Sudden-onset trochlear nerve palsy: clinical characteristics and treatment implications

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PURPOSE To identify clinical characteristics, etiologies, and treatment implications of sudden-onset, acquired cases of superior oblique palsy.

METHODS The medical records of patients diagnosed with trochlear nerve palsy between January 2010 and January 2018 were reviewed retrospectively to identify cases of acquired trochlear nerve palsy with a specific date of onset of acute symptoms or specific causative incident. Patients with congenital palsies or an uncertain date of onset, history of other strabismus, concomitant oculomotor or abducens nerve palsies, or history of strabismus surgery were excluded.

RESULTS Of 214 patients with superior oblique palsy, 23 had sudden-onset, acquired palsies. There were 14 cases of unilateral palsy and 9 cases of bilateral palsy. Patients with unilateral palsy presented with vertical diplopia, while those with bilateral palsy complained of either torsional (4/9 patients) or vertical (5/9 patients) diplopia. The most common etiologies were severe trauma associated with traumatic brain injury, followed by central nervous system neoplasm and stroke. Fifteen patients underwent surgical intervention, 3 of whom required more than one surgery.

CONCLUSIONS Patients with acute superior oblique palsy invariably complain of vertical and/or torsional diplopia. Neuroimaging should be considered in cases of acquired superior oblique palsy without a known traumatic cause. Bilateral cases of acquired superior oblique palsy are more challenging to manage surgically because of symptomatic torsional diplopia. (J AAPOS 2019;23:321.e1-5)

Trochlear nerve palsy represents the most common paralysis of a single cyclovertical muscle. Patients with both decompensated congenital and acquired superior oblique palsies present with varying degrees of cyclovertical diplopia, torticollis, asthenopia, and other nonspecific symptoms.[1] It is generally understood that large vertical fusional amplitudes and chronic head tilt seen on review of old photographs suggest long-standing congenital palsies. Acquired palsies frequently result in acute and debilitating diplopia and torsional symptoms.[2] However, these distinctions are not always apparent, and there may be considerable overlap in presentation for patients in whom the etiology is not clearly congenital or acquired.[3,4]

Acquired superior oblique palsy is usually caused by closed head trauma, vascular injury to the central nervous system (CNS), or brain tumor and may therefore be the presenting sign of underlying neurologic or systemic pathology. Differentiating between congenital and acquired etiology is important in guiding further evaluation of these patients, because a recently diagnosed palsy without a known traumatic etiology could suggest the possibility of intracranial pathology and need for further investigation, including neuroimaging.[5]

The distinction between congenital and acquired superior oblique palsies also affects management and treatment options for patients. Previous reports have suggested that acquired ischemic or idiopathic etiologies of trochlear nerve palsy should be observed without surgical intervention for at least 4-6 months after onset of symptoms because spontaneous improvement can occur. In contrast, decompensated congenital palsies do not improve over time, and surgical intervention can alleviate the patient's symptoms without delay.[5] Additionally, clearly acquired superior oblique palsy due to neurogenic injury presents with different symptoms and motility characteristics, requiring different treatment strategies than congenital palsies. The purpose of the current study was to characterize the clinical characteristics and etiologies of sudden-onset, clearly acquired cases of superior oblique palsy. We studied only cases where the patient could precisely identify the date of onset of their symptoms. This allowed us to examine only cases of acquired palsy without the confounding effects of cases where the onset was more obscure.

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Subjects and Methods

This study was approved by the Indiana University Institutional Review Board and adhered to the tenets of the Declaration of Helsinki.The medical records of patients diagnosed with trochlear nerve palsy between January 2010 and January 2018, identified by database search of ICD-9 and 10 codes (378.53, H49.11-13), were reviewed retrospectively. All patients were examined and treated by physicians of the Pediatric Ophthalmology and Adult Strabismus section of Indiana University Health Physicians at Riley Hospital for Children and Midwest Eye Institute. Cases of acute-onset binocular vertical or torsional diplopia consistent with definitively acquired trochlear nerve palsy with a specific date of onset of acute symptoms were identified. Patients with congenital palsies or an unknown or unclear date of onset of symptoms, history of other strabismus (such as esotropia or exotropia), concomitant oculomotor or abducens nerve palsies, or history of prior strabismus surgery were excluded.

Detailed, deidentified data were extractedfrom the records, including demographics, subjective symptoms, etiology, laterality, visual acuity, ductions and versions, head posture, and results of any neuroimaging. Measurements of deviation in all gazes, and head tilts, when available, documented by alternate prism cover testing and torsional deviation measured by double Maddox rod in primary position were recorded. Finally, surgical interventions and postoperative alignment and torsional deviation when available were also recorded. The data were then carefully analyzed for overall trends in presentation, patterns in etiology, and outcomes of surgical correction.

Results

Of 214 patients diagnosed with trochlear nerve palsy, 23 patients (12 males) met inclusion criteria. These patients were able to identify a specific inciting event or date of symptom onset. Eleven additional patients had acquired palsies but were excluded because of concomitant palsies of other cranial nerves or associated strabismus of other etiology. In these 11 patients, the etiology of multiple cranial nerve palsies was traumatic brain injury associated with motor vehicle accident in 4 patients, CNS tumors treated with surgical resection in 3 patients, ruptured intracerebral aneurysm in 1 patient, cryptococcal meningitis with stroke in 1 patient, and iatrogenic cranial nerve palsies after head and neck surgery in 1 patient. The etiology of multiple cranial nerve palsies in the final patient was unknown.

Eighty patients had clearly congenital palsies, which were defined as a vertical strabismus or torticollis beginning in infancy either by clinical history or examination at an early age. One hundred additional patients had likely congenital palsies. These patients were asymptomatic and not diagnosed with superior oblique palsy at an early age but did show signs of longstanding deviations on initial presentation such as characteristic facial asymmetry, large vertical fusional amplitudes, and longstanding head posture on review of photographic history.

The average age of those with sudden-onset acquired palsies was 45.1 years (range, 7-87 years). Five patients were ≤18 years of age at the onset of their palsy. Etiology of all cases of sudden-onset acquired palsies was trauma in 12 cases, known brain tumor after neurosurgical intervention in 4 cases, and cerebral vascular accident in 3 cases. Two cases were of microvascular etiology, and another case occurred after a viral infection. The etiology of the final case remains unknown (Tables 1 and 2).

The chief complaint of all patients with sudden-onset acquired palsy was acute vertical or torsional diplopia, occurring immediately after the inciting event. All 14 patients with unilateral palsy complained of vertical diplopia, 1 of whom noted vertical and torsional components. No patients with unilateral palsy complained exclusively of torsional diplopia. However, patients with bilateral palsies complained primarily of either torsional diplopia (3/9 patients), vertical diplopia (5/9 patients), or both (1/9 patients). The mean vertical deviation on presentation for all patients with sudden-onset, acquired superior oblique palsy was relatively small, at 6.1^{Δ} of hypertropia (range, 2^{Δ}-20^{Δ}). The mean vertical deviation for unilateral cases was 6.5^{Δ} of hypertropia (range, 2^{Δ}-16^{Δ}), whereas for bilateral cases, this value was also small, at 5.4^{Δ} of hypertropia (range, 2^{Δ}-20^{Δ}). Patients with unilateral palsies had positive Bielschowsky head tilt testing on the ipsilateral side. The majority of bilateral palsies (6/9 patients) had alternating hypertropias in right and left gazes (right hypertropia in left gaze and left hypertropia in right gaze). The remaining 3 of 9 bilateral palsies had excyclotorsion of >15° and bilateral fundus excyclotorsion on fundoscopic examination. Twenty patients had documented excyclotropia measured by double Maddox rod testing, with an average of 12.05° (range, 3°-25°). Two patients had no measurable torsion, and torsional deviation was not recorded for 1 patient. Those with bilateral trochlear nerve palsy had greater excyclotorsion (mean, 15.2°; range, 5°-25°) compared to unilateral cases (mean, 8.9°; range, 3°-20°). On version testing, 17 of 23 patients (74%) showed varying degrees of superior oblique muscle underaction, and 15 (65%)showed only mild inferior oblique overaction. The remaining 8 patients (35%) showed no evidence of inferior oblique overaction.

Sixteen patients had neuroimaging done prior to presentation. Five patients had normal magnetic resonance imaging results (1 patient with unknown etiology, 1 patient with a traumatic etiology after a motor vehicle collision, 2 cases of microvascular trochlear nerve palsy, and 1 postviral case). The other 11 showed structural or traumatic changes consistent with the underlying cause of the patient's palsy. These results included CNS tumors with post-neurosurgical structural changes, and evidence of severe trauma such as intracranial hemorrhage, facial fractures, and stroke. The 7 patients who did not have neuroimaging had a clear, known history of major head trauma that was thought to be the causative event of their palsy and did not undergo further investigative neuroimaging.

Table 1. Unilateral superior oblique palsy

					Presenting alignment				Postoperative alignment		
					Primary vertical deviation				Vertical deviation		
Case	Age of onset, years	Sex	Etiology	Laterality	Type	Amount, PD	Excyclo	Surgery	Type	Amount, PD	Excyclo
1	58	M	Microvascular	R	R-HT	6	12				
2	50	F	Postviral	L	L-HT	14	3	R-IR recession	L-HT	8	1
3	58	M	Unknown	R	R-HT	4	6				
4	74	M	CVA	R	R-HT	6	20	R-IO myectomy	R-HT	0	
5	78	F	Severe head trauma	L	L-HT	3	3				
6	81	F	Microvascular	L	L-HT	2	0				
7	20	F	Severe head trauma	R	R-HT	3	5				
8	18	M	Severe head trauma	L	L-HT	4	15	L-IO recession	L-HT	2	8
9	62	F	Brain tumor/post-neurosurgical	L	L-HT	14	10	L-IO myectomy	L-HT	6	
10	57	F	CVA	L	L-HT	10	10	L-IO myectomy	L-HT	6	
11	49	M	Severe head trauma	L	L-HT	2	0				
12	15	M	Severe head trauma	R	R-HT	4	0	R-IO myectomy	R-HT	0	
13	39	F	Brain tumor/post-neurosurgical	R	R-HT	3	5				
14	25	M	Severe head trauma	R	R-HT	16	0	R-IO recession	R-HT	5	

CVA, cerebral vascular accident; *Excyclo*, excyclotorsion (by double Maddox rod); *HT*, hypertropia; *IO*, inferior oblique; *IR*, inferior rectus; *PD*, prism diopter.

One patient who had a unilateral microvascular trochlear nerve palsy had spontaneous resolution of symptoms and improvement in vertical strabismus from 6^{Δ} of hypertropia to no hypertropia over 4 months. Another patient with post-traumatic superior oblique palsy also had improvement in her symptoms over 8 months following her injury, with associated improvement in her strabismus from hypertropia of 7^{Δ} to 3^{Δ}.

Fifteen patients had undergone surgery at the time of our chart review. All patients reported subjective improvement in their diplopia postoperatively, with a range of follow-up intervals of 1-12 months. Of these 15 patients, 3 required two strabismus surgeries.

Four patients who underwent unilateral inferior oblique myectomy were satisfied with their surgical outcome and did not undergo further surgery. Two patients with unilateral traumatic superior oblique palsy underwent inferior oblique recession, also with satisfactory outcomes.

Four patients with bilateral superior oblique palsy underwent bilateral Harada-Ito procedures with bilateral inferior rectus recessions. This resulted in an overcorrection and subsequent incyclotorsion in 1 patient, who required a second surgery. This patient had a preoperative vertical deviation of 2^{Δ} of left hypertropia in primary gaze and 15° of excyclotropia. Postoperatively, the patient was overcorrected to 4° of incyclotorsion and consecutive 2^{Δ} of right hypertropia, which resolved to no torsion and no vertical misalignment after an advancement of one of the previously recessed inferior rectus muscles. One case had persistent torsional deviation and inferior oblique overaction postoperatively, requiring a second surgery (bilateral inferior oblique myectomies).

Discussion

Our study uniquely describes the etiologies and clinical characteristics of sudden-onset acquired unilateral and bilateral trochlear nerve palsies. Although many patients with a trochlear nerve palsy present with diplopia, it is often difficult to determine whether the underlying cause is decompensation of a congenital condition or an acquired etiology. Our patient cohort included only those patients with acute-onset acquired trochlear nerve palsy. It is noteworthy that whereas patients with congenital superior oblique palsy or imprecise etiology of superior oblique palsy frequently present with vague complaints of visual discomfort, torticollis, or other nonspecific symptoms, all patients in this series presented with a principle complaint of diplopia—either vertical, torsional, or both.

The mean age of onset of acquired trochlear nerve palsy in our study is similar to that reported previously.[6] Of note, 22% (5/23) cases occurred in children (≤18 years of age), including 2 unilateral cases and 3 bilateral cases, with etiologies of trauma or surgery for CNS malignancy.

The most common etiologies of sudden-onset trochlear nerve palsy in our series were severe trauma and traumatic brain injury, as others have reported,[5,7,8] followed by CNS neoplasm and stroke. Microvascular causes were less common in our series (2/23 cases) compared with prior reports.[6] This could be because of the rigid inclusion criteria of unquestionably sudden-onset of symptoms in our study. Park and colleagues[6] assigned a vascular etiology to those patients with trochlear nerve palsy presenting with symptoms of diplopia of variable duration and at least one vascular risk factor without evidence of other etiology. Because evidence of

Table 2. Bilateral superior oblique palsy

				Presenting alignment				Postoperative alignment		
				Primary vertical deviation				Vertical deviation		
Case	Age of onset, years	Sex	Etiology	Type	Amount, PD	Excyclo	Surgery	Type	Amount, PD	Excyclo
1	14	F	Brain tumor/post-neurosurgical	R-HT	3	25	B–Harada-Ito with B-IR recession	R-HT	2	10
2	55	F	Severe head trauma	L-HT	2	5	B-IR recession and B-MR recession	L-HT	2	10
3	68	M	Severe head trauma	L-HT	9	20	B–Harada-Ito with B-IR recession	R-HT	2	0
4	7	M	Severe head trauma	R-HT	20	15	L–Harada-Ito with R-SO tuck	R-HT	12	4
5	87	F	Severe head trauma	L-HT	2	12	None			
6	21	M	CVA	L-HT	5	10	L–Harada-Ito with LLR recession	L-HT	2	3
7	32	M	Severe head trauma	L-HT	2	15	B–Harada-Ito with B-IR recession	R-HT	2	4
8	10	M	Brain tumor/post-neurosurgical	L-HT	2	20	B–Harada-Ito with B-IR recession	R-HT	0	0
9	59	F	Severe head trauma	L-HT	4	15	L–Harada-Ito	L-HT	2	2

B, bilateral; *Excyclo*, excyclotorsion (by double Maddox rod); *HT*, hypertropia; *IR*, inferior rectus; *LR*, lateral rectus; *MR*, medial rectus; *SO*, superior oblique.

decompensated congenital palsies can be subtle, the criteria outlined may overestimate the number of microvascular palsies in that report, as the authors have noted.[6] Additionally, our series includes only 1 case of spontaneous recovery, which differs from Park and colleagues' report, which found a higher rate of recovery of cranial nerve function. This could be due to referral patterns in our area of practice, where acute-onset vascular palsies may be seen and monitored to recovery by adult neuro-ophthalmologists without a need for strabismus surgical evaluation.

All cases of bilateral superior oblique palsy were secondary to trauma, underlying CNS neoplasm, or stroke; all microvascular and unknown causes were unilateral.[9] CNS lesions accounted for 2 cases of bilateral superior oblique palsy and 2 cases of unilateral superior oblique palsy, highlighting the importance of including CNS tumor in the differential diagnosis for both types of acquired palsies. While neuroimaging may not be required in the evaluation of all acquired trochlear nerve palsies, these findings suggest that it would be prudent to obtain neuroimaging in cases with unknown traumatic cause.

Eleven patients in our series were excluded from further investigation because multiple cranial nerve palsies were present. The etiologies of these cases were similar to those of isolated sudden-onset trochlear nerve palsy; without a known underlying cause, such patients warrant further evaluation with neuroimaging.[5]

Consistent with a prior report by Sydnor and colleagues[10] of exclusively traumatic superior oblique palsies, patients with unilateral sudden-onset, acquired trochlear nerve palsy complained of vertical diplopia. Additionally, torsional deviation was larger in patients with bilateral superior oblique palsy than in those with unilateral palsy. The mean torsional deviation in unilateral cases was somewhat higher than that reported by others in acquired palsies; however, our results may be affected by the relatively small sample size[11,12] and the acute nature of these palsies.

Not all patients presented immediately after onset of diplopia; thus a precise time between onset of deviation and surgical intervention is not available in our data set. Nevertheless, the authors' typical practice for cases of acquired cranial nerve palsy would be to wait at least 6-12 months to see whether the deviation resolved spontaneouslyor stabilized before recommending surgery. Surgical management was markedly different in unilateral and bilateral palsies. Of 7 patients with unilateral superior oblique palsy that were managed surgically, 6underwenta weakening procedure (recession or myectomy) on the ipsilateral inferior oblique. Although postoperative torsional data is not available for all of these patients, the patients did well and did not require further surgery. It is noteworthy that inferior oblique myectomy decreased hypertropia only 4^{Δ}-8^{Δ} in patients with acquired superior oblique palsy and mild inferior oblique overaction, regardless of the magnitude of their preoperative hyperdeviation, which is much less than in typical in cases of congenital palsy with larger deviations.

Bilateral trochlear nerve palsy is challenging to manage surgically. Surgical planning must include procedures aimed at reducing significant amounts of excyclotorsion. Of 8 patients with bilateral palsies, 7 underwent Harada-Ito procedures, often combined with inferior rectus recession. All 3 of the 23 cases requiring a second surgery in our series had bilateral palsies. The difficulty in surgical correction of these patients is due to torsional symptoms in addition to vertical deviation. The results of superior oblique surgical procedures in these cases is consistent with the

results reported by Arici and Oguz[13] on pre- and postoperative torsional deviation in patients after inferior oblique versus superior oblique procedures, where more torsional response was observed after superior oblique strengthening than inferior oblique weakening.

As suggested by Gräf and colleagues,[8] inferior oblique surgery may be considered in unilateral cases of trochlear nerve palsy with small vertical deviation and small degrees of torsional deviation and, importantly, no subjective symptoms of torsion preoperatively. When excyclotorsion is significant, as in our cases of bilateral trochlear nerve palsy, successful surgical treatment frequently requires strengthening procedures on the superior oblique.

Our findings are limited by the study's retrospective nature, relatively small sample size, variable follow-up intervals for postsurgical intervention, and lack of ophthalmic examination prior to the occurrence of the palsy. Additionally, the patients in this study came from a single strabismus referral practice, with several surgeons involved; therefore, results may not be representative of other populations.

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