Radiology Quiz

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Clinical History

A 33 year old male was admitted for acute onset of right hemiplegia. He has suffered for 4 years from recurrent episodes of hemiparesia and cranial nerve palsies. The erythrocyte sedimentation rate was raised at 70mm in the first hour. A favorable clinical issue was usually obtained within a few weeks on steroid therapy. The following images show: transverse brain computed tomography scan (CT) before and after contrast injection (Figure 1) performed at the time of the first clinical attack, transverse brain magnetic resonance imaging (MRI) performed at the last clinical onset (3 years after the brain CT).

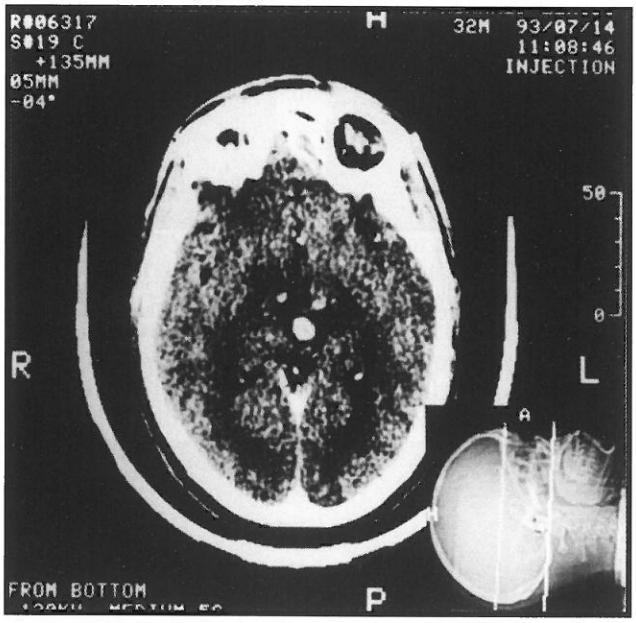

Figure 1 - Post contrast transverse CT scan. Nodular contrast enhancement in both thalamic nuclei.

What are the imaging findings?

What are the diagnostic possibilities?

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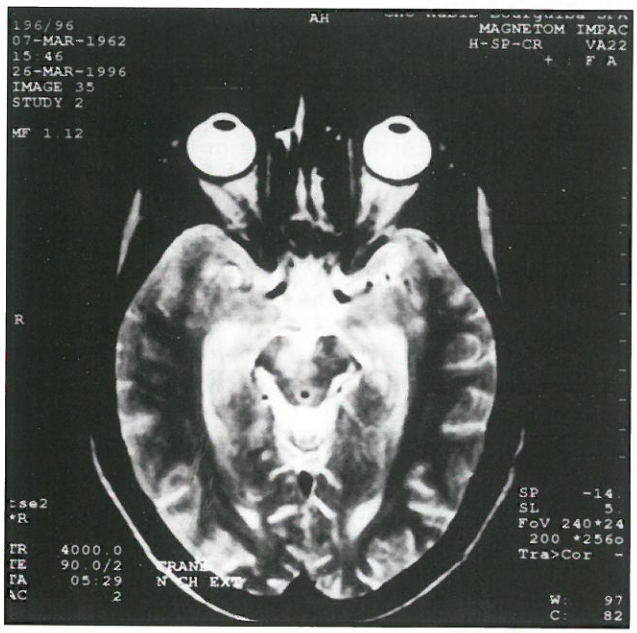

Figure 2 - T2 weighted transverse MR scan of cerebral penduncles. Multiple hypersignal in both penduncles, more on the right side.

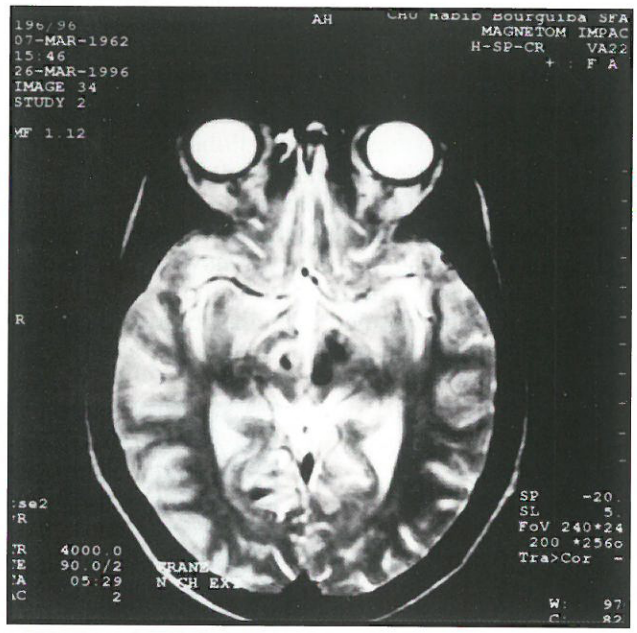

Figure 3 - T2 weighted transverse MR scan of the basal ganglia. Three small nodular areas of low signal corresponding to foci of hemosiderin deposits denoting old hematomas.

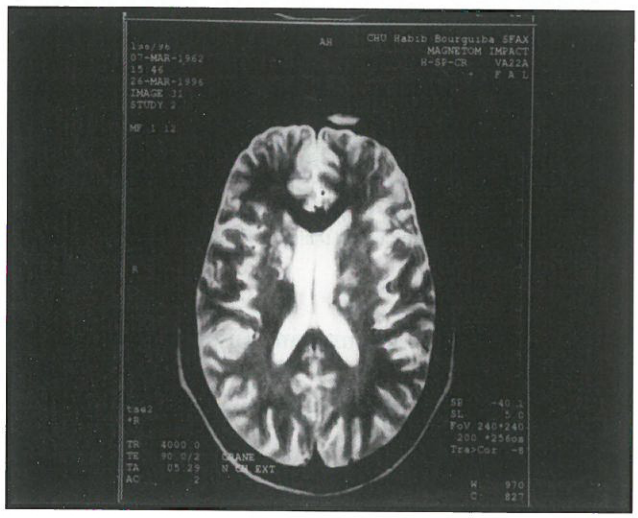

Figure 4 - T2 weighted transverse MR scan of the lateral ventricles. Multiple and bilateral hypersignal within the paraventricular white matter. Moderate diffuse brain atrophy. Diffuse low signal of the rostrum of the corpus callosum, sequeller of previous bleeding.

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Findings

Brain CT scan reveals three nodules strongly enhancing on post IV contrast films, located in both thalamic nuclei (Figure 1). Magnetic resonance imaging study shows multiple anomalies on T2 weighted images (T2W1): multiple foci of hypersignal scattered throughout both cerebral peduncles more on the right side, both capsulo-lenticular and caudate areas, paraventricular white matter and splenium of the corpus callosum, 3 small foci of very low signal located in the upper part of both peduncles more at the left side, and corresponding to deposits of hemisiderin and ferritin related to old hematomas, a diffuse low signal in the rostrum of the corpus callosum indicating a previous diffuse bleeding, a mild cerebral atrophy.

On the basis of the clinical history and the imaging findings, the diagnosis of multiple sclerosis, neurobehcet disease (NB) and neurosarcoidosis may be discussed. But MRI findings are highly suggestive of neurobehcet involvement owing to the predominance of the lesions in the brainstem, basal ganglia and paraventricular white matter. Furthermore on inquiry, the patient has experienced recurrent oral and genital aphtous ulcers and was found to be positive to alloantigen HLA B5, supporting NB disease as the most likely diagnosis. The old areas of hemorrhage and cerebral atrophic changes denote the chronicity of the neuroinvolvement.

Behcet disease is a rare multisystem immune related vasculitis, that is prevalent in Mediterranean countries, Middle East and Japan.[1,2] Central nervous system involvement or neurobehcet is well known to occur in this disorder[3] usually 2 to 4 years after the first attack. But a few cases, NB may be inaugural or revealing the disorder in the condition of lack of the extra neurological symptoms.[4,5] Multiple clinical symptoms are suggestive of the diagnosis, including cranial nerve palsies, speech disorders, cerebellar signs, sensory and motor disturbances, loss of vision, diplopia and nystagmus. Neuropathology of NB consists of a disseminated meningoencephalo-palomyelitis with perivascular cellular infiltration around venules, capillaries and occasionally around arteries.[2,3] These changes may cause infarcts with small necrotic areas around blood vessels, hemorrhage, demyelination and gliosis.[2,3] Brainstem and diencephalon are the main sites of the lesions. In the early stages, lesions may resolve completely or partially even on steroid therapy. As the lesions become old and chronic, gliosis, atrophy and in some cases hemorrhage and thickening of the meninges are observed.[6]

Magnetic resonance imaging is much more sensitive than CT scan for detection of the NB involvement. Two patterns of lesions are observed, parenchymal lesions and dural venous thrombosis.

1) The parenchymal lesions appear as areas of high signal on T2W1 and iso or low signal on T1W1, mostly located in the brainstem, basal ganglia, internal capsules and paraventricular white matter. In the acute phase, the inflammatory process appears as a large lesion with occasionally a significant mass effect. Large contrast enhancement is common in evolutive lesions. In the subacute and chronic stages, some of the involved foci particularly in the brainstem and basal ganglia, tend to acquire a low signal intensity on T2W1, suggestive of magnetic susceptibility effect related to hemosiderin deposits. These findings were reported by few authors.[2] Therefore, our case is very demonstrative and complete, showing multiple foci of hemosiderin deposits (Figure 4) in the basal ganglia, and a diffuse callosal T2 low signal indicating a previous old bleeding. To our knowledge, such phenomens were not previously as clear documented as in our case. Moreover, the presence of hemispheric atrophic changes denotes the chronicity of the illness. 2) The dural venous thrombosis is a frequent manifestation of NB disease. The longitidinal superior and the lateral sinuses are the most commonly involved. Venous angio-MR is actually the most safe and accurate diagnostic modality.

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References

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