

Spasmodic torticollis: a case study

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Torticollis is a term that describes abnormal posturing of the head secondary to the contraction of the neck musculature. Spasmodic torticollis is a rare form of this disorder that has been attributed to disturbances in the extrapyramidal system. It is a form of focal dystonia that primarily affects women in their forties and usually progresses slowly, leading to severe disability. Although the torticollis may be painful, the patient's main preoccupation is usually with the deformity itself. The diagnosis is often delayed because of the unusual clinical presentation and these patients are frequently labelled as neurotic. The following report illustrates a case of torticollis and thoracolumbar scoliosis secondary to dystonia. (JCCA 1992; 36(3):146-151)

KEY WORDS: spasmodic torticollis, scoliosis, dystonia, chiropractic.

Torticollis est un terme qui décrit une posture anormale de la tête suite à la contraction des muscles du cou. Le torticollis spasmodique est une forme rare de cette affection qui a été attribuée à des troubles du système extrapyramidal. Il s'agit d'une forme de dystonie focale affectant principalement les femmes dans la quarantaine et qui progresse habituellement lentement, aboutissant à une infirmité profonde. Bien que le torticollis puisse être douloureux, le patient se préoccupe principalement de la difformité elle-même. Le diagnostic est souvent retardé dû à l'aspect clinique inhabituel et au fait que ces patients sont souvent classés comme névrosés. L'étude suivante présente un cas de torticollis et de scoliose thoracolumbaire suite à une dystonie. (JCCA 1992; 36(3):146-151)

MOTS CLÉS : torticollis spasmodique, scoliose, dystonie, chiropratique.

Introduction

Torticollis is characterized by contraction of the neck muscles causing the head to be tilted or rotated to one side. The term torticollis is only descriptive and should not be considered a diagnosis.¹ One should look for an underlying disease. Torticollis is usually thought of as originating from osseous, ligamentous or soft tissue injury to the neck. But, torticollis may occur secondary to central nervous system diseases causing dystonia of the neck musculature.

Dystonia is a syndrome dominated by sustained muscle contractions causing twisting and repetitive movements or abnormal posture that may be sustained or intermittent. Diffuse diseases

of the brain such as autoimmune, toxic or degenerative disorders can cause a symptomatic form of dystonia. Still, idiopathic dystonias have also been recognized. These can be generalized, like dystonia musculorum deformans, which is an inherited form of dystonia appearing in childhood, or they may be focal, like writers cramp, blepharospasm, and spasmodic torticollis.^{2,3,4}

Spasmodic torticollis is one of the most common forms of focal dystonia and its incidence in the Canadian population is about 1 in 250,000. The disease is twice as common in females and often appears between the 4th and 6th decades with a mean age at onset of approximately 40 years.^{2,5}

Spasmodic torticollis is frequently misdiagnosed. A retrospective study on 266 patients found that the diagnosis had been delayed by an average of 37 months.² The purpose of this case report is to familiarize the reader with this disease and illustrate the problems associated with late diagnosis.

Case report

A 50-year-old woman presented to the Neurology Outpatient Clinic for an evaluation of a left-sided torticollis. There was no family history of a similar problem. Her neck had started twist-

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ing fifteen years ago without apparent reason. Otherwise, the history was unremarkable. Initially, the anomaly was very subtle and barely noticeable, except in periods of stress. In time, it gradually worsened. She had some pain in the right suboccipital region that developed over the last few years. The discomfort was minor and not as disabling as the deformity itself, which caused her to feel unsteady while performing her usual activities. Over the past five years, she noticed a tendency to walk on the toes of her right foot. Throughout the years, she had seen several physicians and specialists regarding these complaints. She had received several courses of chiropractic manipulation and physiotherapy to the neck. She had been told several times that her disorder was psychological and that no treatment would help. The first few years following the onset, she had been on diazepam, but had not received any other medical treatment.

On examination, there was a marked torticollis with tilting of the head to the left and elevation of the right shoulder associated with a moderate right thoracolumbar scoliosis. (Figure 1) The torticollis and the scoliosis could be momentarily corrected by

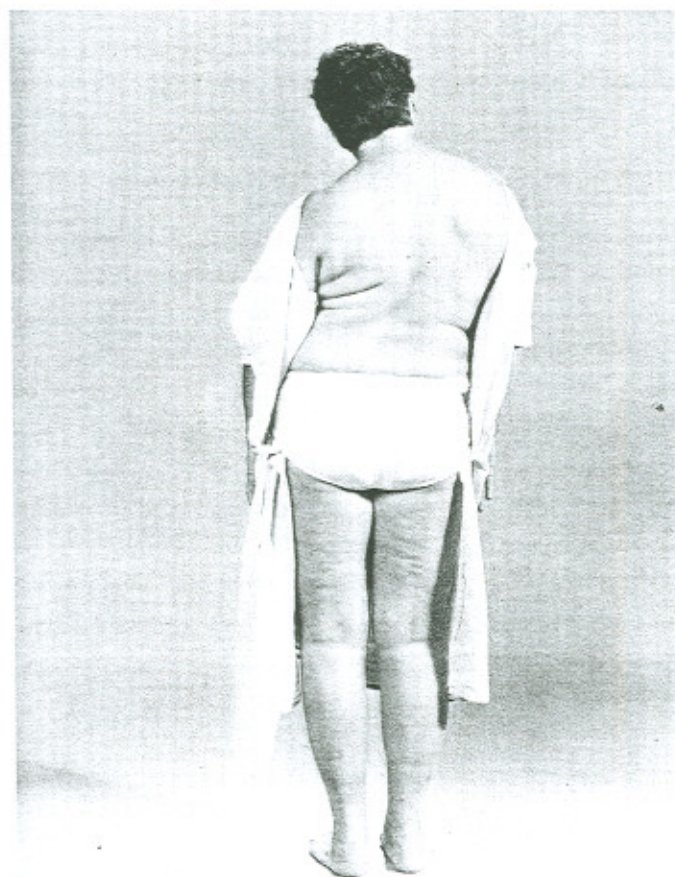


Figure 1 A posterior view of the patient shows the torticollis and convex right thoracolumbar scoliosis.



Figure 2 (a) An anterior view of the head and neck shows the extent of the torticollis.
(b) This picture shows that the patient can momentarily straighten the neck with a conscious effort.

the examiner or by the patient when she made a conscious effort to stand in a normal position. (Figures 2 and 3) Upon forward flexion, the scoliosis almost fully corrected and was not associated with a rib hump. (Figure 4) Head jerks were present and amplified by her attempt at keeping the head in a normal position. The left sternocleidomastoideus was hypertrophied, but not tender. In addition, she tip-toed on the right side when walking. Examination was otherwise unremarkable.



Figure 3 (a) This picture shows the right thoracolumbar scoliosis and torticollis.
(b) The patient can almost fully correct her curve with a conscious effort to maintain herself in a straight position.

The radiological examination, including cervical spine views (Figure 5) and a scoliosis view (Figure 6), showed the postural deformities and degenerative changes in the lower cervical spine. The right thoracolumbar scoliosis measured 32 degrees from T12 to L1. Other studies, including a myelogram and a cervical spine CT scan, were unremarkable.

The neck deformity was considered secondary to an involuntary movement disorder. In the absence of any associated findings, the cervical dystonia was considered idiopathic and recognized as a spasmodic torticollis. Still, the case was slightly unusual because the dystonia showed a tendency for generalization and was affecting the trunk and the right leg. For this reason, she was prescribed a dopaminergic medication, but if no improvement is noted, this medication will be substituted for an anticholinergic agent. If the medical treatment is unsatisfactory, she will be given intramuscular injections of botulinum toxin into the dystonic muscles.

Discussion

The patient with spasmodic torticollis has a unique presentation that should not be mistaken for other forms of this disorder.¹ The cardinal features include neck spasms characterized by forceful contraction that cannot be overcome, or head jerks defined as large amplitude, irregular and sudden involuntary movements.² The dystonia occurs in the absence of neurological deficit and the radiographic findings are usually absent or limited to secondary degenerative changes.⁴

The onset of dystonic torticollis is not usually related to a traumatic injury. The disorder appears insidiously and progresses slowly, but steadily, through stages.⁵ Spontaneous remission occurs in about 12 percent of cases, usually in the year following the onset of the disorder.⁶ Initially, the dystonia is present with activities such as walking and when the patient is under emotional stress. It then intensifies and persists at rest, but can still be overcome momentarily by a conscious effort. The

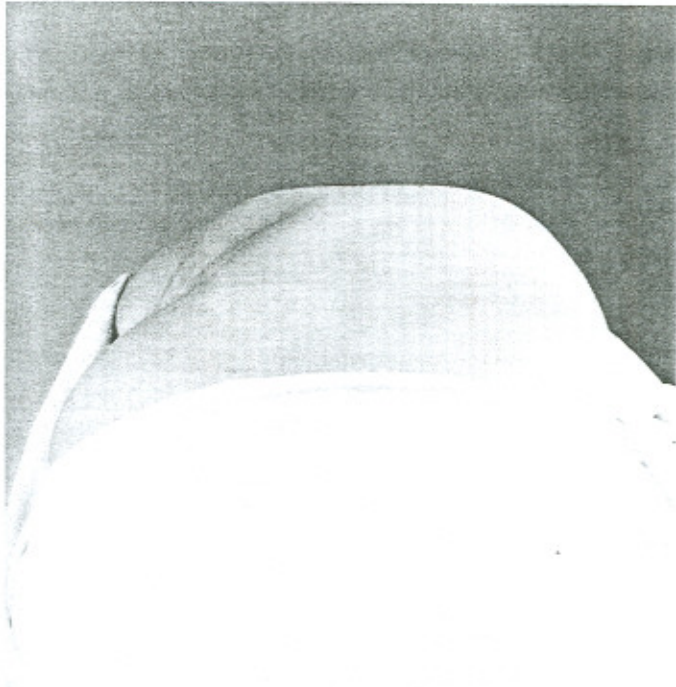


Figure 4 In this picture, the scoliosis almost fully corrects upon forward flexion. There is no evidence of a rib hump.

dystonia then becomes so severe that the patient is unable to keep their head in a normal position without hand support. Finally, the head becomes fixed in an abnormal position. As the severity of the dystonia increases, muscle hypertrophy and pain are more frequent findings.^{4,5}

In one quarter of patients with dystonic torticollis, there is an associated postural tremor of the head or hands, similar to essential tremor. It is characterized by small amplitude, rhythmic oscillations of the head or hands, that increases with action and interferes with activity. The dystonia may also involve adjacent areas such as the shoulder girdle. Other areas like the trunk or limbs can be affected. In addition, the disorder can be generalized.

Several strange phenomena are associated with spasmodic torticollis. The disorder may diminish when the patient is lying down, or when they are distracted while watching television, reading a book, or during other types of activity. Furthermore, a marked torsion of the head can be momentarily corrected if the patient simply places a finger on the rotated chin.

The strange clinical presentation of patients with spasmodic torticollis has contributed to the belief that the disorder is a form of neurosis.^{3,7} Yet, several studies, including a study of 85 patients with spasmodic torticollis, have shown that the incidence of psychiatric disorders at the onset of the disease is no different from normal controls.⁸ Although this disease is commonly misdiagnosed as a psychiatric disorder, the scientific

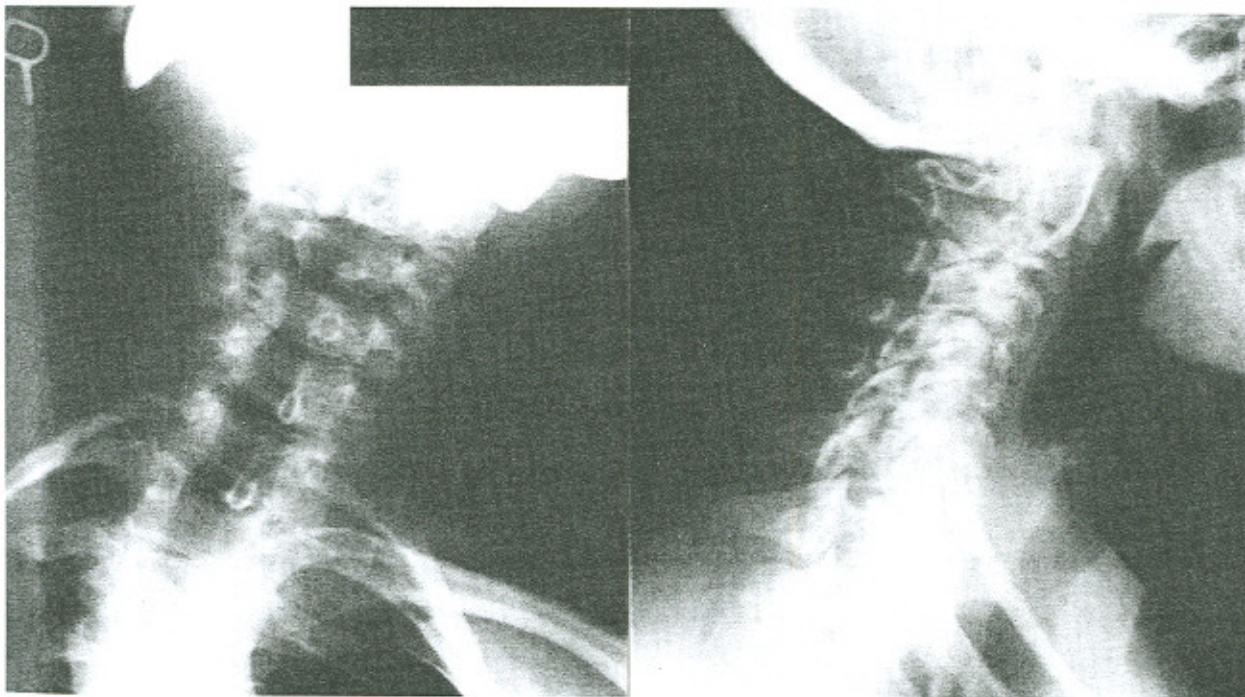


Figure 5 The anteroposterior and lateral cervical radiographic views shows tilting of the cervical spine to the left and degenerative changes involving the lower levels.

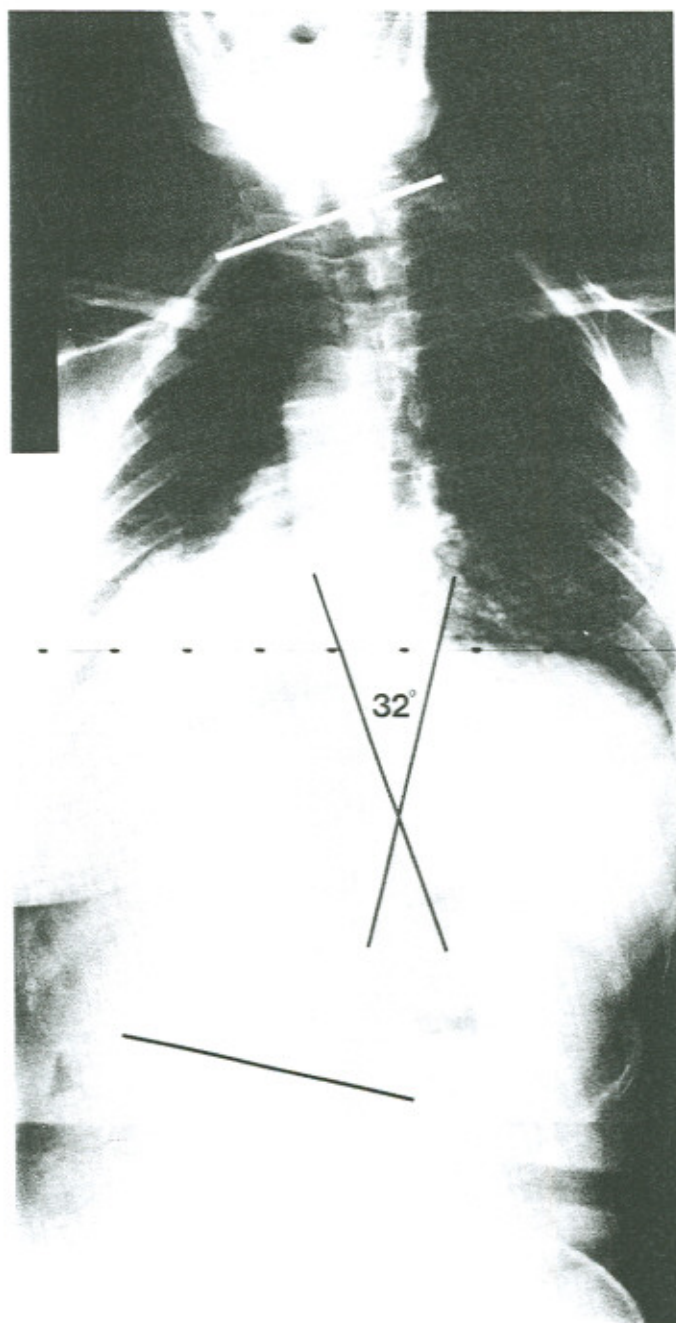


Figure 6 The posteroanterior scoliosis view shows a 32 degrees right thoracolumbar scoliosis measured from T1 to L2.

community now recognizes that it is related to disturbances in the extrapyramidal system.

The extrapyramidal system including the basal ganglia and cerebellum modulates motor activity. The static postural tone provided by the basal ganglia is essential for voluntary movement. For instance, basal ganglia lesions may cause a motor syndrome characterized by involuntary movement, choreoathetosis and dystonia. The features of the motor syndrome closely correspond to those associated with spasmodic torticollis. In spite of this, no distinct lesion or abnormality capable of explaining the symptoms have yet been identified in patients with spasmodic torticollis.⁴

The low rate of spontaneous remission and the paucity of effective pharmaceutical treatment has, until recently, limited the chance of improvement for patients with this disorder. While dopaminergic medication may occasionally be effective with more generalized dystonia, anticholinergic agents are usually more effective.^{3,9} These medications benefit 40 percent to 50 percent of patients, but they need to be given in large doses and are associated with severe side effects.¹⁹

The most effective surgical treatment consists of bilateral sectioning of the rootlets of the 11th cranial nerve, combined with bilateral anterior rhizotomy of C1 to C3, and section of the anterior root of C4 on the side of the more involved sternomastoid muscle.¹¹ The most promising treatment consists of injecting the involved muscles with botulinum toxin, which causes denervation atrophy and correction of the deformity. This treatment is effective in 60 to 87 percent of cases and has very limited side effects.^{10,12}

Conclusions

Spasmodic torticollis is an idiopathic form of cervical dystonia. It is an unusual form of torticollis that usually affects women between the fourth and sixth decades of life. This disorder should not be mistaken for other forms of torticollis, nor should it be considered a form of neurosis. The dystonia is attributed to a disturbance in the extrapyramidal system, yet no definite abnormality has been identified. These patients are usually seen by several physicians prior to diagnosis. Although spinal manipulation and physiotherapy may provide temporary pain relief, medical treatment, such as botulinum toxin injection, is usually necessary to control the dystonia.

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Oh what a feeling!

