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**Original Research Article**

# Contribution of MRI in the Management of Neuro-Behçet, Retrospective Study: About 16 Cases

Oum Rachad Hamdaoui[1*], Achta Adam Fadoul[2], Hmada Sanaa[1], Hasna Tabakh[1], Najwa Touil[1], Omar Kacimi[2], Nabil Chikhaoui[2]

[1]Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco
[2]Departement of Radiology, Ibnou Rochd Hospital, Casablanca, Morocco

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**Abstract:** Neurological damage from Behçet's disease is considered to be a severe neurological manifestation. We have collected 16 cases from the internal medicine, neurology and radiology department of Ibnou Rochd hospital in Casalanca. Our patients have a mean age of 39 years, with male predominance and an average delay of 6.18 years. Headache and central motor impairment were the first telltale signs of neuro-Behçet in 62.5%. The brain scan was carried out in 15 patients, it objectified an ischemic stroke in a single case, poorly systematized hypodensities raising suspicion of thrombophlebitis in 5 cases and an image of venous thrombosis concerning the upper longitudinal sinus in a single case. Brain MRI showed demyelination lesions, of infra-centimeter sizes and variable shapes; in hypo signal T1, hyper signal T2 and T2 FLAIR in 9 patients, of supratentorial seat in 5 cases and lesions in under tentorial in 4 cases with uptake of nodular contrast in 2 cases and annular contrast in only one case. The MRI also highlighted 3 cases of cortical subcortical atrophy on the upper tentorial level, one case of atrophy of the brainstem, 7 cases of venous thrombosis objectified on angio MRI and 2 cases of stroke ischemic. Thus, a pseudo-tumor aspect was reported in only one case. And only one case benefited from a spinal MRI and objectified cervical lesions. These lesions are often associated. The preferred location found in our patients is the brainstem, essentially ponto- mesencephalic. This distribution makes it possible to make the differential diagnosis with other vasculitides and inflammatory diseases.

**Key words:** Imaging - MRI - Neuro-Behçet – Support.



## INTRODUCTION

Behçet's disease is a systemic vasculitis of unknown etiology. It mainly affects men more than women between 20 and 40 years old and it is common in the Far East and around the Mediterranean. Neurological involvement in Behçet's disease is considered to be a serious manifestation that can be life-threatening. It is due to an inflammation which preferentially affects the veins of small and medium caliber and which preferentially concerns the brainstem, the central gray nuclei and the supratentorial white matter without particular predilection for the peri-ventricular regions, as well as the thrombosis of the veins cerebral and dural sinuses.

The objective of our work is to highlight the contribution of MRI in the diagnosis of neurological involvement in Behçet's disease.

## MATERIAL AND METHODS

Our work a study is based on a retrospective review of 16 patients who all benefited from a brain MRI and collected from the internal medicine, radiology departments of Ibnou Rochd hospital in Casablanca over a period of 11 years, between June 2010 and July 2021.

## RESULTS

- Gender: 15 male patients and 01 female patients
- The average age was 39 years.
- Geographical origin: 12 cases of urban origin and 4 cases of rural origin.
- Pathological history:
  - Oral aphthosis: 15 cases
  - Genital aphthosis: 8 cases
  - Eye redness: 6 cases

***Corresponding Author:** Oum Rachad Hamdaoui
Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco

- Neurological: 6 cases
- Pseudo-folliculitis: 4 cases
- Articular: 2 cases
- Vascular:1 case

- The method of installation of neuro-Behçet was progressive in 15 patients while the acute form was found in only one patient.
- Reasons for hospitalization:
  - Headache : 10
  - Motor disorders : 10
  - Speech disorders : 07
  - Visual disorders : 05
  - HTIC syndrome : 05
- Brain CT is performed in 15 patients, normal in 4 cases and pathological in 11 cases
- Appearance and location of lesions on brain CT:
  - poorly systematized hypodensities leading to suspicion of thrombophlebitis: 05 cases
  - systematized hypodensity: only 1 case
  - corticosteroid-subcortical atrophy in the supra-tentorial stage: 03 cases
  - Brain edema: 01 cases
  - Venous thrombosis (superior longitudinal sinus): 01 cases

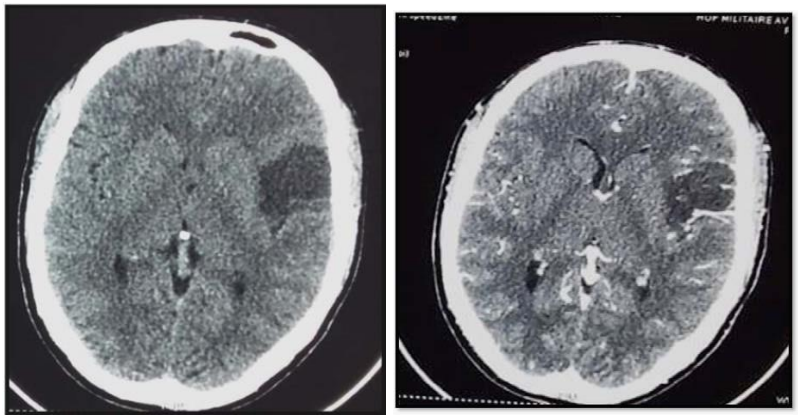

**Figure 1 and 2: Brain CT with and without injection of iodinated contrast product: shows systematized hypodensity at the left fronto-insular level and the external capsule suggesting ischemic stroke in the territory of the left superficial Sylvian artery**

- Brain MRI, performed in 16 patients, was pathological in 11 cases and normal in 5 cases.
  - White matter demyelination lesions, infra centimetric sizes and varying shapes: 09 cases
  - Cerebral thrombophlebitis: 2 cases
  - Arterial ischemia: 02 cases
  - Cerebral atrophy: 04 cases
  - A pseudo-tumor appearance: only one case
  - Only one case underwent a spinal cord MRI and found cervical lesions.

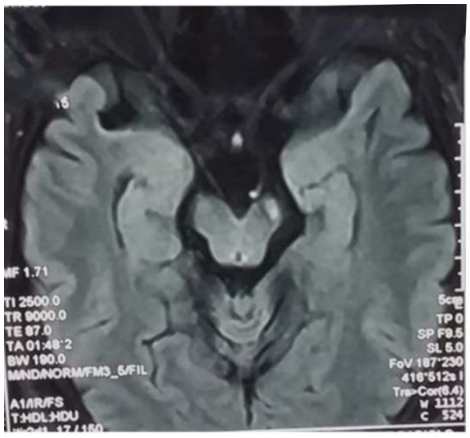

**Figure 3: Cerebral MRI in T2-weighted sequence along an axial plane: shows a hypersignal at the level of the left cerebral peduncle suggesting demyelination lesions of the cortico-spinal bundle**

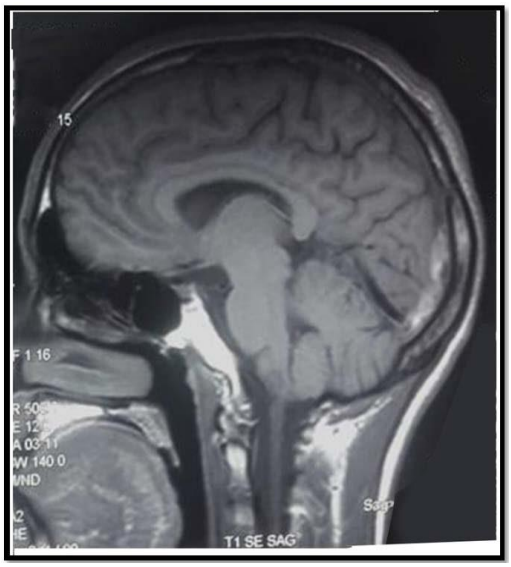


**Figure 4: Brain MRI in T1-weighted sequence according to a sagittal plane: shows a T1 hypointense focus in favor of thrombosis of the superior longitudinal sinus**

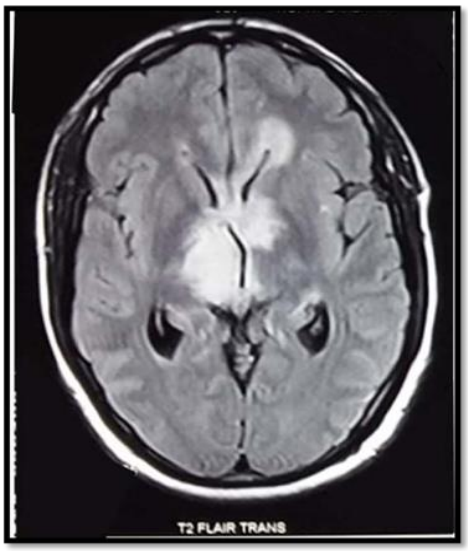


**Figure 5: Cerebral MRI in T2-weighted FLAIR sequence according to an axial plane: shows a range of hypersignal extended to the level of: left caudate nucleus, right capsulothalamic, knee of the corpus callosum, peri-ventricular with a mass effect on the line median taking the aspect of: Pseudo - Tumor**

# DISCUSSION

**General**

Neuro-Behçet is a vasculitis of the central nervous system, with venous tropism, evolves by outbreak interspersed with remission, or gradually.The prevalence of Behçet's disease is different from country to country [5]. It is endemic around the Mediterranean, in Central Asia, Japan and Iran. It affects men more than women with an average age close to 40 years (39.56 years), its frequency is estimated at 22.5%.Neurological involvement in Behçet's disease varies between 4-49% of cases [10]. Neurological manifestations of Behçet's disease can be divided into 2 groups:

1. Parenchymal involvement of the central nervous system (Neuro-Behçet) which includes the brainstem, hemispherical manifestations, lesions of the spinal cord and meningoencephalitis, as well as thrombosis of the dural sinuses and central cerebral veins.
2. Non-parenchymal involvement of the central nervous system (Angio-Behçet) which includes arterial occlusion and arterial aneurysm.

Neuro-Behçet can be inaugural during Behçet's disease with an often progressive onset and an average delay close to 7 years (6.18 years), is frequently revealed by headaches, motor deficit, dysarthria and is often explained by pyramidal syndromes, damage to the cranial nerves and bulbar syndromes [10].

**BRAIN SCAN**

The study was performed without injection of iodinated PDC for spontaneous hypodensities, subarachnoid hemorrhages and after intravenous injection of iodinated PDC for the assessment of cerebral vascularization, showing evidence of cerebrovascular thrombosis [11].

The most frequently encountered lesions are spontaneous hypodensities and parenchymal atrophy. Images of cerebral edema, calcifications and subarachnoid hemorrhage remain rare [5, 6]. Brain lesions are most often unique in 80% of cases and multiple in 20% of cases [12].

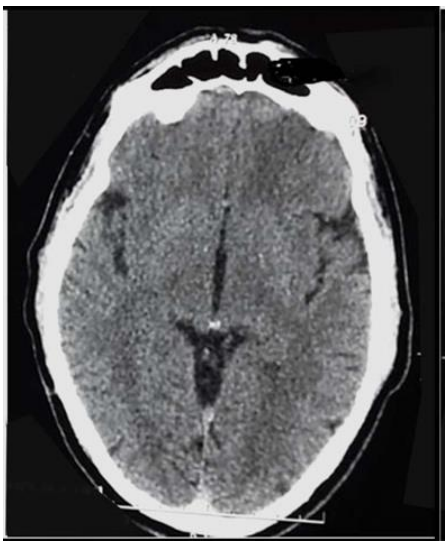

**Figure 6: Brain CT shows a poorly systematized left frontal hypodensity suggesting a stroke of venous origin**

**MRI**

Magnetic resonance imaging is the gold standard in the exploration of neuro-Behçet, whether it is cerebral venous thrombosis or parenchymal lesions [5].

Parenchymal involvement is the most common, accounting for 60 to 81% of all neurological damage. It is dominated by involvement of the brainstem seen in 25 to 60% of cases, followed by involvement of the thalamus, basal ganglia, internal capsule [4, 5, 2] and spinal cord involvement found in 2.5 to 18% of cases [2-7]. These attacks are often associated [7, 8].

The distribution of lesions may be subcortical, periventricular, or at the level of deep white matter. Extensive, confluent lesions associated with lesions of the brainstem, basal ganglia, and / or the internal capsule and very suggestive of neuro-Behçet [2].

Signal abnormalities: The lesions are in T2 hypersignal, variable signal in T1 and are enhanced after injection of gadolinium in 60% of cases. This enhancement is variable (in clods or annular). The confluent and extensive character of T2 hypersignal lesions of the brainstem and the central gray nuclei seems specific to acute neurobehçet damage [8, 7].

Another characteristic of these lesions is their reversibility. They may disappear completely on control MRIs, decrease in size or remain unchanged. No lesion increases in size. Brainstem atrophy characterizes chronic neuro-Behçet. Brainstem atrophy without cortical atrophy is a specific sign of neuro-behçet (specificity 96.5% and low sensitivity).

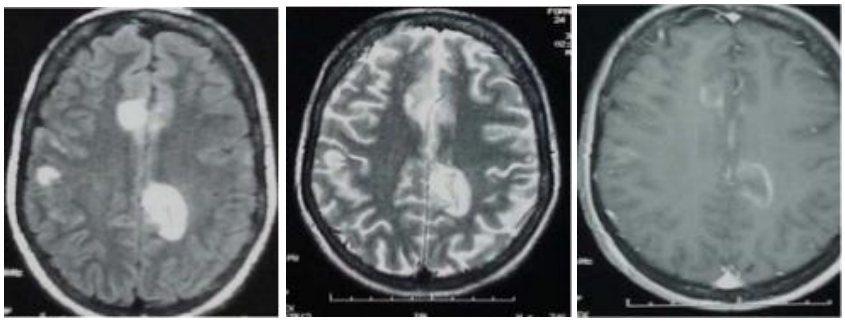

**Figure 7: MRI in axial section shows: A: FLAIR sequence: shows three cortical lesions in hyper signal. B: T2 sequence, the three lesions are in T2 hyper signal. C: T1 sequence with gadolinium injection, shows annular enhancement of the three lesions**

Neuro-Behçet is a vasculitis with venous tropism, hence the predominance of venous thrombosis compared to arterial thrombosis. MRI with angio sequences has a vital role in the diagnosis of these thrombophlebitis. It allows to study: The absence of flow in the thrombosed vessel, The impact on the adjacent cerebral parenchyma and the evolution of the thrombus.

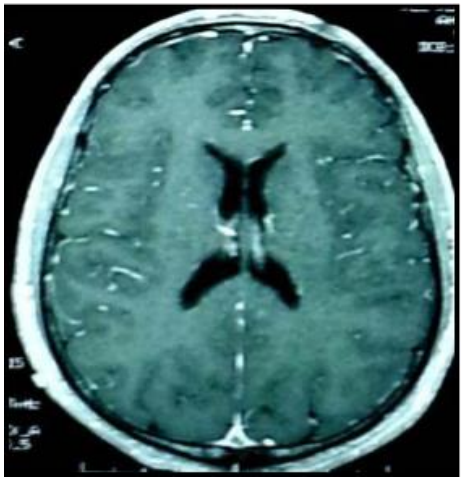

**Figure 8: T1-weighted sequence MRI + GADO along an axial plane: shows a thrombus of the superior longitudinal sinus in delta**

The pseudotumoral form: rare; Extensive lesion with T1 hypointense, T2 hypersignal, surrounded by perilesional edema and heterogeneously enhanced after contrast. The mass effect is not very marked compared to the lesion volume [9].

Meningeal lesions: A thickening with meningeal enhancement is a sign also described in neuro-behçet.

Spinal cord injuries: Spinal cord injury is rare. The sites of predilection are the cervical cord and the thoracic cord. The impairment is typically T2 hypersignal, multifocal and non-contiguous.

**Differential diagnosis**

The differential diagnosis arises with all demyelinating lesions but especially with multiple sclerosis (MS) and systemic lupus erythematosus. Neuro-behçet lesions predominate in the brainstem and are rather anterior; those of MS, rather concern the floor of the fourth ventricle and the middle cerebellar peduncle. The confluent and extensive character remains specific to neurobehçet [5-7].

## CONCLUSION

MRI and angioirm are currently the best method of exploring the neurobehçet. They have a role in the precise characterization of the lesions, thus allowing the positive and differential diagnosis as well as the follow-up of patients under treatment.

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