Research Article

BEHCET'S SYNDROME – An Un Common Disorder

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ABSTRACT

Hulusi Behcet in 1937 first time described Behcet's Disease. It is characterised by oral and genital aphthae, uveitis, skin leisions etc. It is chronic relapsing and affects multiple systems. This is a case of 25 yr old gentleman who developed signs and symptoms of this syndrome and responded well to his medications and discharged at a clinically stable state.

Keywords: behcet's disease, genital aphthae, uveitis, chronic relapsing.

INTRODUCTION

Synonym for BD is Adamantiades – Behcet's syndrome and subdivisions include Neuro-Behcet, Ocular-Behcet, and Vascular-Behcet.[1-4] It is a rare systemic condition of small vessels with mucocutaneous, ocular, vascular, and CNS manifestations. It has unknown etiology but includes genetic predisposition, environmental factors and immunological abnormalities. Ocular apthous ulcers, genital ulcers, ocular leisions, skin leisions occurs frequently and occasionally CVS, CNS, GI tract etc gets affected.[4,5] Triggers by infectious agents can induce inflammatory response in genetically susceptible host. Disease managed with systemic and anti-inflammatory and or immune modulating drugs. [6]

CASE REPORT

25yr old gentleman presented with complaints of erythematous papules which rupture to form ulcer over lower lip 9 yrs ago and then started to develop ulcers over lips, palate, and groin area since 6yrs. Then he had recurrent ulcers over mouth and groin since 3 months .Also had severe body ache and swelling of leg. Had history of redness and pain of both eyes. He visited Dermatology OPD and was started on Wysolone 10mg/5mg and colchicine. He had slight improvement. Since the past 5 days he started to develop pain over the left knee joint, associated with swelling, range of movements were restricted. He visited an orthopedics and was prescribed Vit D supplementation along with other symptomatic measures. He stopped his medications last month then he started having back pain with features of bilateral sacroilitis. MRI Pelvis was done and it provisionally showed no features of the same. HLA B tissue typing was also sent for the patient was treated with Prednisolone 20mg, Etoricoxib 90mg, Sulfasalazine delayed release 1g, colchicines 0.5 g and pantoprazole 40mg. Meanwhile the patient's condition improved was clinically stable and discharged.

MRI Lumbar Spine Plain:

-Both SI joints appear normal with no joint effusion or diastasis.

-Adjacent sacru and ileum show normal marrow signal with smooth articular surface.

-Pelvic viscera within the scan region appear normal.

-Both hip joints appear normal with minimal free fluid in the joint space.

-Screening of lumbar spine did not reveal any obvious pathology.

HLA B Tissue Typing Report

Date	Name & MRD	Sex & Age	HLA-B Type	Source
17-10-16	Mehanas, 1652577	Male, 25	B*15, B*55	Whole blood

DISCUSSION

BD affects the age group 20 to 40. It includes young adults but occur in children also. Recurrent oral ulcers presents earliest in about 47-80% people.[7] Here the patient had ulcer over lower lip nine years ago which later developed over entire lips, palate and groin area since six years. Then he had recurrent ulcers over mouth and groin. Genital ulcers seen in -93% patients. Ocular leisions in retina and uvea found in 30-70% patients.[8] Here the patient had history of redness and pain of both eyes. About 45% patients shows arthralgia or arthritis .Here the patient developed pain over left knee joint, associated with swelling and range of movements were restricted. The patient also had erythematous plaque over left side of face near nasolabial fold with active periphery scaling over scalp and mustache area. The diagnosis based on criteria by ISG (International Study Group) includes recurrent oral ulceration, genital ulceration, eye leisions, and skin leisions. Prevalence in Turkey is 80to 370 cases from 100,000 populations, 10 from 100,000 in Japan.

Increased prevalence among Mediterranean population.[9] 1% patients show pulmonary artery aneurysm which is highly dangerous and can cause even death.[10]

CONCLUSION

Increased morbidity and mortality is reported with BD. Visual loss owing to uve it is and neurological involvement increase morbidity. In the last years, IL-1inhibitors and TNF-alpha inhibitors were reported to be effective in the treatment of BD. Despite all these efforts, we need to develop more standard therapy for BD. There is huge scope for research in this area.

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