEG electrodes, corneal transplant, and pituitary hormones such as growth hormone. A bovine source can also be a cause (infected beef products). The clinical manifestations differ according to the stage of the disease. At onset there is fatigue, visual disturbance, depression, and insomnia. After a few weeks there is rapidly progressive mental deterioration with dementia. As the disease progresses periodic synchronous discharges occur on the EEG and myoclonus may be evident a quite typical finding in CJD. Death usually occurs within 1 year of onset due to a respiratory tract infection. CSF protein 13-3-3 immunoassay is a useful diagnostic test although this protein may be present in other disorders. A brain biopsy is a highly accurate method of diagnosis. On conventional MR high signal intensity in the cerebral cortex and basal ganglia may be apparent on T2 weighted images. Nevertheless with conventional MR imaging findings are difficult to visualize. FLAIR images tend to reveal the cortical abnormalities better.

DWI are the cornerstone for early diagnosis of CJD showing areas of high signal in the cortex and in the basal ganglia and thalamus. These imaging findings are accompanied by decreased ADC values suggestive of restricted diffusion within the tissue due to vacuolisation.

Differential diagnosis include MELAS, venous hypertensive encephalopathy and chronic herpes encephalitis.

At present there is no treatment for CJD but the early changes on DWI may prove very valuable once a treatment becomes available.

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