Pattern of Behcet's Disease in Sohag Governorate

Ass. Prof. Esam Muhammed Abu Al-Fadl[1], Dr. Ahmed Roshdy Al-Aga[2], Manar Gamal 'Muhammad Ameen' Ali[3]

1, Assistant Professor of Physical Medicine, Rheumatology and Rehabilitation, Faculty of Medicine – Sohag University. 2, Lecturer of Physical Medicine, Rheumatology and rehabilitation. Faculty of Medicine, Sohag University. 3, Msc of Physical Medicine, Rheumatology and rehabilitation.Faculty of Medicine, Sohag University

Abstract

Behçet's disease (BD) is a chronic inflammatory disease with multisystem involvement. It presents with remission and exacerbation. The aim of our study was to study the pattern of Behcet's disease among patients attending the Rheumatology and Rehabilitation department, Sohag University. **Conclusion:** Behcet's disease starts frequently around the beginning of the third decade and has a male predominance (70%). The age at diagnosis of Behcet's disease ranged from 19-59 years, with a mean of around 28.8 years. Most of the cases in our study were married (73.3%). It was found that the most common first manifestation in our study group was oral ulcers, followed by ocular manifestations, then CNS manifestations, articular or vascular manifestations and lastly genital ulcer or skin manifestations.

Introduction

Behçet's disease (BD), which was first defined by Hulusi Behçet, a Turkish dermatologist, in 1937, is a chronic inflammatory disease with multisystem involvement **(Behçet 1937)**. It presents with remission and exacerbation of mucocutaneous, ocular, articular, vascular, or gastrointestinal lesions **(Yazisiz 2014)**.

The prevalence of BD varies geographically and the disease is more prevalent in certain groups. It is most common in populations clustered along the ancient Silk Road. Turkey has the highest prevalence (80-370 cases/100000), followed by Asia and the Middle Eastern countries, including Egypt, Palestine, Saudi Arabia and Iran **(Davatchi et al. 2010, Yazici et al. 2010(.**

The age at onset of the disease is usually between 20.8 and 40 years, as it is more common in young individuals. The male-to-female ratio varies regionally. The disease is more common among men in Russia, Saudi Arabia, Iraq, Lebanon, Jordan, Kuwait, Greece, Italy, Turkey, and Iran, while it is more frequent among women in Japan, South Korea, and Israel **(Ambrose and Haskard 2013).**

Ocular involvement in BD includes anterior or posterior uveitis, vitritis, retinal vasculitis, retinal vein thrombosis, corneal ulcers, and retrobulbar neuritis. BD-associated uveitis is defined as chronic and recurrent non-granulomatous panuveitis and retinal vasculitis with a bilateral course. Recurrent episodes may result in permanent vision loss **(Tugal-Tutkun et al. 2013).**

Musculoskeletal disorders are also common in patients with BD. Palindromic asymmetric arthritic exacerbations involving the knee, wrist, and ankle may develop. Chronic erosive arthritis is relatively rare. The incidence

of sacroiliitis has been reported to increase in patients with BD. Due to peripheral arthritis characteristics and sacroiliac joint involvement, BD is evaluated in the spectrum of seronegative spondyloarthropathy **(Tugal-Tutkun et al. 2013(.**

BD-related vasculopathy differs from other vasculitides, due to its pattern of arterial and venous involvement. Venous thrombus may develop. It may present with superficial thrombophlebitis or involve deep veins, as well as the inferior/superior vena cava, the right atrium, or intracranial large sinuses. The major hepatobiliary disease is Budd-Chiari syndrome, which is one of the leading causes of mortality **(Ben Ghorbel et al. 2008).** Unlike other thrombotic events, embolism is not anticipated. Primary thrombosis, which is often accompanied by right atrial thrombi, may occur in the pulmonary artery and its thin branches. In addition, arterial aneurysms are common. Pulmonary artery aneurysms may lead to massive bleeding and a fatal outcome **(Yilmaz et al. 2013).**

Central nervous system (CNS)-related symptoms may develop secondary to vascular events, such as sinus thrombus and intracranial aneurysms. Primary parenchymal involvement including meningitis and encephalitis, mostly in the pons and mesencephalon, is also seen in patients with BD. It is also known as Neuro-BD, accounting for 10% of patients. In addition, longitudinal extensive transverse myelitis (LETM), characterized by spinal cord lesions, may occur **(Yilmaz et al. 2013).**

Patients and Methods

It is A descriptive cross-sectional study that included all BD patients attending Rheumatology Department, Sohag University Hospital , Sohag University, in the period from July 2017 to December 2017.

Patients Criteria

Inclusion criteria: Age > 16 years, Diagnosed as having Behcet's disease. Exclusion criteria: Any other autoimmune rheumatic diseases (e.g. SLE, RA, Vasculitis). And Chronic organ diseases not related to BD (e.g. liver cirrhosis, renal impairment or eye diseases)

All of the cases were subjected to the following: (1) Full history taking including :Personal and demographic data.(2)History of the present illness (including details on the most commonly affected organs by BD).(3)Family history of BD or any other auto-immune disease. (4) Full general and local examination. (5)Behcet's disease scoring. (6) Investigations including Pathergy test, routine investigations and Immune assay.

Ethical Consideration

This research was revised by the Scientific Ethical Committee of Sohag University Hospital and An Informed written consent was taken from all of the cases.

Statistical analysis

Statistical package for social sciences (IBM-SPSS), version 24 IBM- Chicago, USA (May 2016) was used for statistical data analysis. And Data expressed as mean, standard deviation (SD), number and percentage. Mean and standard deviation were used as descriptive value for quantitative data, while number and percentage were used to describe qualitative data.

Results

Table No. (1) show This table shows that the mean age of our study group was around 34 years, with a range from 20-60 years. The majority of our cases were males (70%), with only 9 cases (30%) were females. Most of the cases were married (73.3%), while 23.3% were single and only one case (3.3%) was widow. Regarding occupation, half of the cases were manual workers (50%), 3 cases were drivers and two cases were accountants. The remaining 10 cases were either housewives or non working males. Only one third of the cases came from urban areas, with the remaining two thirds were rural. Also, the socioeconomic status was good in only 3 cases (10%). A little than half of the cases (43.3%) were smokers, mostly cigarette smokers.

Value	Item	
33.97±9.72	Mean±SD	**Age (years)**
30.5(20-60)	Median(range)	
21(70%)	Male	**Sex**
9(30%)	Female	
7(23.3%)	Single	**Marital status**
22(73.3%)	Married	
1(3.3%)	Widow	
8(26.7%)	Housewife	**Occupation**
2(6.7%)	Not working	
15(50%)	Worker	
3(10%)	Driver	
2(6.7%)	Accountant	
10(33.3%)	Urban	**Residence**
20(66.7%)	Rural	
3(10%)	Good	**Socioeconomic status**
27(90%)	Low	
13(43.3%)	Yes	**Smoking / addiction**
17(56.7%)	No	

Table 1. Demographic data of the study group

Value (yrs)	Item	
28.8±9.85	Mean±SD	**Age at diagnosis**
26.5(15-59)	Median(range)	
5.17±4.08	Mean±SD	**Disease duration**
4(0-20)	Median(range)	
11(36.7%)	Oral ulcer	**First organ/system affected**
10(33.3%)	Ocular manifestations	
3(10%)	CNS	
2(6.7%)	Articular	
2(6.7%)	Vascular lesion	
1(3.3%)	Genital ulcer	
1(3.3%)	Skin	

Table 2. Disease history

Table No. (2) shows that the age at diagnosis of Behcet's disease ranged from 19-59 years, with a mean of around 28.8 years. The disease duration showed very wide variation, from zero (diagnosed at time of inclusion in the study) to over 20 years, with a mean of 5.2 years and a high standard deviation

of over 4 years. The most common first manifestation in our study group was oral ulcers (seen in 36.7% of cases), followed by ocular manifestations (33.3%), then CNS manifestations (10%), articular or vascular manifestations (6.7% each) and lastly genital ulcer or skin manifestations(3.3% each). Analysis of oral ulcers in our study group showed that it was present in nearly all of the cases (29 cases, 96.7%).

It was recurrent; painful and progressive in all of the affected cases; acute in 58.6% of them and insidious in the remaining cases. Their shape were rounded in 82.8% of the cases, and oval in the remaining. The most common affected site was the lips; either alone (in 4 cases) or associated with gums or tongue (18 cases). This was followed by the gums and then the tongue. All of the cases complicated with scars, and the number was multiple (>3 ulcers) in the vast majority of cases (86.2%).

Analysis of genital ulcers in our study group shows that it was seen in 25 cases (83.3%). They were progressive and painful in all of the affected cases. They were de novo in only 2 cases, and recurrent in the remaining 23 cases. Half of them (52%) showed acute onset and the other half (48%) had insidious onset. Regarding the shape, 18 cases were rounded (72%) and 28% had oval shape. All of the cases in males were in the scrotum (17 cases), while the female cases were either in the vulva (3 cases) or vaginal mucosa (5 cases). Most of the cases complicated with scar tissue (88%), and most of the cases had only one scar (23 cases; 92%).

Table No. (3) shows that cutaneous manifestations were seen in less than half of the cases (43.3%). Regarding the type of cutaneous manifestations, acniform nodules were the most common lesion detected, either alone (in 6 cases) or combined with erythema nodosum (2 cases); while erythema nodosum alone was seen in 4 cases. The remaining cases showed pseudo-folliculitis.

Also the table shows that the ocular affection was detected in 73.3% of the cases. The most common sole finding was panuveitis (seen in 7 cases, 31.8%), followed by anterior uveitis (27.3%), then posterior uveitis (22.7%). The remaining 4 cases had either panuveitis with complete blindness, blurring with posterior uveitis, hemiretinal vascular occlusion, or intraocular hypertension and posterior uveitis (one case for each finding).

GIT manifestations were seen in only 4 cases of our study group (13.3%). These 4 cases were presented with either gastritis, peptic ulcer, constipation or diarrhea.

CNS manifestations were seen in 20% of our study cases. The most common finding was chronic headache (4 cases, 66.7%), followed by transverse myelitis and quadriparesis (one case each).

It also seen that articular and/or musculoskeletal manifestations were seen in 60% of cases. The most common joints affected were the knees (alone in 7 cases, 38.9% and combined with other joints in another 7 cases). Other joints affected included ankles (9 cases), wrist and hands (7 cases), elbows (4 cases), shoulder (1 case). Plantar faschiitis was seen in one case. Regarding the duration of musculoskeletal manifestations, it ranged from 1-72 months, with a mean of 26 months and a very wide standard deviation of 22 months. These articular manifestations were recurrent in the vast majority of cases (13 cases, 72.2%), and was insidious in 14 cases (77.8%).

9

Vascular manifestations were detected in around one third of cases (9 case, 30%). Most of these cases had DVT (de novo in 4 cases, and recurrent in 2 cases). The other three cases had either thrombosis (2 cases) or vasculitis (one case).

Table No. (4) shows that the chronic diseases and associated disorders were not common in our study population, being seen only in a minority of cases. Hypertension was seen in 2 cases, and each of malar rash, facial palsy, MI, HCV infection were seen in only one case, with non of the cases were diabetic.

Family history was seen in only 2 cases, one case had Behcet second degree relative, and the other had RA second degree relative.

Table No. (6) shows that Pathergy test was positive in only 5 cases (16.7% of cases). Regarding Behcet's disease scoring, it ranged from 4-8, with a mean of 6.03±1.35.

Table No. (7) shows that azathioprine was the most commonly drug used for our patients (93.3%). This was followed by steroids (86.7%), then oracure gel (73.3%), colchicine (70%), bone one (53.3%), hydroxychloroquine (505), calcium supplementations (46.7%), cyclophsosphamide (26.7%), methotrexate (13.3%). On the other hand, PPIs were used for only 10% of cases, artificial eye drops for 6.7%, and only one case used each of tetrcyclin, trental, potassium permanganate and the antibiotic claforan.

Value	Genital ulcer analysis	
25(83.3%)	Present	Present or not
5(16.7%)	Absent	
23(92%)	Recurrent	De novo or recurrent
2(8%)	De novo	
13(52%)	Acute	Onset
12(48%)	Insidious	
25(100%)	Progressive	Course
18(72%)	Rounded	Shape
7(28%)	Oval	
25(100%)	Painful	Painful or not
17(68%)	Scrotum	Site
3(12%)	Vulva	
5(20%)	Vaginal mucosa	
22(88%)	Present	Scars
3(12%)	Absent	
23(92%)	One	Number
1(4%)	Two	
1(4%)	Many	

Value	Oral ulcer analysis	
29(96.7%)	Present	Present or not
1(3.3%)	Absent	
29(100%)	Recurrent	De novo or recurrent
17(58.6%)	Acute	Onset
12(41.4%)	Insidious	
29(100%)	Progressive	Course
24(82.8%)	Rounded	Shape
5(17.2%)	Oval	
29(100%)	Painful	Painful or not
4(13.8%)	Lips only	Site
14(48.3%)	Lips and gums	
4(13.8%)	Lips and tongue	
1(3.3%)	Gums and tongue	
1(3.3%)	Tongue only	
5(17.2%)	Buccal cavity	
29(100%)	Present	Scars
1(3.4%)	Two	Number
3(10.3%)	Three	
25(86.2%)	More than three	

Value	GIT manifestations	
4(13.3%)	Present	Present or not
26(86.7%)	Absent	
1(25%)	Gastritis	Type
1(25%)	Peptic ulcer	

Value	Cutaneous manifestations	
13(43.3%)	Present	Present or not
17(56.7%)	Absent	
6(46.2%)	Acniform nodules	Type
4(30.8%)	Erythema nodosum	

Value	Left manifestation	Left category	Value	Right manifestation	Right category
1(25%)	Constipation		2(15.3%)	Acniform nodules & erythema nodosum	
1(25%)	Diarrhea		1(7.7%)	Pseudo-folliculitis	
Value	**Articular manifestations**		**Value**	**Ocular manifestations**	
18(60%)	Present	Present or not	22(73.3%)	Present	Present or not
12(40%)	Absent		8(26.7%)	Absent	
2(11.1%)	Ankles	Site	6(27.3%)	Anterior uveitis	Type
1(5.6%)	Hand, wrist, knee and ankle		5(22.7%)	Posterior uveitis	
1(5.6%)	Hand, wrist, knee, ankle, shoulder, elbow		7(31.8%)	Panuveitis	
1(5.6%)	Knee and ankle		1(4.5%)	Panuveitis ended with complete blindness	
1(5.6%)	Knee and wrist		1(4.5%)	Blurring and posterior uveitis	
7(38.9%)	Knees		1(4.5%)	Hemi-retinal vascular occlusion	
1(5.6%)	Plantar faschiitis		1(4.5%)	Intraocular hypertension and posterior uveitis	
1(5.6%)	Wrist and ankle		**Value**	**CNS manifestations**	
3(16.7%)	Wrist, elbow, knee and ankle		6(20%)	Present	Present or not
26.2±22.3	Mean±SD	Duration (months)	24(80%)	Absent	
24(1-72)	Median(range)		4(66.7%)	Headache	Type
5(27.8%)	De novo	De novo or recurrent	1(16.7%)	Transverse myelitis	
13(72.2%)	Recurrent		1(16.7%)	Quadriparesis	
4(22.2%)	Acute	Onset	**Value**	**Vascular manifestations**	
14(77.8%)	Insidious		9(30%)	Present	Present or not
			21(70%)	Absent	
			4(44.4%)	DVT (de novo)	Type
			2(22.2%)	DVT (recurrent)	
			2(22.2%)	Thrombosis	
			1(11.1%)	Vasculitis	

Table 4. Other chronic diseases

%	No	Disease
0	0	Diabetes mellitus
6.7%	2	Hypertension
3.3%	1	Malar rash
3.3%	1	Hair falling
3.3%	1	Facial palsy
3.3%	1	Previous myocardial infarction
3.3%	1	HCV infection
3.3%	1	Ecchymotic patches in abdomen & dyspnea grade III & Rt Cataract &tonsilectomy

Table 5. Other diseases in Relatives

%	No	Item
93.3%	28	**No**
3.3%	1	**Behcet's disease in 2nd degree relative**
3.3%	1	**Rheumatoid arthritis in 2nd degree relative**

Table 6. Examination

Value	Item	
5(16.7%)	Positive	**Pathergy test**
25(83.3%)	Negative	
4(13.3%)	4	**Behcet's disease scoring**
9(30%)	5	
4(13.3%)	6	
8(26.7%)	7	
5(16.7%)	8	
6.03±1.35	Mean±SD	
6(4-8)	Median(range)	

Table 7. Therpay taken

%	No	Drug
93.3%	28	**Azathioprine**
50%	15	**Hydroxychloroquine**
26.7%	8	**Cyclopsohphamide**
13.3%	4	**Methotrexate**
70%	21	**Colchicine**
86.7%	26	**Steroids (prednisolone)**
73.3%	22	**Oracure gel**
46.7%	14	**Calcium supplementation**
53.3%	16	**Bone one**
3.3%	1	**Tetracycline**
3.3%	1	**Trental**
10%	3	**Proton pump inhibitors**
6.7%	2	**Artificial eye drops**
3.3%	1	**Potassium permanganate**
3.3%	1	**Antibiotics (claforan)**

Discussion

Behcet's disease is an inflammatory, systemic vasculitis of unknown etiology. It is characterized by recurrent episodes of oral and genital ulcers, ocular and cutaneous lesions, as well as vascular, articular, gastrointestinal or neurological lesions *(Mendes et al. 2009)*. A prevalence of 2.4/ 00,000 cases is estimated in Portugal. Behçet's disease Behçet's disease is a relapsing multisystem polysymptomatic disease with exacerbations and remissions. Mucocutaneous manifestations like oral and genital ulcers, and cutaneous lesions (papulopustular lesions, erythema nodosum-like lesions, cutaneous ulcers, superficial thrombo-phlebitis), are considered the "fingerprint" of the disease, being the most common and often the first signs to appear *(Uva et al., 2013)*.

The mean age of our study group was around 34 years, with a range from 20-60 years. The majority of our cases were males (70%), with only 9 cases (30%) were females. Our results were similar to *Coutinho et al. (2017)* as the average age in their study during diagnosis was 33.45±6.49 years. However, their cases included 5 men (45.45%) and 6 women (54.55%), all were Caucasian. Our cases were somewhat different from those recorded by *Ishido et al., (2017)*where among the total of 6627 Japanese cases, 2651 (40.0%) were men and 3976 (60.0%) were women with a median age of 39 years (interquartile range: 31–50 years). The study of *Pipitone et al., (2004)* done on 137 Italian BD patients showed less prominent male predominance than our study (76 males versus 61 females). However, *Oliveira et al., (2011)* included different population than our study, with a mean age at study of 40.0±10.7 years, with female

usually occurs between the 2nd and 4th decade of life, although it may affect any age, with no difference in the socioeconomic distribution *(Deuter et al. 2008)*. It is discretely more prevalent in males in which, due to an increased risk of ocular, cardiovascular or neurological involvement and an earlier age of manifestation, is associated with a worse prognosis *(Tugal-Tutkun. 2009)*.

predominance, with female-male ratio of 1.2 to 1.

Most of the cases in our study were married (73.3%), while 23.3% were single and only one case (3.3%) was widow. Regarding occupation, half of the cases were manual workers (50%). Only one third of the cases came from urban areas. Also, the socioeconomic status was good in only 3 cases (10%). A little than half of the cases (43.3%) were smokers, mostly cigarette smokers.

In our study the age at diagnosis of Behcet's disease ranged from 19-59 years, with a mean of around 28.8 years. The disease duration showed very wide variation, with a mean of 5.2 years. The study of *Pipitone et al., (2004)*showed similar figures than ours, with mean age at disease onset of 29.6±12.2 years.

An Egyptian study done in Zagazig by *El-Najjar et al., (2015)*showed similar patients' profile compared to ours, with mean age of 31.5±7.1 years (20–44 years) and disease duration of 6.2±3.5 years (2–12 years); also with male predominance of around 63.2%, with male to female ratio of 1.7:1.

Esatoglu et al., (2017) found that male gender and younger age at disease onset are associated with a more severe disease course.

Davatchi et al., (2010) analyzed five nationwide surveys of BD from Iran, Japan, China, Korea, and Germany. The male to female ratios were, respectively

1.19, 0.98, 1.34, 0.63, and 1.40 to 1. The mean age at onset was 26.2, 35.7, 33.8, 29, and 26 years.

The most common first manifestation in our study group was oral ulcers, followed by ocular manifestations, then CNS manifestations, articular or vascular manifestations and lastly genital ulcer or skin manifestations.

The study done by *Al Ghafri et al., (2018)* concluded that oral ulcers were the most common manifestation, followed by genital ulcers, ocular lesions, and arthritis. The frequency of neurological involvement was significantly high.

Pipitone et al., (2004) stated that the most frequent manifestations at disease onset were oral (78.3%) and genital ulcers (29.2%) followed by inflammatory ocular involvement and arthritis. Also, the commonest manifestations were oral aphthae (99.3%), genital aphthae (62.8%), various cutaneous lesions including erythema nodosum (81.8%), and inflammatory ocular disease (60.6%). Panuveitis and posterior uveitis/ retinitis occurred more frequently in males compared with females (28.9% versus 11.5% and 57.9% versus 36.1%, respectively; p < 0.05). 61.6% of their patients were HLA-B51 positive**.**

According to *Oliveira et al., (2011)*, the frequency of disease manifestations was as follows: oral ulcers, 100%; genital ulcers, 93.3%; ocular manifestations, 63.3%; arthritis, 46.7%; cutaneous lesions, 71.7%; positive pathergy test, 22.7%; neurologic involvement, 28.3%; thrombosis, 13.3%; and gastrointestinal involvement, 3.3%.

According to *El-Najjar et al., (2015)*, the frequency of the clinical manifestations was oral ulcers in 84.2%, followed by genital ulcers in 78.2%,

ocular involvement in 73.7%, arthralgia in 26.3%, followed by arthritis of the knees and ankles, neurological, blurred vision, difficult hearing, pulmonary, gastrointestinal, cutaneous and none had peripheral vascular disease.

Davatchi et al., (2010) in their analysis of the five nationwide surveys of BD from Iran, Japan, China, Korea, and Germany found that the major manifestations seen, respectively were: mucous membrane; skin manifestations; erythema nodosum; ocular manifestations.

Analysis of oral ulcers in our study group showed that it was present in nearly all of the cases (29 cases, 96.7%). It was recurrent; painful and progressive in all of the affected cases; acute in 58.6% of them. Their shape were rounded in 82.8% of the cases,. The most common affected site was the lips. All of the cases complicated with scars. Similar to our results, *Coutinho et al. (2017)* found that oral aphthosis appeared as the first manifestation in all patients in their study.

Analysis of genital ulcers in our study group shows that it was seen in 25 cases (83.3%). They were progressive and painful in all of the affected cases. Half of them (52%) showed acute onset. Most of the cases complicated with scar tissue (88%). In study of *Pipitone et al. (2004)*, the mostfrequent heralding manifestations of BD were oral aphthae (78.3%) and to alesser extent genital aphthae (29.2%) which similar to that reported inprevious studies from Germany, Turkey,and Greece *(Zouboulis et al. 2003)*.

The study done by *Ishido et al., (2017)* concluded that ocular lesion was more common in male and genital ulceration was more common in female. Also, ocular lesion, arthritis and vascular

lesions were more frequently observed in elderly patients.

In this study cutaneous manifestations were seen in less than half of the cases (43.3%).

Also the ocular affection was detected in 73.3% of the cases. The most common sole finding was panuveitis (seen in 7 cases, 31.8%), followed by anterior uveitis (27.3%), then posterior uveitis (22.7%). Similarly in study of *Coutinho et al. (2017)* the main ocular manifestation was bilateral panuveitis, which is in agreement with other similar studies *(Tugal-Tutkun. 2009, Tugal-Tutkun et al. 2004)*. *Pipitone et al. (2004)* found in their study that erythema nodosum and inflammatory eye disease to occur more commonly in females and males, respectively.

In our study GIT manifestations were seen in only 4 cases of our study group (13.3%).

Hum et al., (2017) stated that pancreatitis is a rare manifestation of BD that has scarce documentation in the literature, although other GIT manifestations may be much more common.

Articular and/or musculoskeletal manifestations were seen in 60% of our cases. The most common joints affected were the knees (alone in 7 cases, 38.9% and combined with other joints in another 7 cases). Other joints affected included ankles (9 cases), wrist and hands (7 cases), elbows (4 cases), shoulder (1 case). Plantar faschiitis was seen in one case. Regarding the duration of musculoskeletal manifestations, it ranged from 1-72 months, with a mean of 26 months and a very wide standard deviation of 22 months. These articular manifestations were recurrent in the vast majority of cases (13 cases, 72.2%), and was insidious in 14 cases (77.8%).

Vascular manifestations were detected in around one third of our cases (9 case, 30%). Most of these cases had DVT (de novo in 4 cases, and recurrent in 2 cases). The other three cases had either thrombosis (2 cases) or vasculitis (one case). *Coutinho et al. (2017)* reported in their study that in 2 patients, the first manifestation of the disease was branch venous occlusion, and the diagnosis of Behçet's disease was established after 1 and 2 years, respectively.

In this study family history was seen in only 2 cases, one case had Behcet second degree relative, and the other had RA second degree relative. Pathergy test was positive in only 5 of our cases (16.7% of cases). *Davatchi et al., (2010)* in their analysis of the five nationwide surveys of BD from Iran, Japan, China, Korea, and Germany found that laboratory tests are not useful except the pathergy test, which was positive in 54%, 44%, N/A, 40%, and 34% of cases; respectively. ESR was normal in many patients. Diagnosis is based upon clinical manifestations. The International Criteria for Behcet's Disease (ICBD, 2006) may be of help, having a sensitivity of 98.2% and a specificity of 95.6% in Iranian patients.

In our stud, regarding Behcet's disease scoring, it ranged from 4-8, with a mean of 6.03±1.35. The study done by *El-Najjar et al., (2015)* showed that the BDCAF was 3.8±1.3 (2–7).

Azathioprine was the most commonly drug used for our patients (93.3%). This was followed by steroids (86.7%), then oracure gel (73.3%), colchicine (70%), bone one (53.3%), hydroxychloroquine (505), calcium supplementations (46.7%), cyclophsosphamide (26.7%), methotrexate (13.3%). On the other hand, PPIs were used for only 10% of cases, artificial eye drops for 6.7%, and

only one case used each of tetrcyclin, trental, potassium permanganate and the antibiotic claforan. In study of *Coutinho et al. (2017)* topical therapy consisted of corticotherapy, mydriatic, and hypotensors whenever necessary. Systemic treatment included oral corticosteroid therapy in combination with other immunosuppressive adjuvants, with the most commonly used being azathioprine, cyclosporine and colchicine. In 3 patients there was need for biological therapy with infliximab for disease control.

The frequency of each treatment modality in the study of *Oliveira et al., (2011)* was as follows: colchicine, 78.3%; thalidomide, 26.7%; colchicine and penicillin, 21.7%; dapsone, 8.3%; and pentoxyphyline, 8.3%. These treatments were mainly used for mucocutaneous manifestations. Immunosuppressive drugs were prescribed for 70% of the patients, including azathioprine (35.0%), cyclophosphamide (28.3%), cyclosporin A (21.7%), methotrexate (18.3%), and chlorambucil (6.7%). Infliximab was used in 5.0% of refractory patients.

Conclusion

Behcet's disease starts frequently around the beginning of the third decade and has a male predominance. The disease is usually more severe in men than women in Egyptian population.

References

1. Behçet H. Überrezidivieren deAphthose, durchein Virus verursachteGeshwüre am Munde, am Auge und an den Genitalien. Dermatol Wochenschr. 1937;105:1152–1157.
2. Yazisiz V. Similarities and differences between Behcet's disease and Crohn's disease. World J Gastrointest Pathophysio l2014;5(3):228-38.
3. Davatchi F, Shahram F, Chams-Davatchi C, Shams H, Nadji A, Akhlaghi M, et al. Behcet's disease: from East to West. Clin Rheumatol. 2010 Aug;29 (8):823-33
4. Yazici, Y., S. Yurdakul and H. Yazici. Behcet's syndrome. CurrRheumatol Rep 2010;12(6): 429-35
5. Ambrose, N. L. and D. O. Haskard. Differential diagnosis and management of Behcet syndrome. Nat Rev Rheumatol2013;9(2): 79-89.
6. Tugal-TutkunI, Gupta V and Cunningham ET. Differential diagnosis of behcet uveitis. Ocul Immunol Inflamm2013;21(5):337-50.
7. Ben Ghorbel, I., R. Ennaifer, M. Lamloum, M. Khanfir, M. Miled and M. H. Houman. Budd-Chiari syndrome associated with Behcet's disease." Gastroenterol Clin Biol 2008;32(3):316-20.
8. Yilmaz S, Karadag O, Yazisiz V, Altun B, Gezer M, Karaman M, Cinar M, Erdem H, Pay S, Dinc A. Systemic involvements and preferred treatments in a large cohort of Behcet's disease. Rheumatol Int 2013; 33(12):3025-30.
9. Mendes D, Correia M, Barbedo M, Vaio T, Mota M, Gonçalves O, Valente J. Behçet's disease - a contemporary review. J Autoimmun. 2009;32(3-4):178-88.
10. Deuter CM, Kotter I, Wallace GR, Murray PI, Stübiger N, Zierhut M. Behçet's disease: ocular effects and treatment. Prog Retin Eye Res. 2008;27(1):111-36.
11. Tugal-Tutkun I. Behçet's uveitis. Middle East Afr J Ophthalmol. 2009;16(4):219-24.
12. Uva L, Miguel D, Pinheiro C, Filipe P, Freitas JP. Mucocutaneous manifestations of Behçet's disease. Acta Reumatol Port. 2013;38(2):77-90
13. Coutinho I, Diana Silva, Isabel Ferreira, Bruno Grima. Behçet disease: our experience. Rev Bras Oftalmol. 2017; 76 (4): 210-12.
14. Ishido T, Horita N, Takeuchi M, Kawagoe T, Shibuya E, Yamane T,

SOHAG MEDICAL JOURNAL
Vol. 23 No.2 Apr 2019
        Pattern of Behcet's Disease in Sohag Governorate
Manar Gamal.et al

Hayashi T, Meguro A, Ishido M, Minegishi K, et al. Clinical manifestations of Behçet's disease depending on sex and age: results from Japanese nationwide registration. Rheumatology, 2017(Nov); 56 (11): 1918–27.

15. Pipitone N, Boiardi L, Olivieri I, Cantini F, Salvi F, Malatesta R, La Corte R, Triolo G, Ferrante A, Filippini D, Paolazzi G, Sarzi-Puttini P, Restuccia G, Salvarani C. Clinical manifestations of Behçet's disease in 137 Italian patients: Results of a multicenter study. Clinical and Experimental Rheumatology, 2004; 22 (Suppl. 36): S46-S51.

16. Oliveira AC, Buosi AL, Dutra LA, de Souza AW. Behçet disease: clinical features and management in a Brazilian tertiary hospital. J Clin Rheumatol. 2011 Dec;17(8):416-20.

17. El-Najjar AR, Abou El-Soud AM, Amara HA, Diabb MA. Clinical characteristics and disease activity of Behçet's disease patients in Zagazig, Egypt. The Egyptian Rheumatologist. 2015;37(4):191-6.

18. Esatoglu SN, Kutlubay Z, Ucar D, Hatemi I, Uygunoglu U, Siva A, Hatemi G. Behçet's syndrome: providing integrated care. Journal of Multidisciplinary Healthcare. 2017;2017(10):309-19.

19. Davatchi F, Shahram F, Chams-Davatchi C, Shams H, Nadji A, Akhlaghi M, Faezi T, Ghodsi Z, Faridar A, Ashofteh F, Sadeghi Abdollahi B. Behcet's disease: from East to West. Clin Rheumatol. 2010 Aug;29 (8):823-33.

20. Al Ghafri A, Al Wahshi H. Demographic features and clinical aspects of Behçet's disease in Omani patients. Rheumatology Practice and Research, 2018.

21. Zouboulis CC, Vaiopoulos G, Marcomichelakis N. Onset signs, clinical
course, prognosis, treatment and outcome of adult patients with Adamantiades - Behçet's disease in Greece. Clin Exp Rheumatol 2003; 21: S19-26.

22. Hum M, Muhammad N. Mahmood, Vivian Huang, Marlene Dytoc. Pancreatitis as a rare manifestation of Behçet disease.2017;3(6):470-3.

17