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Tremor and Other Hyperkinetic Movements

Articles

Cervical Dystonia Mimics: A Case Series and Review of the Literature

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Abstract

Background: Cervical dystonia is mostly idiopathic in nature. However, a small subset of cases are mimics, leading to diagnostic pitfalls. There is paucity of literature on pseudodystonias affecting the cervical region.

Method: We performed a retrospective review of patients attending a movement disorders clinic over a period of 7 years (2012–2018). Among them, those who were considered to have mimics of cervical dystonia based upon clinical and supportive investigations were included.

Results: Six out of 2,412 patients (0.24%) were diagnosed as cervical dystonia mimics and the causes included isolated neck extensor myopathy (2), craniovertebral junction anomalies (2), sternocleidomastoid fibrosis (1) and post traumatic sequelae (1). Among these patients, three patients had received various treatments for cervical dystonia, including botulinum toxin injections.

Discussion: Mimics of isolated cervical dystonia are rare. A high degree of suspicion and proper diligent clinical assessment assists management and prognostication.

Keywords: Cervical dystonia, pseudodystonia, dystonia mimics, head drop, neck extensor myopathy

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Introduction

The 2013 expert consensus panel by Albanese et al. defined dystonia as "a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both. Dystonic movements are typically patterned, twisting, and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation."[1] The goal of the revised definition was to exclude possible conditions that may mimic dystonias, also known as "pseudodystonias." These pseudodystonias are thought to be caused by etiologies that are presumed to be different from the broader dystonia group.[1] It is critical to diagnose these pseudodystonias, as the therapy and prognostication is significantly different from that of typical idiopathic dystonia. In this article, we review our cases of cervical pseudodystonias along with a review of the published literature.

Methodology

The study involved retrospective analysis of subjects attending a movement disorders clinic over a period of 7 years (January 2012 to December 2018). Clinical records of subjects who were diagnosed to have cervical pseudodystonias based on clinical and imaging findings were included in the study. Patients who had pseudodystonia involving other body parts/segments were excluded.

Results

Six out of the 2412 (0.24%) patients who attended the movement disorders clinic were diagnosed with cervical pseudodystonia (Table 1). These patients were evaluated by health care professionals at various levels, including general physicians and neurophysicians. The causes of cervical pseudodystonia included isolated neck extensor myopathy (n = 2), congenital sternocleidomastoid fibrosis (n = 1), Post traumatic craniocervical junction sequelae (n = 1), and developmental craniovertebral junction (CVJ) anomaly (n = 2). The following two cases illustrate the diagnostic dilemmas, leading to various therapeutic interventions.

Table 1. **Clinical Features and Imaging Findings of Pseudodystonia Subjects**

Video	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
	Video 1	Video 2				Video 3
Age at onset	45	Childhood	6	21	11	24
Current age	48	22	6	53	14	24
Gender	M	M	F	M	M	M
Clinical presentation	Neck posturing with neck flexion	Abnormal posturing of neck since childhood	Abnormal posturing of neck following a trauma	Posturing of neck since age of 21. Increased in the last 5–6 months with paraspinal spasm	Abnormal neck movements	Neck posturing and discomfort in left hand (acute onset)
Clinical findings mimicking dystonia	Antecollis with facial dystonia	Torticollis to the right with minimal retrocollis; left SCM is taut and string like	Severe torticollis with right shoulder elevation	Mild torticollis to the left present with features of antecollis	Jerky neck movements (dystonic jerks vs. akathisia)	Lateral shift to the right with right torticaput
Other neurological /clinical findings	Neck extensor weakness (MRC grade 1/5)	Thin and fibrosed left SCM	None	Head drop with neck extensor weakness (G3+/5)	Dysmorphic features	Jerky tremors of hands, bilateral hand grip weakness
MRI findings	Fibrosis with fatty infiltration of deep cervical muscles	Atrophy and fibrosis of the left SCM. Scoliosis the of cervical spine with convexity to the right side	Subluxation of C2-C3	Deep paravertebral muscle atrophy	Increased Atlanto dental distance of 100 mm with retroversion of odontoid process. Signal change and thinning of cord at C1-C2 level. Hypertrophied anterior arch of atlas, hypoplastic odontoid process	Multiple anomalies in CVJ with atlanto occipital assimilation, partial fusion of C1-C2 vertebrae, hemi vertebra with fusion of right C4-C5 level along with C3-C4 spinal cord hyperintensities
Interventions done before diagnosis	Anticholinergics, Benzodiazepines, Dopamine blocker, Botulinum toxin, none	Two sittings of botulinum toxin injections given. Anticholinergics	None	None	Benzodiazepines	None
Final diagnosis	Isolated neck extensor myopathy	Muscular fibrosis	Posttraumatic subluxations	Isolated neck extensor myopathy	Atlanto axial dislocation	CVJ anomaly
Intervention after diagnosis	Medical management and supportive care	Option for surgical release given. Patient opted for no intervention	Surgical corrections	Medical management and supportive care	Surgical correction	Surgical referral

Abbreviations: CVJ, Craniovertebral Junction; MRC, ; SCM, Sternocleidomastoid Muscle.

Case 1

A 48-year-old gentleman presented with a 2.5-year history of abnormal posturing of the neck. The symptoms started gradually, reaching a peak at around 5 months after onset. He was diagnosed with cervical dystonia and was treated with an anticholinergic, dopa agonists, and benzodiazepines without any benefit. During this course, he underwent repetitive nerve stimulation and magnetic resonance imaging (MRI) of brain, which was normal. An MRI of the cervical spine showed degenerative disc changes without any significant compression of nerve roots or cord changes. Due to persistent symptoms and impaired activities of daily living, he was referred for botulinum toxin injections. There was no significant family history or history of exposure to antipsychotics. He had anterocollis and intermittent lower facial contractions. Passive range of movements of the neck was within normal limits. Power assessment revealed severe neck extensor weakness (Grade 0–1/5 Medical Research Council (MRC)) (Video 1). There were no other associated findings.

Further evaluation showed normal serum creatinine phosphokinase (CPK) (162 IU/dl). An MRI of the cervical muscles showed atrophy, fatty infiltration, and fibrosis of deep cervical muscles, especially at the level of C2, extending above and below. Trapezius and sternocleidomastoid muscles appeared normal (Figure 1A). Electromyogram of the Trapezius and Semispinalis showed very few motor unit potentials (MUP) and no recruitment. Bilateral sternocleidomastoid showed increased spontaneous activity (right > left), with normal recruitment. Clinically, he was considered to have head drop secondary to isolated neck extensor weakness (INEM), mimicking cervical dystonia.

Case 2

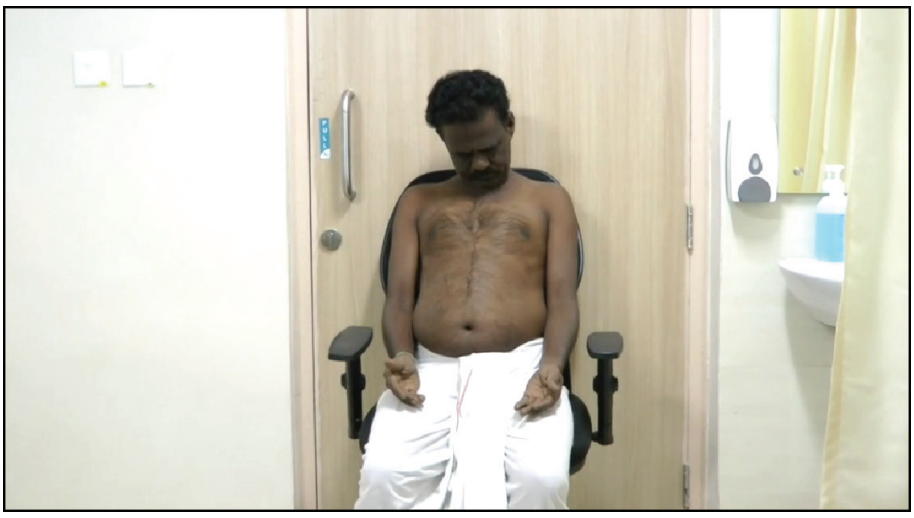
The 22-year-old man presented with a history of abnormal neck posture since early childhood. He was evaluated and diagnosed with cervical dystonia and treated with botulinum toxin injections. Due to limited clinical benefit, he was referred to our clinic. On evaluation, he was noted to have torticollis to the right with mild retrocollis (Video 2). In addition, he was also noted to have a thin and taut left sternocleidomastoid muscle. An MRI of the neck showed atrophy and fibrosis of the left sternocleidomastoid (Figure 1B) along with scoliosis of the cervical spine with convexity to the right side. He was diagnosed with developmental sternocleidomastoid fibrosis mimicking cervical dystonia, and a surgical release was suggested.

Discussion and literature review

Pseudodystonias are a rare group of disorders where appropriate clinical suspicion and evaluation has an impact on treatment and prognostication. Cervical pseudodystonias can be classified further into (1) vascular, (2) musculoskeletal, (3) infections, (4) mass lesions, (5) traumatic, (6) ocular causes (7) otological causes, (8) gastrointestinal causes, (9) psychogenic, and (10) others (Table 2). The aim of the discussion is to highlight potential presentations of cervical pseudodystonia.

(1) Vascular

A number of vascular pathologies can result in true cervical dystonia, majority of them being ischemic or hemorrhagic events of the brainstem, cerebrum, and cerebellum.[2–5] However, vascular anomalies are noted to have abnormal neck posturing due to compensatory mechanisms to aid visual defects.[6] Other vascular causes like aneurysms and arteriovenous malformations (AVM) can also result in dystonia.[6–9] An abnormal neck posture can also be seen as a compensatory mechanism to overcome visual field defects masquerading as cervical dystonia.[6] Spinal epidural hemorrhage secondary to severe hemophilia can present with torticollis.[10] The outcomes are



Video 1. **Video Shows Neck Antecollis along with Lower Facial Movements.** In the second part of the video, clinical examination shows difficulty in neck extension movements against gravity.

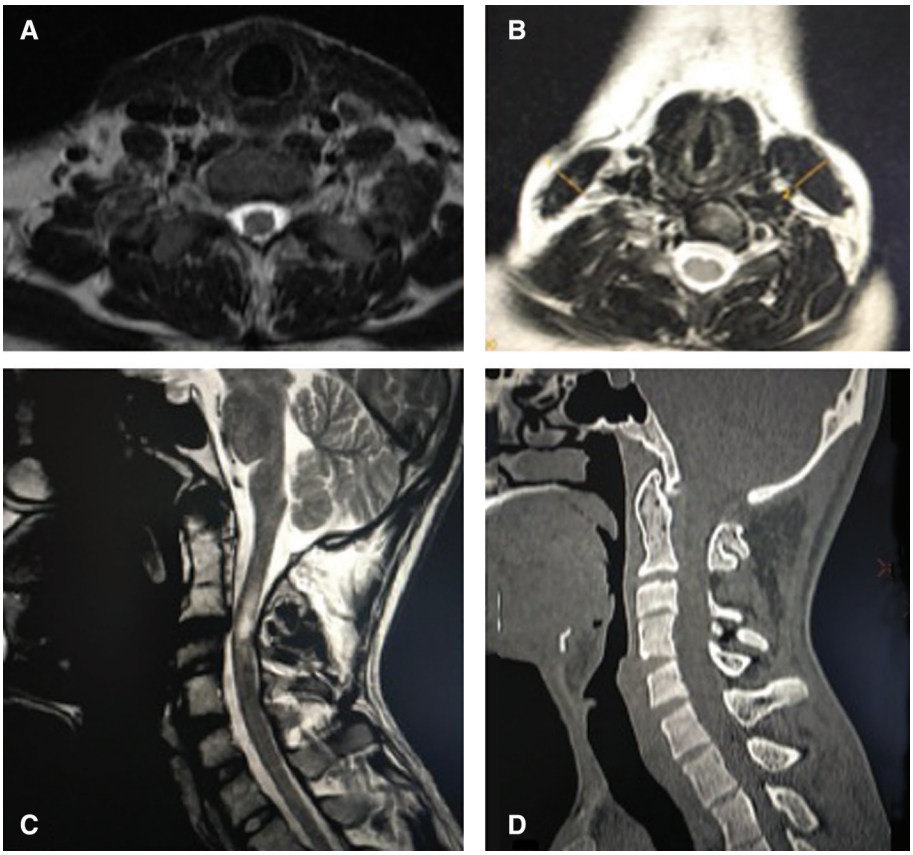


Figure 1. **(A) T2WI Axial Section of cervical Spine Shows Atrophied Deep Paracervical Muscles with Fibrosis and Fatty Infiltration.** (B) T2WI axial section of the Neck showing the atrophied left sternocleidomastoid muscle (yellow line-1) in comparison to the normal right sternocleidomastoid muscle (Yellow line-2). (C, D) T2WI Sagittal MRI (C) of CVJ area and sagittal CT section cranio-cervical junction (D) showing multiple anomalies in CVJ with atlanto occipital assimilation, partial fusion of C1-C2 vertebrae, hemi vertebra with fusion of right C4-5 level along with C3-C4 spinal cord hyperintensities.

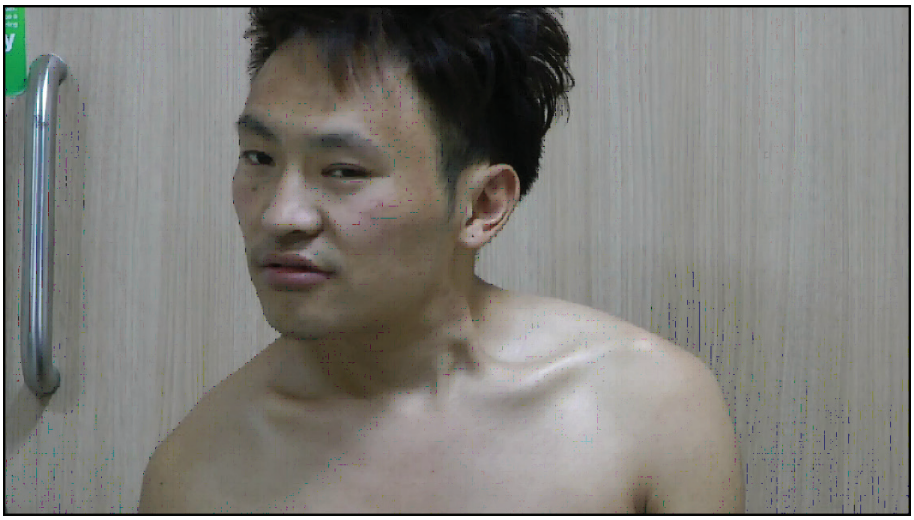
variable, but spontaneous improvement is reported in almost half of the cases.[2–10]

(2) Musculoskeletal

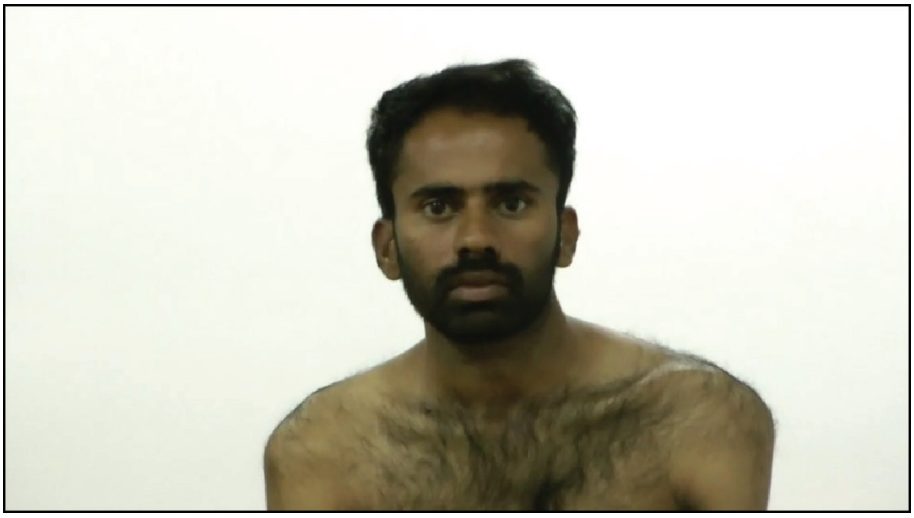
Any anatomical/physiological variations in the cervical musculoskeletal system can present with cervical pseudodystonia. In our series, the possible cause for incorrect diagnosis of cervical dystonia in INEM was additional movements of the facial and neck muscles in an effort to lift the head, giving rise to the interpretation of neck drop as anterocollis. Clinical assessment for muscle strength was the key clinical feature in the diagnosis. Every patient should be evaluated to rule out secondary causes. INEM mimicking cervical dystonia is a rare condition and can pose a diagnostic dilemma. This condition is also known as "dropped head syndrome," which has a wide spectrum of causes.[11] Routine blood, electrophysiological, radiological, and histological investigations help in confirming isolated myopathy. Some of these causes are potentially treatable, including myasthenia, hypothyroidism, hyperparathyroidism, hypokalemia, etc. Management of INEM involves physiotherapy, orthotic collars or braces, and rarely surgical fixation.[12]

Congenital muscular torticollis is another common musculoskeletal cause of cervical pseudodystonia. It is commonly diagnosed during infancy and is caused due to intrauterine malposition or birth trauma. On palpation, the sternocleidomastoid muscle is taut and non-tender. Absence of tremor and sensory trick makes cervical dystonia less likely (see Case 2). There is a restriction in the range of movements of the neck. Some cases resolve spontaneously, while most of them respond well to physical therapy. Physical therapy involves passive stretching and encouraging active movements. Less than 10% of cases require surgery. Early treatment remains the key for better outcomes.[13–15]

Various craniocervical developmental anomalies can also present with cervical pseudodystonia. Klippel–Feil syndrome is a congenital anomaly caused by the fusion of a pair or more of cervical vertebrae. It is characterized by a short neck, torticollis, limited range of movements and low posterior hairline. Symptoms become more pronounced during adolescence or young adulthood. Vertigo, tinnitus, and hearing loss are sometimes associated with this condition. It can masquerade as cervical dystonia, which becomes apparent after radiological evaluation. Treatment is mostly symptomatic. Surgery may be needed if associated



Video 2. **Video Shows Limitation of Rotation of Neck to the Left Along with Right Lateral Shift Torticaput to the Right.** The prominence of sternocleidomastoid at its origin can be easily appreciated on the left side.



Video 3. **Video Shows Right Lateral Shift of Neck Along with Left Shoulder Elevation.** In addition, limitation of rotatory movements of neck to both right and left is shown.

with scoliosis, radiculopathy, or myelopathy. Pseudodystonic posture secondary to Klippel–feil syndrome and diastometamyelia has been reported.[16] Other CVJ anomalies like Chiari malformation and basilar invagination are known to cause torticollis.[17–18] Syringomyelia associated with torticollis is a rare presentation. It requires a high degree of suspicion when head tilt is seen along with sensory loss. Dystonic movements of the rest of the body can also be seen occasionally. An MRI of the spine can confirm the diagnosis. Decompression of the syrinx relieves the symptoms.[19]

Acute, painful torticollis in children with a history of fall or trauma points toward atlantoaxial rotatory subluxation (AARS).[20–22] It can also be due to a local inflammatory process, or connective tissue disorders. A rare case of AARS was seen associated with familial Mediterranean fever (FMF) presenting with torticollis.[23] FMF is an autosomal recessive autoinflammatory condition, presenting with episodes of fever and serositis. Cervical dystonia in ankylosing spondylitis resulting in AARS and craniocervical osseous fusion (CCOF) has been reported.[24] If there is no history of trauma, it is important to look for a history of recent ENT surgeries or any inflammatory conditions of the neck, as it could be Grisel's syndrome, which is a non-traumatic atlantoaxial subluxation resulting from an ongoing local inflammatory process.[25–27] Grisel's syndrome is more commonly seen in children due to laxity of ligaments along with the tenderness of the spinous process, and there may be unilateral occipital pain. A spinal cord injury should be

Table 2. **Cause of potential mimics of isolated idiopathic cervical dystonia Pseudodystonia**

Classification	Causes
Vascular causes	1. Spinal epidural haemorrhage 2. Cerebellar infarct/haemorrhage 3. Lateral medullary infarct 4. Cerebral haemorrhage 5. Bilateral Putaminal haemorrhage 6. Cerebral AVM 7. Unilateral hypoplasia of Internal Carotid artery 8. ACOM 9. Vascular pseudo retrocollis
Musculoskeletal	1. AARS 2. Fibrodysplasia ossificans 3. Acute calcific tendinitis 4. Basillar invagination 5. Chiari 1 malformation 6. Klippel-Fiel syndrome 7. Syringomyelia 8. Diastometomyelia 9. Osteomyelitis 10. Inter vertebral disc calcification 11. Facetal hypertrophy 12. Ankylosing spondylitis 13. Fibromatosis of sternocleidomastoid 14. Nodular fascitis of sternocleidomastoid 15. Congenital oseous c2-c3 synostosis 16. Cervical spondylo discitis 17. Osteoporotic fracture 18. Absent sternocleidomastoid muscle
Infections	1. Septic arthritis 2. Cat scratch disease 3. Tuberculoma 4. Bacterial meningitis 5. Acute febrile torticollis 6. Acute encephalomyelitis 7. Paravertebral Brucellar abscess 8. Cervical epidural abscess 9. Pharyngeal abscess 10. Retropharyngeal abscess 11. Sternocleidomastoid abscess 12. Pyomyositis of paraspinal muscles 13. Lymphadenitis 14. Tuberculosis of bones & joints
Mass / Space occupying lesions	1. Tumor calcinosis of cervical spine 2. Intra thoracic malignancy 3. Posterior fossa tumor 4. Arachnoid cyst 5. Spinal cord ependymoma 6. Cervical osteoblastoma 7. Medullary tumor 8. Posterior glioma 9. Ewing's sarcoma 10. Osteochondroma 11. Giant cell tumor 12. Osteoid osteoma 13. Cervical hemangioblastoma 14. Fibrodysplasia ossificans progressiva 15. Post radiation therapy of carcinoma larynx 16. Cerebellar gangliocytoma 17. Spinal cord astrocytoma 18. Colloid cyst of 3rd ventricle 19. Sternocleidomastoid tumor 20. Cervical eosinophilic granuloma
Traumatic	1. Odontoid fracture 2. Laminar fracture 3. Condylar fracture 4. C1 dislocation with split atlas 5. Fracture of c2 lamina 6. Brachial plexus injury 7. Post trauma foreign body 8. Pneumomediastinum
Ocular Causes	1. Congenital Nystagmus 2. Nystagmus Compensation Syndrome 3. Spasmus nutans 4. Oculomotor apraxia 5. Refractive error 6. Blepharoptosis 7. Superior oblique palsy 8. Abducens Palsy 9. Vertically incomitant horizontal strabismus 10. Duane syndrome 11. Brown's syndrome 12. Double elevator palsy 13. Orbital floor fracture 14. Endocrine ophthalmopathy 15. Congenital fibrosis syndrome 16. Inferior oblique muscle palsy (ocular torticollis)
Otological Causes	1. Acute Mastoiditis 2. Saccular dysfunction 3. Bezold's abscess
Gastrointestinal causes	1. Sandifer's syndrome
Others	1. Multiple sclerosis 2. Acute disseminated encephalomyelitis 3. Idiopathic intracranial hypertension 4. Hypereosinophilic syndrome 5. Widespread nevus spilus 6. Parry-Romberg syndrome 7. Moyamoya disease 8. Behcet's disease 9. Kawasaki disease 10. Goeminne syndrome 11. Langerhans cell histiocytosis 12. Allergy 13. Iatrogenic hypoparathyroidism 14. Congenital muscular torticollis 15. Benign paroxysmal torticollis

Table 2. (*Continued*) **Cause of potential mimics of isolated idiopathic cervical dystonia Pseudodystonia**

Classification	Causes
	16. Grisel's syndrome 17. Complication of ventriculo-peritoneal shunt 18. Familial Mediterranean fever 19. Foreign body
Psychogenic	

Abbreviations: AVM - arteriovenous malformation, ACOM - anterior communicating artery aneurysm, AARS - Atlanto Axial Rotatory Subluxation.

suspected if there are associated neurological symptoms such as unsteady gait or hyperreflexia.[25] Early diagnosis and management is the key. It is usually treated by manipulation under anesthesia and immobilization. For less severe cases, cervical soft collar with rest and analgesia may be sufficient. Delay in the diagnosis can lead to neurological complications and require surgery.[28–30]

Other causes like intervertebral disc calcification, acute calcific tendinitis of longus colli, facetal hypertrophy, and nodular fasciitis of sternocleidomastoid should be considered in the differential diagnosis of musculoskeletal causes of pseudodystonia.[31–34]

(3) Infections

Infections of the upper respiratory tract or soft tissues of the neck can cause torticollis to mimic cervical dystonia. These include cervical adenitis, lymphadenitis, retropharyngeal abscess, and sternocleidomastoid myositis.[35,36] Retropharyngeal abscess is a serious condition presenting with severe pain, fever, and difficulty in breathing and swallowing along with torticollis. It requires immediate treatment with antibiotics, non-steroidal anti-inflammatory drugs (NSAIDs), and sometimes surgical drainage of the abscess. Similarly, paravertebral brucellar abscess and sternocleidomastoid abscess are documented to present as cervical pseudodystonia.[37]

Other reported causes of atypical infections presenting with cervical pseudodystonias include cat scratch disease, encephalomyelitis, bacterial meningitis, neuroborreliosis, tuberculoma, tuberculosis of the bone and joints, septic arthritis, spondylodiscitis, and osteomyelitis.[38–46]

(4) Mass lesions/space occupying lesions

Central nervous system lesions can also cause cervical pseudodystonia, along with symptoms like nausea, vomiting, headache, ataxia, visual disturbances, and cranial nerve deficits. These lesions include posterior fossa and infratentorial tumors (more commonly in the cerebellum, third ventricle, and brainstem) and spinal cord tumors like astrocytomas, medulloblastomas, and ependymomas.[7,35,47,48] Cervical osteoblastoma, hemangioblastoma, osteochondroma, osteoid osteoma, fibrodysplasia ossificans progressiva, giant cell tumor, and Ewings sarcoma can all present with torticollis.[7,49–54] Torticollis in children requires brain and spinal cord imaging to avoid delay in diagnosis, which can be life threatening. Surgical removal of the tumor in most cases results in resolution of the symptoms.

Cervical pseudodystonia has been described in carcinoma larynx (following radiation therapy) due to fibrosis of neck muscles and in a Pancoast tumor of the lung due to possible segmental demyelination of the 11th cranial nerve.[55,56]

(5) Traumatic

Trauma resulting in odontoid fracture, laminar fracture, condylar fracture, osteoporotic fracture, C1 dislocation with split atlas due to various causes can all present as dystonia mimics.[57–61] Appropriate management will alleviate symptoms.

Other traumatic conditions presenting with a similar picture include brachial plexus injury, pneumomediastinum, foreign body, and rarely electrical injury.[62–65]

(6) Ocular causes

Ocular causes like congenital nystagmus, nystagmus compensation (blockage) syndrome, spasmus nutans, oculomotor apraxia, refractive error, blepharoptosis, superior oblique palsy, abducens palsy, vertically incomitant horizontal strabismus, Duane syndrome, Brown's syndrome, double elevator palsy, orbital floor fracture, endocrine ophthalmopathy, congenital fibrosis syndrome, and inferior oblique muscle palsy may present with torticollis called ocular torticollis. Abnormal head position is assumed in order to maintain binocularity and/or to optimize visual acuity. Bielschowsky head tilt test is the primary test in the ocular torticollis workup. Treatment is usually surgical and depends on the underlying cause.[66]

(7) Otological causes

Cervical pseudodystonias secondary to vestibular dysfunctions Contribute to otological causes. These patients have ataxia, vertigo, or nystagmus along with torticollis. Acute mastoiditis, saccular dysfunction, and Bezold's abscess are few examples. This requires further assessment of the vestibular system and management.[67–69]

(8) Gastrointestinal causes

In a child presenting with torticollis or side-to-side head movements associated with vomiting, regurgitation, or epigastric pain, Sandifer's syndrome can be suspected. The symptoms can be intermittent and are associated with meals in most cases. The symptoms are probably due to the patient assuming a position to minimize the painful acid reflux. As the early symptoms resemble dystonia, evaluation is focused on neurological etiology, which is usually normal. Diagnosis is by monitoring esophageal pH and demonstrating reflux. Medical treatment for gastroesophageal reflux usually resolves the symptoms, but sometimes this condition requires surgery.[70,71]

(9) Psychogenic dystonias

Psychogenic dystonia, also known as functional dystonia, is a controversial diagnosis commonly associated with psychiatric comorbidities

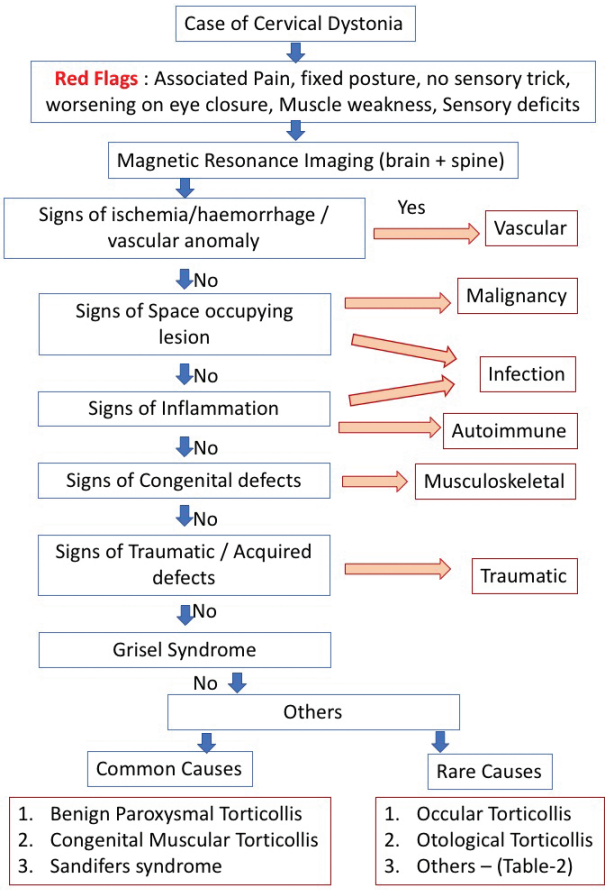


Figure 2. Algorithm for Mimics of Isolated Cervical Dystonia.

like anxiety, depression, and personality disorders. Variability of symptoms suggests a psychogenic cause. The current classification has suggested categorization of psychogenic dystonias as one of the acquired dystonias.[1] Although prognosis can be poor with long-term disability, mainstay of management involves communicating the diagnosis with the patient, physiotherapy, and behavioral therapy.[72,73]

(10) Others

There are various other causes of cervical pseudodystonias, which are either rarely reported or documented as single association. Among these, benign paroxysmal torticollis (BPT) of infancy is a self-limiting condition, characterized by periods of unusual, sustained posture of the head and neck, during which the head tilts to one side. Episodes are often accompanied by marked autonomic features, irritability, ataxia, apathy, and drowsiness. They last several hours to a few days and often recur every few weeks. They subside within the pre-school years. It is essential to recognize this condition and to reassure parents of its benign course and not to be misdiagnosed for other disorders, such as epileptic seizures. BPT of infancy has been linked to CACNA1A mutations and are likely to be associated with familial hemiplegic migraine, episodic ataxia, and paroxysmal tonic upgaze.[74,75] There is no approved medication for the disease. Some studies have demonstrated the uses of cyproheptadine and topiramate for BPT.[76]

Other rare neurological causes presenting as cervical pseudodystonia include idiopathic intracranial hypertension, acute demyelinating encephalomyelitis, multiple sclerosis, Guillain–Barre syndrome, hypereosinophilic syndrome, widespread nevus spilus, Parry–Romberg syndrome, moyamoya disease, atypical Kawasaki disease, Behcet's disease, Langerhans cell histiocytosis, Goldenhar syndrome, Goeminne syndrome, iatrogenic hypoparathyroidism, and allergy.[77–92] These conditions have been reported in isolated case reports.

Conclusion

In any case of presumed cervical dystonia, a diligent clinical evaluation is a major step in diagnosis and management. The examination must include a complete neurological assessment, including the power and range of movements of the neck. It is also critical not to forget the examination of the ocular and vestibular system, and the local examination of the neck for any obvious deformities. Imaging must include an X-ray of the neck for bony deformities and an MRI of the brain and neck to rule out the possibility of tumors and other spinal lesions. We have provided a simple algorithm for workup for suspected cases of cervical pseudodystonia (Figure 2).

In patients with atypical features, a multi-disciplinary approach helps in arriving at an accurate diagnosis (pediatricians, rehabilitation experts, ENT surgeons, ophthalmologists, gastroenterologists, neurosurgeons, movement disorder specialists, and others). Dystonia mimic or pseudodystonia is an under reported entity in our movement disorder literature; this review highlights the varied causes and will help in diagnosing this chameleon-like entity and directing treating physicians toward the right intervention.

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