Thoracic Outlet Compression Syndrome: A Rare Cause of Upper Extremity Symptoms in Children

John Shepherd, MD

Children frequently complain of upper extremity pain often secondary to trauma, yet rarely do they suffer from thoracic outlet compression syndrome (TOCS). Through 1997 the literature contains only 6 reported cases: 4 in the United States, 1 in Turkey, and 1 in Italy.[1–3] Although family physicians need to focus on the common causes of symptoms, we must consider the unexpected possibilities. Fortunately, we can use our knowledge of adult medicine to recognize unusual pediatric presentations.

Case Report

An 11-year-old boy came to the clinic for a fall sports physical examination for football in August of 1997. His medical history was notable only for a 9-year history of maternal concern regarding diminished use of his left hand, specifically the third through fifth digits. He stated that his left upper extremity felt "cold, heavy, and tired," particularly along the medial elbow. He also reported weakness in his left hand.

He was 57 inches tall and weighed 92 pounds. His blood pressure in both upper extremities was 108/60 mm Hg, his heart rate was 86 beats per minute, and his respiratory rate was 20/min. When examined, he had a hard 3 × 5-cm protuberance in his left supraclavicular fossa without appreciable neck adenopathy. Neurologically he was found to have diminished strength in his entire left upper extremity compared with the right, although bilaterally he had symmetrical muscle mass above and below the elbow. During his peripheral vascular examination he had positive Roos (3-minute abduction with exercise) and Wright (hyperabduction) tests and a negative Adson test (monitoring the radial pulse while inhaling and turning extended head toward the involved extremity). The last radiograph of his neck taken in 1996 pictured bilateral large cervical ribs with callus formation on the mid left cervical rib. Electromyographic studies were ordered to rule out other neurologic causes of upper extremity weakness and paresthesias. These studies of both upper extremities showed only prolonged latency in the ulnar sensory distribution of the left hand. The pediatric neurologist, who performed the test, reported decreased two-point discrimination and slightly decreased sensation in the left hand.

A review of the boy's medical records showed that at 3 years of age he saw a provider who noted a prominent left supraclavicular mass and ordered a chest radiograph. The radiology report noted an "unusual anomalous left first rib" with no other abnormalities. The primary provider attributed the neck lump to this anomalous rib.

One and one-half years later, the records indicated continued concern about left hand disuse and weakness. No workup ensued at that time. Five years after that report, a school examination again elicited the same complaint. Although the neurologic examination findings did not appear consistent with the symptoms, another provider referred the now 9-year-old to physical therapy. The physical therapist's evaluation recorded "below average strength in the left hand." The boy pursued physical therapy and occupational therapy for 2 months before "insurance problems" interrupted treatment.

Seven years after the initial radiograph, another provider ordered the 1996 film, which the boy's mother brought to our office. This radiological report noted an "abnormality of the left first rib which appears to articulate with the second rib with some type of pseudoarthrosis. This is not a cervical rib." At this time the boy resumed hand therapy

Submitted, revised, 12 August 1998.
From the Department of Family Medicine, University of Colorado, Denver. Address reprint requests to John Shepherd, MD, Department of Family Medicine, University of Colorado, Health Sciences Center, Campus Box B155, 1180 Clermont Street, Denver, CO 80220.

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and attended 14 sessions. The discharge summary reported that his left upper extremity strength matched the right except for wrist flexion and hand intrinsic function. Decreased hand sensation was also noted.

After his football physical examination, which occurred 1 year after his last therapy, the boy had 2 more months of occupational therapy in an attempt to avoid a possible sports injury resulting from his muscular imbalance. This treatment decreased the symptoms and increased the boy's strength, although his left upper extremity did not equal the right according to the therapist's report. The neurologist who performed the electromyogram then recommended evaluation by a thoracic surgeon because the arterial signs persisted after therapy. The surgeon ordered Doppler studies of the left upper extremity and interpreted the test as showing relatively reduced finger pressures in the left hand and relatively reduced pulse volume recordings in the left forearm, but both well above any suggestion of ischemia.

The combination of clinical and laboratory findings and the incomplete response to therapy lead to the surgeon's recommendation for resection of at least the left cervical rib with intraoperative evaluation of the vascular system. The boy and his family chose this approach. Using an anterior supraclavicular incision, the surgeon removed the left cervical rib and reported obvious compression of the plexus of nerves, but no pressure upon the subclavian vessels. Two months after the operation the boy experienced complete resolution of his pain and paresthesias with gradual increasing strength in his left hand.

Discussion

Since Galen first described the cervical rib in 150 AD and Peet introduced the term thoracic outlet syndrome in 1956, practitioners have long recognized the vulnerability of the upper extremity for neurovascular pressure compromise created by modification of the narrow anatomic space known as the thoracic outlet.[4,5] This region, defined by the anterior and middle scalene muscles and the first rib, can suffer compression by bony and soft tissue impingement. Both the costoclavicular space lateral to the scalene muscles and the subcoracoid space beneath the pectoralis minor muscle insertion into the coracoid process also provide possible sites for compression of the brachial plexus and subclavian vein and artery. Just as in adults, who usually have this problem, children with the complications of thoracic outlet compression syndrome suffer from persistent brachial plexus pressure leading to paresthesias and pain and muscle weakness and atrophy. Venous obstruction, however, which occurs in less than 5% of all adult cases, and arterial damage, such as stenosis and aneurysm formation, which appears in even fewer cases, have not occurred in children.[4]

Although the paucity of cases in the pediatric age group (seven with this report) makes generalization difficult, neurologic symptoms dominate, particularly the complaint of "tiredness, coldness, and heaviness" of the involved extremity. Six of the 7 children had cervical ribs, whereas anatomists have found cervical ribs in only 1% of the population and in 6% to 11% of persons with thoracic outlet compression syndrome.[6] Girls with this problem have outnumbered boys in the US literature, and symptoms have appeared frequently during a time of rapid growth (12 to 13 years).[1–3]

The literature is replete with opinions on the diagnosis and treatment of thoracic outlet compression syndrome, with the neurologists and the surgeons frequently disagreeing.[6,7] The physical examination seems to provide the confirmatory data in all the pediatric cases. In a group of 100 normal subjects, however, false-positive vascular signs existed with 13.5% having a positive Adson test and 57% displaying a positive Wright test or hyperabduction maneuver.[8] In addition, abnormal neurologic findings occurred in 2% of the group after the Adson maneuver and in 16% after the Wright maneuver. Conversely, the electromyogram can give false-negative results because of the possible transient nature of the nerve bundle compression.[8,9] Because US pediatric cases did not show vascular obstruction, vascular studies might also have limited applicability in diagnosing thoracic outlet compression syndrome because their value is not established.

The differential diagnosis in both adults and children includes cervical spine injury as well as cervical disc disease, reflex sympathetic dystrophy, and conversion reaction, with supraclavicular fossa lesions, such as vascular malformations and tumors, unlikely in the pediatric group. Other nerve compression syndromes such as anterior interosseous, pronation, posterior interosseous, radial tunnel,

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carpal tunnel, ulnar tunnel, and cubital tunnel syndromes can also mimic diagnosing thoracic outlet compression syndrome.

Treatment for all age groups includes adequate occupational and physical therapy to correct soft tissue deficits by improving posture, increasing posterior cervical strength, and stretching anterior neck muscles, and, in children, time to allow growth and possible enlargement of the thoracic outlet.[10,11] If this conservative approach fails to alleviate the symptoms after the recommended 2 years, then surgery is recommended to remove the cervical rib or the first rib and release the scalene muscles. This procedure has successfully treated two of the previous four US pediatric cases and appears to have relieved the problem in this case.[1] Seventy percent of adults note improvement with surgery, but only 20% to 30% become completely asymptomatic.

While a rare finding, thoracic outlet compression syndrome does occur in children, and family physicians should consider this diagnosis in pediatric upper extremity complaints. Consistent and persistent nerve root symptoms with or without vascular findings support the diagnosis of thoracic outlet compression syndrome. Of course, as with most diagnoses, unusual problems become more apparent with additional information and time. The Marcel Proust quote that "the real voyage of discovery consists not in seeking new landscapes, but in having new eyes" describes the daily diagnostic dilemmas faced by family physicians.

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