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ORIGINAL ARTICLE

Frequency of Behçet's disease among a group of visually impaired adults

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Abstract

Background Behçet's disease (BD) is one of the reasons of acquired visual impairment among young adults. Ocular involvement is a significant cause of disability in BD. The objective of this study is to assess the prevalence of BD among a group of adults who has visual impairment.

Methods Ankara Metropolitan Municipality Education and Technology Center is one of the official institutions which records and follows the demographic data of visually impaired people in Turkey. In November 2014, there were 675 visually impaired people recorded at this center. Medical history was taken from 294 adults by phone in November and December of 2014. Participants were asked if the visual impairment had been either acquired or congenital. If the patients had BD or suspicious BD, they were recalled for detailed investigation which would be carried out by an internist, a rheumatologist and an ophthalmologist.

Results Two hundred thirteen of 294 (72.4%) visually impaired adults were male. One hundred nine of 294 (37.1%) had acquired visual impairment. Six (5.5%) of those 109 patients had BD. Overall prevalence of BD among study group was 2.04%.The median age of people with BD was 35 years. The median age at BD diagnosis was 16.5 years and the median duration from diagnosis to visual loss was 2.5 years.

Conclusion BD is still one of the causes of acquired visual impairment in Turkey. In this study, BD prevalence among a visually impaired adult group was 2.04%. BD accounted for 5.5% among adults who had acquired visual impairment. In a study of 1965, BD prevalence among people with acquired blindness was 12%. However, this study was conducted at pre-immunosuppressive period. Our prevalence is obviously lower than those studies. Extended population-based studies are needed for population estimations.

Keywords Acquired visual impairment · Behçet's disease · Blindness · Ocular involvement · Visual impairment

Introduction

In general terms, visual impairment can be defined as total or severe reduction in vision. Worldwide, visual loss or blindness is major organ dysfunction and related with physical and mental disability. In addition, blindness is closely associated with increased mortality [1]. In 2012 World Health Organization (WHO) estimated that there were 285 million visually impaired people globally. Thirty-nine million of those were blind and 246 million had low vision. The most frequent causes of visual impairment were uncorrected refractive errors (43%) and cataract (33%), respectively [2]. According to Turkey Disability Survey in 2002, it was estimated that the crude frequency of visually disabled people among Turkish population was 0.6%. Majority of them (76%) had acquired visual impairment [3].

Behçet's disease is a multisystemic vasculitis that involves several organ systems including the eyes. It is well known that BD may cause acquired visual loss at especially young adults [4]. Approximately 50–70% of patients with BD have ocular involvement [4, 5]. In BD, visual loss can be prevented by appropriate treatments such as azathioprine, interferon alpha, and anti-tumor necrosis factor treatments [6]. Up to now, there is no study assessing the prevalence of BD among blind population in Turkey. The objective of our study is to assess the prevalence of BD among a visually impaired adult group.

The study is presented in poster sessions of EULAR Congress-ROME 2015 and 16th National Rheumatology Congress-ANTALYA 2015

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Methods

Ankara Metropolitan Municipality Education and Technology Center for Blinds is one of the official institutions which records and follows the demographic data of visually impaired people. Visually impaired adults and children have the opportunity of being a member of this center. When they become a member, the center offers exceptional technological facilities, including talking books and Braille printers.

In November of 2014, there were 675 visually impaired (blindness or low vision) people recorded at this center. Telephone numbers of 131 people could not be available. Of 544 people, 250 were excluded from this study. These 250 patients could not reach by phone (at least two calls), did not want to attend study or were younger than 15 years old. Medical history was taken from 294 adults by phone.

Participants were first asked if the visual impairment had been either acquired or congenital. People with acquired visual impairment were ascertained by standard questionnaire for BD. By this questionnaire, people were searched for the presence of the diagnostic criteria proposed by International Study Group for Behçet's disease (ICBD). Patients who have a score ≥ 4 points is classified as having BD [7]. If the patients had BD or suspicious BD, they were recalled for detailed investigation which would be carried out by an internist, a rheumatologist and an ophthalmologist. The SPSS version 15.0 (SPSS, Inc., Chicago, Illinois) is used for descriptive statistics of BD.

At the beginning of the phone interviews, verbal informed consents were obtained from everyone attending the study. This study has been approved by Hacettepe University Ethics Commission (Approval number: GO 16/287 - 23). Survey was conducted in accordance with the Declaration of Helsinki.

Results

Study population

In total, 213 of 294 (72.4%) visually impaired adults were male. One hundred nine of 294 (37.1%) had acquired visual impairment. Six (5.5%) of 109 patients had BD (Fig. 1). Overall prevalence of BD among visually impaired people was 2.0%.

Demographic and clinical features of Behçet's disease with visual loss

Disease characteristics of six patients with Behçet's disease can be seen in Table 1. Five of six patients had already had BD diagnosis and had taken immunosuppressive drugs. These five patients rejected further investigation in outward clinics. They were questioned for diagnostic criteria and disease characteristics by phone. One of 6 patients did not know his diagnosis but was meeting the diagnostic criteria of ICBD. He came to our hospital and was examined in our rheumatology clinic. He was precisely diagnosed as BD (fifth patient in Table 1). The median age of people with BD was 35 years (IQR 27–47 years) The median age at BD diagnosis were 20 ± 8 years and 16.5 years (IQR 15–23 years), respectively. The

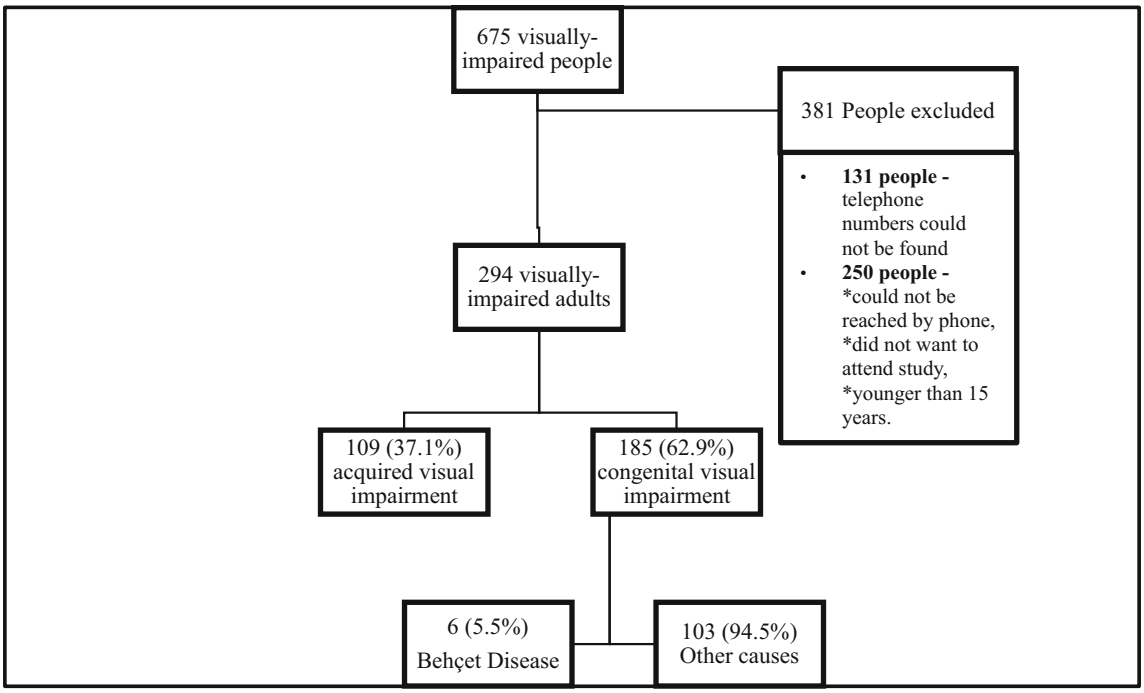

Fig. 1 Proportions of people in the study group

Table 1 Disease characteristics of six patients with Behçet's disease

	Age	Gender	Oral ulcer	Genital ulcer	Blindness	Uveitis	Ant. uveitis	Post. uveitis	Pathergy test	Skin inv.	Arthralgia	Arthritis	Neurological inv.	DVT	IBH
1st patient	64	Male	+	+	+	+	–	+	+	+	+	–	–	–	–
2nd patient	27	Male	+	+	+	+	+	+	+	–	+	+	–	–	–
3rd patient	24	Male	+	+	+	+	?	?	?	+	–	–	–	–	–
4th patient	42	Male	+	+	+	+	?	?	+	–	+	+	+	–	+
5th patient	47	Male	+	+	+	+	?	?	?	+	+	+	–	–	–
6th patient	28	Male	+	–	+	+	–	+	?	+	–	–	–	–	–

DVT deep venous thrombosis; *IBH*, inflammatory bowel disease; "+", present; "–", absent "?", unknown

median duration from diagnosis to visual loss was 2.5 years (IQR 1–7 years). All patients were blind. Uveitis location was known on three patients, one had panuveitis, two patients had only posterior uveitis.

Discussion

BD frequency among acquired visual loss patients was 5.5% in Ankara, capital city of Turkey. In general, BD is rarely seen worldwide. However, it is more common among residents of Old Silk Road and BD seems to be most common in Turkey with different estimated prevalence of 80–370 cases per 100.000 [8]. Ocular involvement is an important cause of disability in BD [4]. Approximately 50–70% of patients with BD have ocular involvement [4, 5]. In an international collaborative study from 14 countries, evaluated eye involvement in BD and overall blindness prevalence among BD patients with ocular lesions was 23% [9]. There are certain differences among countries, for instance visual loss prevalence was more than 30% in India, Iran, Japan, and Australia, on the other hand, almost 9% in Italy. In Turkey, visual loss prevalence among BD was 16.5%. One of the biggest tertiary center in Turkey, Istanbul University, found that, visual loss was almost 30% in male BD patients [10]. All these results demonstrated that, visual loss in BD is an important health care issue in certain countries such as Turkey.

Most of the people recorded in this center were male; therefore, all of our cases were male. Male BD patients have poor prognosis when they have major organ involvement such as eye and vascular. Male BD patients had more frequent visual loss in other series [11–13]. Similarly, all of the patients were male and were diagnosed between second–fourth decades. Interestingly, the median age from diagnosis to visual loss was 2.5 years. Overall visual loss survival was 6% at 1 year, 17% at 5 years, 25% at 10 years, and 29% at 20 years. According to this results, first decade is critical period for the prevention of visual loss in BD.

In an old Japanese study at 1965, BD prevalence among people with acquired blindness was 12% [14]. Our prevalence is obviously lower than this study. However, this Japanese study was conducted at pre-immunosuppressive period. Indeed, at the mentioned study, they compared visual loss survival before and after 1990. In this center, immunosuppressive treatments were usually started after 1990. After 1990, 5 years' visual loss survival (16 vs 26%) was significantly less in male BD patients.

The major limitation of our study was lack of complete eye examination of all BD patients. Although, we do not have any doubt about diagnosis because of past medical history, we could not demonstrate objective eye involvement. However, it is a big drawback that such a country like Turkey has no data on prevalence of BD among blind or visually impaired population. Our study is the first survey viewing BD frequency among a visually impaired adult group. We cannot estimate the proportions per population with our small sample size. Therefore, community-based surveys among visually impaired people are needed for population estimations.

Conclusion

In conclusion, BD is still one of the causes of acquired visual impairment in Turkey although immunosuppressive treatments are used in routine practice. In this study, BD accounted for 5.5% among adults who had acquired visual impairment. Extended population-based studies are needed for population estimations.

Compliance with ethical standards

At the beginning of the phone interviews, verbal informed consents were obtained from everyone attending the study. This study has been approved by Hacettepe University Ethics Commission (Approval number: GO 16/287 - 23). Survey was conducted in accordance with the Declaration of Helsinki.

Conflict of interest The authors declare that there is no conflict of interest.

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