Cervical Spine Dysmorphism in Congenital Muscular Torticollis

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Background: Congenital muscular torticollis is a common childhood musculoskeletal anomaly that might result in permanent craniofacial deformity, facial asymmetry, and changes in the cervical vertebrae, if not treated during early childhood. Although there have been many studies on cervical vertebral changes, their onset in children has not been previously studied.

Methods: Fifteen patients (aged <8 years) with a confirmed diagnosed of torticollis were included. Three-dimensional computed tomography scans were obtained, and segmentation of the cervical vertebrae was done. Division of the atlas and axis across the midsagittal plane was done to compare the anatomical changes. The volumes of each halves of the atlas and axis were measured.

Results: An apparent change was observed in the axis of the vertebral column when compared with that of the skull. There were progressive anatomical changes affecting the upper cervical vertebrae, which started to develop around the age of 8 months and became more evident in older children. The axis vertebra was the first to be affected. Rotational and bending deformities were the most likely changes to occur. Pearson correlation analysis showed a statistically significant trend in the volume and height changes for both halves of the atlas and axis ($P < 0.001$ and $P < 0.001$).

Conclusions: Children with untreated congenital muscular torticollis show progressive anatomical changes of the cervical vertebrae which started at the age of 8 months. The severity of the deformity increased with the advance of age as well as with the severity of sternocleidomastoid tightness, which might result in permanent deformities.

Key Words: Cervical spine, congenital muscular torticollis, segmentation, vertebrae

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Despite torticollis is a symptom of various underlying diseases. Congenital muscular torticollis (CMT) represents the most common cause of neck tilting that is detected at, or immediately after, birth. Congenital muscular torticollis occurs due to a pathological shortening of the sternocleidomastoid muscle (SCM).[1] It is the third most common childhood congenital musculoskeletal anomaly with a reported incidence ranging from 0.3% to 3.92%,[2,3] and a male-to-female ratio of 3:2.[4] According to the presence of a mass or tightness, muscular torticollis is classified into 3 main groups.[5]

Long-standing CMT can lead to progressive craniofacial deformities and asymmetry.[6] Moreover, positional plagiocephaly is frequently associated with CMT.[7,8] Progressive pain and limitation of neck movement were also reported in many cases.[9]

The integrity of the cervical spine is essential for craniofacial stability and movement.[10] Many authors had reported on the anatomical changes in the cervical spine of patients with CMT[11,12]; however, there have been no reports about the detailed anatomical changes and on the onset of these anatomical changes. Thus, this study was directed toward exploring the presence of such changes in children under the age of 8 years, and toward detecting the onset of such changes.

PATIENTS AND METHODS

Fifteen patients with a diagnosis of CMT showing a clinically apparent tightness of the SCM, which had been ultrasonographically confirmed, were included in this study. Their ages ranged from 4 months to 8 years, and the male-to-female ratio was 9:6. Demographic distribution is described in Table 1. None of the patients had received physiotherapy before the time of presentation, which allows us to assess the vertebral changes in the absence of any treatment modalities that might affect the deformities itself. Other causes of torticollis had been excluded to confirm the diagnosis of CMT.

Because of previously reported cervical spine changes in older patients,[11,12] three-dimensional (3D) computed tomography (CT) scans of the skull and cervical spine were obtained routinely for all patients at the time of presentation for research purpose. The CT scans were obtained by using a multidetector-row helical CT scanner (Somatom Sensation 64, Facial 1/1 mm pac, matrix size 512 × 512; Siemens Medical Solutions, Erlangen, Germany). Analyze Direct AVW (Mayo Clinic, Rochester, MN) software was used for vertebral segmentation, which allows creating a separate 3D reconstructed module for each vertebra, hence allowing to translate and rotate each vertebra freely.

The midsagittal plane was established across each vertebra to assess any asymmetry or rotational deformity. Furthermore, the atlas and axis vertebrae were divided across this plane, and the volume for each half was obtained to confirm the volumetric difference. Because of the anatomical differences between the atlas and the axis, the midsagittal plane had to pass through the anterior and posterior tubercle of the atlas, and through the center of the odontoid process, the body, and the spinous process. For the remaining vertebrae, the midsagittal plane passed through the center of the body to the spinous process. The vertical height of each vertebra was compared across both sides of the vertebral bodies to assess the bending deformities.

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TABLE 1. Demographic Distribution of the Patients

Age	Sex	Affected Side
5 mo	F	Lt
5 mo	M	Rt
5 mo	F	Lt
8 mo	F	Rt
8 mo	F	Rt
8 mo	M	Rt
12 mo	F	Rt
13 mo	M	Rt
21 mo	F	Rt
24 mo	M	Rt
28 mo	F	Lt
3 y	M	Rt
4 y	M	Rt
5 y	M	Rt
8 y	M	Rt

Both vertebrae were initially segmented, divided, and measured at the same threshold to avoid any bias that might result from threshold changes. Moreover, the positions of both vertebrae were adjusted to be vertically and horizontally aligned before division. Volume measurement was done by using the volume render measurement tool in Analyze Direct AVW software.

Paired *t* tests were used to compare both the volume and height changes across both halves of the atlas and axis. Pearson correlation analysis was used to detect the relation between the volume and height changes. All data analyses were performed with standard software (SPSS for 7 Windows v20.0; IBM Corp., Armonk, NY). *P* values of <0.05 were considered to indicate a statistical significance.

RESULTS

The posterior view of the skull and cervical vertebra showed a change in the axis of the cervical spine in comparison with that of the skull, whereas the atlas vertebra seemed to follow the axis of the skull. The lateral view showed apparent changes in the vertical height, especially for the atlas and axis, across both sides of the spine (Fig. 1). These discrepancies were confirmed by the volume differences across each halves of the atlas and axis vertebrae.

Infants aged <1 year showed no gross changes besides minimal rotational changes of the axis across its laminae, and minor changes of the superior articular facet of the atlas on the affected side (Figs. 2 and 3). On the contrary, older patients showed progressive changes in the rest of the vertebrae. The third cervical vertebra was the most frequently affected region after the axis and atlas. In all patients, there were mild deformities of the vertebrae from C4 to C6, with

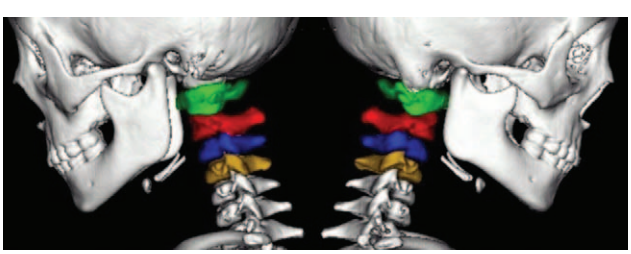

FIGURE 1. Lateral view of segmentation of the upper 4 cervical vertebrae in a 5-year-old patient, showing a difference in the vertical height of the upper 4 cervical vertebrae.

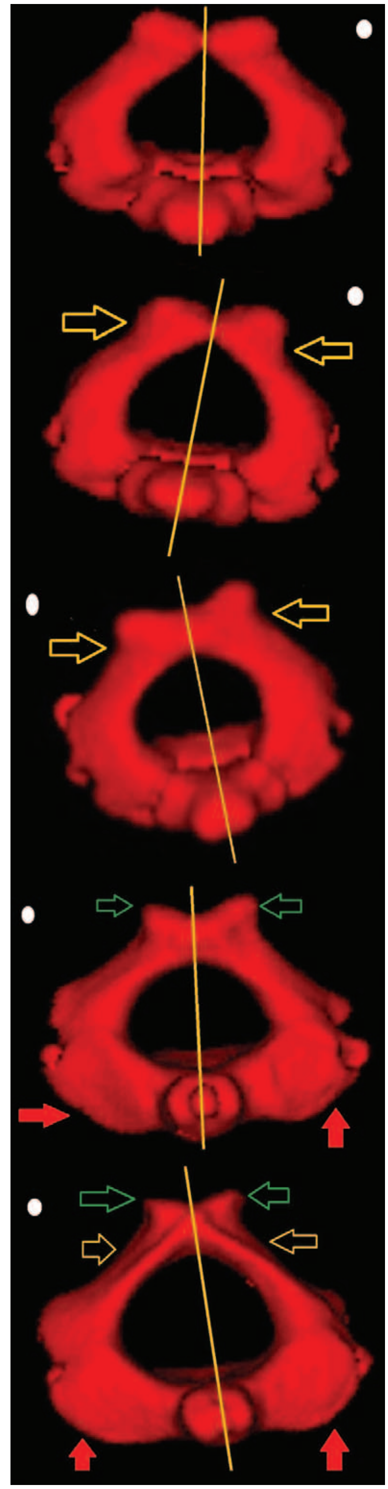

FIGURE 2. Top view of the axis vertebra of 5 different patients aged 5 months, 8 months, 2 years, 4 years, and 8 years, sequentially (right-sided congenital muscular torticollis [CMT] in the upper 2 patients and left-sided CMT in the lower three patients) with the midsagittal line, showing symmetry of either sides of each vertebra. In the patient aged 5 months, the sides are symmetrical. On the contrary, there are progressive changes in the laminae (yellow arrows), the transverse process (green arrow), and the shape and slope of the superior articular facet (red arrow) in older children.

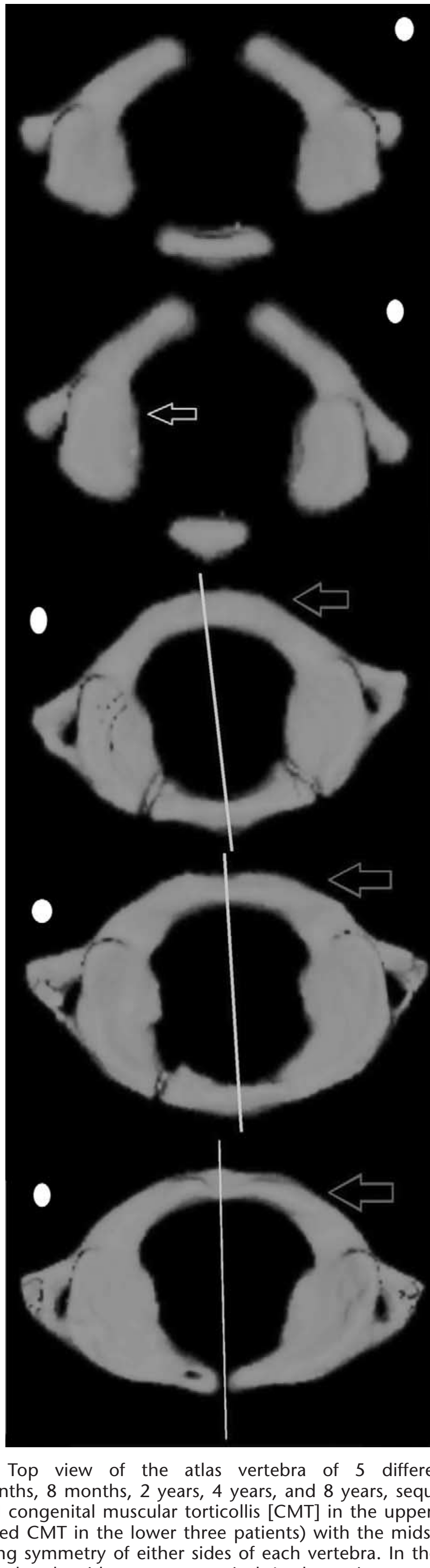

FIGURE 3. Top view of the atlas vertebra of 5 different patients aged 5 months, 8 months, 2 years, 4 years, and 8 years, sequentially (right-sided congenital muscular torticollis [CMT] in the upper 2 patients and left-sided CMT in the lower three patients) with the midsagittal line, showing symmetry of either sides of each vertebra. In the patient aged 5 months, the sides are symmetrical. In the patient aged 8 months, there are minor changes in the superior articular facet on the affected side (top arrow). Older children show mild progressive changes in the laminae (bottom 3 arrow), the transverse process, and the shape of the superior articular facet.

alteration of the shape and direction of their spinous processes. These changes were more clinically apparent in older children. The seventh cervical vertebra was almost normal without any apparent-deformities.

The axis vertebra was the first to be affected. The changes started at the age of 8 months. Almost all of vertebral components exhibited changes. Tilting of the odontoid process was observed, as shown in Figure 4. Moreover, there were significant changes in the superior articular facet in terms of shape, slope, and size. It became larger and steeper, as shown in Figure 4. Gradual alterations of the direction and curvature of the lamina were also observed with increasing age of the patients. The convexity of the nonaffected side and the concavity of the affected side became clearer from the top view, as shown in Figure 2.

The changes of the atlas vertebrae were milder than those of the axis. There were minimal progressive rotational deformities across the midsagittal plane, and its anterior arch showed apparent changes, as shown in Figure 4 Furthermore, there were changes in the direction of the atlas transverse processes that resembled a bending deformity to match the tilted skull base, as shown in Figure 5.

None of the volume changes or height differences across both halves of the atlas or axis showed any statistically significance. For the volume changes, the P value was 0.323 and 0.082 for the atlas and axis, respectively. For the height changes, the P value was 0.154 and 0.101 for the atlas and axis, respectively. However, the Pearson correlation analysis showed a statistically significant trend in the volume and height changes.

DISCUSSION

Congenital muscular torticollis is known to be a curable childhood condition. It can be cured completely without any functional or cosmetic residuals with early diagnosis and appropriate management during the first year of life. Although many hypotheses have been proposed to explain the shortening of the SCM, the actual cause is still unclear.[12] Endomysial fibrosis and muscle fiber atrophy are the constant pathological findings.[13] Macdonald classified muscular torticollis into 2 main groups according to the presence of a mass or tightness; type I includes patients with palpable pseudotumor or a sternocleidomastoid tumor, whereas type II includes patients with tight SCM without any palpable masses.[14,5] A third group has been recently distinguished: postural torticollis without any masses or tightness.[3]

The main lines of treatment include physiotherapy, active home exercises, gentle manual stretching, and vigorous manual myotomy, whereas surgical intervention is always reserved for resistant cases when physiotherapy failed to correct the head tilting.[5] Generally, the success of physiotherapy depends on the patient's age. Earlier ultrasonography studies demonstrated that the degree of fibrosis is proportionate to the age of patients when untreated: the older the child, the more fibrosis was likely to present.[15]

In our previous study on neglected long-standing untreated adult cases of CMT (the article had been accepted for publication by the *Journal of Craniofacial Surgery* in the upcoming issues under the number of SCS-16-0457R2), we reported apparently permanent changes affecting the cervical spine in the form of bending and rotational deformities that were maximum at the level of the atlas and axis. These changes resulted in residual head tilting despite adequate treatment in those patients. Most of these changes were also detected in the children in this study. This result also corroborates that of Chen and Ko, who reported that there were morphological changes that include both the odontoid process and the superior articular facet of the axis in their study on 18 children >6 years.[12]

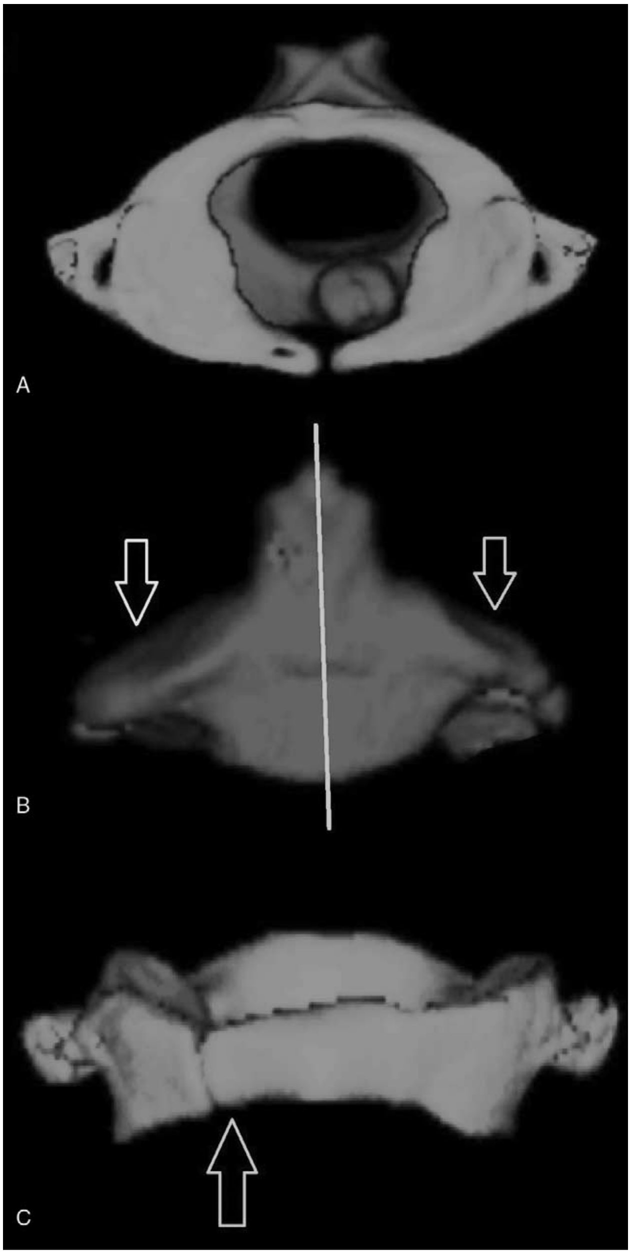

FIGURE 4. (A) Top view of both the atlas and axis vertebrae of an 8-year-old boy, showing the relation between the atlas and axis with a tilted odontoid process toward the unaffected side. (B) Anterior view of the axis vertebra of a 5-year-old patient, showing changes in the slope of the superior articular facet (arrows). The difference of the vertical height on both sides is also evident. (C) Anterior view of atlas vertebra of a 5-year-old patient, showing apparent change in the anterior arch (arrow).

The atlas vertebra, unlike other vertebrae, does not have a body or spinous process. It is formed by 2 lateral masses that are connected by anterior and posterior arches. It articulates with the occipital condyles through the atlanto-occipital joint.[16,17] The limited mobility across this joint might explain the bending deformity that made the atlas match the tilted skull base. Moreover, the passive movement of the atlas vertebra through the muscles that act on the head might explain the rotational deformity noticed in this study.[18] This result is similar to the result of Ozer et al, who reported

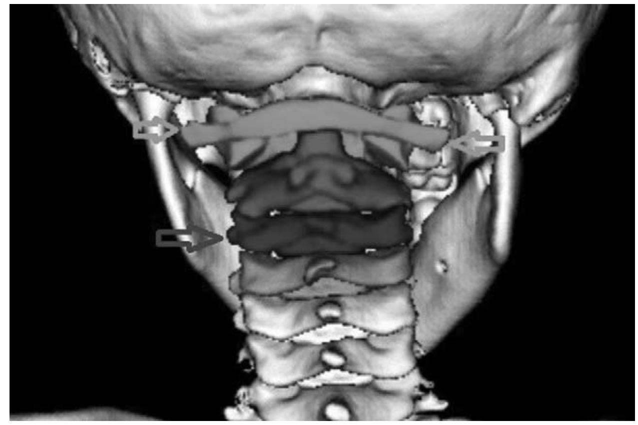

FIGURE 5. Posterior view of the skull and cervical spine of a 5-year-old patient with segmentation of the upper 4 cervical vertebrae, showing changes in the level and direction of the transverse process of the atlas vertebrae (top arrows), as well as show changes in the shape and height of the third cervical vertebra (bottom arrow).

the presence of rotational deformities of the atlas vertebra in their 3D-CT investigation of craniofacial and cervical spine anomalies in 6 patients with CMT ranging from 2 to 26 years old.[11]

Concerning the axis, the unique superior articular facet of the axis permits rotatory movement of the head around the median atlanto-axial joint, and also allows weight transmission through the lateral atlanto-axial joint.[19] In this study, the observed changes in the slope and shape of the facet might be due to the unequal weight transduction through the lateral atlanto-axial joints. These findings also matched the results of Oh and Nowacek, who reported the same radiographic abnormalities in the axis vertebra and the lateral atlanto-axis joint.[20] Furthermore, the lack of typical restraints to the rotatory movement of the atlas makes it susceptible to rotation deformities.[21]

Concerning other cervical vertebrae from C3, normally, the superior articular facets of the third vertebra and the inferior articular facets of the axis that form the C2–3 joint also have different architectures and different mechanisms of action during axial rotation.[22] This might lead to changes in the direction of weight transduction across the C2–3 junction, thus decreasing the risk of exposure to abnormal weight forces in cases of head or neck tilting. From the third to the seventh vertebrae, the saddle-shaped intervertebral joint in association with the free rocking movement of the vertebrae across 2 different planes[10] might also play a role in the weight distribution across the vertebrae, thus decreasing the susceptibility to deformities. This compensatory mechanism might be the cause of the radiological appearance of secondary scoliosis, as the muscle release leads to its correction, as indicated by Chen and Ko.[23]

Although our current study showed that there is no statistically significant difference in the volume and height themselves across each halves of the atlas and axis, this result might be attributed to the small sample size. However, for the trend of the changes, Pearson correlation analysis of both variants showed statistically significant trends between volume and height for the atlas and axis.

Although there are multiple reports with a similar anatomical finding,[11,12] the onset of these morphological changes had not been clarified yet. The results of this study suggest that the onset of deformable changes in the cervical spine might start at the age of 8 months. As a definite finding, we can suggest that the axis vertebra is the first to be affected. In addition, the patient age plays an important role in the severity. As the patient ages without treatment, the severity of the deformities, as well as the volume differences, might gradually increase.

In the present study, we could not reveal whether these changes are reversible or not, after adequate management, owing to the lack of long-term follow-up of the study group. However, Chen and Ko reported that even the tilting of the odontoid process was improved; however, the deformity of the superior articular facet of the axis persisted after the surgical treatment of children >6 years.[12] Furthermore, Canale et al reported residual facial asymmetry in patients >4 years.[1] This previous suggestion was further enforced by the results of our previous study on neglected long-standing untreated adult cases of CMT. These findings suggest that already started or long-standing deformities would be impossible to return to the normal symmetrical anatomy. Therefore, more investigations on the reversibility of these changes are warranted. Until that time, we suggest, on the basis of our results as well as the previous reports, that early intervention (especially in physiotherapy-resistant cases and type I and II CMT) should be considered for resistant patients to physiotherapy to avoid the development of potentially permanent cervical deformities.

CONCLUSIONS

Progressive vertebral changes along the cervical spine were observed in children with untreated CMT, which started around the age of 8 months. These changes were more clinically significant at the level of the atlas and axis vertebrae, and decreased gradually toward the seventh vertebra. The severity of the changes increased gradually with aging as well as the severity of SCM tightness.

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