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*Case Report*

# An Upper Digestive Hemorrhage Revealing Behçet's Disease: About a Case

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## ABSTRACT

### Summary

Behçet's disease is a systemic vasculitis of unknown etiology, characterized by oral and genital aphthosis and uveitis. The most common gastrointestinal location is the colon and the ileocecal region. The diagnostic criteria were established in 1990 by a classification of the International Study Group for Behçet's Disease. The digestive involvement of Behçet's disease has many similarities with inflammatory bowel diseases (IBD), Crohn's disease in particular. We report the observation of a 34-year-old patient with no previous pathological history, admitted for digestive bleeding. Attention was paid to the presence on clinical examination of a cerebellar syndrome, mouth ulcers in the oral cavity, aphtoïd ulcerations in the peri-anal region, contemporary with pseudo folliculitis of the back. This clinical picture suggests a priori Behcet's disease; a pathergy test was carried out, which came back positive, as well as specific biological, radiological and endoscopic explorations then confirming the diagnosis of neuro-Behcet.

**Keywords:** Digestive hemorrhage, mouthulcers, pseudo-folliculitis, cerebellar syndrome, neuro-Behcet

## 1. Introduction

Behcet's disease is a systemic vasculitis of undetermined etiology, described by the Turkish dermatologist Hulusi Behçetin 1937. The hypothesis of the occurrence of this disease would be an inflammatory response induced by environmental factors or infectious agents. The diagnosis is essentially clinical. The neurological damage is called neuro-Behçet, this name was made by Cavara and D'Ermoin 1954. The neurological and ocular damage makes the disease serious.

## 2. Observation

Patient D.A. aged 34, chronic smoker with a history of allergy to wool, consulted for minor hematemesis in three episodes over 3 successive days with dizziness, balance disorder and profound asthenia. In-depth questioning reveals a notion of repeated oral ulcers > 3 times/year.

The clinical examination found: A conscious, uncooperative patient, presenting dysarthria, afebrile, with stable hemodynamic constants, BMI: 24.

On oral examination, we found Perlèche of the corners of the lips and a sore on the inner side of the lower lip.

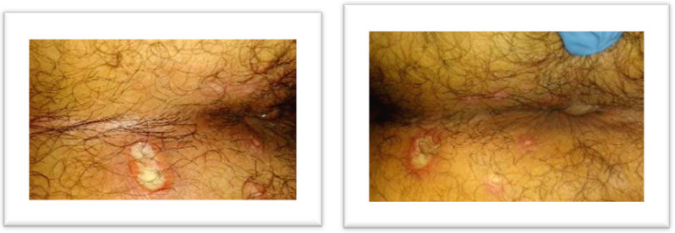
The abdominal examination was unremarkable. Proctological examination reveale dpainfulperi anal aphthoid ulcerations (Figures 1 and Figure 2). Examination of the genital tract showed a genital ulcerscar at the scrotal level.

Examination of lymph node areas, cardiovascular and pleuropulmonary examinations were unremarkable.

The joint examination motivated by the knee pain did not find any sign of knee arthritis. The neurological examination revealed a cerebellar syndrome (cerebellar ataxia, pendulumosteotendinous reflexes on the left, diminished on the right).

Skin examination revealed pseudo-folliculitis and acneiform nodules on the back. Pathergy test carried out +++: This is a hypersensitivity to the skin points, the injection is done on the anterior surface of the for armsubcutaneously and/or intra venously.

The reading is taken after 48 hours, this test reproduces pseudo-necrotic folliculitis which results in the appearance of a pustule with aredareola (circle) reflecting the inflammation figure.



**Figures 1&2:** Perianal aphthoid ulcerations.

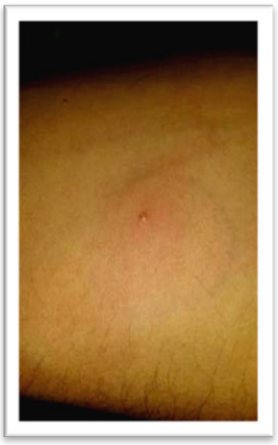


**Figure 3:** Pathergy test.

The biological assessment carried out on the patient revealed neutrophilic polynucleosis. No inflammatory syndrome: CRP negative. Negative HIV, HBV, HCV, HSV, CMV viral serologies. HLA B51 typing results inconclusive.

Morphological assessment Eso-gastro-duodenal fibroscopy (FOGD) was performed revealing congestive gastro-bulbitis, without bleeding marks (Figure 4).

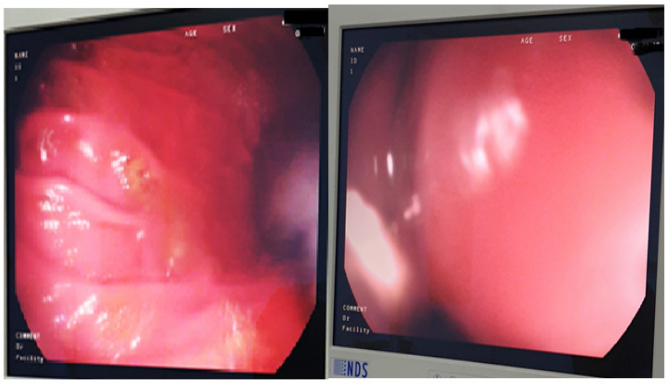


**Figure 4:** FOGD endoscopic appearance of a congestive gastro-duodenal mucosa.

Duodenal biopsies looking for associated IBD were performed. The anatomo-pathological study found a histological appearance in favor of subacute duodenitis. Recto-sigmoidoscopy: revealed erosive recto-sigmoiditis.

Rectal and sigmoid biopsies performed. (**Figure 5**). A complete colono-scopy performed under-premedication: absence of specific IBD lesions. Pathological study: non-specific subacute recto sigmoiditis.

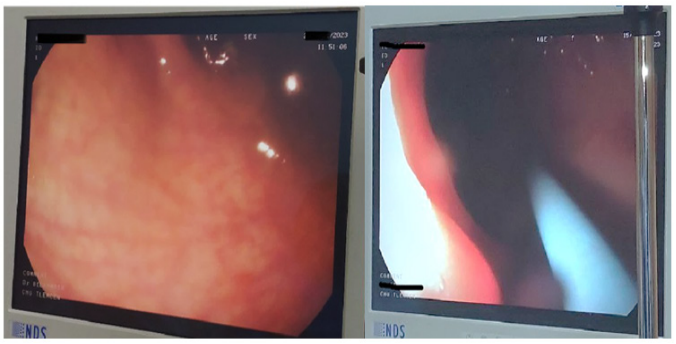


**Figure 5:** Rectoscopy: Image A congestive rectal mucosa, image B sigmoidbiopsy.

Front and profile radiograph of the knee without abnormalities (Figure 6A, Figure 6B).

Telethorax: Normal.

An ophthalmological examination including avisualcuity test, a slit lamp examination and fundus examination were carried out, concluding that there were no signs of uveitis.

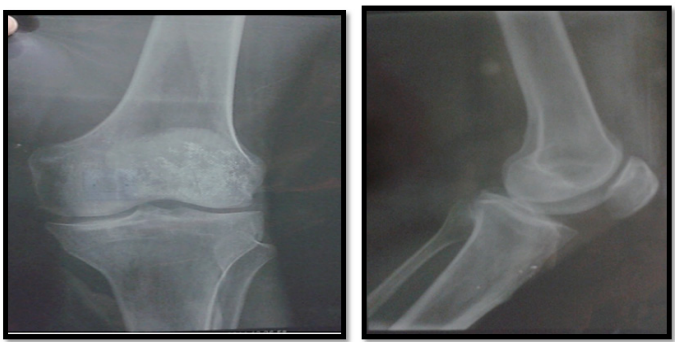


**Figure 6:** Image 6A x-ray of the normal side of the knee image 6B: x-ray of the normal side of the knee Brain MRI: hyper intense lesions suggesting, depending on the clinical context (sudden onset), acute encephalomyelitis or neuronitis. Behcet's disease cannot beruled out.

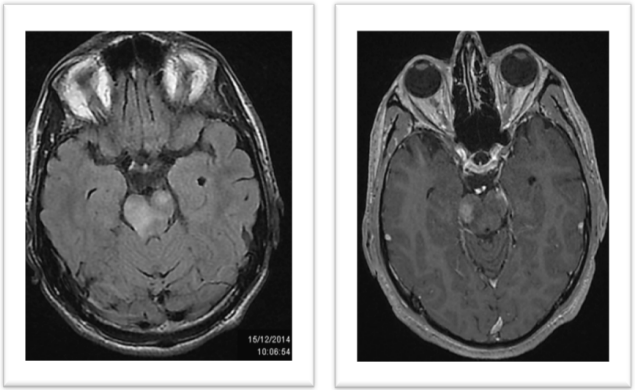


**Figure 7:** Magnetic radiation imaging (MRI) sagittal section; A) before injection; B) after injection.

The spinal MRI showed a dorsal intramedullary plaque mainly facing D8, discreetly enhanced after the gadolinium injection, probably linked to his neuro Behcet.

A lumbar puncture with biochemical and bacteriological study was performed.

Results: 12 white elements/mm3

Leukocytes: 29 elements/mm3

Lymphocytes: 80%

PNN: 20%, absence of red blood cells

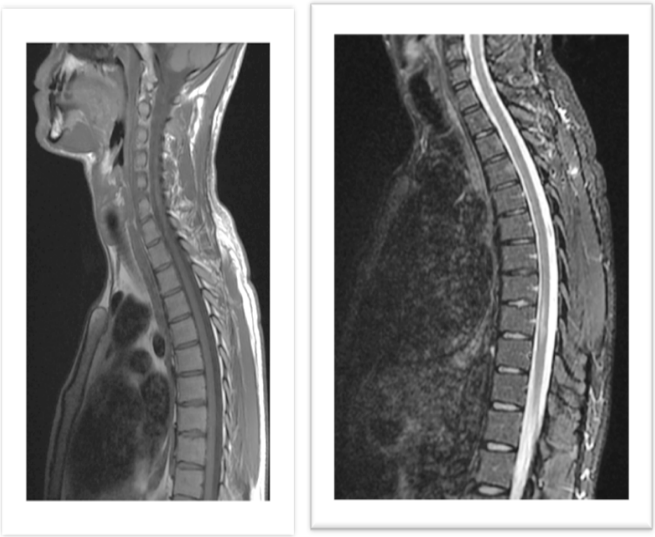


**Figure 8:** MRI in sagittal sections shows an enhancement of a medullary plate next to D8.

Treatment and evolution

Symptomatic treatment:

-Aspégic mouth wash.

-Colchicine 01 mg 2 times a day

Corticosteroid bolus 500 mg/day for 3 days.

Oral relay 01 mg/kg of Prednisone (70 mg/day) with supplementation of calcium, potassium, proton pump inhibitors, salt-free diet and physical activity.

Good clinical-biological evolution under corticosteroid therapy.

The patient has been on Imurel for four years, given the discovery of spermogram disorders preventing the use of Cyclophosphamide.

## 3. Discussion

During Behçet's Disease (BD) we note an increased frequency of HLA-B51 Ag, but does not constitute a diagnostic criterion.

- Recurrent genital ulcerations or scarringlesions.

- Eye lesions: Uveitis, vasculitis.

- Skin lesions: Erythemanodosum, pseudo folliculitis, papulo-pustularlesions, acneiform nodules.

- Positive pathergic skin test.

Skin involvement: Represented mainly by mouth sores (98% of cases), which can affect the inner side of the lips, the inner side of the cheeks, the gingivo-labial furrow, the edge of the tongue, the floor of the mouth, the palate, tonsils and pharynx, but canals occur on: the skin, the esophagus, the stomach and the intestine, exceptionally leading to perforations and ulcerations of the anal verge. The clinical symptoms related to digestive damage in Behçet's disease are varied and includ eanorexia, nausea and vomiting, dyspepsia, diarrhea and abdominal pain. Endoscopy canreveal segmental mucosal inflammation and aphthoid ulceration smostoften affecting the ileum, cecum and ascending colon[1,2]. Stenosis are rare, but transmural inflammation and fistulas are frequently observed.

Joint damage 50-65%: early, sometimes inaugural which can precede other manifestations by several years, it affects the large joints of the lower limbs, with a recurrent and asymmetrical character and has several aspects.

Ocular damage: Anterior, posterior uveitis and other ocular lesions: rare conjunctival ulcer, episcleritis, keratitis. The prognosisissevere.

Neurological damage 15-20%: neuro-Behçet: the frequency of neurological damage during Behçet's disease is of the order of 5%, this damage occurs around the thirties in the 4 to 5-years following the first signs with a discreet male predominance in whom the attack appears more severe. In 7.5% of cases it initiates the first attack and in 3% of cases it precedes the other signs by 1 to 10 years[3,4].

It involves two main mechanisms: macro vascular venous damage (exceptionally arterial) and parenchymal damage. In a clinical study with long-term follow-up of 25 patients, the frequency of cerebral thrombo-phlebitis was 8% and the cause of a third of the neurological manifestations observed[5].

Magnetic resonance imaging and magnetic resonance angiography are the best exploration methods[6]. The involvement of cerebral arteries is exceptional. These may be an eurysms[7] which are symptomatic or responsible for stroke.

Parenchymal involvementis diffuse, although with a predilection for the brainstem; it is generally severe and has a poor prognosis[4,8]. Cognitive disorders mainly affecting memory have been noted in the absence of neurological damage[9]. Myelitis[10], Parkinson's syndrome, internuclear ophthalmoplegia, mutism, attacks of ataxia or dysarthria, sensory disorders, abnormal movements have been reported. The CSF is most often abnormal but does not rule out an infection in the acute phase, especially since a fever is possible. Hyper cytosisis the rule.

Protein levels are high and indicate damage to the blood-brain barrier. Peripheral damage is exceptional with a frequency of 2 to 6% of cases of neuro-Behçet, it manifests as polyneuritismultineuritis or polyradiculoneuritis[11,12]. MRI is the reference examination for diagnosis and progressive monitoring[6]. Digestive damage: the symptoms are nonspecific, particularly digestive bleeding. Biologymakeslittle contribution, inconstant inflammatory syndrome. Treatment: It aims to initially combat inflammatory lesions with strong corticosteroid therapy 01 mg/kg/day, possibly initiated by boluses of methyl-prednisolone (usually 01 g 03 days in a row) and to prevent the usual relapses during the reduction. corticosteroids by the addition of an immunosuppressive treatment (Azathioprine[13] 2.5 mg/kg/day or Cyclophosphamide intra-venously 1 g every 4 weeks, see interferon Alpha[14].

The decision to choose the treatment is easy when the diagnosisis certain, otherwise in the face of particularly recurrent mening oencephalitis and when any other etiology is reason ably ruled out, trial corticosteroid therapy is worth trying under the cover of probabilistic anti-infective treatment. .

The duration of treatment is poorly codified. Immunosuppressants are offered for a minimum duration of 2-years, corticosteroid therapy is gradually reduced. The neurological prognosis is severe with up to 28% recurrence

and 14% disability at 3-years for KIDD[15] and 50% dependent and death for Akman-demir[3,4], this progressive risk makes the treatment imperative and rapid implementation.

## 4. Conclusion

Perianal aphthosis is rare and must rule out IBD. A complete clinical examination is essential for any digestive symptom because it can reveal a systemic disease whose spontaneous prognosis sun favorable but can be improved by early diagnosis and treatment.

## 5. Conflicts of Interest

The authors declare no conflict of interest.

## 6. Author Contributions

All authors approved the final version of the manuscript.

## 7. Consent for Publication

Written informed consent to publish this information was obtained from. The parent of the study participant.

## 8. Source of Funding

None.

## 9. Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

## 10. References

1. H Yazici, H Pazarli, C G Barnes, et al. (1990) A controlled trial of azathioprine in Behçet's syndrome. N Engl J Med, 322: 281-285.
2. G Akman-Demir, P Serdaroglu, B Tasçi, (1999) Clinical patterns of neurological involvement in Behçet's disease: evaluation of 200 patients. The Neuro-Behçet Study Group. Brain, 122: 2171-2182.
3. B Wechsler, M Vidailhet, JC Piette, et al. Cerebral venous thrombosis in Behçet's disease: clinical study and long-term follow-up of 25 cases. Neurology, 42: 614-618.
4. R Monastero, Cecilia Camarda, Carmela Pipia, et al. (2004) Cognitive impairment in Behçet's disease patients without overt neurological involvement. J Neurol Sci, 220: 99-104.
5. G Akman-Demir, S Bahar, O Coban, et al. (2003) Cranial MRI in Behçet's disease: 134 examinations of 98 patients. Neuroradiology, 45: 851-859.
6. (1990) Criteria for diagnosis of behçet's disease. International study group for Behçet's disease, Lancet, 335: 1078-1080.
7. O Kizilkilic, S Albayram, I Adaletli, et al. (2003) Endovascular treatment of Behçet's disease-associated intracranial aneurysms: report of two cases and review of the literature. Neuroradiology, 45: 328-334.
8. JC Nichols, A Ince, L Akduman, et al. (2001) Interferon-alpha 2a treatment of neuro-Behcet disease. J Neuroophthalmol, 21: 109-111.
9. S. Gerber, A Biondi, D Dormont, et al. (1996) Long-term MR follow-up of cerebral lesions in neuro-Behçet's disease. Neuroradiology, 38: 761-768.
10. https://link.springer.com/article/10.1007/s12157-008-0047-8
11. M. Waldner, C Schimanski, W Klaas, et al. (2006) Massive hemorrhage in a 28-year-old male patient with intestinal Behçet's disease. Z Gastroenterol, 44: 1047-1051.
12. M Calgüneri, Ahmet Mesut Onat, M Akif Oztürk, et al. (2005) Transverse myelitis in a patient with Behcet's disease: favorable outcome with a combination of interferon-alpha. Clin Rheumatol, 24: 64-66.
13. https://www.em-consulte.com/article/105434/neuropathie-peripherique-au-cours-de-la-maladie-de
14. DP Kidd (2015) Rituximab is effective in severe treatment-resistant neurological Behçet's syndrome. J Neurol, 262: 2676-2677.
15. Bitik B, Ucar M, Tezcan ME, et al. (2013) Transverse myelitis in Behçet's disease: A series of four cases and review of the literature. Clin Exp Rheumatol, 31: 20-24.