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Assessment of Health Related Quality of Life (HRQoL) in Patients with Oromandibular Dystonia

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Abstract

Objectives: There is paucity of literature regarding health related quality of life in Oromandibular dystonia (OMD) especially from India. This study assessed HRQoL in its global and disease specific aspect by previously validated instruments in patients with OMD. *Method:* Subjects with OMD as well as age and gender matched healthy controls were enrolled from Movement Disorder OPD and botulinum toxin clinic. Uneducated patient, those could not read questionnaires, cases that had associated other neurological or debilitating systemic disorders, secondary / pediatrics dystonias, pregnancy or received botulinum toxin within 6 months or underwent surgical treatment were excluded from the study. Each patient filled SF-36 (HRQoL), BDI (Beck Depression Inventory for depression) and Oromandibular dystonia rating (OMDRS for Disease Severity Scale). *Results:* 42 pts of OMD were enrolled. There was no significant difference in demographic details between patients with OMD and control. Compared with controls, OMD patient group suffered from statistically significant impaired global health related quality of life (SF 36) in all domains ($p < 0.05$). More than 60% of patients with OMD had depression (compared to <25% of controls) out of whom 11% had moderate to severe depression (compared to 3.6% of controls). 4.7% of OMD patients had minimal severity scale scoring while 76.2% had moderate, 19.1% had severe disease scoring. *Conclusion:* This study clearly demonstrated that patients with OMD suffered from significant impairment in HRQoL as compared to controls. Higher proportion of patients with OMD suffered from moderate to severe depression compared to their control.

Keywords: OMD = Oromandibular dystonia; HRQoL = Health related quality of life; SF -36 = 36 item short form health survey; BDI = Beck's depression inventory; BRS: Blepharospasm rating scale; OMDRS = Oromandibular dystonia rating scale

Introduction

Dystonia is defined as a "neurologic syndrome characterized by involuntary, sustained, patterned contractions of opposing muscles, causing twisting and repetitive movement or abnormal postures"[1,2] may be associated with tremor (dystonia tremor) or myoclonus (myoclonic dystonia). Oromandibular dystonia is a focal dystonia characterized by forceful contractions of the face, jaw, and/or tongue causing difficulty in opening and closing the mouth, often affecting chewing and speech.

Primary dystonia is one of the most prevalent movement disorders. Worldwide prevalence of focal dystonia varies from 3 to 73 per 1,00,000 population from various studies[3-5] In only Indian community based study, crude prevalence rate of focal dystonia is 43.9 per 1,00,000 population.[6] In majority of studies cervical dystonia (CD) and blepharospasm (BSP) are found to be common dystonia accounting for about 75% of cases with primary focal dystonia. Indian study by Das et al., shows that writer's cramp and blepharospasm are the most common focal dystonia.[6] Oromandibular dystonia (OMD) is found to be a less common form of focal dystonia.

Very little is known about impact of OMD on quality of life. In most cases life expectancy is not reduced; however; it may be responsible for considerable morbidity in terms of pain, low self-esteem, depression, embarrassment and poor social interaction. Health-related quality of life (HRQoL) is a multi-dimensional concept that encompasses the subjective assessment of the impact of illness or treatment across the physical, psychological, social and somatic domains of functioning and the well being.[7]

There is paucity of literature regarding health related quality of life in focal dystonia especially from India. Little is known about the clinical and demographic factors associated with poor HRQoL and depression in patients with OMD.

Since most of the oromandibular dystonia (OMD) have a lifetime visible chronic disability, it is important to identify the factors that influence quality of life in these patients to optimize the goal directed therapy. This study was conducted to assess HRQoL in its global and disease specific aspect by previously validated instruments in patients with OMD.

Materials and Methods

The study was performed between Sep 2016 and Aug 2018 in GSVM, Medical college, Kanpur, India. Subjects with Oromandibular dystonia (OMD) as well as age and gender matched healthy controls were enrolled from movement disorder clinic, Department of Neurology, GSVM, Medical college, Kanpur. All the patients aged > 15 years with clinical diagnosis of OMD by Neurologist, were screened for the enrollment in the study. Uneducated patient, those could not read questionnaires, cases that had associated other neurological or debilitating systemic disorders, secondary/pediatrics dystonias, pregnancy or received botulinum toxin within 6 months or underwent surgical treatment were excluded from the study. Ethical clearance was taken from institutional ethical committee (IEC) of All India Institute of Medical Sciences, New Delhi. Written informed consent was taken after explaining nature and need of the study. Finally study patients were enrolled after fulfilling the inclusion and exclusion criteria.

Oromandibular dystonia is a focal dystonia characterized by forceful contractions of the face, jaw, and/or tongue causing difficulty in opening and closing the mouth, often affecting chewing and speech.

Study questionnaire: After enrollment, demographic and clinical details of cases were noted down in a preset form designed for the study. Each patient filled SF-36 (for HRQoL), BDI (Beck Depression Inventory for depression). OMD severity scale was filled by investigator during same session.

Global HRQoL: SF-36^{R} (short form 36) is acceptable, internally consistent, valid and reliable measure of the health status of patients[8,9] SF36R a 36 item, self report generic measure that provides a profile assessment of health-related quality measuring[9] multi-item variables, which includes physical functioning (PF) 10 items; role limitation due to physical problem (RP, 4 items), bodily pain (BP, 2 items), general perception of health (GH, 5 items), vitality (VT, 4 items), social functioning (SF, 2 items), role limitation due to emotional problem (RE, 3 items) and mental health (MH, 5 items). A score ranging from 0 (worst health) to 100 (best health) is generated for each domain/ subscale.

Disease severity scales: In patients with OMD, oromandibular dystonia rating sacle (OMDRS) was used.

Statistical analysis: Comparison between all of the variables described earlier for patients vs. control was carried out using *t*-test and X (chi-square) test for continuous and categorical variables, respectively. Association between SF 36 subscales and variables addressing disease duration, age of onset and severity were evaluated by Pearson correlation coefficients. *p* values of 0.05 or less (2-sided) were considered statistically significant. All data were analyzed using SPSS 12 software.

Results

Demographic characteristics: This study included 42 patients of OMD. Demographic details of these patients are described in Table 1. There was no significant difference in demographic details between patients with OMD and controls.

QOL characteristics: Compared with controls OMD patient group suffered from statistically significant impaired global health related quality of life (SF36) in all domains ($p < 0.05$).

Patients with OMD had significantly worse BDI (mean 13.0 ± 8.7 vs 6.38 ± 5.8, $p < 0.001$) score as compared to control group (Table 2). More than 60% of patients with OMD had depression (compared to < 25% of controls) out of whom 11% had moderate to severe depression (compared to 3.6% of controls)(Table 2). 4.7% of OMD patients had minimal severity scale scoring while 76.2% had moderate, 19.1% had severe disease scoring.

There was no correlation of SF36 with age ($p > 0.1$) except mental health ($p = 0.018$) where younger patients performed better. Two domains of SF36, general health and mental health, showed significant correlation with education ($p < 0.05$), educated patient performed better in these domains. No correlation found between SF36 and marital status of patients, duration of disease and age at onset ($p > 0.1$). Male patients scored statistically better in vitality than the females ($p = 0.043$) but in other domains no significant difference was observed between two genders.

No correlation of BDI scores and disease severity score with any demographic or disease duration variables found ($p > 0.1$ in all). There was no statistically significant correlation of disease severity with any domains of SF36 or BDI.

Table 1: Demographic Characteristics among patients with Oromandibular dystonia and OMD- controls

Demographic Characteristics	OMD ($n = 42$)	Control ($n = 55$)	Significance p value
Age mean (years), (SD)	51.9 (10.2)	50.9 (10.6)	0.819 (NS)
Age groups, %			
1) <39 yr	9.5	12.7	0.711 (NS)
2) 40–49 yr	31	32.7	
3) 50–59 yr	31	36.4	
4) 60–69 yr	23.8	12.7	
5) >70 yr	4.8	5.5	
M:F (n)	28:14	34:21	0.391 (NS)
Education, (%)			
1) ≤ 12th standard	33.3	23.6	0.059 (NS)
2) Graduate	38.1	61.8	
3) Postgraduate or higher	28.6	14.5	
Marital status, (%)			
1) Married	97.6	94.5	0.416 (NS)
2) Non married	2.4	5.5	
Current employment status, (%)			
1) Currently employed	50	69	0.21 (NS)
2) Retired	16.7	1.8	
3) Housewives	33.3	29	
Age of onset mean (years), SD	48.3 (9.7)	NA	
Age of onset groups			
1) ≤ 39 yr	16.7	NA	
2) 40–49 yr	35.7		
3) 50–59 yr	38.1		
4) 60–69 yr	9.5		
5) >70 yr	0		
Duration mean (years), SD	3.7 (3.07)	NA	
Duration groups			
1) ≤2 yr	33.3	NA	
2) 3–5 yr	54.8		
3) >5 yr	11.9		

Table 2: QOL Characteristics among patients with Oromandibular dystonia and OMD- controls

	OMD	Controls	*p* value
SF 36 subscale, mean (SD)			
(1) Physical functioning (PF)	71.4 (22.8)	80.4 (19.0)	0.037
(2) Role physical (RP)	48.6 (32.8)	71.7 (24.9)	<0.001
(3) Bodily pain (BP)	63.32 (28.6)	81.8 (18.5)	0.001
(4) General health (GH)	42.3 (19.1)	70.4 (21.0)	<0.001
(5) Vitality (VT)	52.4 (20.2)	64.6 (22.8)	0.007
(6) Social functioning (SF)	62.9 (18.4)	79.5 (15.6)	<0.001
(7) Role emotional (RE)	51.5 (29.7)	80.5 (26.4)	<0.001
(8) Mental health (MH)	59.7 (16.8)	72.6 (18.2)	0.001
BDI mean (SD)	13.0 (8.7)	6.38 (5.8)	<0.001
BDI groups, No, %			
1) 1–10: Normal	38.1	74.5	
2) 11–16: Mild mood disturbance	35.7	16.4	
3) 17–20: Borderline depression	14.3	5.5	
4) 21–30: Moderate depression	4.8	3.6	
5) 31–40: Severe depression	4.8	0	
6) over 40: Extreme depression	2.4	0	
OMDRS severity scales mean (SD)	14.1(3.1)	NA	
OMDRS groups, %			
1) Minimal ≤ 25%	4.7	NA	
2) Moderate 26%–50%	76 2		
3) Severe 51–75%	19.1		
4) Very severe > 75%	0		

Table 3: Reviews of studies of BS and SD

SN	Author, Year	n	Scale used	Comments
Oromandibular dystonia				
No study found which address issue of HRQoL and depression in patients of OMD who have not received botulinum toxin in last 6 months.				
Oromandibular dystonia and effect of botulinum toxin				
1	Bhattacharyya N, Tarsy D. 2001[16]	SD: 18 OMD: 5 Total: 23	GBI (Glas gow benefit inventory)	No general or disease specific quality of life scale used. OMD/SD: Significant benefit with Btx treatment with Btx for these conditions is effective on the basis of quality of life criteria
2.	Allyson D et al. June 2017[17]	OMD = 10, 10 controls	American Speech-Language-Hearing Association's Quality of Communication Life Scale	Study was designed to assess impact of botulinum toxin on communication related QoL in OMD patients.
3.	Nastasi L et al. Sep 2016[18]	OMD = 30	A disease-specific questionnaire, the oromandibular dystonia questionnaire-25 (OMDQ-25)	Study was designed to assess impact of botulinum toxin on communication related QoL in OMD patients.

(BDI = Beck's depression inventory, BS = blepharospasm, BRDS: Blepharospasm Rating/Disability Scale, Btx: botulinum toxin, CD = cervical dystonia, GBI: Glasgow Benefit Inventory, HFS: hemifacial spasm, NEI-VFQ; national eye inventory- visual function questionnaire, OMD: Oromandubular dystonia, QoL= quality of life, SD: spasmodic dysphonia, Pts= Patients)

Discussion

This study was designed to provide information regarding impact of Oromandibular dystonia (OMD) (botulinum toxin naïve patients) on QoL consisting of physical, psychological and social aspects of life. As effect of botulinum toxin usually last for 2–4.5 months[10–15], all the patients who received boulinum toxin within 6 months were excluded from study.

HRQoL is a tool to assess impact of disease and treatment on QoL. Studies regarding impact of

OMD on HRQoL are very limited in numbers[16–19] Table 3. There is no study in patients with OMD from India or Asia. Most of the previous studies were designed to see the impact of botulinum toxin on improvement in HRQoL and found that effect was mild and not satisfying.[16–18] In most of previous studies sample size of patients was very small.

This study clearly demonstrated that patients with OMD suffered from significant impairment in HRQoL as compared to controls. In SF-36, patients with OMD had impaired HRQoL in all the 8 domains. A recent meta-analysis by Ayisha G. et al.[19] showed that only 3 studies addressed HRQoL in patients of OMD. These 3 studies were aimed to see the effect of BTX on HRQoL in Total 72 patient of OMD.[16–19] Significant effect of BTX is found in all the 3 studies. Studies on HRQoL in other focal dystonia shows similar finding.[14,15,19] Some studies showed worse score in patients of blepharospasm (other focal dystonia) in all eight domains of SF-36 (compared to normal population) Muller J et al.[14]

Higher proportion of patients with OMD suffered from moderate to severe depression compared to their control. Higher incidence of depression could be due to impaired mental and social life and disfigurement. No study addressed the issue of depression in patients of OMD. Although depression is also found in patients with other focal dystonias like Blepharospasm, Scheidt CE, et al.[14,15]

There was negative correlation of mental health with age domain in OMD patients, concluding that younger patients performed better. Male patients with OMD scored mildly better than females in vitality. This may reflect facial disfigurement and feeling of being ugly hampering the social life in females. Positive correlation of educational status was seen with domains of general health and mental health of SF-36 in OMD. This reflects that educated patients can understand, accept and handle disorders, better than less educated patients.

There was no correlation of disease severity or depression (BDI) with age, gender, marital status, occupational status, education, age at onset and duration of disease in patients with OMD. On the contrary there was no statistically significant correlation of disease severity with HRQoL, or depression in patients of OMD suggesting that severity of disease is not the only factor which determines QoL. Other factors like, educational level, duration of disease or age contribute greatly to QoL.

Conclusion

The present study's result regarding HRQoL in its global and disease specific aspect provides further evidence for profound impact of OMD on physical, psychological and social aspect of quality of life. This study indicates that psychological counseling of patients, their family members and treatment aiming to treat depression may be a part of comprehensive treatment approach for these patients. Treatment should also improve quality of life of patients which is found lacking in several treatment trials of botulinum toxin[16,17] indicating requirement of much more broad and comprehensive approach.

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