CLINICAL PRACTICE

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Behçet disease – Case presentation

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Abstract: *Behcet's disease is a rare and poorly understood condition with multiple systemic manifestations. The disease causes inflammation in blood vessels throughout the body which leads to numerous symptoms that may appear and disappear unpredictable.*
Case presentation
A 33 year old woman was admitted (interned) to our clinic. Her family medical history reveals – multiple strokes (father) and autoimmune thyroiditis (sister).
The onset of her symptomatology was in 2013 and it consisted of fever (39-40 Celsius degrees) sicca syndrome and persistent headaches, recurrent oral and genital ulcerations.
An important event is essential to be mentioned -the patients has suffered an episode of upper gastrointestinal bleeding (hemoglobin has dropped to 2.5g/dl which has led to cardiac arrest-resuscitated).
The lab tests showed: C3 hypocomplementemia, Anti-centromere antibodies (-), anti-b2gp1 antibody (+), lupus anticoagulant (+), U1RNP(-).
We started to administrate cyclophosphamide to the patient, thus his condition has improved after the 2nd dose.
At the 12-month evaluation we were able to see a significant clinical and biological improvement.
Conclusions
In order to be able to talk about "evidence based medicine" for the management of Bechet's disease, a large number of clinical trials is required to provide to the attending physicians the necessary data for diagnosis and treatment.

INTRODUCTION

Behcet disease is a rare condition with a sinusoidal evolution which can affect almost all the organs and human systems, determining multiform symptoms with a variable severity. The medical practitioner has at his disposition a diversity of therapeutic options (from symptomatic therapy to a major systemic immunosuppression); at this moment we can't discuss about fundamental therapeutics protocols and unanimously recognized.

Behcet disease is an inflammatory systemic condition with an uncertain etiology. In the disease pathogenesis both environmental and genetic factors are involved like for the majority autoimmune diseases. This disease has been worldwide described but the maximum prevalence is registered in Middle East, adjacent zones of Mediterranean Sea and Asia

Considering the potential muco-cutaneous, articular, ophthalmic, neurologically, etc. damages, the interdisciplinary collaboration becomes more important. Due to the lack of specific guidelines for the diagnosis and treatment, the therapeutic approach of

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individuals' cases still remains dependent on the doctor treating experience or on the clinics where these complex cases are monitored.

CASE PRESENTATION

A 33 year old woman was admitted to our clinic presenting severe muscle weakness and joint pain.

Her medical history has revealed that in the last years the patient was admitted several times in the Infectious Diseases department with persistent fever and severe headaches.

Her family medical history has revealed – multiple strokes (father) and autoimmune thyroiditis (sister).

The onset of her symptomatology was in 2013 and it consisted of fever (39-40 degrees Celsius) sicca syndrome and persistent headaches. As the disease was progressing, our patient presented oral ulcerations, odynophagia, muscle weakness and blindness of the left eye.

The onset of the decreased visual acuity started with the right eye. An ophthalmologic examination has revealed uveitis and chorioretinitis.

Considering an inflammatory demyelinating disease (multiple sclerosis), the neurological exam suggested a cranial magnetic resonance imaging (which has revealed optic nerve edema) and anti-aquaporin 4 antibodies screening (which was negative). The patient treatment has started on Corticosteroids (methylprednisolone in pulse therapy) resulting a slight improvement of visual acuity, persistent joint pains, xerophtalmia.

An important event is essential to be mentioned -the patients has suffered an episode of upper gastrointestinal bleeding (hemoglobin drops to 2.5 g/dl which leads to cardiac arrest – resuscitated).

When the patient was admitted to our clinic the symptoms become more aggressive; the patient present extreme fatigue (with orthostatic intolerance), recurrent oral and genital ulcerations, odynophagia (due to the pain caused by oral and esophageal ulcerations she was not able to solid food or liquids) and blindness of the left eye.

Figure1: Atrophy of retinal layer

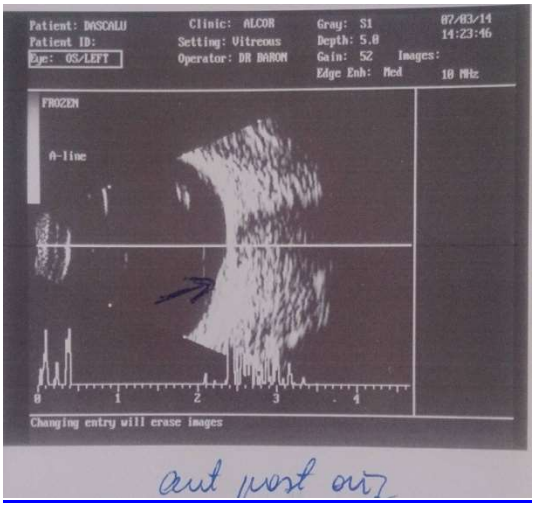
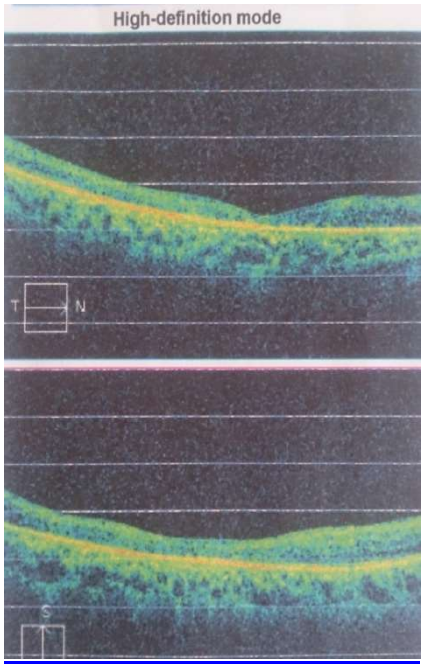


Figure 2: FOS – thicknesses of retinal layer



Despite the muscle weakness and severe fatigue that the patient suffered from throughout this whole period, muscle enzymes was never elevated.

Nevertheless we continued our investigations with an electromyogram. The EMG showed acute polymyositis aspect and sensory polyneuropathy lesion.

Then neurological exam showed spastic paraplegia, left brachial plegia and visual acuity loss. The brain MRI revealed a lacunar ischemic stroke in the vertebrobasilar territory.

The ophthalmological exam showed optic nerve atrophy.

We were facing with a patient, having a chronic, inflammatory multisystem condition of unknown etiology. The blood tests revealed nothing but a persistent inflammatory syndrome (ESR = 80-100 mm/h); the immunology tests didn't provide enough information either (C3 hypocomplementemia, Anti-centromere antibodies (-), anti-B2GP1 antibody (+) lupus anticoagulant (+), U1RNP (-), anti-aquaporin 4 antibodies (-) HLA B51 (-), Leiden V factor (+), ANA (+).

However we were in possession of his clinical examination highlights: chronic fatigue, recurrent mucosal ulceration, neurological symptoms, bilateral posterior uveitis, inflammatory arthritis various symptoms that we were able to identify as features of Behcet's disease.

By the time an accurate diagnostic could be established, the patient had already suffered some severe life threatening complications: upper GI bleeding with severe anemia that led to cardiac arrest, severe sepsis caused by infected decubitus injuries and medullar aplasia caused by high dose of immunosuppressive medication.

Behcet disease – generally accepted therapeutic objectives:

1. It's essential having a clear diagnostic and well founded as well as a clear setting of a lesional exact balance, with the progressive potential appreciation of the identified lesions;

2. The remission induction and maintenance;

3. The prevention of irreversible organs lesions (for example the ophthalmological one), which tends to appear early, especially in some problematic groups like young men;

4. The articular and muco-cutaneous exacerbations prevention can significantly improve the quality of life;

5. The multisystem involvement imposes for most cases interdisciplinary implication.

We want to underline the importance of having an accurate and precise diagnostic before starting a treatment, but in this particular case, a rapidly evolving disease worsened by the upper mentioned events, we decide that it was extremely necessary for the patient to start a remissive treatment. We initiated a moderate dose of cyclophosphamide (800 mg – 15 mg/kg bolus intravenous) and a low dose of oral corticosteroid (methylprednisolone 8 mg).

A significant improvement was observed after the second course of cyclophosphamide. The patient has regained the strength and muscular tonus, neurological symptoms has disappeared and she did no longer mucosal ulcerations.

Although the inflammatory ocular symptoms were suppressed, the significant loss of visual acuity persisted (it was caused by initial optical neuritis).

At the 12-month evaluation we were able to see a significant clinical and biological improvement. The patient is now able to do her daily activities therefore her lifestyle has improved.

The brain MRI shows normal brain parenchyma, normal vascular structures and normal blood flow.

Ophthalmological exam: minor restant vitrean abnormalities.

CONCLUSIONS

In the past decade we have witnessed a significant improvement in the therapeutic approach of Behcet's disease by optimal use of the old available drugs and also by the development and clinical use of new therapeutic solutions.

Behcet's disease still remains a multidisciplinary challenge for attending physicians.

Available data in the medical literature consists mainly in clinical cases and series of clinical cases with a limited statistic value.

In order to be able to talk about "evidence based medicine" for the management of Bechet's disease, a large number of clinical trials is required to provide for the attending physicians the necessary data for diagnosis and treatment algorithms.

Abbreviation

[1] U1RNP – Anti-Ribonucleoprotein

[2] EMG – Electroyography

[3] ESR – Erythrocyte Sedimentation Rate

[4] HLA B51 – Human leukocyte antigen B51

[5] ANA – Anti-nuclear antibody

[6] GI – Gastrointestinal

[7] MRI – Magnetic resonance imaging

[8] B2GP1 – Beta 2 Glycoprotein 1

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