Neuro Behçet and Pregnancy: a Case Review

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ABSTRACT

Behçet’s syndrome is a multiple system chronic inflammatory disorder, characterized by the recurrence of genital and oral ulceration, and in most cases, iridocyclitis. The causes of this disease are largely unknown, but are related to autoimmune factors, and can usually lead to thrombotic complications and different sorts of vasculitis. Behçet’s syndrome flares sometimes occur during pregnancy, and thus to complications for to the mother and the fetus. Neuro Behçet is a rare and fatal complication that occurs many years after the installation of the main symptoms. Our case is about a 33 years old patient diagnosed with Behçet disease five years ago, under colchicine, who was affected with lateral cerebral thrombosis. Pregnancy associated with Neurobehçet needs to be carefully planned and followed up with strict checkups of the mother and the fetus. The treatments have to be adjusted to avoid any malformations to the fetus and complications to the mother.

Keywords: gynecology obstetrics, Behçet, Neurobehçet, Systemic Vasculitis

1. INTRODUCTION

Behçet’s disease is a multi-systemic inflammatory Vasculitis, which combines triple symptom complex of recurrent oral and genital aphthous ulcers and uveitis. It is more common in the Middle East and the Mediterranean area. Neuro-Behçet develops over a time interval after the onset of Behçet disease, the neurological involvement can be severe, and its evolution can be fatal(1).

2. CASE REPORT

The patient is a 33 years old, diagnosed with Behçet disease five years ago under colchicine, with a hospitalization two years ago in a neurology department for intracranial hypertension, during which an ophthalmological checkup revealed a papillary edema, and a brain MRI revealed a lateral sinus thrombophlebitis. The immunological test of antinuclear antibody was negative; a Neuro-Behçet was then diagnosed. The patient received a bolus of solumedrol and anti-coagulation with Low-Molecular-Weight Heparin that was relayed by an oral corticosteroid and Anti-vitamin-K.
The patient is a Gravida 2 Para 0. G1: abortion, G2 presented at the 17th week of gestation, with a normal clinical examination with no aphthous, or any eye or neurological complaints, except for a genital ulcer scar on labia minora. A prenatal assessment and serology of the mother were without peculiarities, antepartum fetal morphological ultrasound at 34 weeks of gestation (Figure 1), associated with a Doppler was realized showing no anomalies. A complete eye examination in the ophthalmology department showed no abnormalities. The patient was kept under corticosteroid and Anti-vitamin-K up to 35 weeks of pregnancy, with a decreasing corticosteroids dose.

Fig. 1: Ultrasonography at 34 Weeks showing no morphologic anomalies, with normal obstetrical Doppler

At 40 weeks of gestation and five days, the patient went into spontaneous labor, but underwent cesarean section because of suspicion of acute fetal distress (fig 2) and gave birth to a newborn female weighing 2750 g with Apgar score of 9.

Fig. 2: Extern electronic fetal monitoring showing variable decelerations
The post-partum was uneventful; patient had preventive anticoagulation, followed by colchicine treatment.

3. DISCUSSION

Behçet’s is a disease affecting young women between the ages of 20 and 30 years. Pregnancy is often associated with a flare in 20% of the cases. In the post-partum remission, rates are up to 61% with an exacerbation of about 17% [1]. Also, severe neurologic or vascular disease flares have been reported, which raise the question about the influence of Behçet disease on the outcome of pregnancy [2]. Few studies on Behçet’s disease have been done. However, some authors state that complications such as high blood pressure, gestational diabetes, prematurity, infection, premature rupture of membranes, Embolic accidents, miscarriage [3-4], are more important after the diagnosis. But according to other authors, the course of Behçet disease is variable. Flare, remission can vary to each person or even in different pregnancies in the same patient [5]. The Exacerbation itself occurs most of the time in the form of painful genital aphthosis [5,6]. During her pregnancy, the patient in our case study was in remission, and the examination was normal, only showing an old genital ulcer scar.

High thromboembolic complications could be explained by the vascular impairment of Behçet’s disease. Mostly described affections are brain thrombophlebitis, pulmonary emboli, thrombosis of the vena cava, and a Budd-Chiari syndrome [7].

The main base of treatment of Behçet’s disease during pregnancy is a regulation of the use of immunosuppressive drugs, which can be toxic to the fetus. Corticosteroids such as prednisone and prednisolone may be used without special restrictions to treat vasculitis in pregnant and breastfeeding women since the fetus is exposed to 10% of the maternal dose [8]. Azathioprine (Imurel®) is an immunosuppressant that can be used during pregnancy with the dose of 2,5mg/day [14]. However, it is noted that an increased risk of maternofetal infection (especially cytomegalovirus) due to immunosuppression induced by the treatment [10,11]. Colchicine does not seem to cause or increase any malformations [12]. The used dose is 0.5-2mg/day depends on the weight and age of the patient [13]. Other treatments may be used, such as infliximab, (Remicade®) and interferon alpha-2 especially for the recurrent cases that resist to other treatments, which must be used carefully [15].

For our part, we support what has been proposed by the Le Thi Huong [16]. The usual treatment of this Vasculitis is aspirin and colchicine that is essential to be continued during pregnancy and also while breastfeeding. If the patient is already under effective anticoagulation due to a vascular impairment of Behçet’s disease, a relay by Low-Molecular-Weight Heparin (in addition to aspirin and colchicine) is envisaged. Obstetric follow-up has to be done monthly, using ultrasound check. If a Systemic breach of the disease appears with no history of vascular injury, in addition to aspirin and colchicine, a preventive dose treatment of LMWH for about six weeks has to be proposed.

Concerning the way of delivery in the literature, there are no described specific recommendations; it is left to the hand of the obstetrician according to the clinical condition of the mother and fetus.

4. CONCLUSION

The Neurobehçet associated with pregnancy is rarely described in the literature. Behçet’s disease can compromise fetal growth. The recognition of this disease allows us to prevent prenatal complications and maternal complications associated with the disease.

REFERENCES
