

SYMPTOMATIC CERVICAL RIB IN CHILDHOOD*

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The term "thoracic outlet syndrome" (TOS) was first popularized by Rob and Standeven¹ in 1958, as a collective term including some previously described syndromes such as scalenus anticus, cervical rib, costoclavicular, hyperabduction and shoulder-girdle compression syndromes.

Symptomatic TOS is common in females (female/male ratio: 1.5/1) and encountered especially between the third and fifth decades of life²⁻⁴. It is quite unusual in the first decade of life and commonly indicates the presence of a congenital fibromuscular band or a cervical rib⁵.

Case Report

H.K., a 6-year-old girl, was referred to the Hacettepe Children's Hospital with swelling in the left supraclavicular region and complaints of fatigue in the left arm while playing with her friends. The swelling had increased during the preceding year. Numbness and paresthesia of the left arm on elevation were also described.

Tests for the thoracic outlet syndrome performed during the physical examination were non-contributory. Neither vascular nor muscular disturbances were noted. X-rays revealed a complete left cervical rib that was connected to the first thoracic rib (Figures 1 and 2).

The patient underwent an operation on February 27, 1981, whereby the left cervical rib was removed completely using a supraclavicular approach. The scalenus anticus

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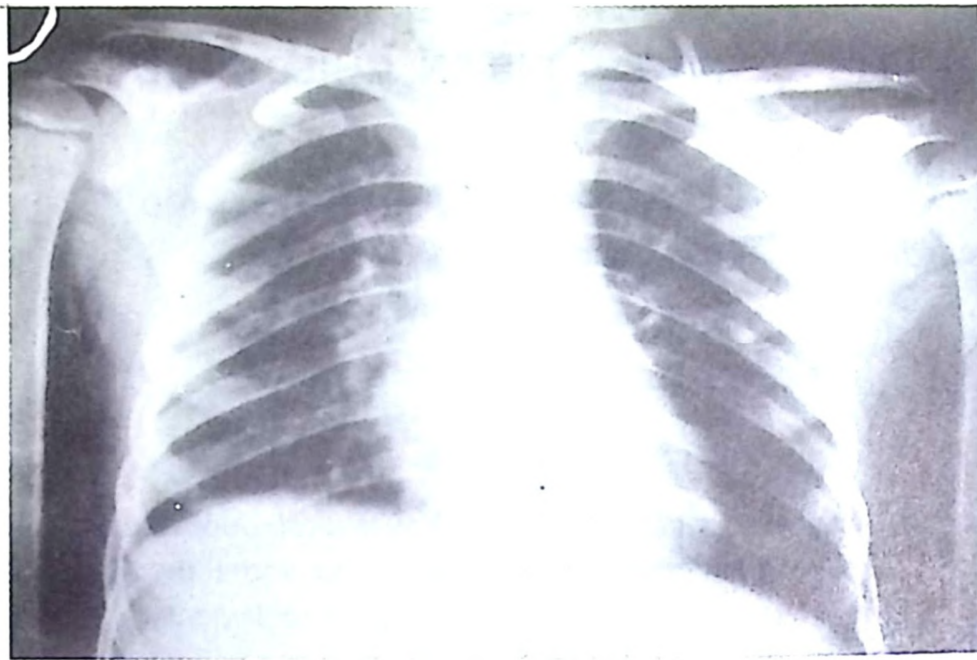


Figure 1.

A complete cervical rib on the left side is shown.

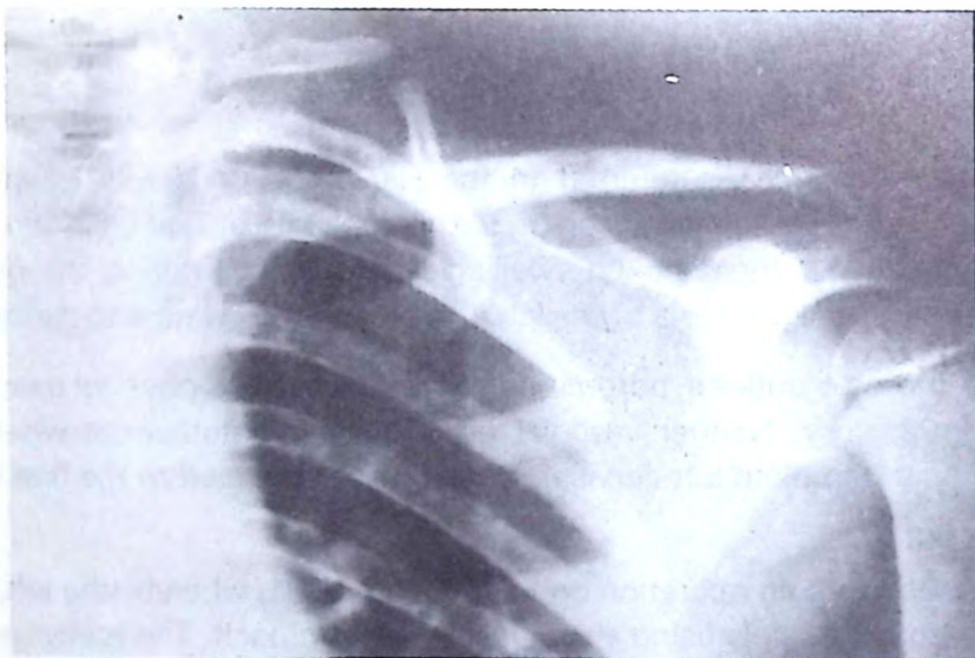


Figure 2.

Close-up view of the left cervical rib.

muscle was also divided at its insertion on the first thoracic rib. Upon pathological examination, osteochondroma of the cervical rib was found.

A follow-up examination 9 months after the operation showed complete recovery with no recurrence of the preoperative symptoms.

Discussion

Dale and Lewis⁴ cite five factors mentioned by Lord and Rosati as causing compression of the thoracic neurovascular bundle:

1. Congenital anomalies,
2. Static size and shape of compressing structures,
3. Dynamic changes with motion,
4. Trauma (fractures, automobile whiplash accidents, etc.),
5. The aging processes of arteriosclerosis and muscle atrophy.

Roos⁵ pointed out the predisposing underlying anatomic abnormalities in cases with TOS which are not present in the general population.

Compressing elements consist mainly of bony structures (cervical rib, clavicle, bifid first rib, and fusion of the first and second ribs), muscles (scaleneus anticus, subclavius, and pectoralis minor), and fibromuscular bands³.

A cervical rib is said to be present when the transverse process of the seventh cervical vertebra extends beyond the process of the first thoracic vertebra⁵. Cervical ribs may be unilateral or bilateral; unilateral ones are commonly on the left side. A complete cervical rib is a bridge-like bony structure connecting the seventh cervical transverse process and the first thoracic rib. Incomplete or short cervical ribs have congenital ligamentous bands attaching the cervical rib anlage to the first thoracic rib. These bands are not visible on x-rays, because they contain no calcium⁵. Cervical rib is said to exist in 1% of the general population and to be symptomatic in only 10% of the involved subjects⁴.

Symptomatology in TOS is outlined in Table I²⁻⁵. Symptoms are not severe in the majority of the cases, and only a small number of individuals with TOS display incapacitating symptoms requiring surgical intervention. Roos⁵ evaluated more than 2300 cases of TOS and operated on only 776 of them (33.7%). Mildly and moderately symptomatic patients respond well to avoidance of aggravating activities, physical therapy and pain medication⁵.

Numerous clinical tests have been described for TOS which are not always helpful for a correct diagnosis⁵. Common lesions to be ruled out consist mainly of cervical root pressure by a disc, arthritis, shoulder bursitis, upper extremity vascular occlusions and the carpal tunnel syndrome⁴. Spinal cord tumors, upper mediastinal tumors (such as lymphoma) and, rarely, neurofibromatosis should also be considered in differential diagnosis in the pediatric age group.

According to the most recent knowledge concerning TOS, many investigators have accepted the following²⁻⁵:

1. Diagnosis is chiefly based upon history of the patient, because physical signs are inconsistent.

2. Angiography is not always necessary, since major vascular problems are unusual.
3. Electromyography is not critical in diagnosis.
4. The majority of patients respond well to non-operative measures, and only a small number of individuals require surgical intervention.
5. The operation of choice, when needed, is resection of the first thoracic rib via a transaxillary approach⁶, removal of the cervical rib when present, plus complete excision of all anomalous fibromuscular tissue around the neurovascular bundle.

Roos⁵ describes the first operation in 1861 for a well-documented case of TOS, removal of a cervical rib by Coote at St. Bartholomew's Hospital in London.

A supraclavicular approach provides easy access for resection of the cervical rib. Results of the operations for TOS are satisfactory; symptoms are completely or partially relieved in 92% of those who have the operation, and poor results are obtained in only 8% of the cases³. Postoperative scar tissue may cause recurrence of the original symptoms in rare instances (0.3% in a large series)⁵, which may require re-operation.

TABLE I

Signs and Symptoms in Thoracic Outlet Syndrome*

I. Compression of the Brachial Plexus:

1. Pain (neck, shoulder, arm)
2. Numbness
3. Paresthesia
4. Weakness
5. Muscular atrophy of the hand

(Numbness and paresthesia occur commonly along the distribution of the ulnar nerve, to the fourth and fifth fingers.)

II. Arterial Occlusion:

1. Pain, weakness, coldness and easy fatigability of the arm
2. Dampened or absent radial pulse
3. Ulceration and gangrene
4. Reynaud's phenomenon

III. Venous Obstruction:

1. Aching, tired arm with swelling
2. Cyanosis and edema
3. Distended collateral veins (around the shoulder and anterior chest wall)
4. Effort thrombosis (Paget-Schroetter syndrome)

* A great number of cases present primarily with neurological symptoms. Major vascular signs and symptoms are very infrequent.

Summary

A six-year-old girl with symptomatic left cervical rib is presented, where complete recovery was observed after removal of the aberrant rib via a supraclavicular approach. It is also of interest that pathological examination of the resected rib revealed osteochondroma. This is an unusual case of thoracic outlet syndrome, since symptomatic cervical ribs occur mostly between the third and fifth decades of life and are quite uncommon in the first decade.

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