A 14-YEAR-OLD BOY WITH NEUROGENIC THORACIC OUTLET SYNDROME:  
A CASE REPORT

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ABSTRACT

Background: Thoracic outlet syndrome (TOS) is a complex condition that involves the neurovascular bundle located between the scalene triangle and the inferior border of the axilla. TOS represents a wide variety of clinical symptoms and there is no single test available for establishing the definitive diagnosis. This case report aims to report the clinical history, clinical features, imaging findings, and treatment of a patient with neurogenic TOS secondary to suspected cervical muscle compression due to cervical dextroscoliosis, accompanied by patent ductus arteriosus.

Case Presentation: A 14-years-old boy reported progressive weakness in his right hand. The patient also complains of headaches and tiredness every time after exercise since 3 years ago and progressive loss of body weight. Physical examination revealed a lower motor neuron type of superior paraparesis with gradual...
progressive onset on the right upper limb, accompanied by weakness of the deltoid, biceps, triceps, and finger muscles. Adson’s test of the right upper limb showed positive results. MRI and CT scan of the spine showed a cervical dextroscoliosis causing a mild bilateral neural stenosis and minimal right thoracic scoliosis. A continuous murmur grade 3/6 intercostal space II-III was heard on heart auscultation. Echocardiography showed a patent ductus arteriosus. The patient was scheduled for electromyography and nerve conduction velocity test.

**Discussion:** Neurogenic TOS is a chronic compressive brachial plexopathy where the brachial plexus is pressed anteriorly and upwardly by the cervical rib arising elongated transverse process of C7, with the symptoms characterized by pain, tingling, numbness, or motoric weakness in the upper or lower arm. Multiple tests and examinations are needed to establish a definitive diagnosis. Treatment available for neurogenic TOS included conservative treatment with physiotherapy for posture and shoulder movement correction.

**Conclusion:** TOS is a condition that could cause a variety of symptoms. Appropriate investigations and multiple diagnostic tests are needed for establishing the diagnosis. Patient’s history, clinical presentation, and examination could help eliminates the differentials, with imaging and electrophysiological testing helps identify the etiology.

**Keywords:** Thoracic outlet syndrome, patent ductus arteriosus, scoliosis
INTRODUCTION

Thoracic outlet syndrome (TOS) is a complex condition that involves the neurovascular bundle located between the scalene triangle and the inferior border of the axilla. The clinical manifestations of TOS are based on the structures that are affected, including pain, paresthesia, numbness, motoric muscle weakness in the upper limb, swollen or cyanotic upper limb, enlarged veins in the shoulder, cold and pale hand, or pain elicited by overhead arm motions. TOS could often be confused with other conditions that have similar presentation such as brachial plexopathy, cervical radiculopathy, and complex regional pain syndrome.

TOS has most consistently been diagnosed based on a complete history, clinical presentation, physical examination, and radiographic imaging studies with electrophysiological investigations. Currently, there is no standard approach for establishing the diagnosis of TOS. While efficacy of conservative treatment for TOS is still weak due to lack of evidence. This case report aims to report the history, clinical features, imaging findings and treatment of a patient with neurogenic TOS secondary to suspected cervical muscle compression due to cervical dextroscoliosis, accompanied by patent ductus arteriosus.

CASE REPORT

A 14 years-old boy was admitted to Nusa Tenggara Barat Hospital with weakness in the right hand since 2 months before admission. The nature of the weakness was progressive. The patient also complains of headaches and tiredness every time after exercise since 3 years ago and progressive loss of body weight (2.2 kg in 3 months). The patient’s vital signs are within normal limits. Physical examination found a lower motor neuron type of superior paraparesis with gradual progressive onset on the right upper limb, accompanied with weakness of the deltoid, biceps, triceps, and finger muscles. Adson’s test of the right upper limb showed positive results. Neurological examination revealed an increase in the right biceps and triceps reflexes.

Spinal palpation revealed right cervical scoliosis on the level of C3, with MRI imaging showing a cervical dextroscoliosis, hypertrophy of the facet joint, and thickened of flavum ligament, causing a mild bilateral neural stenosis (Figure 1). CT scan of the thorax showed minimal right thoracic scoliosis at the level of Th2 (Figure 2).

Thorax physical examination revealed a thrill on palpation at the fifth intercostal space on the left midclavicular line. First and second heart sounds were normal; a continuous murmur grade 3/6 intercostal space II-III was heard. Echocardiography showed a patent ductus arteriosus 4 mm in diameter with a left to right shunt and LVEF 78%.

The patient was diagnosed with thoracic outlet syndrome at the level of interscalene triangle and was prescribed with methylprednisolone 3x24 mg, furosemide 1x20 mg, and captopril 3x62.5 mg.
For further management, the patient was scheduled for an electromyography and nerve conduction velocity (EMG-NCV) test.

Figure 1. Cervical vertebra MRI showed cervical dextroscoliosis with mild bilateral neural stenosis.

Figure 2. Thoracal CT Scan showed a minimal right thoracic scoliosis at the level of Th2.

DISCUSSION

TOS is a complicated condition that affects the neurovascular bundle located between the scalene triangle and the axilla’s inferior border. TOS could be classified into 2 major types based on the structure affected, neurogenic thoracic outlet syndrome, a chronic compressive brachial plexopathy where the brachial plexus is pressed anteriorly and upwardly by the cervical rib arising elongated transverse process of C7, that accounts to 90-95% cases, and vascular thoracic outlet syndrome that consists of venous thoracic outlet syndrome and arterial thoracic outlet syndrome which comprises 3-5% and 1-2% cases, respectfully.5
TOS symptoms could be grouped as neurologic and vascular symptoms. Neurologic symptoms are more common in patients and are characterized by pain, tingling, numbness, or motoric weakness in the upper or lower arm. Vascular symptoms caused by vascular obstruction that manifested as cyanosis, pallor, coldness, and early fatigability, hence clinical presentation becomes a critical part of diagnosing TOS.² Due to the non-specific nature of this condition, the diagnosis of TOS should be accompanied with several differential diagnoses. Imaging tools such as MRI and EMG are useful modalities in ruling out the causes and therefore eliminate the differentials.⁶

Our patient had more dominant neurological than vascular symptoms. Patients with this symptom often varied, where some patients could experience mild discomfort while others experienced chronic pain. Patients with true or classic neurogenic TOS are usually younger, skinny, athletes that utilize scalene muscle, and Galliat-Summer hand, which is a sign of abductor pollicis brevis muscle atrophy and lesser atrophy of the interosseous and hypothenar muscles.⁷ Our patient only has movement restriction in the arm, that if not treated properly could progress into this condition.

Adson test on the patient revealed a positive result showed by a reduction of the radial artery pulsation when the right arm was extended, abducted, and externally rotated. This test is a part of classic provocative maneuvers for TOS, where studies have shown a sensitivity of 72%, specificity of 53% and 77% false positive; therefore, this tests may be unreliable since ischemic symptoms vary depending on chronicity and duration of ischemia, and so clinicians needed further examinations for diagnosing the condition.⁸

Spine physical examination revealed right cervical scoliosis at the level of C3, which was confirmed by MRI imaging, and causes a mild bilateral neural stenosis. Thoracal CT scan also revealed right thoracal scoliosis at the level of Th2. Based on the literatures, pediatric patient should be referred to a specialist if the scoliosis curve exceeds 10 degrees in patients younger than 10 years old, or more than 20 degrees in patients older than 10 years old, with atypical features or related to a back pain or neurologic disorders.⁹ Our patient experienced a neurological deficit which result in the suspicion of TOS due to structural anomalies on the cervical vertebra bone, transverse process of C7, scalene muscle hypertrophy, fibrous tissue and sibson’s fascia anomaly.¹⁰

Cardiac physical examination revealed a continuous murmur grade 3/6 intercostal space II-III with echocardiography revealed a 4 mm patent ductus arteriosus with a left to right shunt and 78% LEVF. PDA is one of the most common congenital heart diseases which 90% of the cases are usually closed spontaneously. The failure of closure causes left to right shunt under systemic pressure, where this condition becomes symptomatic when pulmonary vascular pressure and resistance increases.¹¹
The patient was diagnosed with thoracic outlet syndrome at the level of interscalene triangle secondary to suspected cervical muscle compression due to cervical dextroscoliosis, based on the diagnostic algorithm which consists of clinical features, positive provocative maneuver tests and confirmation from x-ray, CT scan, and MRI imaging. The patient was scheduled for EMG-NCV, where studies have found that slow conduction velocity in patients with TOS could be manifested to cutaneous nerve. EMG result indicates a lower brachial plexopathy, cervical anomaly, and C7 transverse process elongation.  

Effective management of TOS requires early recognition and elimination of other differential pathologies. Usually, initial management of neurogenic TOS consists of conservative treatments, while arterial and venous TOS most often needed surgical intervention due to physical therapy not being helpfull.  

The patient was prescribed with methylprednisolone for improving neurologic outcome. Furosemide was given for maintaining the patency of ductus arteriosus, and captopril for reducing the aorta pressure and systemic vascular resistance, also reducing the pressure on the left and right atrium in patients with congenital heart disease. The patient currently has physiotherapy scheduled, with the aim for back and scalene muscles movement correction, in the hopes of improved posture. Although few studies have shown an improvement with conservative care, the results vary. The uncertain diagnostic criteria and the lack of a “gold standard” limits the generalizability and clinical utility of the studies.

**CONCLUSION**

TOS is a condition that could cause a variety of symptoms. Appropriate investigations are needed to evaluates the patient’s condition and multiple diagnostic tests are required for establishing the diagnosis. Patient’s history, clinical presentation, and examination could help eliminates the differentials, with imaging and electrophysiological testing allowing for helping identify the etiology. Effective management requires early recognition and is tailored to the patient’s condition.

**REFERENCE**

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