CADASIL

MRI Films and results

For CADASIL Together We Have Hope Non Profit Organization (Foundation) to assist doctors in understanding CADSAIL
Figure 1. MRI in two CADASIL patients
MRI in two patients with cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy illustrating the variable extent of lesions seen on T1-weighted (A, C) and T2-weighted (B, D) images. (A, B) A 30-year-old woman with migraine with aura (Rankin Scale disability grade, 0; Mini-Mental State Examination score [MMSE], 30). (C, D) A 57-year-old woman with a history of migraine with aura, stroke, and cognitive decline (Rankin Scale disability grade, 3; MMSE score, 24).

MRI: cerebral micro-bleeds - caution in the use of anti-coagulants

Dichgans et al. Stroke 2002; 33:67-71
Fig. 1. Neuroimaging studies. Axial T2-weighted MR images (A, B, C and D) show 1) diffuse confluent ischemic changes in periventricular and subcortical white matter, 2) lacunes in the basal ganglia, thalamus, and brainstem, and 3) abnormal white matter hyperintensities in both temporal pole areas (arrow). A diffusion-weighted MR images (E) shows high signal intensity, suggestive of recent infarction, in the right corona radiata. FDG-PET scans (F, G, H, and I) obtained with the same angle and slices as in MRI show an abnormally decreased glucose metabolism bilaterally in fronto-parieto-temporal cortices, basal ganglia, and thalamus, more markedly in the right hemisphere than in the left.
Male has been diagnosed with CADASIL since 1997 –
Results of three MRI’s

MRI - February 5th, 2004
JACKET NO.1 01204.
Male – 47.5 age - MRI results 2/5/04 - Open field strength MRI of the brain performed with and without gadolinium.
FINDINGS; Comparison to November of 2001 ARE MRI of the brain. Again identified is much greater than expected white matter signal hyperintensity, both supficial and deep white matter, confluent, anterior and posterior cerebral hemispheres, extending to the level of the convexity superiorly, and inferiority to the level of the occipital and temporal horns. There is cortical medullar junction involvement, without clear evidence of full thickness cortical signal abnormality. These findings are nonspecific. Differential considerations include severe advanced small vessel ischemic change secondary to hypertension and/or diabetes or other etiology of vasculopathy, history of severe watershed ischemic injury, chronic demyelination, as well as other less likely etiologies of white matter disease.
Clinical correlation is requested. Signal hyperintensity in the brainstem is greater than expected, amorphous and predominantly in the pons and to a lessor extent the medulla, without mass effect. None of the abnormal signal in the brainstem or supratentorial white matter has mass effect. The ventricles are normal. There is remarkably little atrophy for the degree of white matter disease, which was present on the comparison study, and has not changed significantly in the interval in the supratentorial white matter, although there has been a significant increase in abnormal signal involving the brainstem, particularly the pons. In light of this interval change, other etiologies of the abnormal signal need to be considered including encephalitis (including ADEM) as well as central pontine myelinolysis. However, both of these possibilities are thought less likely given the chronic appearance of the supratentorial white matter signal abnormality, as well as the lack of any mass effect. No pathologic enhancement of brain parenchyma or extraaxial space is identified. No pathologic extraaxial fluid or mass. Flow voids intact. No focal mass effect. Basal ganglia and thalami intact. Mastoid air cells are free of disease. paranasal sinuses are free of air-fluid levels with circumferential moderate inflammatory change right maxillary antra. Mild-moderate inflammatory left maxillary antra and scattered ethmoid air cell opacification, left more than right is present. Additional, mild mucoperiosteal thickening of left more than right frontal sinus is present. A small amount of inflammatory change left $phenoid sinus is present. Orbits are unremarkable. Calvarium, skull base. sella and pituitary gland are unremarkable.
IMPRESSION:
1. INTERVAL INCREASE (SINCE NOVEMBER 2001) IN BRAINSTEM SIGNAL ABNORMALITY, WITH NO INTERVAL CHANGE IN THE AMOUNT OF WHITE MATTER SIGNAL HYPERINTENSITY OF SUPRATENTORIAL BRAIN PARENCHYMA. BOTH THE BRAINSTEM AND SUPRATENTORIAL WHITE MATTER HAS MUCH GREATER THAN EXPECTED SIGNAL ALTERATION WITHOUT MASS EFFECT NOR PATHOLOGIC ENHANCEMENT. DIFFERENTIAL CONSIDERATIONS ARE DISCUSSED ABOVE. CLINICAL CORRELATION IS REQUESTED. GLIOSIS IS THOUGHT MOST LIKELY RADIOGRAPHICALLY.
2. NO CORTICALLY BASED ISCHEMIC INJURY DETECTED.
3. MULTIFOCAL PARA NASAL SINUS INFLAMMATORY CHANGE WITH NO AIR- FLUID LEVELS.
Male has been diagnosed with CADASIL since 1997 –

**MRI May 10, 2000**

RESULTS FOR MRI DONE ON 5-10-00 AND BELOW THIS IS
THE RESULTS OF THE FIRST MRI.
Exam Date: 5-10-00
MRI of the Brian without and with contrast:
Indication: 43 years old male with CADASIL (cerebral autosomal dominant arteriopathy with strokes, eye) and leukoencephalopathy who has dementia, headaches, double vision and eye pain; evaluate for acute infarct.
Procedure: At 1.5 Tesla, T1 weighted sagittal, oblique axial diffusion weighted, Flair and T2 weighted axial, T2 weighted coronal and post contrast T1 weighted axial images were obtained.
Observations: There are moderately severe patchy areas of abnormal signal intensity in perventricular and subcortical white matter bilaterally in a relatively symmetric fashion. No abnormal enhancement is identified accompanying these lesions. There are a few small foci of abnormal signal in the mid aspect of the upper pons in the central aspect of the upper pons which may represent additional small vessel disease. No other areas of abnormal signal intensity are noted. No abnormal contrast contour without evidence of mass effect or shift. No abnormal extra axial collections are identified. Diffusion weighted images show one are of mild hyperintensity, less than would be expected for an acute infarct within high fronto-parietal subcortical white matter. This probably represents a T2 effect.
There is mild mucosal thickening noted in the right frontal sinus in both maxillary antra. The other paranasal sinuses appear well aerated the grossly clear. The orbits and mastoids appear grossly unremarkable. The 7th and 8th nerve complexes appear grossly intact.
Impression:
Moderate severe leukoencephalopathy in a relatively symmetric pattern with some small areas of abnormal signal in the central aspect of the mid to upper pons. Diffusion weighted images who one area of borderline positively for infarct within high fronto-parietal subcortical white matter as discussed. I do not believe that this presents an acute or subacute infarct.
No mass, cortical infarct or hemorrhage is seen. 5-10-00 1:10 PM Film # - 70058642262
Male has been diagnosed with CADASIL since 1997 –

MRI – Male – 40 year old – date of MRI December 7, 1996
with Gadolinium Enhancement Results

DIAGNOSIS: Extensive abnormally in both temporal poles and in deep and subcortical white matter in both hemispheres is not diagnostic in appearance. Some considerations would include post traumatic gliosis, Multiple infarctions such as might be associated with Vasculitis, or unusual or opportunistic infection. The appearance would be Atypical for demyelinating disease.

Comment: A total of seven sequences utilizes protein density, T2, and inversion recovery images, and T1 images obtained before and immediately following the intravenous infusion of Magnevist. An CT scan of November 23, 1996 was reviewed. Extensive, confluent regions of abnormally increased T2 intensity are present in deep white matter in both cerebral hemispheres, contiguous with the lateral ventricles. Smaller areas of abnormal intensity are present in subcortical white matter in the temporal and parietal regions bilaterally. Ill-defined areas of abnormal intensity involve both gray and white matter in the temporal poles. Multiple small rounded foci increased intensity are present in the lenticular nuclei bilaterally, and in the head of the right caudate nucleus. Some of these larger lesions display diminished intensity on TI weighted images. No abnormal enhancement is demonstrated. The brain stem and cerebellum appear normal in intensity. No mass effect is demonstrated. The internal carotid and basilar arteries appear grossly patent. Minimal mucosal thickening is present in the ethmoid sinuses bilaterally and in the left frontal sinus. A 1.5 mm ovoid structure in the inferior right maxillary sinus may represent lobulated tissue in the floor of the right maxillary sinus may represent mucosal thickening or small cysts or polyps. No abnormalities visualized in the posterior nasopharynx or skull base. The pituitary is normal. Orbits and petrous ridges are symmetrical and normal in appearance. DD 12/9/96