Cardiovascular Manifestations Of Behçet's Disease

Thesis

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List of Abbreviations

Abbreviation	Term
ACA	Anti-cardiolipin antibody
AF	Atrial fibrillation
ALT	Alanin transaminase
AR	Aortic regurge
AST	Aspartate transaminase
AZA	Azathoiprine
BD	Behçet's disease
BS	Behçet's syndrome
BC	Before cencery
BCS	Budd-chiari syndrome
BUN	Blood urea nitrogen
C	complement
CBC	Complete blood count
CNS	Central nervous system
CSA	Cyclosporin-A
CRP	C-reactive protein
CSF	Cerebro spinal fluid
CT	Computarized tomography
DVT	Deep venous thrombosis
E/A ratio	Ratio of mitral inflow velocity
ECG	Electrocardiogram
EULAR	European League Against Rheumatism
EF	Ejection fraction
ESR	Erythrocyte sedimentation rate
FDA	Food and drug administration
FV	Factor five
GN	Glomeriolonephritis

Hcy	Homocystein
HLA	Human leukocyte antigen
HDL-C	High density lipoprotein cholesterol
HSP	Heat shock protein
HSV	Herpes simplex virus
IG	Immunoglobulin
IL	Inter leukien
IOP	Intra ocular pressure
INF	Interferon
IVC	Inferior vena cava
ISG	International study group
IVSD	Interventricular septum diameter
KD	Kilo Dalton
LAD	Left atrial diameter
LDL-C	Low density lipoprotein cholesterol
LL	Lower limb
LV	Left ventricle
LVEDD	Left ventricular end diastolic diameter
MHC	Major histocompatibility complex
MI	Myocardial infarction
MIC	Major histocompatibility complex class I chain
MR	Mitral regurge
MVP	Mitral valve prolapse
MTX	Methotrexate
NB	Neuro- Behçet's
NORD	National Organization for Rare Disorders
NSAIDs	Non steroidal anti-inflammatory drugs
PAP	Pulmonary artery pressure

PG	Pyoderma gangreosum
PMNLS	Polymorph nuclear lymphocytes
PWP	P-wave dispersion
RV	Right ventricle
SVC	Superior vena cava
TC	Total cholesterol
TNF	Tumor necrosis factor
TR	Tricuspid regurge
UK	United kingdom
UL	Upper limb
USA	United stats of America
VLDL-C	Very low density lipoprotein cholesterol

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Introduction

Behçet's disease (BD) was first described in 1937 by a Turkish dermatologist Dr. Hulusi Behçet from Istanbul as a triad of manifestations consisting of oral aphthae, genital ulcers, and hypopyon uveitis **(Ustun, 2002).**

Behçet's disease typically arises in young adults, although childhood-onset BD has also been reported. The disease can affect both genders and has been observed all over the world, but the largest numbers of cases have been reported from Japan, the Middle East, and Mediterranean countries (**Sevda and Cengiz, 2006).**

The etiology of Behçet's disease is unknown, although genetic susceptibility, autoimmunity and environmental factors are suspected **(Faruk et al., 2007).**

Nonspecific vasculitis may affect any artery or vein and involve both microcirculation and macrocirculation (**Athanassios et al., 2006).**

The vascular involvement may consist of thrombophlebitis, deep vein thrombosis, and arterial obstruction. Aneurysms, particularly of the pulmonary arteries, may also develop **(Ignatios et al., 2004).**

Involvement of the heart is called cardio-Behçet's disease. Endocarditis, myocarditis, pericarditis, acute myocardial infarction and aneurysms, intracardiac thrombus, endomyocardial fibrosis, valve involvement, conduction system disturbance, dilated cardiomyopathy, ventricular arrhythmias, and sudden cardiac death may be seen in this disease. Left ventricular systolic dysfunction is rare but diastolic dysfunction is common. Of significance, mitral valve prolapse and dilatation of the proximal aorta were observed in 50% and 30% of patients with BD, respectively **(Atzeni et al., 2005).**

Aim of the study

To search for the different cardiovascular manifestations of Behçet's disease and its relations to disease activity and morbidity.

Behçet’s disease

Behçet's disease is a chronic, relapsing, multisystemic, and inflammatory disease. It causes mucocutaneous, ocular, vascular, articular, gastrointestinal, urogenital, pulmonary, and neurological abnormalities. Etiopathogenesis is still a subject of study. Infectious agents, immune mechanisms, and genetic factors are held responsible. However, Behçet's disease is described as a type of systemic vasculitis **(Celenk et al., 2008).**

Historical back ground

Behçet’s disease (BD) owes its name to the Turkish physician professor Huluci Behçet, who, in 1937, described the classic trisymptom complex of hypopyon, iritis, and orogenital aphthosis **(Hirohataetal and kikuchi, 2003).**

The Greek physician Adamantiades had reported the disease 6 years earlier to Professor Huluci Behçet, accounting for the alternative Synonym Adamantiades Behçet's disease. However, the first description probably predates both of these by 2500 years, when Hippocrates of Kos (460–377 BC) describes an endemic disease in Asia Minor characterised by “aphthous ulcerations,” “watery ophthalmies of a chronic character, which destroyed the insight of many persons,” and “large herpetic lesions.” no further reference to the disease is

encountered in the medical literature until the early 1900s, when the classic trisymptom complex was again described in Europe. At first, this complex was considered to be a manifestation of syphilis, but in 1937 Behçet's proposed a separate disease entity, which, by 1947 had gained widespread international recognition **(Verity et al., 2003).**

Naming

Some workers consider Behçet's disease as a specific disease and should be accepted as such. But others, du to lack of a definitive diagnostic test, the variability in prevalence, incidence of the condition and its constituent manifestations, they preferred the terminology of Behçet's syndrome. **(Barnes and Yazici., 1999)**

Synonyms:

- Adamantiades- Behçet's syndrome
- Behçet's disease (BD)
- Huluci Behçet's syndrome
- Oculo-Bucco-Genital syndrome
- Tourain's Aphthosis
- Triple symptom complex of Behçet's

(Sakane and Peng, 2000).

Epidemiology

Prevalence

It is highest in Turkey, the countries bordering the Mediterranean, the Middle East, Far East, and Japan, where it is 1 in 1000. It occurs less frequently in northern Europe, United stats (U.S), United kingdom (U.K), where it is 1 in 100,000 **(Zouboulis et al., 1999).**

BD has also been reported in many countries such as Israel (120/100,000), Germany (2.26/100,000), Iran (16.7/100,000) **(Al-Otaibi et al., 2005),** Iraq (17/100,000) **(Al Rawi and Neda, 2003),** and Spain (0.66/100,000) **(Gonzalez-Gay et al., 2000).** In Alexandria-Egypt, the prevalence is 7.6 patients per 100.000 inhabitants **(Zouboulis, 1999).** Of interest, it was recently found that the frequency of BD in the Afro-Caribbean population of Guadeloupe (French West Indies) was the least 3/100,000 **(Lannuzel et al., 2002).**

Age

Behçet's disease usually presents in adulthood and is uncommon in children, about 2-3% of all affected individuals; childhood-onset develops between 7 and 13 years of age **(Kone-Paut et al., 2002).**