Magnetic resonance imaging and Neuro-Behcet Disease

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ABSTRACT

Behcet disease is a rare multi-system immune related vasculitis which may involve the central nervous system, usually during the course of the illness. Magnetic resonance imaging is currently the modality of choice for the detection of the parenchymal lesions in their different phases, as well as the dural sinus venous thrombosis.

Keywords: Neuro-Behcet disease, magnetic resonance imaging.

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Behcet disease (BD) is a relapsing and remitting systemic autoimmune vasculitis of unknown etiology,[1] first described by Hulusi Behcet in 1937.[2] This inflammatory disorder is characterized by the classic triad of uveitis, recurrent oral and genital ulcers. Other clinical symptoms include arthritis, thrombophlebitis, skin and neurological signs.[3] The peak onset of the disease is in the 3rd decade, and males are 3 to 4 times more affected than females.[3,4] Mediterranean countries, Japan, China and Iran are the most affected areas. Neurologic involvement or Neuro Behcet (NB) had been reported in variable proportions (5 to 49%) and represents a major poor prognosis factor of the disease outcome.[4,5] It occurs mostly 2 to 4 years after the initial clinical onset[4] but may be inaugural in almost 5% of the cases or reveals the disease when the extra neurological signs are discrete. Sigal[6] mentioned that BD may present with neurologic symptoms before the mucocutaneous and ocular lesions become apparent.[7,8] Neurologic manifestations are non-specific and include loss of vision, diplopia, nystagmus, cranial nerve palsies, speech disorders, cerebellar signs, cerebral and spinal sensory and motor disturbances.[4,9,10] The predominant histopathological findings are meningoencephalitis and vasculitis affecting mainly the small and medium size vessels causing focal necrosis, hemorrhage, demyelination and gliosis.[4,11] During the acute inflammatory process, edema and cellular infiltration may be intense mainly in the brain stem.[3] Brain computerized tomography (CT) scan and magnetic resonance imaging (MRI) are very helpful for the diagnosis. Magnetic resonance imaging had been proved much more sensitive compared to CT findings. When the lesions are seen on CT scan, they are found larger and more widespread on MRI pictures.[7] Neuro Behcet involvement may determine the cerebral parenchymal lesions, dural, sinus thrombosis, or both.

Parenchymal lesions are mostly located in the brain stem (**Figure 1a**), basal ganglia, internal capsules (**Figure 1b & 1c, Figure 2**) and periventricular white matter. The parenchymal distribution of the lesions in NB appears to support the hypothesis of small vessel vasculitis, mainly venular involvement.[12] The lesions are nodular,

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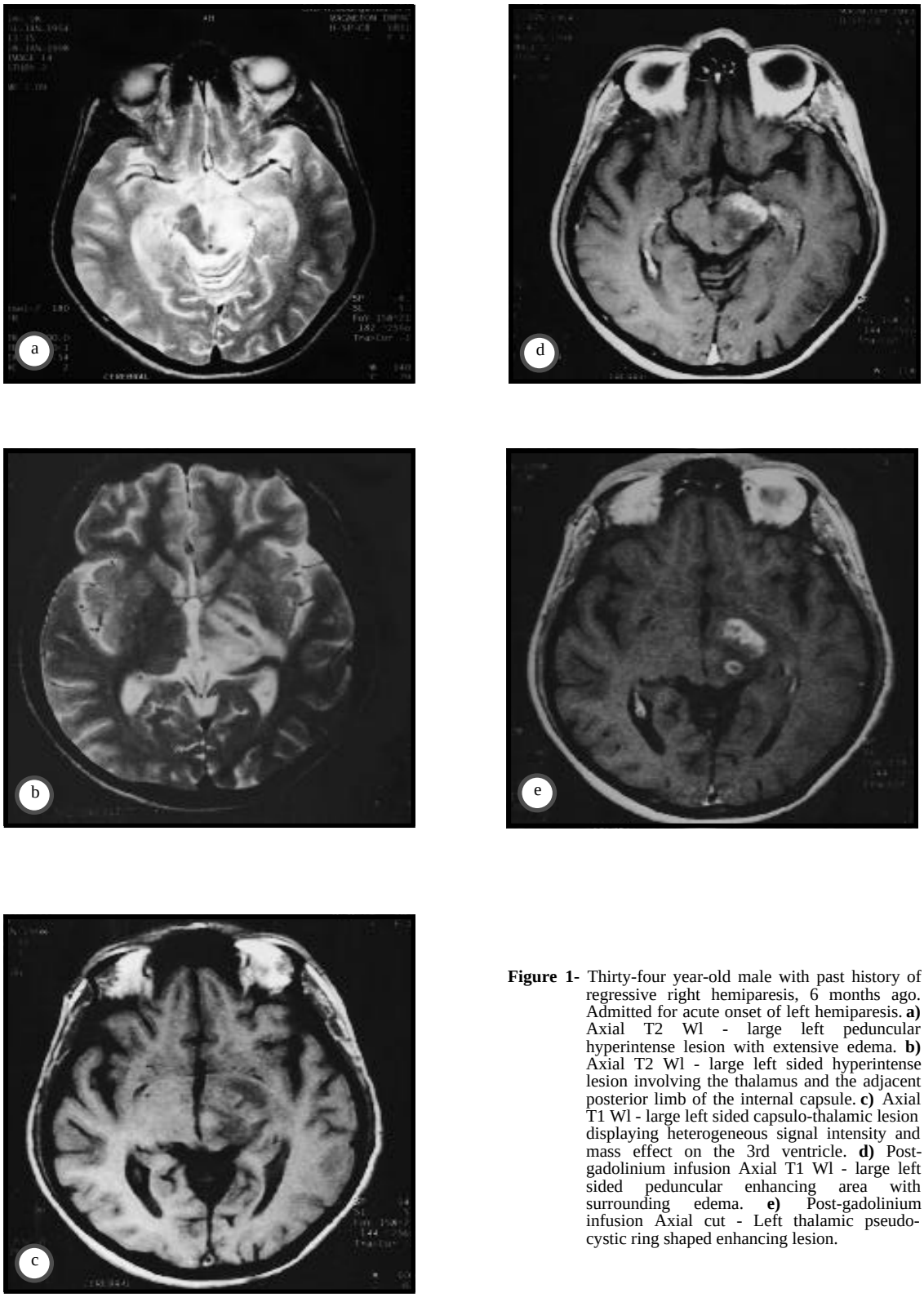

Figure 1- Thirty-four year-old male with past history of regressive right hemiparesis, 6 months ago. Admitted for acute onset of left hemiparesis. **a)** Axial T2 Wl - large left peduncular hyperintense lesion with extensive edema. **b)** Axial T2 Wl - large left sided hyperintense lesion involving the thalamus and the adjacent posterior limb of the internal capsule. **c)** Axial T1 Wl - large left sided capsulo-thalamic lesion displaying heterogeneous signal intensity and mass effect on the 3rd ventricle. **d)** Post-gadolinium infusion Axial T1 Wl - large left sided peduncular enhancing area with surrounding edema. **e)** Post-gadolinium infusion Axial cut - Left thalamic pseudo-cystic ring shaped enhancing lesion.

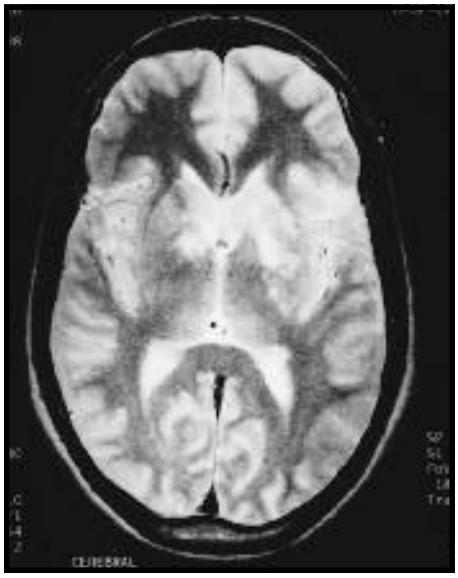

Figure 2 - Thirty-two-year-old patient presenting with sudden left sided hemiparesis. Axial T2 Wl, bilateral multiple tiny hyperintense lesions involving the anterior limb of the internal capsule, and the lenticular nucleus, bilaterally but more on the left side.

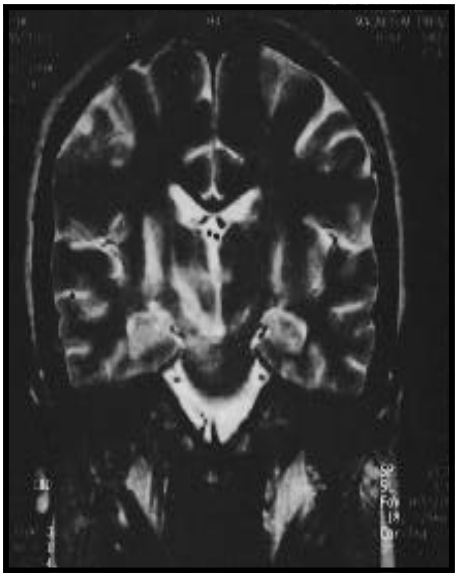

Figure 3 - Twenty-three-year-old female patient, with Behcet disease. Admitted for acute onset of the left upper limb monoplegia. Coronal T2 WI, multiple hyperintense linear lesions involving the right capsulo-thalamic area and basal ganglia bilaterally more on the right side.

multiple and confluent, shown as areas of hypersignal on T2 weighted image (T2 WI) and iso or low signal on T1 weighted image (T1 WI). The white matter lesions are distant from the ventricular wall, and usually associated with the brain stem and basal ganglia involvement.[13,14] Tah et al,[4] described a linear T2 WI high signal intensity along the posterior limb of the internal capsule **(Figure 1b and Figure 3)** and reported it as a specific finding especially if associated with the brain stem and basal ganglia involvement. The fast fluid attenuated inversion recovery (f Flair) is more sensitive than the conventional or fast spin echo T2 weighted MRI, for detecting the brain lesions particularly in the periventricular area.[15] In the acute phase, parenchymal contrast enhancement is frequently noted indicating a breakdown of the brain blood barrier.[3] Three patterns of contrast enhancement are seen in the acute lesions: intense and homogenous enhancement, “Pseudo-cystic” ring shaped enhancement surrounding central hyposignal intensity and peripheral contrast enhancement with irregular margins[12] (**Figure 1d & 1e**). Occasionally a significant mass effect may be observed. Often, the parenchymal lesions regress spontaneously or after steroid therapy. Gerber et al[3] reported one observation with disappearance of the contrast enhancement within an acute parenchymal lesion, after 8 days of steroid therapy. As previously evoked by Weschler et al,[7] the decrease in size of the enhancing lesions observed in the early stage, spontaneously or after steroid therapy, denotes a reversible breakdown in the blood brain barrier rather than gliosis or infarction. With time, the acute lesion decreases in size and in most cases a "sequellar" signal abnormality persists. In the sub-acute and chronic stages, foci of hemosiderin and ferritin deposits may be observed particularly within the brain stem and basal ganglia, appearing as deep low signal nodules in both T1 and T2 weighed sequences.[2] Brain stem atrophic changes appear in the last stage and are characteristic of NB disease. Cerebral venous thrombosis is observed in 35% of patients with NB disorder, and seems to present better prognosis than the parenchymal involvement.[7] The sagittal superior and the lateral sinuses are the most commonly involved. Its pathogenesis still remains unclear. It may precipitated in some cases by high fibrinogen levels,[11] but in many cases the exact mechanism is unknown. A combination of endothelial injury and defective fibrinolytic activity are then invoked.[16] The venous magnetic resonance angiography is a secure and reliable diagnostic modality of the dural sinus thrombosis, showing good correlation with conventional angiography in the detection of dural sinus thrombosis.[7] The differential diagnosis with multiple sclerosis, neurosarcoidosis and neurolupus, may be difficult when the extra neurological features of BD are absent or discrete. In these circumstances, the pattern of lesion distribution observed in MRI might help to differentiate NB disease from other vasculitides, as well as from the inflammatory demyelinating diseases such as multiple sclerosis.

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