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Cerebral Venous Thrombosis in Behcet's Disease about 24 Patients

Oumerzouk J[*1], Louhab N[2], Chraa M[2] and Kissani N[2]

[1]Neurology Department, Military Hospital of Marrakech, Avicenne, Morocco
[2]Neurology Department, Mohamed VI Hospital, Marrakech, Morocco

[*]**Corresponding author:** Oumerzouk J, Military Hospital Avicenne, Marrakech, Morocco 10100, Tel: 00(212)614134817, E-mail: tamamro@yahoo.fr

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Abstract

Introduction: Cerebral venous thrombosis (CVT) is the most common manifestation of angiobehçet and may be superficial and /or deep localization.

The aim of our cohort is to study the clinical and radiological features of CVT associated with Behcet's disease in our population and to compare it with previous studies.

Materials and Methods: We report a retrospective study of 24 cases of CVT secondary to Behçet's disease, collected between 1999 and 2019 in the neurology department of military hospital Mohamed V (Rabat), military hospital Avicenne (Marrakech) and Mohamed VI hospital (Marrakech). The diagnosis of Behçet disease was retained in all cases according to the International Study Group Criteria for Behçet diseases 2014. All the patients received antithrombotic and immunodulatory treatments.

Results: Clinical manifestations were dominated by motor deficit in deep localizations and intracranial hypertension in superficial locations. Motor signs and symptoms, were the most common findings during the disease course of our patients (87,5%). The diencéphalo-mesencephalic syndrom consisting in hemiparesis, hemiataxia and /or movement disorders, ophthalmoplegia were found in 18 patients, whereas, intracranial hypertension was only noted in 5 cases. Brain MRI showed hyperintension lesion in T2-weighted images interesting the diencephalo-mesencephalic region in 15 cases, and capsulo-diencephalic region in 3 cases. AngioMRI and conventional angiography performed in our patients confirmed the predominance of deep venous thrombosis in our patients (18 cases), unlike previous studies.

Discussion: Comparing to previous studies, the frequency of the vascular form of BD, mainly represented by CVT in our series was much higher (87,5%) and motor signs and symptoms, were the most common findings during the disease course of our patients (87,5%), in agreement with previous reports. Magnetic resonance angiography showed 4 cases of sagittal sinus thrombosis and 3 cases of lateral sinus thrombus. However, the benefit of conventional angiography was undeniable in our cohort, by confirming the diagnosis of BVR thrombosis in 75% of our patients.

Conclusion: In our series, the prognosis of the 6 patients presenting with superficial sinus thrombosis, was favorable after anticoagulation and corticosteroids, with moderate persisting signs and/or symptoms in 4 patients and complete recovery in 2 patients. However the persisting sequelas were serious in the 18 presenting deep parenchymal lesions.

Keywords: Cerebral Venous Thrombosis; Behçet's Disease; Angio-Behçet; MRI

Introduction

Behçet's disease (BD) is a chronic multisystem vasculitic syndrome. Cerebral venous thrombosis (CVT), are the most common vascular complication, which can be superficial and /or deep localization. Advances in cerebral imaging have allowed earlier diagnosis and more aggressive therapy, which improve the prognosis of this disease.

The aim of our cohort is to evaluate the clinical and radiological features of CVT associated with Behcet's disease in our series and to compare it with previous studies.

Materials and methods

This is a retrospective study of 24 cases of CVT secondary to Behcet's disease, collected at the neurology department of the military hospital Mohamed V (Rabat), military hospital Avicenne (Marrakech) and Mohamed VI hospital (Marrakech), between January 1999 and December 2019. The diagnosis of BD was retained in all cases according to the International Study Group Criteria for Behçet diseases 2014 (Table 1). The 24 patients underwent complete neurological and physical examinations. All patients underwent brain CT scan which was practiced in the first 48 hours (Figures 1,2 and 3). Brain MRI was pathological in all cases, and was supplemented by a conventional angiography in 20 cases (Table 2). The laboratory screening included a cytochemical study of cerebrospinal fluid (CSF), immunological (antinuclear antibodies, native anti-DNA, antiphospholipid, anti-SSA, -SSB, and anti-RNP, etc..) and inflammatory tests (CRP, orosomucoid, haptoglobin, fibrinogen).

Patients	Age	Gender	Recurrent oral aphthous, ulceration	Recurrent genital ulceration	1.Erythema nodosum 2.Pseudofolliculitis 3.Papulopustular eruption 4.Acneiform nodules 5.Pathergy test (PT) positivity	Anterior or posterior uveitis	Other clinical signs
1	39	F	+	+	+ PT	+	-
2	28	M	+	+	+PT	-	-
3	44	M	+	+	+PT	-	arthralgia
4	34	M	+	+	+PT, pseudofolliculitis	-	-
5	46	M	+	+	+PT, pseudofollicultis, erythema nodosum	+	-
6	36	M	+	+	+PT	-	arthralgia
7	24	M	+	+	+PT	-	-
8	31	M	+	+	+PT, erythema nodosum	-	arthralgia
9	36	M	+	+	+PT	-	-
10	33	M	+	+	+PT	-	-
11	34	M	+	-	+Pseudofolliculitis	+	-
12	29	M	+	+	+PT	+	-
13	20	M	+	+	+PT	-	-
14	36	M	+	+	+PT	+	-
15	41	F	+	+	+Pseudofolliculitis	-	-
16	36	M	+	+	+PT	-	-
17	28	F	+	+	+Pseudofolliculitis	-	
18	33	M	+	+	-	+	-
19	31	M	+	-	+Erythema nodosum	-	-
20	21	M	+	+	+ Pseudofolliculitis	-	-
21	33	F	+	+	+ Pseudofolliculitis	-	-
22	39	F	+	+	+ Pseudofolliculitis	+	-
23	23	M	+	+	+ Pseudofolliculitis	-	-
24	52	M	+	+	+PT	+	-

Table 1: Clinical data of 24 patients presenting with cerebral venous thrombosis secondary to Behçet's disease

Patient	Cerebrospinal fluid (CSF)	Brain CT scan	Brain Angio-MRI	Conventional angiography	Trea Treatment	Outcomes
1	Protein=normal 8 lymphocyts/ mm³	Normal	Hyperintense aspect on T2- and Flair- weighted images localized in left diencephalo-mesencephalic region	Left basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids	Moderate right hemiparesis
2	Normal	Right thalamo-mesencaphalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in right diencephalo-mesencephalic region	Right basal vein of Rosenthal (BVR) thrombosis +radiological signs of cerebral vasculitis	Anticoagulants Corticosteroids	Left hemiparesis
3	Protein=0,68g/l 12 lymphocyts/ mm³	Normal	Hyperintense aspect on T2- and Flair- weighted images localized in right capsulo-diencephalo-mesencephalic region	Right basal vein of Rosenthal (BVR) Thrombosis	Anticoagulants Corticosteroids Endoxan	Moderate left hemiparesis, dysarthria,
4	Protein=0,62g/l 5 lymphocyts/ mm³	Left capsulo-diencephalo-mesencephalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in left capsulo-diencephalo-mesencephalic region	Left basal vein of Rosenthal (BVR) thrombosis + radiological signs of cerebral vasculitis	Anticoagulants Corticosteroids Endoxan	Right hemiparesis, hemiataxia, dysarthria
5	Protein=0,6g/l 8 lymphocyts/ mm³	Right capsulo-thalamo-subthalamic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in right diencephalo-mesencephalic region	Right basal vein of Rosenthal (BVR) thrombosis + radiological signs of cerebral vasculitis	Anticoagulants Corticosteroids Endoxan	Severe left hemiparesis, III nerve palsy
6	Protein=0,8g/l 12 lymphocyts/ mm³	Normal	Hyperintense aspect on T2- and Flair- weighted images localized in right diencephalo-mesencephalic region	Right basal vein of Rosenthal (BVR) thrombosis + right lateral sinus thrombosis	Anticoagulants Corticosteroids Endoxan	Left hemiparesis, left hemihypoesthesia, hemiataxia, III nerve palsy
7	Normal	Normal	Hyperintense aspect on T2- and Flair- weighted images localized in right subthalamo-mesencephalic region	Right basal vein of Rosenthal (BVR) Thrombosis	Anticoagulants Corticosteroids	Left hemiparesis, right hemiataxia, dysarthria, dysphagia
8	Protein=0,4g/l 15 lymphocyts/ mm³	Left capsulo-thalamo-subthalamic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in left diencephalo-mesencephalic region	Left basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids Endoxan	Right hemiparesis, left hemiataxia, dysarthria
9	Normal	Normal	Hyperintense aspect on T2- and Flair- weighted images localized in right diencephalo-mesencephalic region	Right basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids	Left hemiparesis, left hemiataxia
10	Protein=normal 18 lymphocyts/ mm³	Normal	Hyperintense aspect on T2- and Flair- weighted images localized in right capsulo-thalamo-subthalamic region	Right basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids Endoxan	Severe left hemiparesis, left hemiataxia, right III palsy
11	Protein=0,9g/l 40 lymphocyts/ mm³	Bilateral capsulo-thalamic hypodensity	Bilateral hyperintense lesions on T2- and Flair-weighted images localized in capsulo-thalamic region	Left basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids Endoxan	Left hemiparesis, right hemiataxia,
12	Protein=0,5g/l 1 lymphocyt/ mm³	Right capsulo-thalamo-subthalamic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in right capsulo-thalamo-subthalamic region	Right basal vein of Rosenthal (BVR) thrombosis + radiological signs of cerebral vasculitis	Anticoagulants Corticosteroids	Left hemiparesis, left hemiataxia

13	Normal	Hyperdense superior sagittal sinus After injection of contrast: empty delta sign	Superior sagittal sinus thrombosis	Superior sagittal sinus thrombosis	Anticoagulants Corticosteroids	asymptomatic
14	Protein=0,1g/l 480 lymphocyts/ mm^3	Left capsulo-mesencephalo-diencephalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in left capsulo-mesenphalo-diencephalic region	Left basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids Endoxan	Right hemiparesis, left hemiataxia, left III palsy, dysarthria
15	Protein=normal 135 lymphocyts/ mm3 (40% lymphocyts)	Left capsulo-mesencephalo-diencephalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in left capsulo-mesenphalo-diencephalic region	Left basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids Endoxan	Right hemiparesis, right hemihypoesthesia, left hemiataxia
16	Normal	Brainstem hypodensity	Left lateral sinus thrombosis	Left lateral sinus thrombosis	Anticoagulants Corticosteroids	Mild right hemiparesis, diplopia, dysarthria and dysphagia
17	Normal	Hyperdense superior sagittal sinus After injection of contrast: empty delta sign	Superior sagittal sinus thrombosis + right lateral sinus thrombosis	-	Anticoagulants Corticosteroids	Asymptomatic
18	Normal	Hyperdense superior sagittal sinus After injection of contrast: empty delta sign	Superior sagittal sinus thrombosis	-	Anticoagulants Corticosteroids	Blurred vision
19	Protein=1g/l 55 lymphocyts/ mm^3	Left capsulo-mesencephalo-diencephalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in left capsulo-mesenphalo-diencephalic region	Left basal vein of Rosenthal (BVR) thrombosis	Anticoagulants Corticosteroids Endoxan	Right hemiparesis, right hemiataxia, right III nerve palsy
20	Protein=0,1g/l 16 lymphocyts/ mm^3	Hyperdense superior sagittal sinus After injection of contrast: empty delta sign	Superior sagittal sinus thrombosis	-	Anticoagulants Corticosteroids Endoxan	Headache, epilepsy, blurred vision
21	Protein=normal 65 lymphocyts/ mm^3	Right mesencephalo-diencephalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in right mesenphalo-diencephalic region	Absence of venous or arterial thrombosis	Anticoagulants Corticosteroids Endoxan	Left hemiparesis, right hemiataxia, right III nerve palsy
22	Protein=normal 19 lymphocyts/ mm^3	Mesencephalo-diencephalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in mesenphalo-diencephalic region	Absence of venous or arterial thrombosis	Anticoagulants Corticosteroids Endoxan	Tetraparesis, pseudobulbar syndrom
23	Protein=0,6g/l 64 lymphocyts/ mm^3	Mesencephalo-diencephalic hypodensity	Hyperintense aspect on T2- and Flair- weighted images localized in mesenphalo-diencephalic region	Absence of venous or arterial thrombosis	Anticoagulants Corticosteroids Endoxan	Tetraparesis, pseudobulbar syndrom
24	Normal	Brainstem hypodensity	Right lateral sinus thrombosis	-	Anticoagulants Corticosteroids	Mild left hemiparesis, diplopia

Table 2: Paraclinical data, treatment and outcomes of 24 patients presenting with cerebral venous thrombosis in the setting of behçet's disease

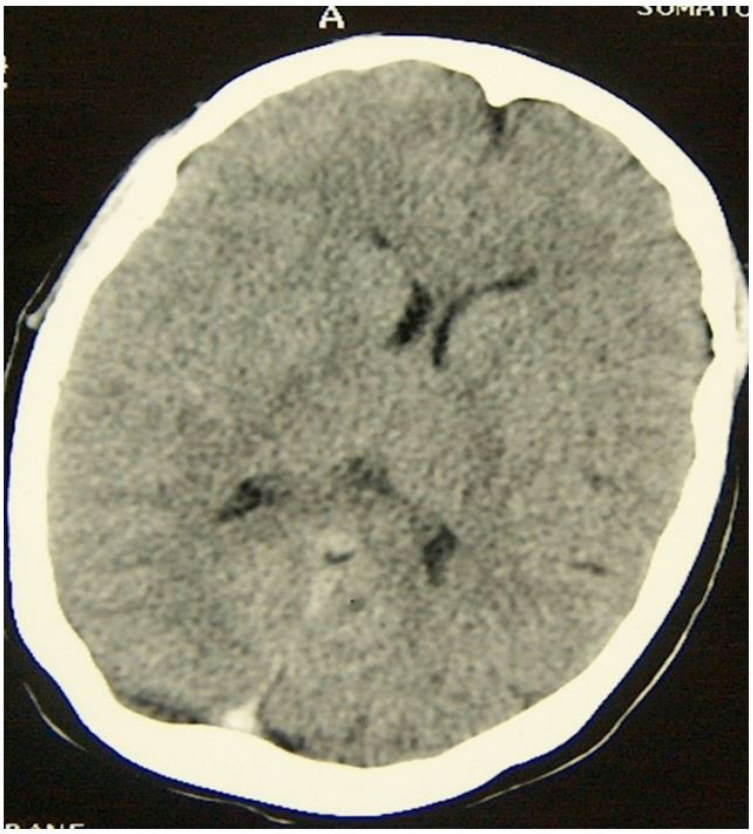

Figure 1: Brain CT scan without injection of contrast showing a spontaneous hyperdense superior sagittal sinus with diffuse cerebral edema

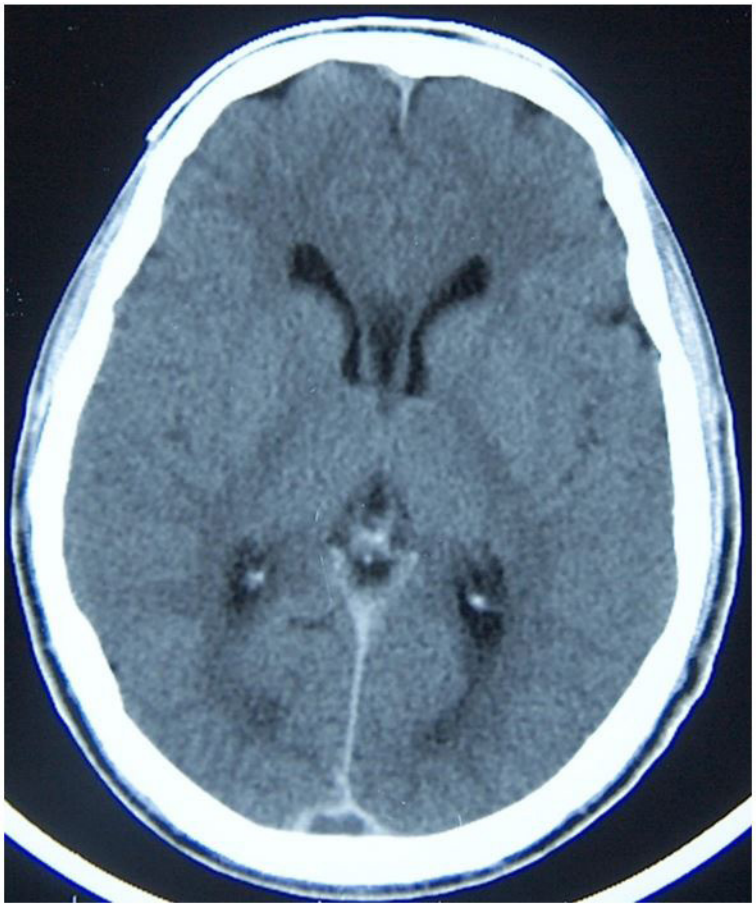

Figure 2: Brain CT scan after injection of contrast objectifying the empty delta sign

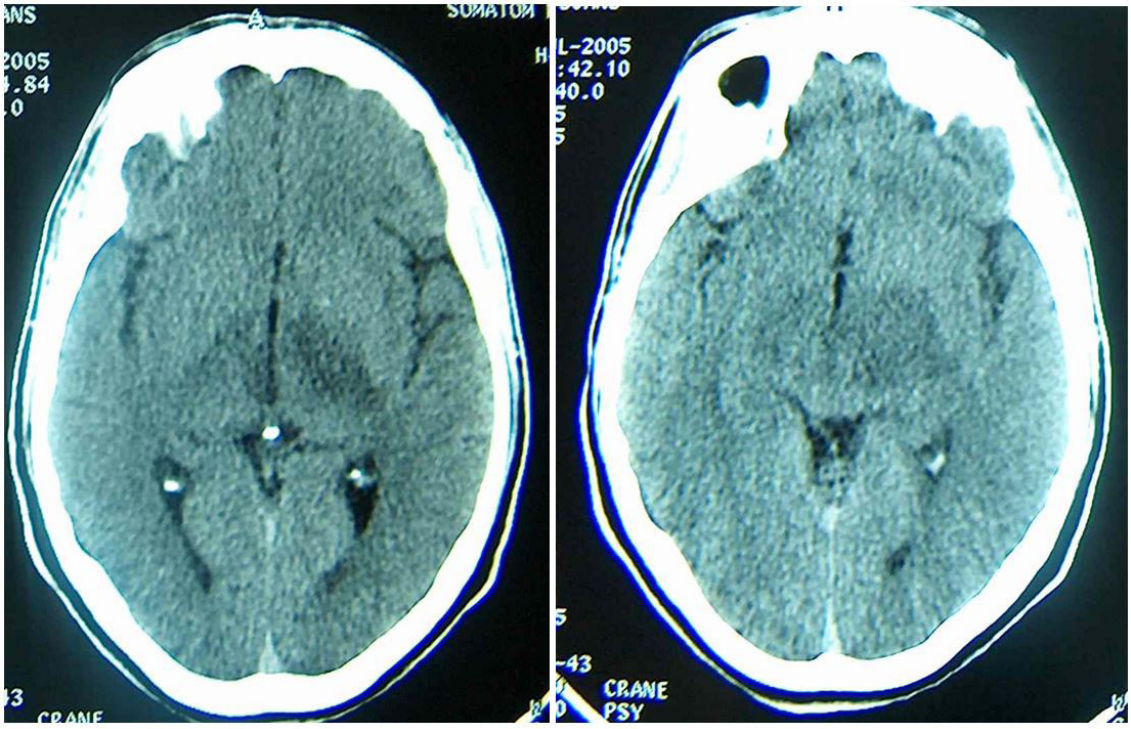

Figure 3: Brain CT scan without injection of contrast showing a left diencéphalo-mesencephalic hypodensity

All our patients received symptomatic treatment (antiepileptic drugs; preferably sodium valproate in cases of seizures, and intracranial hpertension management), antithrombotic treatment (Calciparin which was relayed by anti –K vitamin for 2 years on average) and etiological treatment, based on corticosteroids, in 10 cases of superficial CVT and deep CVT without inflammatory CSF (oral prednisolone at a dose of 1mg/kg/j to be reduced gradually until the lowest effective dose) and Immunosuppressants (cyclophosphamide; Endoxan^{R}) in 14 cases of deep CVT with inflammatory CSF (given as a bolus periodically at 600 mg/m^2 body surface area every 2 months for 2 years).

Results

Epidemiology

We noted a predominance of CVT for patients between 30 and 40 years old. Our series is also characterized by male predominance, and the distribution by age according to sex did not find any significant differences.

Clinical aspects

	Deep CVT	Superior sagittal sinus (SSS) thrombosis	Lateral sinus (LS) thrombosis
Number of cases	18	4	3
Hemiparesis	16	0	3
Tetraparesis	2	0	0
Hemiataxia	12	0	1
III nerve Palsy	7	0	3
Headache	5	4	0
Choreoathetoid movements	2	0	0
Pseudobulbaire syndrome	7	0	0
Consciousness Disorders	4	0	0
Fever	3	0	0
Seizures	0	3	0
Intracranial hypertension	0	4	1

Table 3: Distribution of clinical signs depending on the location

Neurological signs	Number of patients
Hemiparesis	19
Tetraparesis	2
Hemiataxia	13
III nerve Palsy	10
Choreoathetoid movements	2
Pseudobulbaire syndrome	7
Consciousness Disorders	4
Fever	3
Seizures	3
Headaches	9
Intracranial hypertension	5

Table 4: Neurological signs in 24 patients presenting with Behçet's syndrome

The diencéphalo-mesencephalic syndrom consisting in hemiparesis, hemiataxia, ophthalmoplegia and /or movement disorders were found in 18 patients, whereas, intracranial hypertension was only noted in 5 cases (Tables 3 and 4). Motor signs and symptoms, were the most common finding during the disease course of our patients (87,5%) (Tables 3 and 4).

Paraclinical aspects

The analysis of the brain MRI results of our series showed in terms of venous impairment, isolated superior sagittal sinus (SSS) thrombosis in 3 cases, concomitant SSS and right lateral sinus (LS) thrombosis in one case, and isolated LS thrombosis in 2 cases.

CVT results in a spontaneous hyperintense aspect of the sinus on MRI and absence of sinus visualization on angiographic images. Concerning parenchymal ivolvement, brain MRI shwoed hypointense lesions inT1- and hyperintense in T2-weighted images, interesting the diencephalo-mesencephalic region in 15 cases, and capsulo-diencephalic region in 3 cases (Figures 4,5,6,7 and 8).

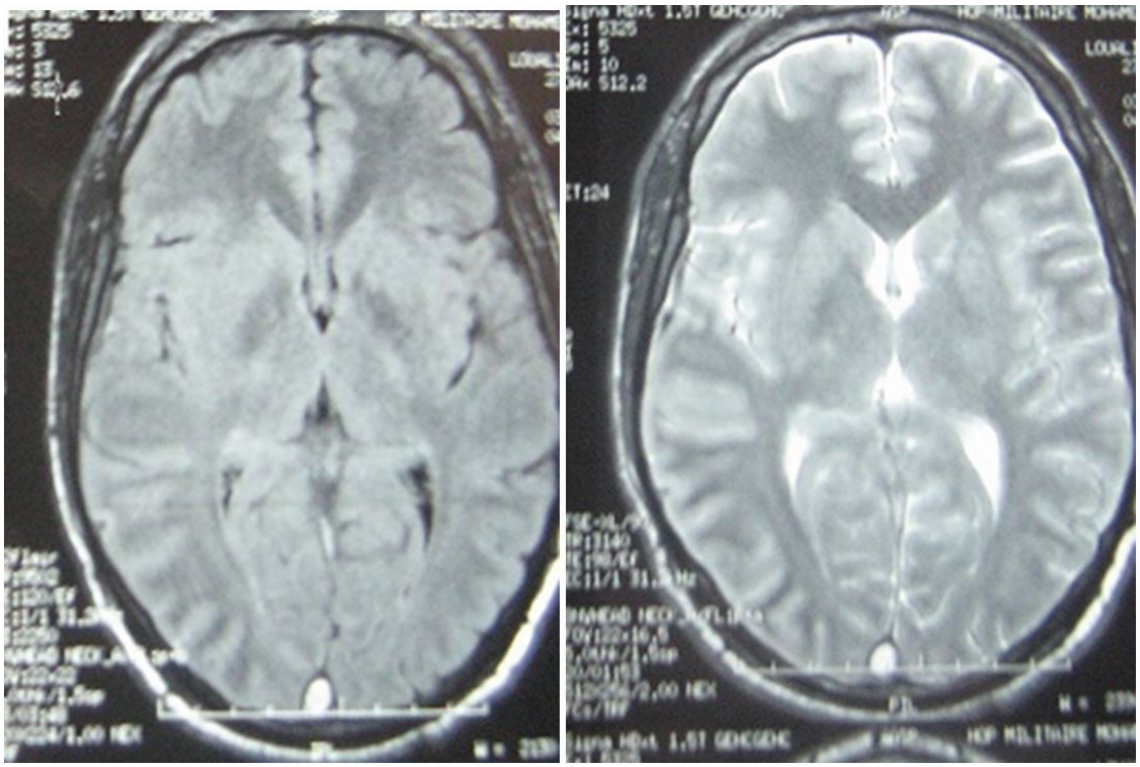

Figure 4: Axial brain MRI, T2 and FLAIR weighted images showing hyperintensity of the superior sagittal sinus

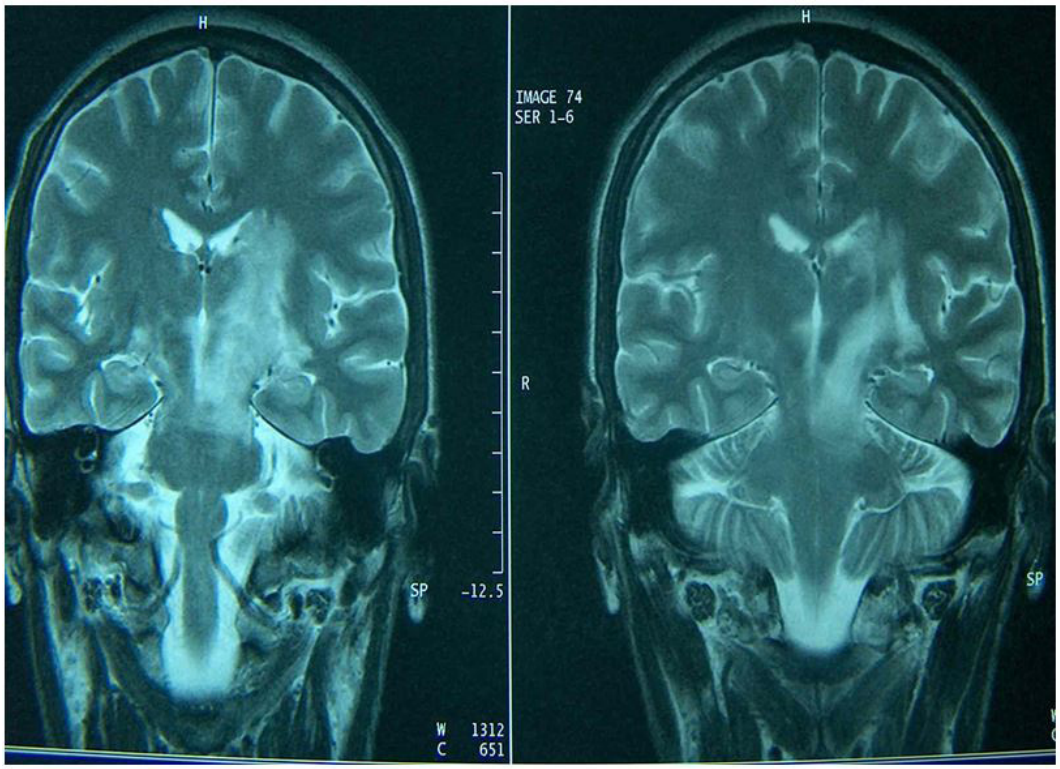

Figure 5: Coronal brain MRI, T2-weighted images, showing a hyperintense lesion in thalamo-midbrain regions

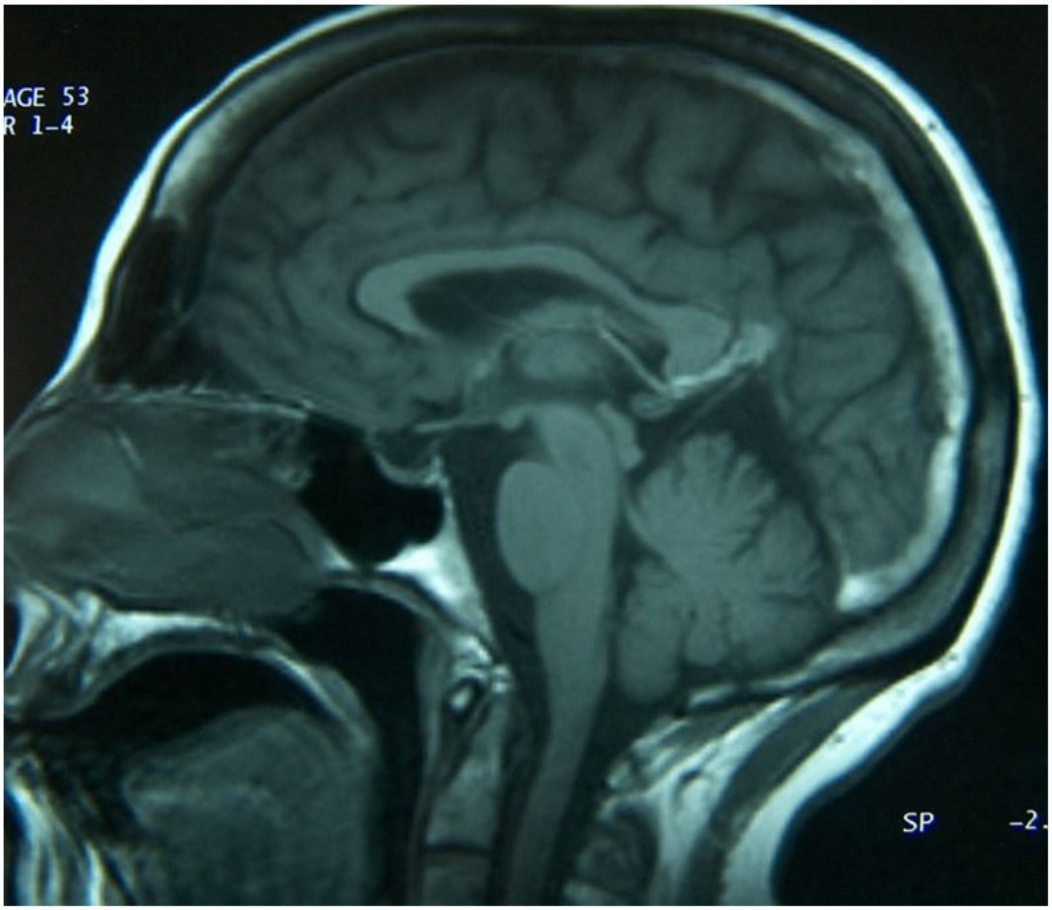

Figure 6: Sagittal brain MRI, T1weighted images, showing a spontaneous hyperintensity of superior sagittal sinus

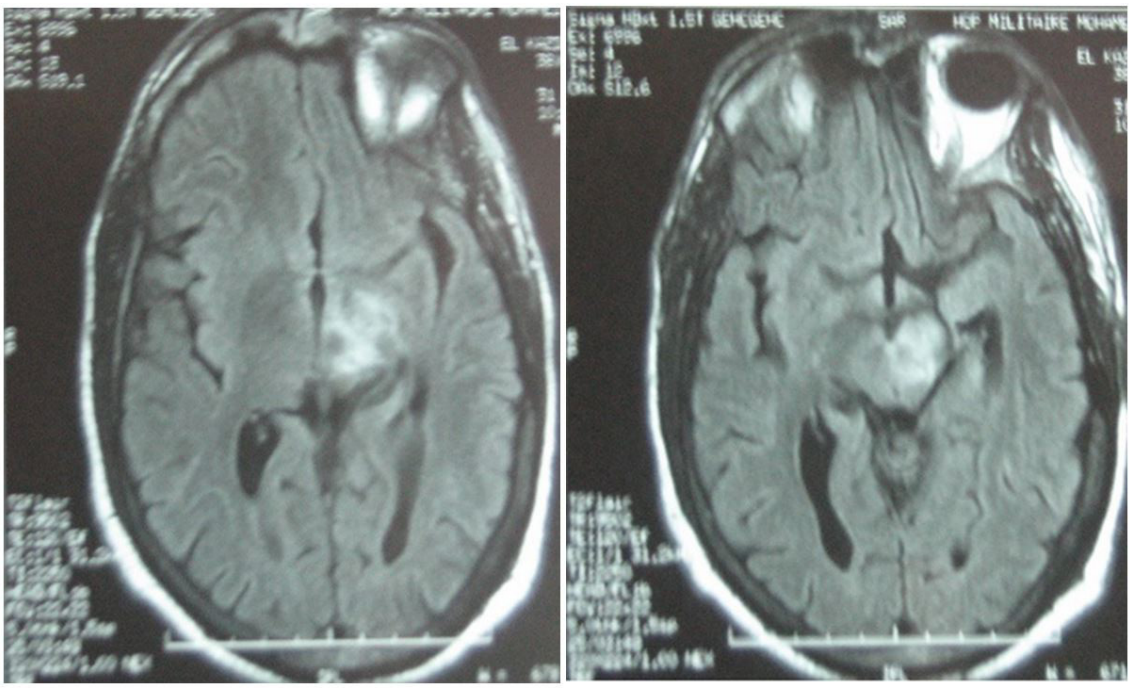

Figure 7: Brain MRI axial section in Flair sequence showing a left midbrain-diencephalic high signal

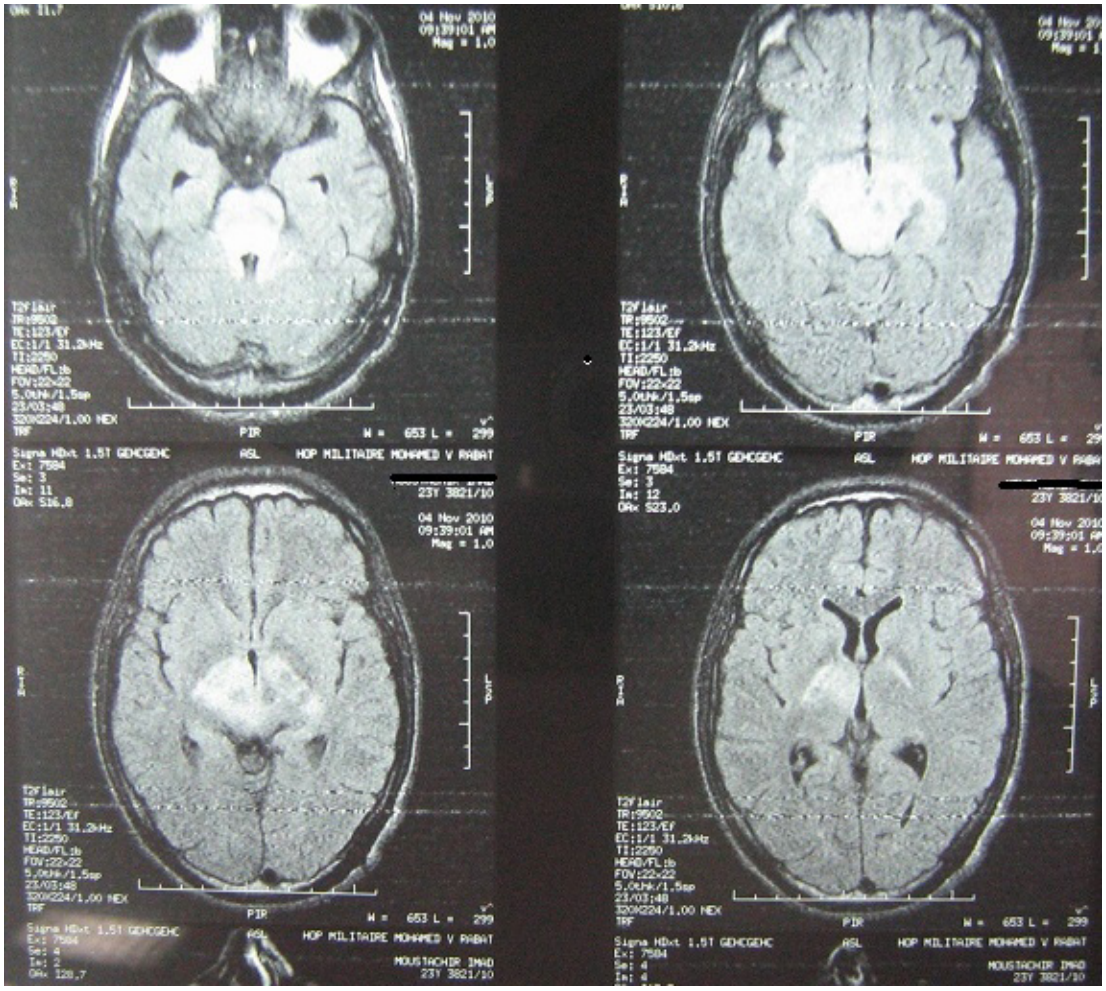

Figure 8: Brain MRI, axial section, Flair weighted images showing bilateral extended hyperintense lesions in midbrain-diencephalic regions

Conventional cerebral angiography currently supplanted by MRI angiography (Figures 9 and 10), is used in the absence of certainty image. In our study, we used conventional angiography in 20 cases and showed basal vein of Rosenthal (BVR) thrombosis in 15 cases, associated with right LS thrombosis in one case (Figures 11).

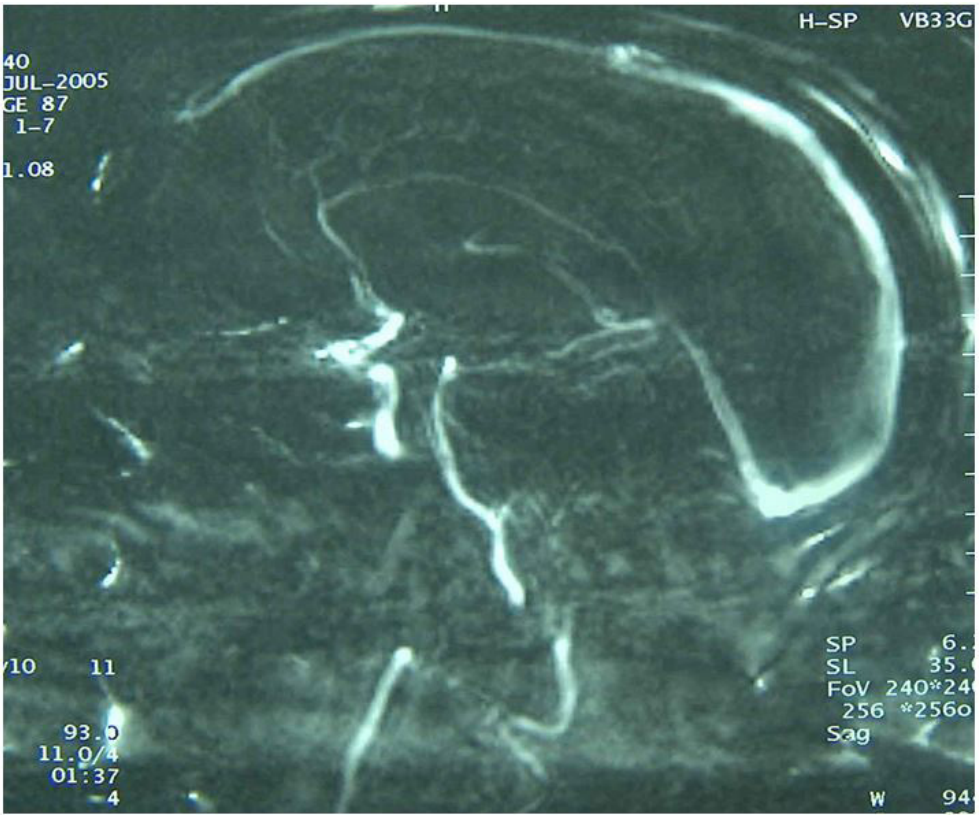

Figure 9: MR angiography with venous phase showing a lack of visualization of the basal vein of Rosenthal

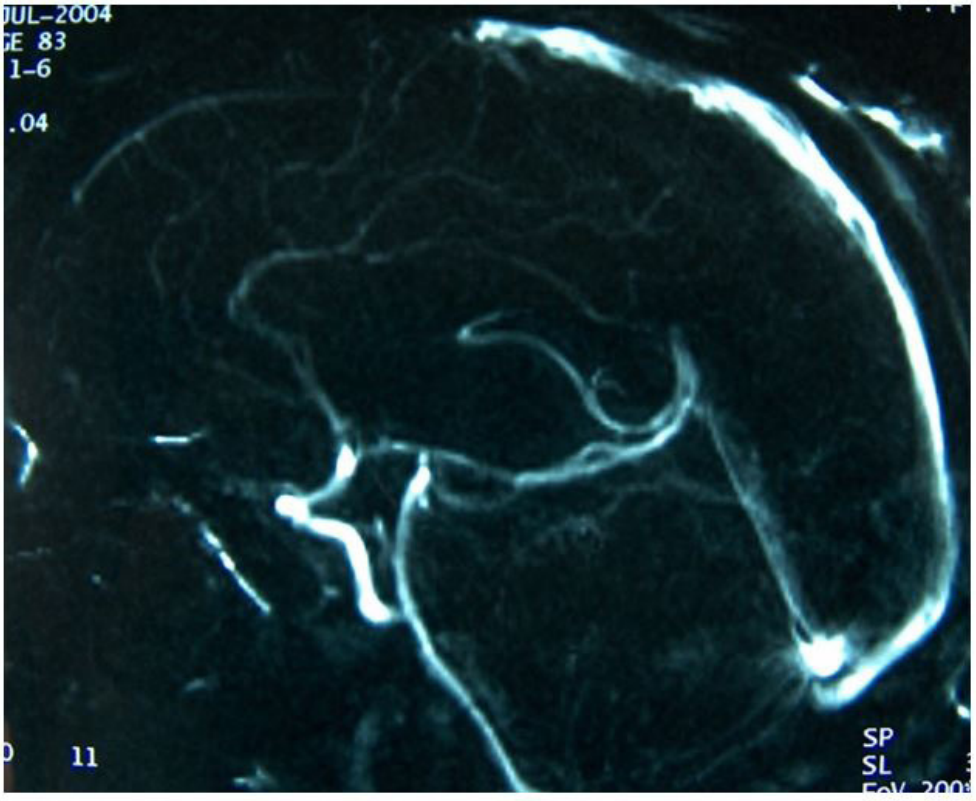

Figure 10: MR angiography in venous phase showing non visualization of the superior sagittal sinus

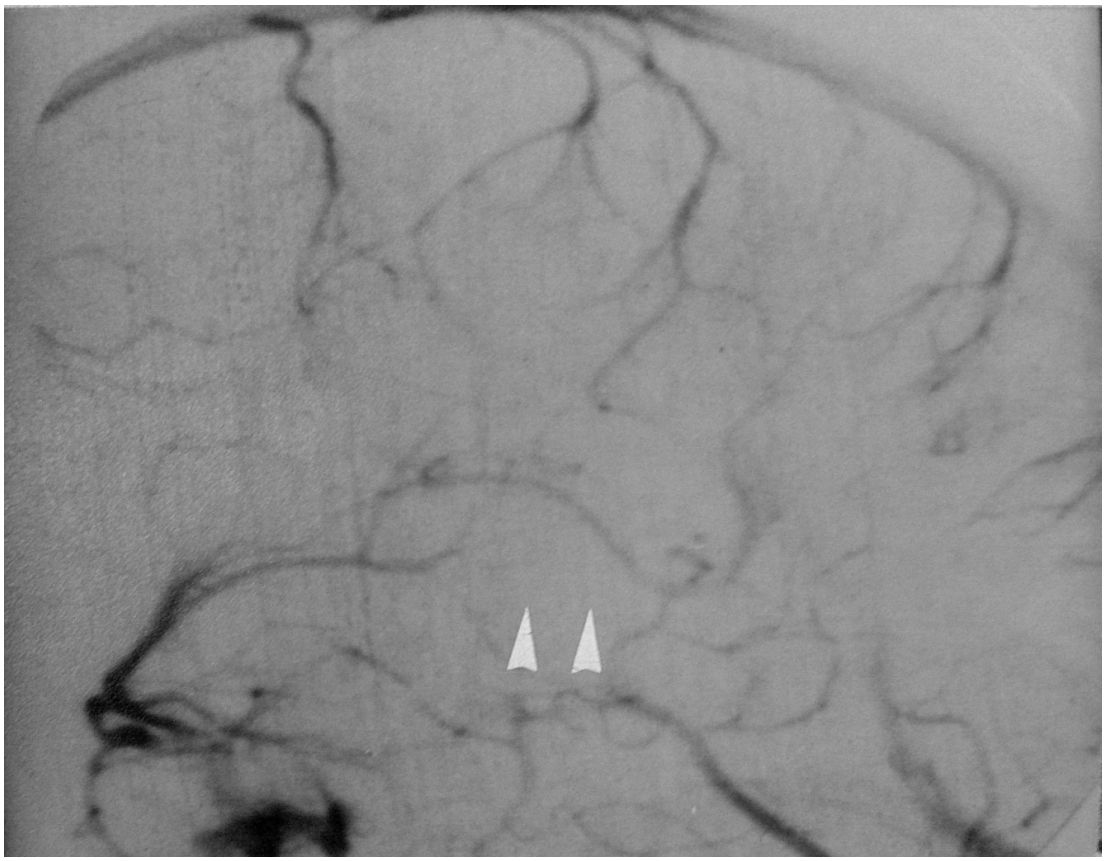

Figure 11: Cerebral angiography (venous time) showing a lack of basal vein of Rosenthal visualization

Lumbar puncture showed normal CSF in 8 cases, lymphocytic hypercellularity in 14 cases, CSF protein level no more than 1 gramme in 8 cases and a high CSF protein level without cellular reaction in 2 cases. The fundus revealed uveitis in 8 cases, and papilloedema associated with intracranial hypertension in 4 cases.

Topographic diagnosis

Our series is individualized by the predominance of deep vein thrombosis in 18 cases (Tables 4 and 5).

	Saadoun[40] (n=64)	Yesilot [110] (n=36)	Our series (n=24)
Sudden onset	35	5,5	83,3
Progressive onset	64	88	16,6
Headache	96,9	100	37
Focal motor deficit	12,5	6	83,3
Consciousness disorders	62	-	16,6
Superior sagittal sinus	64,1	52,7	16,6
Lateral sinus	61	61	12,5
Deep CVT	6,3	0	75

Table 5: Comparison of clinical characteristics and location of the CVT in the different series (%)

Evolution after treatment

In our series, the prognosis of the 6 patients presenting with superficial sinus thrombosis, was favorable after anticoagulation and corticosteroids, with moderate persisting signs and/or symptoms in 4 patients (mild hemiparesis, headache, diplopia, blurred vision, seizures) and complete recovery in 2 patients. However the persisting sequela were serious in the 18 presenting deep parenchymal lesions (moderate to severe hemiparesis, tetraparesis, III nerve palsy, hemiataxia, dysarthria, dysphagia, peudobulbar syndrome) (Table 2). Furthermore, the persistent sequelae, were mainly represented by partial motor deficit in 20 cases.

Discussion

Behçet's disease (BD) is a chronic relapsing multisystem vasculitis which the diagnosis is retained according to the International Study Group Criteria for Behçet diseases 2014. The pathophysiology of the disease involves the role polymorphic inflammatory infiltrate that predominates in the media and adventitia of large, medium and small vessels [1,2].

Sign/Symptom	Points
Ocular lesions	2
Genital aphthosis	2
Oral aphthosis	2
Skin lesions	1
Neurological manifestations	1
Vascular manifestations	1
Positive pathergy test	1

International Criteria for Behcet's Disease – point score system: scoring 4 indicates Behcet's diagnosis

Neurological manifestations account for 10 to 40% of all disease complications. The neurological complications involoving central nervous system (CNS) are classified into two main patterns, a parenchymal pattern "neuro-Behcet's syndrome" (NBS), and "vasculo-Behcet", with non-parenchymal involvement, described as thrombosis of large venous sinuses, large artery occlusion, aneurysm, and hemorrhage. This classification represents 2 groups with different clinical, aetiopathological, therapeutic, and prognostic features [2].

CVT is the most common manifestation of BD, 6 to 45% of all neurological complications. In Morocco, there was a low prevalence of about 3.8 and 4.4% and there is a male predominance in CVT occurring during BD [3,4]. The frequency of "vasculo-Behcet" in behçet disease with neurological involvement is estimated between 40 to 45%, according to the results two important studies (Farah *et al.*, 1998; Al-Araji A *et al.*, 2003). However the frequency of the vascular form, mainly represented by CVT in our series was much higher (87,5%) [3-6].

The CVT is a real chameleon, the time between first signs of Behcet's disease and CVT installation is variable with an average of 5 years. The mode of symptoms onset is also variable. It can be brutal with clinical signs within 48 hours or progressive [4-6]. In our series, the onset was brutal in 83,3% of our patients, comparing with the progressive onset in series of Saadoun *et al.* (64%) and Yesilot *et al.* (88%) (Table 5). Furthermore, the symptoms are variable according to the location, extension, etiology, age of the patient and the interval time between the onset of symptoms and diagnosis time.

In most cases, CVT affects simultaneously several sinuses and/or cerebral veins. The involvement of the superior sagittal sinus is the most common finding (64,1% in Saadoun *et al.* series; 52,7% in Yesilot *et al.* series; 16,6% in our series) and is often associated with other sinus thrombosis, in particular the lateral sinus, or cortical veins. Thrombosis of lateral sinuses is as common (61% in Saadoun *et al.* series; 61% in Yesilot *et al.* series; 12,5% in our series), but the involvement of the cavernous sinus is less frequent. Cortical vein thrombosis often present a mixed picture, and deep CVT are usually severe with usually sudden onset and rich clinical presentation [4,5].

Deep CVT usually reveal the disease, and the BVR thrombosis causing diencéphalo- mesencephalic syndrome is a singular clinico-radiological syndrom which is evocative of angio- Behçet. In our series, we found deep CVT proportion of 75%, which is very different from previously published data (6.3% in Saadoun, and 0% in Yesilot *et al.* series). Similar constations were noted for dural sinus thrombosis. The proportion of superior sagittal sinus was 16,6% in our series (64,1% in Saadoun, and 52,7% in Yesilot *et al.* series), while the proportion of lateral sinus thrombosis is 12,5% (61% in Saadoun, and 61% in Yesilot *et al.* series). This discrepancy between results is probably explained the role of the genetic and epigenetic and environmental factors in variation of disease phenotypes between different populations.

In vasculo-Behcet, major cerebral vessels parietal lesions caused by vasculitis of the vasa vasorum cause a media degeneration and thrombosis that play an important role than changes in hemostasis. There are mainly thrombosis of cerebral veins and dural sinuses (6 to 45%). Superficial CVT cause intracranial hypertension with papilloedema in the fundus and neurological deficits in

the case of venous infarction. Deep CVT (Internal cerebral vein and BVR) cause brainstem synd, pyramidal and/or thalamic synd. However, arterial thrombosis or aneurysms are much rare [2,4,5]. Motor signs and symptoms, were the most common findings during the disease course of our patients (87,5%), in agreement with previous reports [3,6,7].

According to Shahien R and al, parenchymal central nervous system pattern represent 60 to 81% of all neurological involvement [3]. It is generally believed that the underlying pathogenesis in parenchymal CNS involvement is small vessel vasculitis. Inflammation (lymphocytes and macrophages) affecting preferentially the small and medium-size veins, interesting preferably the brain stem, basal ganglia region, supratentorial white matter (SB) and the meninges. The neuro-Behcet's disease (NBD) usually shows three clinical patterns, brainstem syndrome, meningencephaloomyelitic syndrome and an organic confusional syndrome. However, several studies showed that parenchymal involvement of CNS in behcet's syndrome might not be part of a more generalised vasculitis localised in the CNS system. However, Hadfield *et al.* described the postmortem examination of a patient with neurological disease who died after a protracted course. Inflammatory cells had accumulated in the central nervous system, but no evidence of bone fide vasculitis was seen. Nevertheless, in our study we found 15 cases of BVR thrombosis causing diencéphalo- mesencephalic syndrome. Therefore, parenchymal form of BD with similar lesions, can represent in one case "neuro-Behcet's syndrome" (NBS), and "vasculo-Behcet" in other, which make difficult the presumption of its underlying pathogenesis and its classification [7,10,11].

Most of the patients with major vessel disease had disease in their major veins (superficial thrombophlebitis=32%). Some observations demonstrated that CVT usually started nearly a decade earlier than the parenchymal involvement in BD. These data suggest that the CVT in BD may have a different pathogenic mechanism from the parenchymal type. Therefore, it was proposed that there is a mechanism in some patients with BD that make them more prone to thrombosis in veins of all sizes, including dural sinuses [7,9,12,13].

The CT scan remains the initial examination for descrambling. Cranial MRI and MRV are very helpful in categorising the neurological involvement. Both should be used routinely in cases of suspected neuro-Behcet's disease, especially MRV, which may direct the attention to the possibility of CVT requiring anticoagulation. It may also reduce the need of more invasive neuroimaging in suspected cases of CVT [2,5,7-9]. In our series, magnetic resonance angiography showed 4 cases of sagittal sinus thrombosis and 3 cases of lateral sinus thrombus. However, the benefit of conventional angiography was indeniable, by confirming the diagnosis of BVR thrombosis in 75% of our patients, who were initially classified as "parenchymal form".

The treatment CVT in the setting of BD is based on three parameters; symptomatic treatment, long term antithrombotics (heparin followed by oral anticoagulation) and the control of BD by corticosteroids and/or immunosuppressants. It is well established that prognosis of non- parenchymal impairement (CVT) is better than parenchymal one, and functional recovery capabilities are greater in CVT than in arterial stroke [8,9,13]. The other poor prognosis factors are both clinical (old age, presence of focal signs or coma, hemorrhagic infarction) and radiological (positive delta sign on CT, deep venous system or posterior pit veins thrombosis).

Conclusions

BVR thrombosis causing a diencéphalo- mesencephalic syndrome is a clinicol- radiological aspect evocative and singular of angio-Behçet. Thanks to advances in imaging, in particular angiography and angio-MRI, diagnosis of CVT has become inescapable. A therapy combining anticoagulation and immunomodulatory treatments has remarkably improved the prognosis of this disease but with persistence of sequelae dominated mainly by the motor deficit, headache and seizures.

Conflict of interest

The authors declare that they have no conflict of interest

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