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Torticollis in infants and children: a report of three cases

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Three cases of torticollis are recorded, one of a child with congenital muscular torticollis and two of infants with acquired torticollis caused by neurogenic tumours. All were treated by chiropractors before the correct diagnosis was made. The differential diagnosis of torticollis in infants and children is important in clinical practice.
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KEY WORDS: Torticollis, child, infant, differential diagnosis, chiropractic, manipulation.

Trois cas de torticolis chez des enfants ont été rapportés. Un enfant souffrait d' un torticolis congénital et les deux autres, de tumeurs neurogènes. Ils ont tous été traités par des chiropracticiens avant l'établissement du bon diagnostic. Le diagnostic différentiel est important chez les enfants, pour l'exercice clinique.
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MOTS CLÈ: Torticolis, enfant, diagnostic differentiel, chiropraxie, manipulation.

Introduction

An infant or child presenting with torticollis, or wryneck, should always raise suspicion of underlying pathology. Though most frequently a benign condition, torticollis can be the first indication of a more serious disorder.[1,2] Its persistence or its association with other objective findings should prompt an aggressive search for the cause.

The etiology of torticollis is typically divided into congenital and acquired conditions.[3] An indepth discussion of causes is beyond the scope of this paper, however both McDaniel et al.[4] and Kiwak[5] have published excellent reviews.

It is important to recognize the various causes of torticollis and to make a definitive diagnosis as soon as possible. Failure to recognize serious underlying pathology or structural deformity may delay proper treatment.[6] The following three cases are examples of congenital and acquired torticollis that presented to chiropractors' offices.

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Case presentations

Case 1

An eight-year-old girl was seen in the Orthopedic Outpatient Clinic at the University Hospital for her torticollis. She was the product of a normal pregnancy and breech delivery, and at birth it was noticed that she had a torticollis. She had received physical therapy for this in the past, but the mother felt that it was never completely corrected. There was no family history of spinal deformity. She had been otherwise a fit and healthy child.

On examination the torticollis was quite obvious, with contracture of the left sternocleidomastoidius (SCM) muscle. There was subtle facial asymmetry. Movements of her neck were restricted by twenty-five percent in left rotation and right lateral bending. Neurological examination was normal and there were no other remarkable physical findings.

Anterposterior (A-P) and lateral X-rays were taken of her cervical spine (figure 1. a and b). They show the cervical spine listing to the right with the head tilted to the left. There was also a loss of the normal lordosis. An A-P open mouth view of the atlantoaxial joint was unremarkable (not shown). There was no suggestion of any congenital anomaly.

This girl's torticollis was thought to be a congenital muscular torticollis. It was recommended that she undergo surgery to have the SCM muscle released and lengthened at its upper and lower ends. This was to be followed by exercises to prevent recurrence of the deformity.

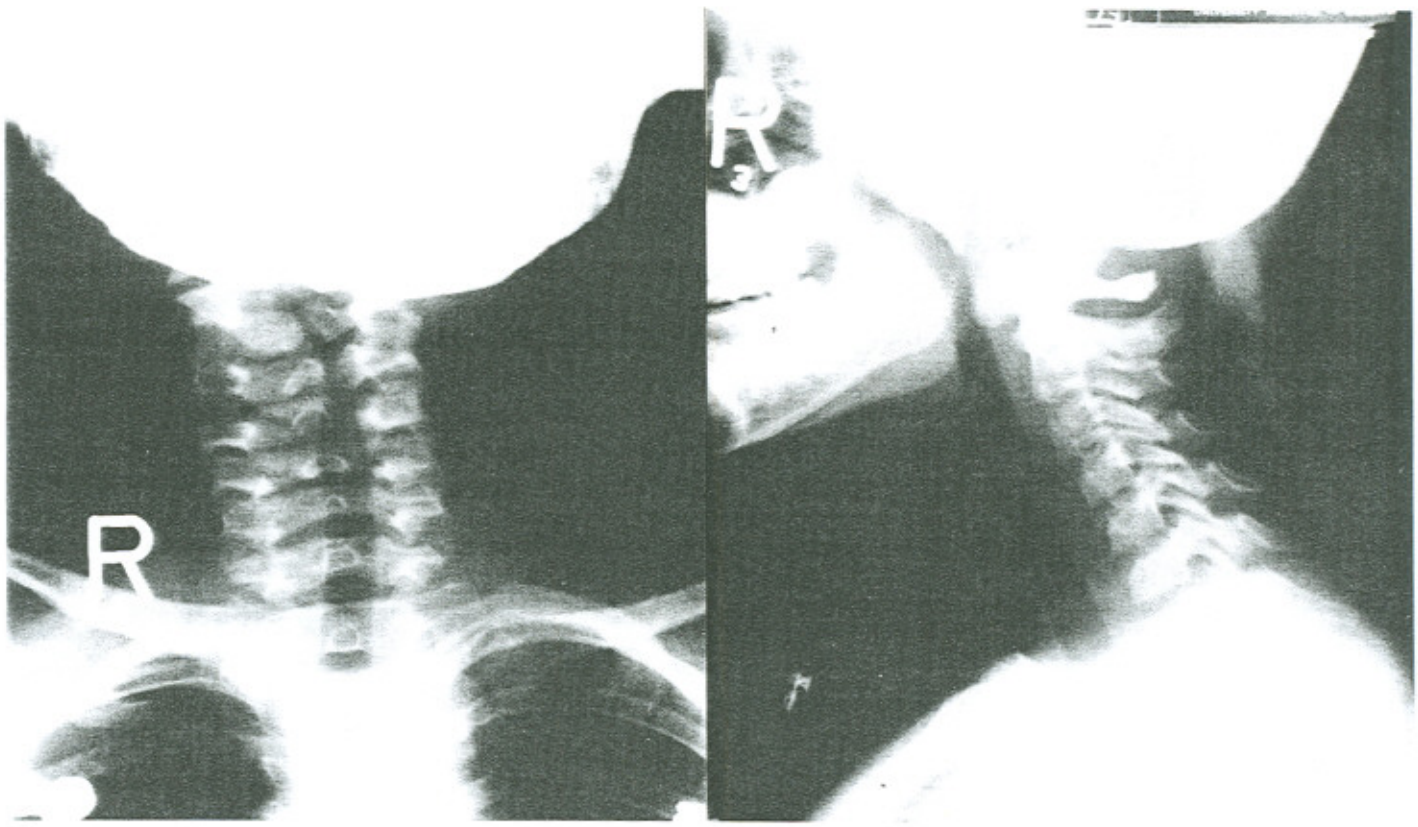

Figure 1. (a and b) Anterposterior and lateral radiographs of the cervical spine of Case 1. There is a right lateral list of the cervical spine and a left tilt of the head. There is also a loss of the normal cervical lordotic curve.

Upon considering the treatment options, the mother chose to seek an opinion from a chiropractor. The chiropractor began treatment involving manipulations and stretches to the neck.

Four months after the initial consultation the child was seen for follow-up. The mother seemed quite convinced that the manipulations had helped, but on examination the facial asymmetry was still present and the ranges of motion remained the same. The mother again refused the relatively simple surgical procedure and decided to continue with the chiropractic treatments. Unfortunately, the child was reaching an age where the facial asymmetry would become permanent.

Four months later the child was again seen. The mother was insistent that the torticollis was less severe, but she was not completely satisfied with the appearance. On examination, there was still contracture of the left SCM muscle with restriction of neck motion and facial asymmetry. It was pointed out to the mother that even with surgery, the facial asymmetry would not likely reverse at this late stage. Despite this, she still requested the operation.

A surgical release of both ends of the left SCM muscle was performed. The procedure was completed without complication and she underwent an uneventful post-operative recovery. Physical therapy was initiated the day after surgery. The mother was shown how to do these exercises and the patient was discharged with instructions to continue the exercises on a regular basis.

The child was reviewed at three weeks and again at four months post-operatively. Although mild facial asymmetry was persistent, the range of motion of the cervical spine was symmetrical and within normal limits.

Case 2

A fifteen-month-old girl presented to the Orthopedic Outpatient Clinic at the University Hospital for evaluation of her torticollis. She was the product of a full-term pregnancy and an uncomplicated delivery, and had been through normal developmental milestones. There was no history of fever or sickness.

The infant had been well until nine weeks before presentation when she sustained a fall. She was admitted to another hospital and was investigated for neck stiffness. These investigations were unremarkable. She was then treated by a chiropractor, with some improvement in neck movements, until approximate-

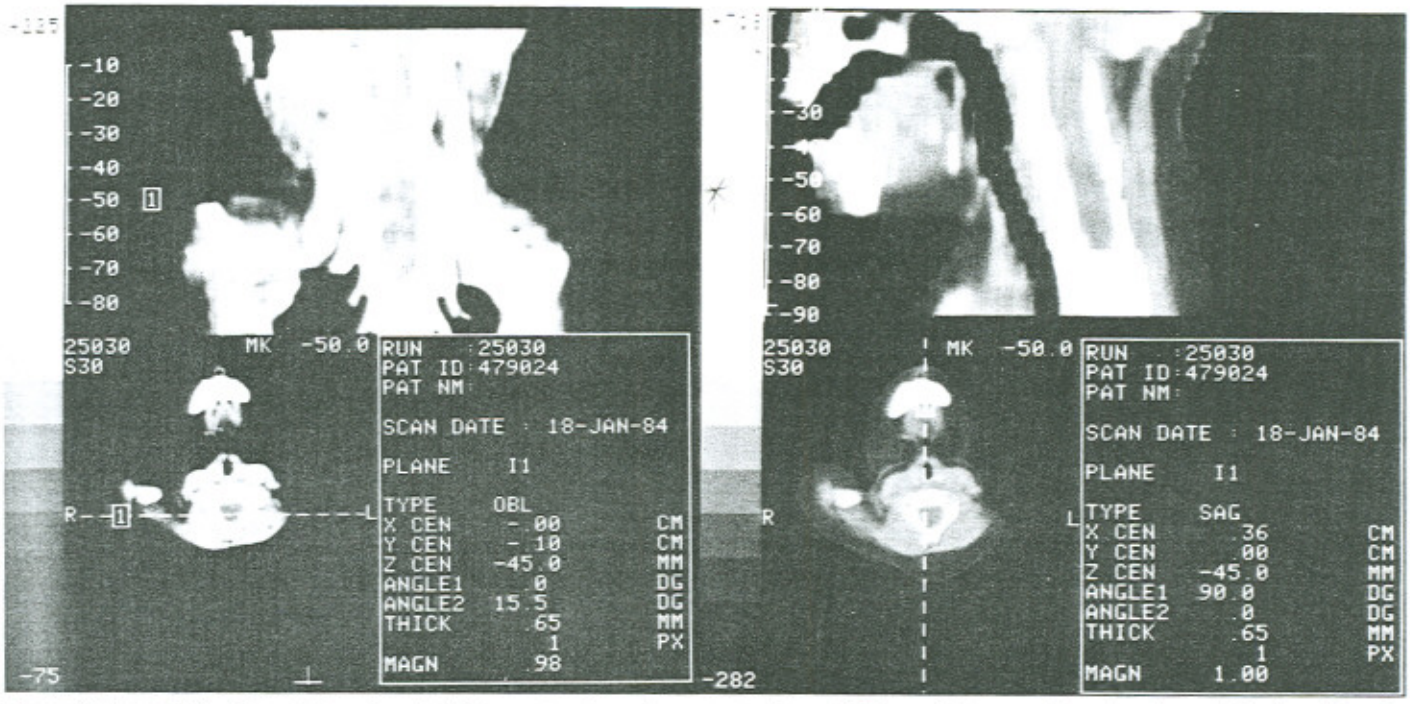

Figure 2. (a and b) Coronal (a) and sagittal (b) reconstructions of a contrast-enhanced CT scan of the upper thoracic and cervical spine of Case 2. There is complete obstruction of the contrast medium due to gross spinal cord enlargement with no flow proximal to the T1 level. In the lower cervical region no contrast medium can be seen, but the cervical spinal canal is enlarged. These findings are consistent with an intramedullary expansile lesion.

ly two weeks prior to presentation. At that time she fell backwards, hitting her head and neck. Following this she progressively became more irritable and refused to move her head and neck. The mother also noted that she began to fall with increasing frequency.

On examination the child was irritable and uncooperative. She did not have a low hairline, short neck or any observable deformity suggesting Klippel-Feil syndrome. There was a slight increase in the cervical lordotic curve and she walked with her shoulders in an elevated position and with a stooping posture. She refused to flex or extend the neck and would rotate to the left and right only about ten degrees. Palpation of her cervical spinous processes seemed to cause pain. The thoracic and lumbar spine exam and the examination of the hips were unremarkable. Neurological exam revealed that the cranial nerves were all intact. Signs of a Horner's syndrome were not present. She moved all limbs well and could walk without assistance. Her tendon reflexes were all present, symmetrical and normal. Sensation could not be adequately assessed.

X-rays of her neck were unremarkable, ruling out fracture or congenital anomaly. To rule out the possibility of neoplasm, she was admitted to hospital and a CT scan was arranged. A contrast-enhanced CT scan of the upper thoracic and cervical spine was obtained (figure 2. a and b). At about T1 there was complete obstruction due to enlargement of the spinal cord. In the lower cervical region no contrast medium could be seen, but the spinal cord and cervical spinal canal were enlarged. There was no evidence of any abnormality at the craniocervical junction. These findings were consistent with an intramedullary expansile lesion such as ependymoma or astrocytoma, although syringomyelia could not be absolutely ruled out. Schwannoma or meningioma could also be considered, but not typical for this age group.

To investigate further, laminectomies were carried out at the C6, C7, and T1 levels and the dura was opened. The spinal cord was swollen. An ultrasound study revealed a solid mass that blended into the spinal cord tissue. The decision was made not to remove the mass due to spinal cord involvement. A small incision was made along the dorsal surface of the spinal cord and some greyish material herniated through the opening. This tissue was taken for biopsy and the pathologist reported a Grade II (low grade, relatively low risk for malignancy) astrocytoma of the spinal cord (figure 3). Because of the swollen and distended spinal cord, fascia had to be used to seal the dura.

At discharge the patient had mild weakness of her right arm, but was able to walk without support and use both upper limbs well. Tendon reflexes were slightly more brisk in the right arm than the left. Neck movements returned, but still remained

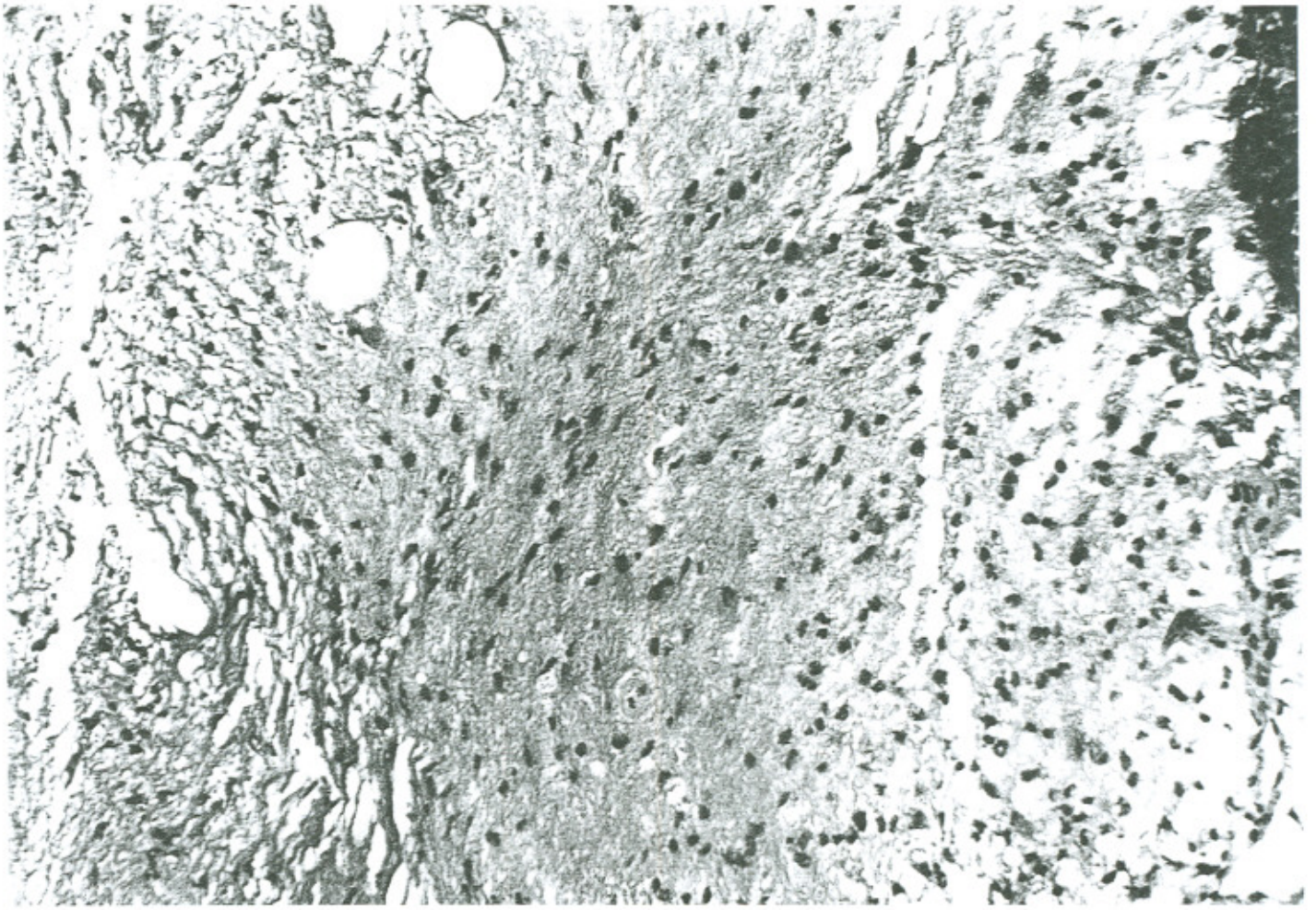

Figure 3. Pathology of the spinal astrocytoma in Case 2. Evenly distributed fibrillary astrocytes can be seen against a background of neuroglial fibrils and microcysts. No mitotic figures are apparent (H&E, × 240).

limited, particularly in rotation to either side.

A post-operative course of radiation therapy was performed, which was tolerated well by the patient.

On last review, four years and four months post-operatively, the child was doing well, with apparently a complete recovery from her cervical cord astrocytoma. On examination, she was short for her age and she had a slightly short neck. Cervical spine ranges of motion were within normal limits and there was no neurological deficit. She is presently undergoing review every six months.

Case 3

A five-month-old boy was taken to the University Hospital for evaluation of a mild torticollis. Sixteen days prior to admission he fell onto a carpeted floor from a height of approximately twelve inches. Since that time he would not sit up and his head remained tilted to the left. The boy had been seen by a chiropractor who had been manipulating his neck. Past history revealed that he was the product of a full-term, normal pregnancy and delivery, and was reaching appropriate developmental milestones. There was no history of fever, sickness or irritability.

On examination the head was tilted to the left and rotated to the right. The left clavicle was noted to be somewhat irregular in the middle third, but there were no further signs of external injury. Neurological examination was unremarkable.

Laboratory investigations, including ESR, WBC and differential were unremarkable. X-rays of the cervical spine, clavicles and chest were normal. (Clavicular fracture can be a cause of torticollis.)

All investigations were negative and no conclusive diagnosis

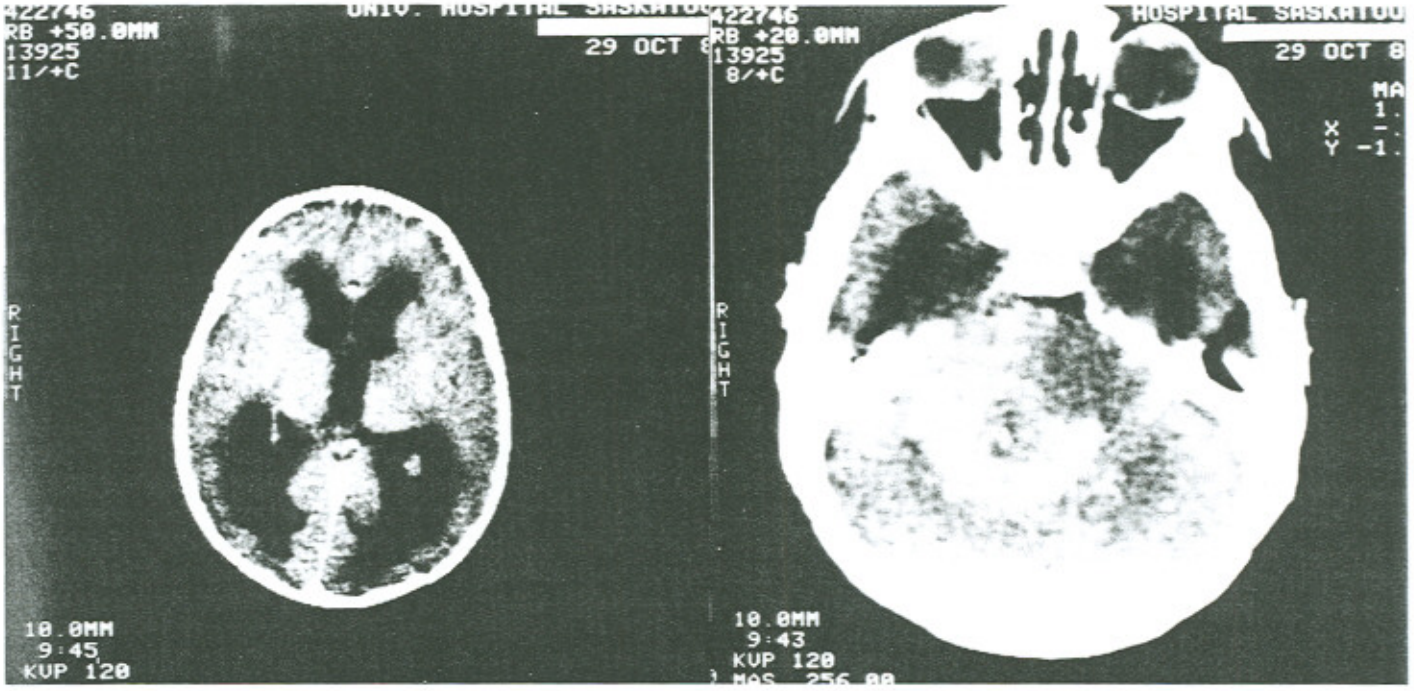

Figure 4. (a and b) Selected cuts of a plain and contrast-enhanced CT scan of the head. (a) The plain CT shows enlargement of the posterior aspects of the lateral ventricles, indicating obstructive hydrocephalus. (b) With contrast, a cut through the posterior fossa demonstrates a radiodensity slightly to the right of midline, consistent with an infiltrating primary neoplasm of the cerebellum.

was reached. It was thought that the torticollis may have been caused by soft tissue injury due to the fall. He was discharged after three days in hospital and the parents were advised to bring him in for review in four weeks time, or earlier if symptoms persisted or became worse.

He was readmitted two days later when the parents stated that immediately following his discharge, he began to vomit periodically, would not eat and became increasingly irritable. He would also have attacks of arching his back and crying. The torticollis had become more noticeable. There was still no history of fever and no weight loss.

On examination he appeared pale and lethargic, but was afebrile. He was comfortable in the prone position, but would become very irritable with screaming and crying attacks and typical opisthotonus on any handling. He had good head control, but while pulling to a sitting position, the head lagged behind with a left torticollis, tipping to the left and remaining in extension. He did not move his eyes from side to side. His pupils were equal and reactive to light and there was no papilledema. He moved all limbs with a slight tremor, from time to time involving his hands. No decerebrate movements of the limbs were noted. Tendon reflexes were brisk and extensor plantar responses were noted. No clonus was present. Ear, nose and throat exams were unremarkable.

A plain and contrast-enhanced CT scan of the head from base to vertex was obtained. Selected cuts are shown (figure 4. a and b). The plain CT showed enlargement of the posterior aspects of the lateral ventricles, indicating obstructive hydrocephalus. On a contrast-enhanced view, an infiltrating tumour in the posterior fossa was visualized. This was thought to be consistent with a primary neoplasm, most likely an ependymoma, neuroblastoma, or embryonal cell tumour.

He was booked for surgery the next day, but on the following morning he went into cardiorespiratory arrest. There was no sign of respiration or heartbeat, and the pupils were fixed and dilated. He died after unsuccessful attempts to resuscitate him. The parents refused to consent to an autopsy.

Discussion

Torticollis can be associated with a wide variety of childhood illnesses.[7,8] The possibility of serious underlying pathology should always be considered, especially in infants or children with persistent pain or deformity. Although numerous causes of torticollis have been documented, see table 1, in many cases the underlying disorder is never found. The clinician's primary concern is to understand the natural history of the disorder and differentiate the benign torticollis from torticollis secondary to a structural deformity or a pathological lesion.

Congenital torticollis

Congenital muscular torticollis is reported as the third most common paediatric orthopaedic anomaly next to congenital dislocation of the hip (CDH) and clubfeet.[9] The reported incidence varies from 0.4% to 1.9%. It is commonly associated with primiparous births and complicated labour and delivery, although the etiology and pathogenesis are not fully understood.[10] A palpable, non-tender, soft enlargement that is attached to or located within the body of the SCM muscle is usually noticed shortly after birth. The muscle itself is shortened and, if shortening persists while growth continues, the torticollis becomes more striking. Craniofacial asymmetry may result if the contracture is not corrected.

The classical clinical presentation is an infant or child with positional deformity of the head and neck (the head bent towards and rotated away from the side of contracture), with marked restriction of neck movements. Craniofacial asymmetry is almost always present. Up to 20% of children with congenital muscular torticollis have CDH.[11]

The treatment of congenital torticollis depends on the age of presentation. Conservative care (daily stretching exercises, and specific positioning and handling instructions) is possible if treatment begins before one year of age.[9] Patients with less than thirty degrees restriction of rotation respond better.[9,12] If these conservative measures are unsuccessful by 18–24 months of age, surgery is recommended, as no other form of therapy has been shown to reduce the deformity. Relatively safe and simple procedures have been described.[12,13] The surgery should be performed prior to school age. An acceptable cosmetic result, however, can be obtained as late as 12 years of age.[14] Asymmetry of the skull and face will correct as long as adequate growth potential remains after the SCM contracture is released.

Congenital anomalies of the cervical vertebrae, collectively referred to as the Klippel-Feil syndrome, may also cause a persistent torticollis. Although classically described as a clinical triad of low posterior hairline, short neck, and limitation of head and neck movement, many of the mildly involved patients do not have all of these clinical features.[15] Approximately 20% of patients with cervical anomalies have facial asymmetry, torticollis, or webbing of the neck.[15]

Congenital vertebral anomalies commonly co-exist with other system defects. Urinary tract anomalies, hearing loss, congenital heart disease, and vertebral artery compromise are commonly found in association with Klippel-Feil syndrome. Many of these hidden abnormalities may be far more detrimental to the patient's well-being than the obvious deformity of the neck.

Torticollis due to bony anomalies can usually be distinguished clinically from other causes of torticollis in children.[3] Unlike muscular torticollis, the deformity may not be noticeable until later in childhood. With time, the deformity becomes more evident and more rigid as the child gradually loses the ability to compensate with motion in the unaffected segments of the cervical spine. The SCM on the side of head tilt is not contracted or in spasm, and there is no history of recent throat infection or trauma. Radiographs are useful in this diagnosis although they may be difficult to interpret until an older age (12–18 months) is reached.[16]

The treatment of torticollis due to bony anomalies is palliative, with aims at reducing the pain associated with the deformity. Some authors feel that early surgical intervention can reduce the amount of deformity in congenital torticollis,[16] although non-operative measures are usually preferred and safer.[3] There is little that can be done to achieve correction of the congenital deformity in the adult.

Acquired torticollis

There are a wide variety of acquired causes of torticollis in infants and children (table 1). Transient muscle spasm, cervical adenitis, and atlanto-axial rotary displacement are some of the most frequently encountered,[1] although more serious and life-

TABLE 1
CLASSIFICATION OF TORTICOLLIS

Congenital
Congenital muscular torticollis
Klippel-Feil syndrome, Sprengel's deformity
Congenital articular and ligamentous lesion
Arnold-Chiari malformation, spina bifida

Acquired

Traumatic
Subluxations, dislocations, fractures

Infection/inflammatory
Upper respiratory tract infections, cervical adenitis
Retropharyngeal abscess, osteomyelitis, tuberculosis
Rheumatoid arthritis

Neoplasm
Neurogenic: posterior fossa, spinal cord, vestibular system
Vertebral column: osteoid osteoma

Neurogenic
Syringomyelia
Ocular dysfunction, bulbar palsies
Dystonic syndromes

Idiopathic
Atlantoaxial rotary displacement
Joint dysfunction

Miscellaneous
Benign paroxysmal torticollis
Spasmodic torticollis
Toxic (drug-induced)
Functional

Other
Gastrointestinal disturbances (Sandifer's syndrome)
Intervertebral disc calcification

threatening disorders can cause the deformity. The acquired causes of torticollis have varying natural histories depending on the underlying disorders. Determining the cause of the deformity is essential before treating the patient.

Neurogenic causes of torticollis in children are not common, but can be life-threatening.[1,17] Posterior fossa tumours, as in Case 3, are thought to be the most frequently encountered neurogenic cause.[18] Tachdjian, in a study on intraspinal tumours, found that torticollis was a common clinical sign, being present in 18% of those studied.[2] It was the third most common presenting complaint next to leg weakness and back pain.[2] Long-standing histories of such symptoms as root pain, back pain, and neck stiffness are typical histories of children with intraspinal tumour.[9] Other long-standing complaints that should raise suspicion of neurogenic tumour include headache, hearing loss, vestibular dysfunction (nausea and vomiting), and abnormalities of vision. Intramedullary gliomata were the most common pathological type of intraspinal tumour found in Tachdjian's study.[2] Neurofibromata, Schwannoma and meningioma, common in adults, were rare.

The diagnosis of intraspinal or posterior fossa tumour is confirmed with imaging techniques. Plain films may reveal widening of the spinal canal. Myelography can confirm the extent of intraspinal occlusion and may help to localize the lesion. CT scanning, with and without contrast, can be of great benefit, as is demonstrated in Case 3. Magnetic resonance imaging (MRI) allows visualization of the neural structures within the spinal canal or cranium, and is the imaging modality of choice in neurogenic soft tissue lesions.

The treatment of torticollis caused by intraspinal or posterior fossa tumour is surgical, occasionally supplemented with radiotherapy.[2] The location, extent of tissue removal, and type of tumour dictate the prognosis in these cases. Recurrence rates vary depending on the type of tumour.

Benign causes of torticollis, such as atlantoaxial joint dysfunction, usually resolve quickly with appropriate treatment. If childhood torticollis is long-standing, resistent to treatment, or progressive, the clinician should carefully search for more serious causes of this disorder. In some cases, delay of the diagnosis can result in permanent disability or even death.

Conclusion

Torticollis in an infant or child can be an indication of serious underlying pathology. The diagnosis depends on a complete history, including pre- and peri-natal periods, and an extensive clinical examination, possibly including diagnostic radiology.

Chiropractors should be aware of the various causes of torticollis in children. Identifying the underlying cause is essential before embarking on efforts to treat the deformity. When a full evaluation rules out a structural or neurologic cause in torticollis, conservative therapy can be safely instituted.

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