Thoracic outlet syndrome in adolescents: a case series

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ABSTRACT

Objective. Thoracic Outlet Syndrome (TOS) is caused by a compression of the brachial plexus and the subclavian vessels in their passage to the upper limb. It mostly occurs in women aged 20-50, so it is infrequent in children. We present our results in the diagnosis and management of pediatric TOS.

Materials and Methods. Retrospective study of patients diagnosed with TOS between December 2017 and June 2018. Clinical, radiological, surgical, and evolution variables were assessed.

Results. Five TOS were diagnosed in 4 patients – one TOS was bilateral. Mean age at diagnosis was 12.5 years (7-15), and there was a delay in diagnosis of 153 days (10-36). TOS was either venous (3) or neurogenic (2). Patients presented with pain (5/5), edema (4/5), hypoesthesia (3/5), decreased strength (3/5), and cervical pain (2/5). One patient presented with sport-related pain. Neurophysiological study was normal in three cases. Two patients presented bone anomalies at CT-scan. Three surgeries were performed in two patients using the supraclavicular approach with resection of the anomalous first rib and scalenectomy. One patient refused surgery, and another patient remained expectant without reappearance of symptoms. Postoperative follow-up was 9 months (6-12), with progressive improvement of symptoms.

Conclusions. TOS may occur in adolescents in the form of upper limb pain and edema. Imaging tests are recommended to detect abnormal anatomical structures. The supraclavicular approach represents a safe and effective technique in decompressing the thoracic outlet.

KEY WORDS: Thoracic outlet; Adolescents; Cervical rib; First rib resection.

SÍNDROME DEL OPÉRCULO TORÁCICO EN ADOLESCENTES: SERIE DE CASOS

RESUMEN

Objetivo. El síndrome del opérculo torácico (SOT) está causado por una compresión del plexo braquial y vasos subclavios en su paso hacia la extremidad superior. Patología típica de mujeres entre 20 y 50 años, que es infrecuente diagnosticar en niños. Presentamos nuestros resultados en el diagnóstico y tratamiento del SOT pediátrico.

Material y métodos. Estudio retrospectivo de pacientes diagnosticados de SOT entre diciembre 2017 y junio 2018. Se analizaron variables clínicas, radiológicas, quirúrgicas y de evolución.

Resultados. Cinco SOT fueron diagnosticados en cuatro pacientes, uno de ellos bilateral. La edad media al diagnóstico fue de 12,5 años (7-15) y hubo una demora en el diagnóstico de 153 días (10-360). SOT venoso (3) y neurológico (2). Presentaron dolor (5/5), edema (4/5), hipoestesia (3/5), disminución de fuerza (3/5) y dolor cervical (2/5). Una paciente presentaba dolor asociado al deporte. El estudio neurofisiológico fue normal en tres casos. Dos pacientes presentaron anomalías óseas por TAC. Se realizaron tres intervenciones quirúrgicas en dos pacientes por abordaje supraclavicular realizando resección de la primera costilla anómala y escalenectomía. Una paciente rechazo la intervención y en otra se mantuvo en una actitud expectante sin reaparición de los síntomas. Seguimiento posoperatorio de 9 meses (6-12) con mejoría progresivas de los síntomas.

Conclusión. El SOT puede darse en adolescentes siendo el dolor y edema de la extremidad superior lo más específico. Se recomienda la realización de pruebas de imagen para detectar estructuras anatómicas anómalas. El abordaje supraclavicular se presenta como una técnica segura y eficaz en la descompresión del desfiladero torácico.

PALABRAS CLAVE: Opérculo torácico; Adolescentes; Costilla cervical; Resección primera costilla.

INTRODUCTION

Thoracic outlet syndrome (TOS) is a condition of the cervical vascular nervous bundle widely known and described in the adult population. There are a dozen clinical cases and case series in the pediatric population, the

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Table 1. Clinical findings.

	Age at diagnosis (years)	*Diagnostic delay (days)*	*Sex*	*Side*	*Type*	*Clinical signs*	*CT-scan*	*Treatment*	*Follow-up (months)*
1	13	360	W	Left	N	Pain, edema, cyanosis, hypoesthesia with thermal changes	Abnormal scalene muscle insertion	Surgical	6
2	7	10	W	Left	V	Sudden episodes of pain, edema, paresthesia	1st and 2nd left rib fusion	Conservative	9
3	15	90	W	Bilateral	V	Bilateral cervical pain, edema, cubital hypoesthesia, decreased strength	Abnormal bilateral thoracic rib implantation	Surgical	12
4	14	90	W	Left	V	Pain + edema	/	Conservative	9

W: woman; N: neurogenic; V: venous.

largest one being Maru et al.[1]. The axillary-thoraco-cervical region is a complex anatomical region hosting the vascular nervous bundle of the upper limbs. Three different anatomical narrowing sites can be identified. They include, from proximal to distal: the upper thoracic opening, the scalene hiatus, and the costoclavicular space[2]. Bundle compression at any of these sites generates a single symptomatology encompassed within TOS. The compressive effect can be due to the presence of: a) bone malformations (prominent cervical rib or transverse apophysis), b) fibrotic bands, or c) muscle hypertrophies involving either the scalene, the subclavian, or the lower pectoral muscles. Symptoms vary according to the element of the bundle involved. TOS can be divided into two large groups: neurogenic and vascular. Neurogenic TOS typically occurs in the adult population and represents 90% of cases. It is associated with a repeated use of the upper limb in individuals participating in occupational or leisure activities. It also occurs following traumatic lesions or associated with predisposing anatomical factors. It causes muscle weakness, paresthesia, and pain. Complementary test findings are usually unspecific. In children, vascular compressions are mostly caused by bone malformations occurring at puberty. Arterial compression brings about muscle coldness and weakness, whereas venous compression causes edema, skin de-coloration, and pain. Radiological studies are diagnostic in most cases. In clinical practice, diagnosis proves complex, since it typically occurs in adolescents with unspecific symptoms associated or non-associated with sports. In addition, specialists are not familiar with this pathology. Definitive treatment is surgical, with various approaches described and success rates of around 100% in arterial TOS, 95% in venous TOS, and 90% in neurogenic TOS after 6 months [1]. We present a review of our experience in TOS diagnosis and treatment in pediatric patients.

MATERIALS AND METHODS

A retrospective review of cases from our healthcare facility between December 2017 and June 2018 was carried out. Patients under 18 years of age with TOS-suggestive symptoms and exploration, positive Wright's and/or Adson's provocative tests, and characteristic radiological and neurophysiological findings such as presence of bone abnormalities were included. Demographic data, risk factors, clinical signs, complementary tests, intra-operative findings, and long-term consultation follow-up were collected. Imaging studies were interpreted by an expert radiologist in pediatric thoracic pathologies. Owing to the small number of patients, data are presented with descriptive statistics measures.

RESULTS

Five TOS were diagnosed, one of them bilateral, in four female patients with a mean age at diagnosis of 12.5 (7-15) years (Table 1). Four TOS were venous and one TOS was neurogenic. All patients had been assessed at least by one specialist other than the pediatric surgeon –in some cases three–, which led to a delay in diagnosis of 153 (10-360) days. In patient 1, symptoms started with physical activity. None of the patients had previous trauma history or suspicious data of thrombophilia. Regarding special patient features, patient 2 had KBG syndrome, characterized by facial dysmorphia, upper central incisor macrodontia, skeleton abnormalities (costovertebral primarily), and delayed development, whereas patient 3 had maternal history of bilateral arterial TOS operated on when she was young. The most frequent symptom was pain (5/5), followed by edema (4/5), hypoesthesia (3/5), decreased strength (3/5), and cervical pain (2/5). Provocative tests, such as Wright's

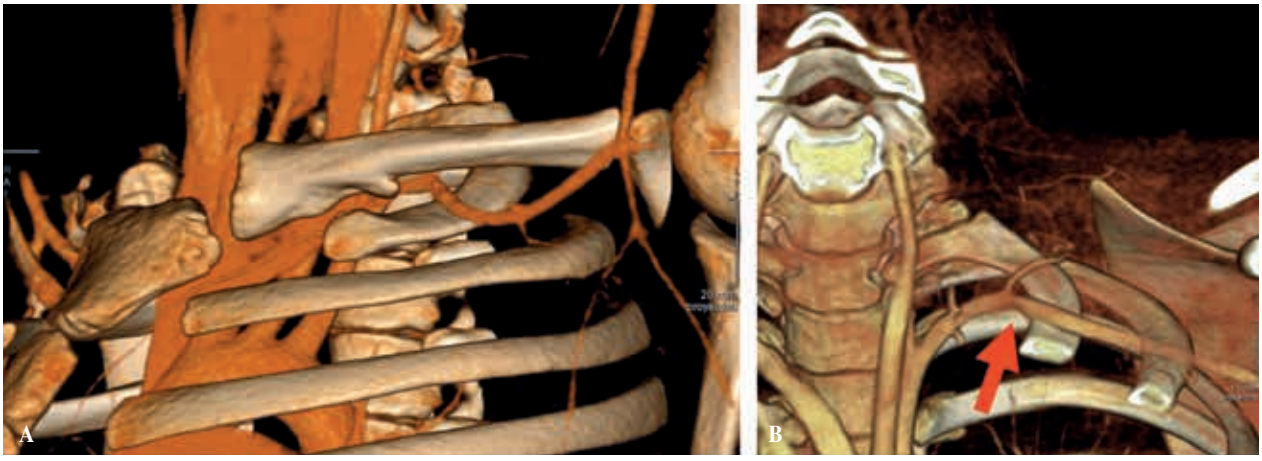

Figure 1. A) and B) 3D vascular and bone reconstructions. Clavicular bone spur. Red arrow: left subclavian artery decompression point.

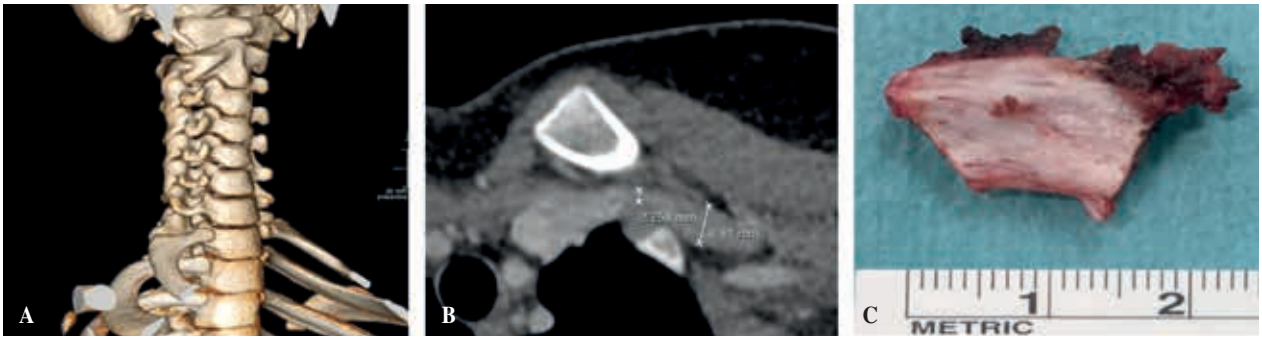

Figure 2. A) 3D axial skeleton reconstruction B) CT-scan axial slice. Subclavian vein sharpening in its passage through the costoclavicular space. C) Costal resection specimen.

test –hyperabduction of the upper limb– and Adson's test –aimed at detecting differences in radial pulse at abduction, extension, and external rotation while the head is being turned to the ipsilateral side– were negative. Doppler ultrasound tests were performed to check for vessel permeability, and a CT-angiography was carried out to analyze the relationship between structures (Figs. 1 and 2). In the neurophysiological study, a mild chronic neurogenic lesion was found at C8 and D1 levels in patient 1, with no other lesions being found in the remaining patients. Conservative treatment consisting of conventional analgesia and rehabilitation was initiated in all patients. Two patients were refractory to treatment, so surgical treatment was decided upon, using a supraclavicular approach under neurophysiological control. Patient 1 had an abnormal insertion of scalene muscles, which were hypertrophic and associated with a bone spur on the internal clavicular side. Patient 2 had a bilateral cervical rib with abnormal insertion. In both cases, first rib resection and scalenectomy were performed. Mean hospital stay was 6.3 (5-8) days without remarkable intercurrent events, except for paresthesia in the cubital region which progressively disappeared. After an external consultation follow-up of 9 (6-12) months, patients undergoing surgery now have the same activity levels they used to before symptom onset and are asymptomatic, with increased muscle strength, even though they occasionally have sporadic paresthesia or edema not limiting their day-to-day life.

DISCUSSION

Typically, our idea about TOS has been based on adult historical series. The first description dates back from the early 20th century. Later, in 1935, Wyllie published the case of a 11-year-old girl with prominent transverse apophysis causing hypertrophy and abnormal insertion of the scalene muscles[3]. It was referred to as *Scalenus Anticus Syndrome* back then. Twenty years later, Rob et al. described it as TOS with venous compression clinical signs[4]. Owing to its anatomical peculiarities, this outlet is the most frequent vascular and nervous entrapment site. The cervical rib (CR)

is an anatomical variant described by Galeno and Vesalio arising from the seventh cervical vertebra as a transverse apophysis and extending beyond the first dorsal vertebra with a freely ending extremity or connected with the first rib. Tomaszewski et al. published a 141-study meta-analysis on cervical rib prevalence and its association with TOS[5]. Prevalence was 1.1% in the healthy population, with a prevalence 25 times higher in TOS patients (29.5%). Only 1 out of 4 had this anatomical variant, but there can be other variants such as first rib fusion in the context of KBG syndrome, such as in patient 2.

Lin et al. published the largest case series of pediatric patients. Of 68 patients, 39 (57%) had venous TOS, 21 (31%) had neurogenic TOS, and 8 (12%) had arterial TOS. 25% developed venous TOS clinical signs associated with sport activities or lesions as a result of repeated and vigorous movements of the limb involved. 4% participated in school music bands or played an instrument. In our series, one patient played basketball, and another patient played the guitar. Similarly to the other case series, incidence was much higher in adolescent women. Therefore, it can be stated that repeated limb movements, together with marked growth peaks like the one occurring in adolescent women, can cause any type of vascular TOS.

Diagnosis is one of the most confusing aspects owing to how little familiar we are with the syndrome and how great the variety of diagnostic procedures available is. There is great controversy regarding provocative maneuvers, which were not positive in any of our patients. Hixson. published a 3-article systematic review on the diagnostic accuracy of these maneuvers[6]. With a grade B evidence, they recommended all patients undergo Wright's test, Halsted's test, Cyriax release test, and supraclavicular pressure test. They advised against Adson's test since it is positive in 50% of the healthy population[7]. Current sensitivity and specificity levels of radiological tests allow these maneuvers to be ruled out. Vascular TOS diagnosis is based on the level of obstruction to dynamic flow as detected by CT-scan or MRI, as well as detection of venous thrombosis[8]. Neurogenic TOS is more challenging as imaging tests or nerve conduction studies associated with electromyography have little sensitivity. Hong et al. published a series of neurogenic TOS cases only[9]. Given that vascular TOS diagnostic tests were little performant, they proposed an exclusion diagnosis based on a combination of persisting and limiting pain associated with numbness or paresthesia exacerbating with arm movements, significant sport history, and simple cervical X-ray showing bone abnormalities.

TOS baseline treatment is conservative and based on rest, posture corrections, and limitation of activities or behaviors potentially worsening symptoms. Drug therapy based on non-steroid analgesics, muscle relaxants, or event intra-muscular injections of local anesthetics or corticoids can also be indicated[10]. Rehabilitation is regarded as the first treatment step, but there is little evidence. No randomized studies are available, and most of the knowledge is gathered from adult population articles. In true cases of neurogenic TOS, Ferrante et al. claimed that there is no use case for conservative therapy, since the denervation process occurs at the beginning of the clinical picture[11]. In addition, Brooke et al. stated that approximately 60-70% of patients successfully responded to conservative therapy[12]. Two of our patients improved with conservative management, but the other cases with more remarkable symptoms were refractory and required surgery. It would not be imprudent to start with rehabilitating therapy for 3 months and, if it does not prove to be effective, resort to surgical treatment –the only therapy that has demonstrated to be effective with full evidence. The ideal approach in surgical treatment remains an issue of discussion. We advocate the supraclavicular approach, since it allows for an adequate exposure of the arterial and venous component, as well as a better visualization of the CR. Other physicians and healthcare facilities prefer the transaxillary approach. For instance, Gloviczki et al.[13] claim that, when facing a similar structure exposure, the transaxillary approach is preferable in young women as it avoids scars. However, a customized assistant or retractor are necessary to raise and hold the limb, and in case of vascular complications, vessel control is more difficult.

The thoracoscopic approach has been proposed as an alternative to traditional techniques[14]. Its main advantage lies in the amplification of the image, which allows for a better vascular control and visualization of cervical soft tissue abnormalities. Apart from typical complications, chylothorax and pneumothorax can also occur[15]. Currently published series are small and only available in adults, so application in pediatric patients would be experimental. Robotic surgery is an even more novel technique[16]. A series of 67 neurogenic TOS patients undergoing robotic resection of the first cervical rib has been recently published, with results comparable to those of the thoracoscopic approach[17].

We are aware of the limitations our study has. Since it is a retrospective study, patient selection and treatment are biased. The small number of patients and the short follow-up period only allow us to detect trends.

CONCLUSIONS

TOS has been widely studied in adult patients, but we are little familiar with it in pediatric patients. Patients with vascular or nerve symptoms in the upper limbs associated with a puberty growth peak should undergo a CT-scan to rule out TOS. If analgesia- and rehabilitation-based conservative treatment does not prove effective, surgical treatment and the supraclavicular approach stand as a safe and effective alternative, with a short-term symptom control.

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