

A case of Behecet's Disease in pregnancy

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Abstract

Behcet's disease is a chroinc multisystem relapsing disorder that affect the mucocutaneous surface. Though it is rare in pregnancy, One should keep the possibility of this rare disease in mind when pregnant pateint with rash and genital ulcers is seen. We wish to share our experince of managing a woman who was diagnosed as having Behcet's disease when she was 16 – year-old.

INTRODUCTION

Behcet's Disease, also known as Behcet's syndrome, is a rare, chronic, autoinflammatory disorder of unknown origin. Its manifestations are thought to be caused by vasculitis resulting in damage to blood vessels throughout the body. The disease is named for the Turkish dermatologist, Dr. Hulusi Behcet, who in 1937, described a triad of oral ulcers, genital ulcers and ocular inflammation. Although Behcet's Disease is recognized worldwide, prevalence is highest in countries in the eastern Mediterranean, the Middle East, and East Asia, along the ancient Silk Road.

Behcet's Disease tends to develop in young adults, typically in their 20's and 30's, but patients of all ages and both sexes may be affected. The disease is most prevalent in Turkey (approximately 400 cases for every 100,000 individuals) [1].

Case report

A 25 – year – old primigravida presented at 36 weeks of gestation with joint pain and painful mouth ulcers .She was diagnosed when she was 16 – year- old she was on Cholchicine 0.5 mg TDS.On examination , multiple , bluish-red , non tender nodules of varying sizes (0.4 – 2 cm) were seen on arms and legs ; bilateral axillary, and inguinal non – tender lymphadenopathy , also multiple ulcers with irregular margins over buccal mucosa. No abdominal pain and no PV bleeding .

Pateint was started on tab Erythromycin 500 mg QID & Aspirin 100mg Antepartum foetal surveillance was done using ultrasound and Umbilical artery S/D ratio were normal .Pateint went into spontaneous labour at 40 weeks and had an uneventful vaginal delivery of femal baby weighing 3 kg with Apgar score of 9 at 5 minutes.Her postpartum period was uneventful with healing of ulcers over the next two weeks.

DISCUSSION

Behcet's disease is a multisystem, relapsing, chronic vasculitic disorder with mucosal inflammation with clinical features of mucocutaneous lesions, ocular, articular, urogenital, pulmonary and neurologic involvement. It usually begins between the age of 20-30 years. The aetiopathogenesis of the disease remains unknown, though the most likely hypothesis is that of an autoimmune reaction. The basic pathologic process is vasculitis. It has been linked to HLA-B5 and HLA-DR5 allogins [2]. The various infectious agents implicated are HSV-1[3], Streptococcus sanguis and E.coli [4]. To confirm the diagnosis, at least two of the following must also be demonstrated:

- Recurrent painful genital ulcers that heal with scarring
- Ophthalmic lesions, including anterior or posterior uveitis, hypopyon, or retinal vasculitis
- Skin lesions, including erythema nodosum-like lesions, pseudofolliculitis, or papulopustular or acneiform lesions
- Positive results from pathergy skin testing, defined as the formation of a sterile erythematous papule 2 mm in diameter or larger that appears 48 hours following a skin prick with a sharp sterile needle (22-24 gauge [a dull needle may be used as a control])

Considering the above diagnostic criteria, case presentation often includes the following characteristics:

- Multiorgan system involvement, often beginning with mucocutaneous involvement and usually sparing the liver, kidneys, and heart
- Age of 25-35 years at onset
- Organ-specific manifestations characterized by exacerbations and a relapsing/remitting course

The course of disease in pregnancy and postpartum is variable with remission noted in some and activation in others [5]. Even different pregnancies have different outcomes in each patient [6]. During preganacy, there is an increase in the vascularity and hyperaemia of the skin and muscels of perineum and vulva, which leads to aggrevation of genital ulcers in pregnancy.

Also, increase of estrogen levels leading to decrease in cellular immunity may also aggravate the disease during pregnancy. There is currently no cure for Behcet's disease, but a number of treatments are available to help relieve many symptoms of the condition and reduce the risk of serious complications.

CONCLUSION

Behcet's disease with pregnancy is rare and thus the clinician is likely to miss it . One should keep the possibility of this rare disease in mind when pregnant pateint with rash and genital ulcers is seen. Pregnant women with behcet's disease is to be considered at high risk for foetat growth restriction and foetal compromise , and hence should have appropriate foetal surveillance . Early recognition of this disease and timely intervention can prevent antenatal complication as well as late complication associated with the disease.

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