Congenital Muscular Torticollis

--- Analysis of 17 Cases ---

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**ABSTRACT**

Our clinical observation are based on a series of 17 cases with muscular torticollis operated on since 1955 at the Department of Orthopaedic Surgery, Yonsei University College of Medicine, Seoul, Korea.

1. In a series of 4 infants and 13 older children with congenital torticollis, a sternocleidomastoid tumor was detected clinically in 4 cases less than 5 months of age and in 5 older patients.
2. The age at the onset of symptoms ranged from one week to 5 years, with an average duration of symptoms of nine and a half years.
3. Results of surgical correction in this series were uniformly good.
4. Secondary changes in these cases of congenital torticollis were improved only when the surgical treatment was given while there was still growth potential.
5. In our series of cases, there was no congenital deformity other than torticollis.
6. On microscopic examination of the pathologic specimen, there was no evidence of hemorrhage or pyogenic infection.

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**INTRODUCTION**

Torticollis may be the result of an irregular contraction of the muscle.

The term torticollis is derived from the Latin words tortus, meaning "twisted", and collum, meaning "neck". Synonyms include wry neck, stiff neck, caput obstipum, crooked neck, and twisted neck.

Most congenital types include those resulting from muscular, osseous, or neurogenic factors, while the commoner acquired types are those resulting from infectious, neoplastic, traumatic, hysterical, or paralytic factors.

Congenital muscular torticollis is a distinct entity, the primary pathologic characteristics of which are limited to the sternocleidomastoid muscle.

Associated deformities of the face, head, ear, and cervical spine are secondary in character, resulting from an abnormal position of the head both prior and subsequent to birth. Muscular torticollis may be present at birth or many manifest itself first on about the tenth to the fourteenth day, at which time the appearance of a hard, fusiform swelling is noticed in the sternocleidomastoid muscle.

The swelling or "tumor", as it is commonly called, usually increases in size for two to four weeks until it reaches the size of pigeon egg. It then begins to regress and may disappear completely in from five to eight months.

The muscle at this time feels short and fibrous, and is noncontractile. The torticollis become increasingly severe, and the head tilted toward the affected side and the face toward the opposite side.
This position may disappear only to reappear later at a period of rapid growth. As a result of the head being held more or less constantly to one side, and the imbalance of forces and muscle pull exerted on the head, the skull becomes foreshortened in the oblique fronto-occipital diameter on the side opposite the neck deformity.

The level of the eye changes, the mastoid process becomes elevated on the affected side.

A cervico-dorsal scoliosis may develop with concavity toward the involved side.

**Theories of Etiology**

Various theories of etiology have been reported and discussed.

Some theories of etiology of muscular torticollis have been discussed in an excellent manner in the writing of Fitz Simmon (1933) and Chandler and Altenberg (1944). However, no single theory seems adequate to explain the findings in congenital muscular torticollis.

The intrauterine theory regards the deformity of muscular torticollis as the result of abnormal pressure, position, or trauma to the head, neck, and particularly the sternocleidomastoid muscle during intrauterine life.

The hereditary theory has been adopted by many writers. But only scattered reports of a familial pattern of this deformity have been found.

The neurogenic theory has been discarded for the most part and has not been associated with muscular torticollis in the literature of recent years, because there is little evidence to support a denervation of the sternocleidomastoid muscle.

The theory of infection is based mainly on the pathologic picture as seen on section, because the microscopic picture is not wholly unlike that of infectious myositis.

Organisms have never been cultured and animal inoculation has failed.

The theory of birth trauma applies to a large number of cases of muscular torticollis, as the trauma causes almost complete derangement of the structure of the muscle.

This theory has evoked the most discussion with reports ranging from absence of birth trauma in an entire series to a report of 32 hematomas in 35 cases of congenital torticollis.

Some authors report no hemosiderin in the section of invaded muscle, and others found it in as many as 25 percent of cases.

Chandler (1948) discounts the hematoma theory as the source of the tumor as he feels the microscopic studies warrant the diagnosis of fibroma.

The theory of venous occlusion during delivery as the etiology of muscular torticollis was first advanced by Middleton (1930). But this theory does not explain the more common finding of a tumor at two to four weeks of age with no abnormality at birth. Also, venous occlusion has not been demonstrated in dissection or microscopic examination.

The theory of arterial occlusion or ischemia during delivery is hardly acceptable as the primary etiologic agent of muscular torticollis, because the fibroed muscle of torticollis does not resemble ischemic muscle.

**Clinical Observation and Analysis of Cases**

Our clinical observations are based on a consecutive series of 17 patients with muscular torticollis operated on since 1955 at the Department of Orthopaedic Surgery, Yonsei University College of Medicine, Seoul, Korea.

A detailed study of seventeen cases occurring in infancy and childhood disclosed the following information.

The age of symptom onset of torticollis ranged from 1 week to 5 years, with an average duration of symptoms of nine and a half years. This reflects a relatively late discovery and treatment in our series.

The age distribution at the time of operation ranged from one month to 27 years of age, with
Table 1. Age at Symptom Onset in 17 Cases

<table>
<thead>
<tr>
<th>Onset</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 week</td>
<td>9</td>
</tr>
<tr>
<td>2 weeks</td>
<td>2</td>
</tr>
<tr>
<td>4 weeks</td>
<td>2</td>
</tr>
<tr>
<td>5 weeks</td>
<td>2</td>
</tr>
<tr>
<td>14 weeks</td>
<td>1</td>
</tr>
<tr>
<td>5 years</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>17</strong></td>
</tr>
</tbody>
</table>

Table 2. Age Distribution in 17 Cases at the Time of Operation.

<table>
<thead>
<tr>
<th>Age in years</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>under 1</td>
<td>4</td>
</tr>
<tr>
<td>2—4</td>
<td>2</td>
</tr>
<tr>
<td>over 5</td>
<td>11</td>
</tr>
</tbody>
</table>

Table 3. Sex, Side involved and Birth presentation in 17 Cases

<table>
<thead>
<tr>
<th>Sex</th>
<th>Male (41.4%)</th>
<th>Female (58.8%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Side</td>
<td>Right (70%)</td>
<td>Left (30%)</td>
</tr>
<tr>
<td>Presentation</td>
<td>Vertex (70%)</td>
<td>Breech (30%)</td>
</tr>
</tbody>
</table>

Table 4. Secondary Changes in the Series of 17 Cases

<table>
<thead>
<tr>
<th>Secondary changes</th>
<th>Right</th>
<th>Left</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asymmetry of face and skull</td>
<td>3</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Cervico-Dorsal scoliosis</td>
<td>3</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Elevation of the shoulder</td>
<td>4</td>
<td>1</td>
<td>5</td>
</tr>
</tbody>
</table>

an average age of nine and a half years.

Four patients have been operated on under one year of age.

Of this series, seven were boys (41.2%) and ten girls (58.8%).

The right side was involved in 12 (70%), the left in 5 (30%). This figure did not correspond to those of other writer's.

In no case was bilateral involvement of the sternocleidomastoid muscle found.

Birth histories were important, but they were sketchy, especially in older patients. The birth histories in those cases in which operation was performed under one year of age were more accurate.

Presentation and delivery were cephalic in 12 cases (70%).

Of these, 2 cases had a history of difficult labor.

Breech presentation occurred in five cases (30%).

In none of these individuals was there any evidence of trauma in the neonatal period.

A familial history of torticollis, emphasized by some writers, did not appear to be important in our series of cases.

A tumor within the sternocleidomastoid muscle was detected by a physician in four cases less than one year of age.

In older patients, five had a history of sternocleidomastoid tumor in infancy.

Secondary changes of muscular torticollis were present in patients over seven years of age as outlined in table-4.

Asymmetry of the face and skull were found in four cases. Cervico-dorsal scoliosis and elevation of the shoulder and clavicle on the affected side were found in five cases with preponderance to the right side (3:2, 4:1).

No associated pathologic condition other than the clinical findings of torticollis was seen.

Operation

There are two generally accepted methods of treatment for torticollis, exercise and surgery.

In a child less than one year of age with a
tight sternocleidomastoid and head tilt with or without a fusiform tumor, a trial of conservative measures is recommended by some authors.

The mother is instructed in manipulations of the child’s head to stretch the sternocleidomastoid muscle.

Proposed indications for operation have been persistence of tumor at three to five months of age, increasing deformities with or without a mass, and persistence of deformity after one year of age.

Torticollis is a serious matter in older cases and radical measures should be resorted to early in the disease.

In our series of cases, all patients were treated surgically. A transverse incision was made in the lower skin crease just above the clavicle. The platysma muscle was split by blunt dissection. The tumor or tight muscle was then separated from adjacent structures on all surfaces by blunt dissection with a small curved forceps. The sternal and clavicular portions were isolated, a small forceps was passed beneath the muscle which was divided by multiple transverse incisions until freed from clavicle and sternum. The tumor or tight fibrosed muscle was then elevated from the wound. Spinal accessory nerve was isolated and protected well. All bleeding points were ligated.

Tumor resection was carried out in four cases in this series. Myectomy was carried out in seven cases and the rest of the patients received tenotomy or myotomy after which the tight structures were allowed to retract away from the original site of attachment. The wound was closed by deep sutures and the skin by interrupted silk.

To obliterate any dead space, a pressure dressing was applied.

Postoperative Care

The stitches were removed on the 7th or 10th day after surgery. After surgery in four cases under one year of age, the infant’s head was maintained in an overcorrected position with a sandbag for about one week.

Halter traction was carried out in five cases older than two years of age.

A Calot Jacket cast maintaining overcorrection was worn for three to four weeks in eight cases of older patients. This was followed by active or passive gentle stretching exercise of the muscle, performed by the patient’s parents three times daily for a few weeks.

Pathology

Excised tumors in four cases of this series consist of fusiform masses, including part or all the muscle belly of the sternocleidomastoid, usually in its middle third. It was firm in consistency and separated easily from adjacent structures. Cross section revealed a whitish, fibrocartilaginous, and glistening surface.

In none of these specimens was there anything to suggest hemorrhage or pyogenic infection.

The pathologic process appears to be that of the replacement of muscle tissue by fibroblasts. In older patients, the tumor had disappeared already, leaving a firm, elastic, and tendon-like band, with varying degrees of degenerative change of striated muscle.

RESULT

In this series of cases, 16 patients have recovered from the wry neck deformity following surgery. Only one showed residual limitation of rotation of the neck and it was corrected by reoperation performed 10 months later.

Secondary changes in muscular torticollis including asymmetry of the face and skull have improved in some cases less than 12 years of age, but in the older patients recovery was
Table 6. Result of Treatment in 17 Cases

<table>
<thead>
<tr>
<th>Deformity</th>
<th>Degree of recovery</th>
<th>No. of case</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tilting of head</td>
<td>Complete</td>
<td>16</td>
</tr>
<tr>
<td></td>
<td>incomplete</td>
<td>1</td>
</tr>
<tr>
<td>Asymmetry of face</td>
<td>Improved</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>not improved</td>
<td>2</td>
</tr>
<tr>
<td>Cervico-dorsal</td>
<td>Improved</td>
<td>2</td>
</tr>
<tr>
<td>scoliosis</td>
<td>Sl. Improved</td>
<td>3</td>
</tr>
<tr>
<td>Elevation of</td>
<td>Improved</td>
<td>4</td>
</tr>
<tr>
<td>shoulder</td>
<td>Sl. Improved</td>
<td>1</td>
</tr>
</tbody>
</table>

incomplete or did not occur.

DISCUSSION

Various theories of etiology of sternocleido-mastoid tumor have been reported but no single theory seems adequate to explain the findings in congenital torticollis.

Chandler (1948) stated that the tumor seen in this condition in early infancy is not a hematoma at birth but a fusiform fibroma found within the substance of the sternocleidomastoid muscle, and intrauterine position, trauma at birth and pre-existing changes of the involved muscle are etiologic factors in muscular torticollis.

A Dutch surgeon, Isacius Minnius, performed tenotomy for torticollis in 1685. He is believed to be the surgeon to obtain operative correction of this condition.

Considerable variation in approach to the treatment of congenital torticollis is noted in the literature, including myorrhexis, myotomy or tenotomy of two heads of the sternocleidomastoid.

External pressure on the sternocleidomastoid to split and tear the muscle and fibrous tissue was called myorrhexis.

Later, resection of the tumor with additional fibrous muscle was advocated and standardized by Chandler and Altenberg (1944) particularly for patients with pronounced deformities.

First, one must rule out cervical spine abnormalities, Klippel-Feil syndrome, spasmodic torticollis, auditory disturbances and ocular abnormalities.

Armstrong and Pickrell (1965) stated that in an older child with wry neck deformity, operation affords the only opportunity for improved range of motion and normal position of the head. Our experience for older cases were more or less comparable to those of other writers.

Many writers stated that secondary changes of muscular torticollis will ultimately disappear if operative correction is carried out while there is still growth potential, and therefore operation should be performed as early as possible.

REFERENCES


