Original Article

Ptosis associated with Monocular Elevation Deficiency

Saemah Nuzhat Zafar, Ayesha Khan, Nadia Azad, Mahmood Ali, Samina Naseer, Sarah Iqbal
Al-Shifa Trust Eye Hospital, Jhelum Road, Rawalpindi, Pakistan.

Abstract

Objective: To document the various clinical features of ptosis associated with monocular elevation deficiency (MED) seen in patients, presenting to the Paediatric and Strabismus Unit, over a period of 2 years.

Methods: All patients seen with monocular elevation deficiency presenting to the Strabismus Clinic from January 2006 to December 2007 were examined and evaluated for presence of associated ptosis, jaw winking phenomenon and pseudoptosis. Patients having aquired causes of monocular limitation in elevation were excluded.

Results: A total of 22 patients having MED were seen. Out of these 50% were males (N=11) and 50% females (N=11). Twelve (54.54%) had MED in the left eye and 10 (45.45%) had MED in the right eye. Ptosis was present in the eye affected with MED in 16 (72.72%) patients. Pseudoptosis was seen in 4 (18.18%) patients whereas no associated ptosis was noticed in 2 (9.09%) patients. Jaw winking phenomenon was present in 9 (40.90%) which comprise almost half (56%) the MED cases with ptosis.

Conclusion: Careful clinical assessment for ptosis, pseudoptosis and jaw winking phenomenon before forced duction test, can help in planning the correct order of surgical management of patients having monocular elevation deficiency. The patient needs to be counseled regarding the multiple surgeries required according to associated clinical features present with MED (JPMA 59:522; 2009).

Introduction

Monocular elevation deficiency (MED) is characterized by congenital unilateral defect in elevation of the eye, both in abduction and adduction (Figure). Saccadic velocity measurements and forced duction tests reveal both restrictive and paretic etiology so that a tight inferior rectus and superior rectus palsy may co-exist. In addition, clinical picture of MED may be due to a supra nuclear problem.[1] Association of Marcus Gunn jaw-winking phenomenon, congenital ptosis and MED indicates congenital misdirection syndrome involving the oculomotor nerve.[2] As with congenital fibrosis syndrome abnormal development of cranial nerves may be the cause of tight inferior rectus. Variable associated clinical features are seen with MED which require different approach to management of the condition. For patients requiring surgical intervention it is important to document and analyze pre-surgical data and counsel the patient about possibility of multiple surgeries.

This study was done to document the various clinical features of ptosis associated with MED, presenting to the Paediatric and Strabismus Unit of Al-Shifa Trust Eye Hospital, Rawalpindi.

Methods

Twenty two patients of MED observed over a period of 2 years from January 2006 to December 2007 were included in the study. Informed consent was obtained from the patients. History of presenting complaints, prescription of glasses, patching, trauma or previous ocular surgery was recorded. Past

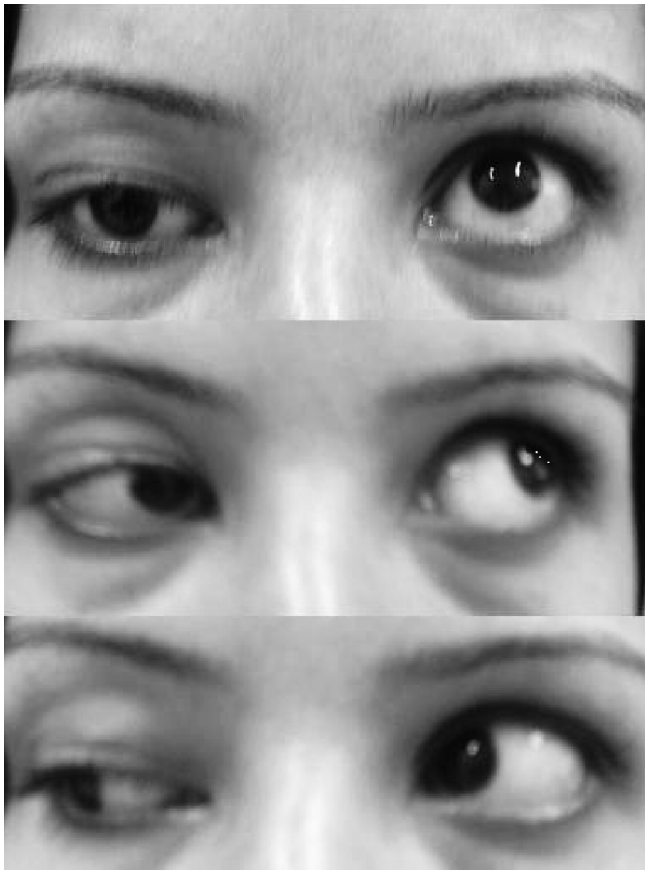

Figure: Right monocular elevation deficiency with ptosis.

medical history, family history and birth history were noted. Visual acuity was recorded by Snellen or EDTRS chart and Lea symbols where appropriate. Cycloplegic refraction in children and autorefraction in adults was performed. Anterior segment examination by slit-lamp biomicroscopy was conducted. Examination of the fundus was done after dilating the pupil. Compensatory head posture and results of cover test for near and distance were recorded. Prism cover test was performed in different gazes. Extra ocular movements were assessed for any under or over action of muscles. Versions and ductions were recorded on a diagram depicted by 0 for no restriction to -4 for very severe limitation in the direction of action of the muscle. Along with orthoptic assessment of monocular elevation deficiency presence of ptosis, pseudoptosis and jaw winking phenomenon was noted. Jaw winking was noted as mild, moderate and severe. Severe jaw winking included visible sclera above the superior limbus during the wink. Marginal reflex distance, palpebral fissure height, levator function, Bell phenomenon and any change in postion of the lid on fixing with the eye with MED was noted in case of ptosis. Patients requiring surgical management were counseled about possibility of multiple surgeries according to clinical assessment. Patients with acquired onset of monocular elevation deficiency, history of trauma, myasthenia gravis, thyroid eye disease, third nerve palsy, Brown syndrome and orbital diseases were excluded. Fisher's exact test was used to calculate significance.

Results

Age range of 22 MED patients was 5 to 26 years (mean 13.37 ± 5.5 years). Half of the patients with MED were male (N=11) and half female (N=11). Twelve (54.54%) had MED in the left eye and 10 (45.45%) had MED in the right eye. Ptosis of varying severity was present in 16 (72.72%) patients. Pseudoptosis was seen in 4 (18.18%) patients. Two (9.09%) patients had no associated ptosis. Jaw winking phenomenon was present in 9 (40.90%) which is almost half (56%) the MED cases with ptosis in this study (Table).

Table: Demography of patients with right MED and ptosis.

Case No	Age (years)	Sex	MED	Ptosis	Jaw-Winking
1	7	M	R	+	-
2	15	F	L	+	+
3	19	F	L	+	-
4	8.6	M	R	+	+
5	20	F	R	+	+
6	20	F	R	-	-
7	16	M	R	+	+
8	6	M	L	Pseudo	-
9	5	M	L	+	-
10	7	F	L	-	-
11	14	F	L	Pseudo	-
12	13	M	R	+	-
13	8	F	R	+	-
14	22	F	L	Pseudo	-
15	26	M	L	Pseudo	-
16	16	M	R	+	+
17	13	F	L	+	+
18	13	M	L	+	+
19	9	M	L	+	-
20	10	M	R	+	+
21	12	F	R	+	-
22	13	F	L	+	+

MED= Monocular Elevation Deficiency, M= male, F= female, += present, -= absent, Pseudo= pseudoptosis.

Discussion

Mild to moderate homolateral ptosis is frequently observed in patients with MED.[3] Along with ipsilateral ptosis, hypotropia and chin elevation is often seen in these patients.[4] True congenital ptosis is described in 25% of cases whereas pseudoptosis is seen mostly in all patients having large hypotropia.[5] The difference in right and left eye involvement as well as gender distribution is not statistically significant in the present study. Presence of ptosis and pseudoptosis compared to no ptosis was statistically significant (P= 0.0045) in our patients. Due to this fact cosmetic correction of the drooping lid is the usual presenting feature apart from the problem related with ocular deviation, limited elevation and compensatory head posture. True ptosis was present in 29% and Marcus Gunn jaw winking phenomenon was present in 2 cases (7%) in one study which included 28 patients.[6] Ptosis noticed in our study (73% of 22 MED patients) was not quite statistically significant but jaw winking noted in our patients (41%) was statistically significant compared with the above mentioned study. Patients of MED having congenital ptosis showed Marcus Gunn jaw winking phenomenon in 25% cases.[2] A study of 11 MED patients showed pseudoblepharoptosis in all the patients.[7] This is not statistically significant compared to our study. Marcus Gunn jaw winking phenomenon was present to a varying severity in almost half (56%) of the MED patients having ptosis which is higher than reported in other studies. Managing MED patients includes treatment of amblyopia, compensatory head posture, hypotropia and any horizontal deviation if present. Ptosis surgery in MED is done after correction of hypotropia which depends on results of forced duction test.[8-11] Assessment of pseudoptosis is essential in management as it is corrected once the hypotropia is taken care of.[9] Pseudoptosis was seen in 4 (18%) patients in the present study and the patient and parents were cautioned that ptosis correction by surgery may be required after correcting hypotropia. Residual hypotropia after initial Knapp's procedure for MED may improve with time.[10] Management strategy for residual hypotropia after Knapp's procedure includes recession of ipsilateral inferior rectus or recession of superior rectus of

the other eye. If only inferior rectus has been recessed, partial horizontal rectus muscles vertical transposition can be done.[11] Bell phenomenon is impaired in patients with restriction having infranuclear etiology.[12] Ptosis needs to be under corrected in such cases as there is risk of corneal exposure.[13] Bell phenomenon may improve after hypotropia and pseudoptosis are corrected by Knapp procedure. Correction of ptosis and pseudoptosis includes muscle transposition operation, levator resection or frontal suspension surgery.[14] In Marcus Gunn jaw winking phenomenon levator disinsertion and sling surgery may be required. Dissociated vertical deviation (DVD) has been described in association with monocular elevation deficiency.[15] DVD was not seen in our patients. Hypotropia and associated ptosis may lead to amblyopia and loss of binocularity.[6] Patients generally present early due to the congenital onset of MED. In the present study however a 26 year old patient presented for the first time mainly for a cosmetic correction of the condition. Post operative complications such as under correction, over correction, diplopia, anterior segment ischaemia, horizontal tropia and lower lid retraction can be anticipated by carefully recording the preoperative findings with photographic record. Surgeries can hence be planned accordingly.

Conclusion

Differentiation of MED on the basis of paretic or restrictive etiology, presence of ptosis, pseudoptosis and jaw winking phenomenon is important for appropriate surgical planning. Association of jaw winking phenomenon seen in our MED patients is more than reported in other studies. The present observational series is part of an ongoing interventionl study of patients with MED.

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