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Original Article

Clinical Characteristics of Abnormal Postures of the Head and Neck Caused by Unilateral Superior Oblique Palsy

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Objective To present the clinical characteristics of abnormal posture of the head and neck (APHN) caused by unilateral superior oblique palsy (USOP) for differential diagnosis of childhood APHN.

Method The medical charts of children who came to Department of Physical Medicine and Rehabilitation, Ajou University Medical Center from 2003 to 2008 for APHN were reviewed with special reference to ocular causes of APHN. Children who showed a positive Bielschowsky's head tilt test were strongly suspected to have USOP. In-depth ophthalmic evaluations were done after 6 months of age to identify USOP for those children. The clinical characteristics of children with USOP were compared with those of children with congenital muscular torticollis.

Results Sixteen children were diagnosed with APHN related to ocular causes. Seven children out of 16 (43.8%) had APHN secondary to USOP, which was the most common ocular cause of APHN. The initial clinical presentation of 7 children with USOP was contralateral laterocollis toward the shoulder on the non-USOP side at a mean age of 15.57±12.55 months; USOP was diagnosed at a mean age of 19.07±11.29 months. APHN was aggravated by staring at objects, and craniofacial asymmetry was not evident at this age compared to children with congenital muscular torticollis.

Conclusion The clinical characteristics of USOP were presented, which showed laterocollis toward the shoulder of non-USOP. Differential diagnosis of APHN is critical for proper management for APHN and high index of suspicion for USOP by non-ophthalmologic physicians could make earlier diagnosis of USOP possible.

Key Words Torticollis, Superior oblique palsy, Strabismus

INTRODUCTION

When an infant comes to the hospital for abnormal posture of the head and neck (APHN), making an accurate differential diagnosis is necessary for providing the proper treatment. Congenital muscular torticollis (CMT) from a shortened ipsilateral sternocleidomastoid (SCM) muscle is the most common cause of APHN. For this diagnosis, identifying the thickened SCM muscle and the limitation of motion with a physical examination is necessary. The simultaneous difference between the bilateral SCM muscles' thickness can also be confirmed for making the diagnosis using ultrasound or other imaging modalities.[1,2] Various diseases besides CMT can

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lead to APHN, so making an accurate differential diagnosis is necessary for beneficial management. It is known that ophthalmological problems and not CMT cause 23% of APHN cases,[3] thus, ocular problems are the second most important part of differential diagnosis for APHN besides CMT. Hypertropia caused by unilateral superior oblique palsy (SOP) is the most well-known cause of ocular APHN.[4,5] There have been some reports of unnecessary stretching exercise using a neck splint and/or resection surgery to the SCM muscle when an incorrect diagnosis is made and the actual cause of APHN is due to ocular problems.[5,6] A belated diagnosis of ocular APHN can lead to secondary complications such as spinal scoliosis.[7-9] A confirmative diagnosis of an ocular cause leading to APHN belongs to the ophthalmological domain, yet well-informed non-ophthalmologists should be aware of the clinical features and physical findings of ocular APHN to refer the patient to an ophthalmologist when necessary. This will enable a timely diagnosis and proper treatment of ocular APHN.

We analyzed the clinical features of children who were diagnosed with unilateral SOP. This study intended to provide the clinical features that can be used by non-ophthalmologists to identify unilateral SOP.

MATERIALS AND METHODS

Diagnosing APHN from ocular causes

The subjects of this study were children who came to Ajou University Hospital, The Center for Torticollis and the Department of Physical Medicine and Rehabilitation and were diagnosed with ocular APHN. These subjects were below 18 years old and were seen at the hospital from January 2003 to April 2008. We conducted a retrospective study by reviewing the medical records. Todescribing the various shapes of APHN, we defined the following: torticollis as face rotation to the right or left side on a horizontal plane, and laterocollis as left or right tilting of the head and face on the coronal plane while the earlobe gets near the ipsilateral shoulder (Fig. 1).[2,10-12] We defined children with APHN that was not caused by CMT as those who 1) showed no visible shortening of the SCM muscle and no meaningful difference (less than 2 mm) between the SCM muscles with diagnostic ultrasound, and 2) showed ophthalmological problems such as strabismus, nystagmus, and ptosis during the physical examination. These children were referred to pediatric ophthalmology for the diagnosis of ocular APHN.

Diagnosis and clinical investigation of APHN from unilateral SOP

Among the children diagnosed with ocular APHN, we suspected unilateral SOP when laterocollis was the major symptom and a positive Bielschowsky head tilt test was shown (Fig. 2).[13] We sent the children after six months of age to a pediatric ophthalmologist for consultation for diagnosing unilateral SOP. At the age of six months or later, we performed the Bielschowsky test using toys or singing so that the child would not cry. If the Bielschowsky test was negative, we performed the exam again 2 months later. When the Bielschowsky test was positive, a referral to pediatric ophthalmology was made. For the cases of CMT, ipsilateral shortening of the SCM muscle can cause both ipsilateral laterocollis and contralateral torticollis, which together act simultaneously; rotation of the head subsequently causes the ear of the ipsilateral side to locate anteriorly and not in the coronal plane (Fig. 3). However, when laterocollis alone is present, there is no cervical rotation on the horizontal plane, and both ears are located on the same coronal plane. This is a differential point to distinguish different types of APHN (Fig. 4).

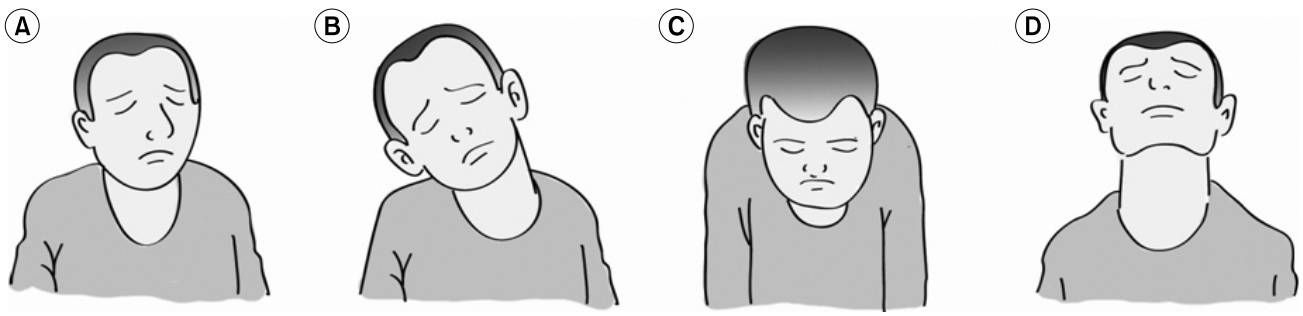

Fig. 1. Abnormal postures of the head and neck. (A) Left torticollis, (B) Right laterocollis, (C) Anterocollis, (D) Retrocollis.

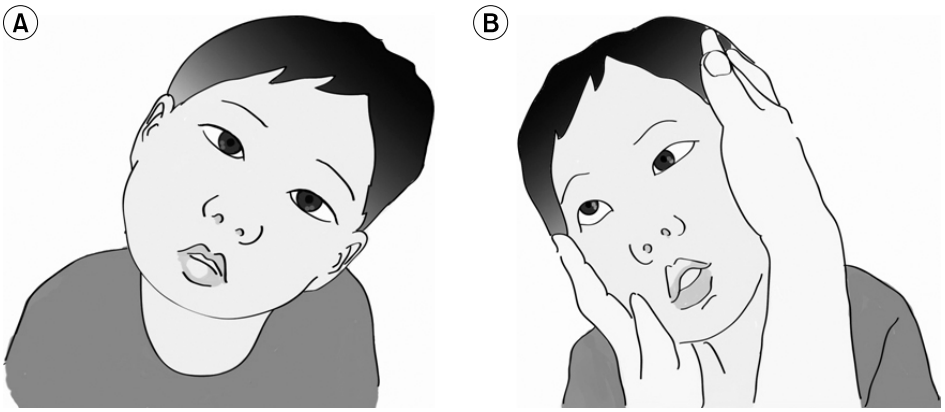

Fig. 2. Positive Bielschowsky head tilt test for a child who has right superior oblique palsy. (A) He has left laterocollis aggravated by staring at objects. (B) Tilting the head toward the shoulder on the affected side induces intorsion of the affected eye. Due to right superior oblique palsy out of two intortors (superior oblique muscle and superior rectus muscle), this maneuver results in upward deviation of the right eyeball caused by the right superior rectus muscle, which functions as both an intortor and an elevator of the eyeball.

Fig. 3. A child who has congenital muscular torticollis due to shortening of the left sternocleidomastoid muscle. She shows ipsilateral laterocollis and contralateral torticollis. Note that the left ear is located anterior to the right ear due to right torticollis.

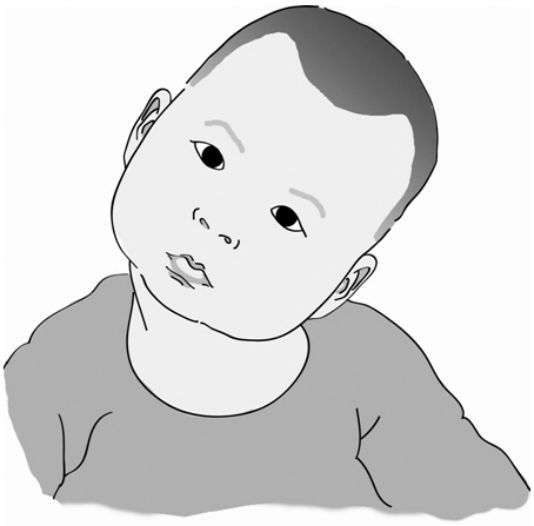

Fig. 4. A child who has right superior oblique palsy shows left laterocollis. Note that both ears are located on the same coronal plane.

The medical records of children with SOP-related APHN were reviewed for clinical features, such as age at first visit, features of the head and neck posture, the range of motion on cervical rotation, and plagiocephaly. Plagiocephaly was determined when there was skull asymmetry on visual inspection of the external appearance.

For the ocular APHN group, a neurodevelopmental examination, the Bayley Scales of Infant Development, brain MRI, chromosomal study, and metabolic disease screening were all done for identifying any accompanying neurodevelopmental disease, such as Down syndrome, intellectual disability, and spastic cerebral palsy.

Comparative analysis between APHN from ocular problems and CMT

In the CMT group, the subjects were children diagnosed with CMT from January 2003 to December 2006. The CMT showed a head tilt to the shortened muscle side and rotation to the contralateral side. CMT was diagnosed when the diseased SCM was thicker (more

than 2 mm) than the healthy side on the transverse view of an ultrasound examination. The age at the first visit, the feature of APHN, the range of cervical rotation, and the presence of plagiocephaly were investigated by reviewing the medical records. These features were analyzed and compared with APHN from unilateral SOP.

Statistics

Statistical analysis was taken with SPSS 11.5. The Mann-Whitney test was done for comparisons between unilateral SOP and CMT. Statistical significance was defined as p-values < 0.05.

RESULTS

Causes of APHN from ocular problem

Sixteen of the 510 children were diagnosed with ocular APHN. Among them, seven (43.8%) were diagnosed with unilateral SOP (Table 1). One of the other 9 children had unilateral ptosis, another had Moebius syndrome accompanied by unilateral sixth, seventh, and eighth cranial nerve palsy. Two had Down syndrome, another two had severe intellectual disabilities, and the remaining three were identified as having spastic cerebral palsy. These seven children showed nystagmus (two), esotropia (three) and refractory error (two), and the APHN might have been related to these problems.

Table 1. Ocular Causes of Abnormal Postures of the Head and Neck (n=16)

Ocular cause	Number of subjects
Unilateral superior oblique palsy	7 (43.8%)
Unilateral ptosis	1 (6.2%)
Moebius syndrome	1 (6.2%)
Ocular abnormalities associated with neurodevelopmental disorders (2 Down syndrome, 2 intellectual disability, 3 bilateral spastic cerebral palsy)	7 (43.8%; 2 nystagmus; 3 esotropia; 2 refractory error)

Clinical features of APHN from unilateral SOP

The clinical features of the seven APHN children with unilateral SOP are listed on Table 2. There were 4 boys and 3 girls; two had right unilateral SOP while 5 had left unilateral SOP. The children with unilateral SOP were suspected of having APHN at the age of 3 or 4 months with the beginning of good head control. The first visit was at the average age of 15.57±12.55 months. Their average age at the time of diagnosis age was 19.07±11.29 months. The 71 CMT children had their first visit to the hospital at an average of 1.90±2.72 months. A statistically significant difference was seen between the unilateral SOP children and the CMT children for the time of first visit ($p < 0.05$). Thus, the unilateral SOP children visited the clinic 13.69 months later than did the CMT children. For the cases of APHN with unilateral SOP, laterocollis on the coronal plane to the contralateral side of the involved eye was noted and could minimize the superior oblique muscle's intortor action. In other words, the right SOP palsy children showed laterocollis to left side and the left palsy children showed laterocollis to the right (Fig. 4). Because there was no shortening of the SCM muscle,

Table 2. Comparison of Clinical Characteristics between Abnormal Postures of the Head and Neck (APHN) due to Unilateral Superior Oblique Palsy (SOP) and APHN due to Congenital Muscular Torticollis

Clinical characteristics	APHN due to unilateral SOP (n=7)	APHN due to congenital muscular torticollis (n=71)
Age at the first visit*	15.57±12.55 months old	1.90±2.72 months old
Type of APHN	Laterocollis toward the shoulder without SOP	Ipsilateral laterocollis toward the shortened SCM and contralateral torticollis
Rotation of the neck at the first visit (mean±SD°)*	Full rotation toward both right and left sides (90.00±0)	Limited rotation toward the shortened SCM (57.80±16.33)
Number of subjects who showed aggravation of APHN when staring at objects*	7 (100%)	0 (0%)
Number of subjects with plagiocephaly*	0 (0%)	33 (46.48%)
Positive Bielschowsky test*	7 (100%)	0 (0%)

SCM: sternocleidomastoid muscle
*$p < 0.05$

torticollis was not visible, and both ears could be seen in the front view (Fig. 4). On the contrary, unilateral shortening of the SCM muscle caused ipsilateral laterocollis and contralateral torticollis in the CMT cases, which makes the chin rotate on the horizontal plane. The degree of torticollis was 57.80±16.33. Consequently, specific findings were observed in which the contralateral ear was not seen due to rotation (Fig. 3). The unilateral SOP children showed laterocollis of 36.43± 9.00 degrees (from 30 to 55 degrees) at the age of 3 to 4 months. All seven children exhibited aggravation when seeing a picture book or toys during physical examination; additionally, no improvement of laterocollis was observed after 6 months of age. However, all 71 CMT children did not show aggravation with looking at a picture book or toys. All the SOP children had no plagiocephaly. Thirty-three of the 71 CMT children (46.48%) showed plagiocephaly on the first visit ($p < 0.05$). The APHN children with unilateral SOP showed normal development in all development domains. These children showed a positive Bielschowsky head tilt test with the above mentioned features. Their diagnoses were confirmed with pediatric ophthalmology consultation. All of these children had no trauma history, so their APHN seemed to be congenital. On brain MRI, no SOP defects were observed. Three of them underwent surgery for unilateral SOP; since then, improvement of their head and neck posture have been shown. The other 4 were planning to undergo surgery.

DISCUSSION

This study reports the clinical features of children with APHN due to unilateral SOP, and these children were seen at Ajou University Hospital, the Center for Torticollis and the Department of Physical Medicine and Rehabilitation. Shin and colleagues reported that the first ophthalmology visit by 55 children with APHN was done at the age of 4 (0.7-9 years).[6] The current study shows that the diagnosis of unilateral SOP was done at 19 months of age, which was 29 months earlier than that of the previous study. This indicates that an early diagnosis is possible when non-ophthalmologists have the information about the clinical features of unilateral SOP, and early consultation is done.

An accurate differential diagnosis is essential for the proper management of infants with APHN. Ballock and Song reported that 81.8% of APHN was due to CMT and 18.4% was non-CMT among 288 children who came to the department of pediatric orthopedics because of APHN. Among the non-CMT group, 23% was ocular APHN.[3] Thus, an ocular cause has to be kept in mind when the differential diagnosis of APHN does not suggest CMT.[2,3,6,14-17] Paralytic strabismus decreased eyesight from refractory error, and nystagmus can cause ocular APHN.[2,3,6,14-17] Hypertropia with unilateral SOP is known to be the most common problem causing ocular APHN.[4,5] In a Korean study, 40% of the cases of ocular APHN were related to strabismus, and 73% of the cases of strabismus were related to SOP.[6] The superior oblique muscle is a kind of external ocular muscle that is innervated from the trochlear nerve. The muscle originates from the annulus of Zinn and inserts into the upper portion of the eyeball via the trochlea. The main function of the superior oblique muscle is intorsion of the eyeball with simultaneous depression and adduction. The superior rectus muscle also functions as an intortor, yet it causes elevation and abduction synchronously. Depression of a paralyzed eye in a child with SOP triggers intorsion, andactivation of the superior oblique and rectus muscles as the intortors contract, but the superior oblique muscle cannot act due to palsy. Another intortor (the superior rectus muscle) then causes intorsion, with synchronous elevation and abduction of the eyeball, which elicits hypertropia. This phenomenon is called the positive Bielschowsky head tilt test[4,13,18] and is indicative of unilateral SOP (Fig. 2). In this study, the SOP children did not show shortening of the SCM muscle nor a difference in muscle thickness on ultrasound examination, laterocollis of more than 30 degrees, aggravation when looking at a picture book or toys, no improvement according to growth and development, and positive findings on the Bielschowsky head tilt test. The confirmative diagnosis of SOP can be performed by ophthalmologists; the details and specialized knowledge of SOP is out of the scope of this study. However, we reported the clinical features that are suspicious for APHN by non-ophthalmologists. This may allow for the early diagnosis of ocular APHN together with ophthalmic consultation. In APHN children with unilateral SOP, laterocollis against the paralyzed eye was shown, and this posture can minimize the action of the superior oblique muscle as an adductor. Previous studies have named ocular APHN with unilateral SOP as ocular torticollis, but clinically, laterocollis is more dominant than torticollis. Thus, we use the terminology of ocular APHN or ocular laterocollis.

The causes of SOP have been classified as congenital and acquired. Congenital causes were reported in 40-75% of SOP cases, and these are congenital laxity of the superior oblique tendon, congenital defect of the superior oblique muscle, and trochlear nerve paralysis.[4,8,19-22] Infant SOP without a history of trauma, such as was seen in this study, was thought to be congenital problem. Holmes and colleagues reported that the incidence of ocular movement related to III, IV and VI nerve palsy was 7.6 of 100,000 children below 18 years old, and IV nerve palsy (trochlear nerve palsy) was the most common problem at 36% of cases.[23] Stretching exercise to the SCM muscle is not helpful for APHN children with unilateral SOP. Caution is needed when trying to correct laterocollis using a neck splint because this aggravates diplopia.[5] For the treatment of SOP, myectomy, recession, or anterior transposition of the overactive inferior oblique muscle is performed. In some children, strengthening of the superior oblique muscle is performed.[19,24] Disappearance of the APHN after surgery was reported in 70% of the cases. Lau et al. reported that children with sustained APHN after surgery underwent an operation at 95.9 months as compared to 79.9 months for 32 improved children with congenital SOP. That was a significant delay of operation time. They suggested that the remaining cases of APHN after surgery were due to contracture of the SCM muscle, so they emphasized an early diagnosis and early operation for congenital SOP.[25,26]

Madigan and Zein reported that there was a different mechanism of craniofacial asymmetry in unilateral SOP, due to unilateral facial growth retardation; in patients with CMT, this was due to anterior transition of pressure from the unilateral occipital bone from the limitation of cervical rotation.[5] Delayed treatment for SOP may bring about secondary skeletal abnormalities, such as facial asymmetry, plagiocephaly, and scoliosis. So, making the early differential diagnosis of APHN due to ocular causes is very important.[7-9,26-28] Han and Cho reported that when the head tilt was more than 10 degrees, facial asymmetry was shown in more than 80% of SOP children and the children of SOP who were below 1 year old exhibited facial asymmetry at about 2 years of age.[8] In this study, the age at the first visit was 15.57±12.55 months, and the age at the time of SOP diagnosis was 19.07±11.29 months. That was a relatively early diagnosis. At time of the diagnosis, there was no plagiocephaly.

Another frequent cause of ocular APHN besides unilateral SOP is nystagmus. It is known to make up 20% of ocular APHN. Refractory error may also cause APHN, and the most common cause is astigmatism.[21] In this study, we found a case of Moebius syndrome that showed laterocollis accompanied with unilateral hearing loss, unilateral facial nerve palsy, and lateral rectus paralysis.[29] One case had unilateral ptosis related to ocular APHN with accompanying amyblyopia.[30]

An accurate differential diagnosis for APHN is essential for proper treatment. Making the differential diagnosis between torticollis and laterocollis is important. It has to be kept in mind that laterocollis has no indication for stretching or surgery to the SCM. For a patient with ocular laterocollis, caution is needed not to correct the laterocollis using a neck splint because it may deteriorate the diplopia. Unilateral SOP, which is a major cause of ocular APHN in infants, shows contralateral laterocollis as the major symptom, and this may be worsened when looking at an object. In patients with ocular laterocollis due to unilateral SOP, the clinical features like non-remarkable craniofacial asymmetry, no limitation of the cervical range of motion, and no torticollis help to differentiate this from CMT. Ocular laterocollis due to unilateral SOP may be diagnosed by the Bielschowsky head tilt test and ophthalomology consultation.

CONCLUSION

This study provides the clinical features that non-ophthalmologists need to suspect unilateral SOP by analyzing the clinical findings of APHN as compared to those of unilateral SOP. An early diagnosis of unilateral SOP combined with consultation with the ophthalmology department is recommended for proper treatment.

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