Ultrasound in Med. & Biol., Vol. ■, No. ■, pp. 1–10, 2016

0301-5629/$ - see front matter

http://dx.doi.org/10.1016/j.ultrasmedbio.2016.06.005

● *Original Contribution*

ULTRASONOGRAPHIC IDENTIFICATION OF FIBROMUSCULAR BANDS ASSOCIATED WITH NEUROGENIC THORACIC OUTLET SYNDROME: THE "WEDGE-SICKLE" SIGN

Zsuzsanna Arányi,* Anita Csillik,* Josef Böhm,† and Thomas Schelle‡

*MTA-SE NAP B Peripheral Nervous System Research Group, Department of Neurology, Semmelweis University, Budapest, Hungary; †Neurologische Praxis, Dr. Friedrich Behse/Dr. Josef Böhm, Berlin, Germany; and ‡Department of Neurology, Städtisches Klinikum Dessau, Dessau-Rosslau, Germany

(*Received* 28 *March* 2016; *revised* 31 *May* 2016; *in final form* 2 *June* 2016)

Abstract—Thoracic outlet syndrome (TOS) is a disorder characterized by compression of the lower trunk of the brachial plexus, most often in association with anomalous congenital fibromuscular bands in the scalenic region. Early diagnosis is important, because the neurologic deficit associated with TOS may be irreversible. Using high-resolution ultrasound, we investigated 20 consecutive patients with clinical signs suggestive of TOS (all females, average age: 40.4 ± 14.9 y) and 25 control patients. In 19 patients, we identified a hyper-echoic fibromuscular structure at the medial edge of the middle scalene muscle, which indented the lower trunk of the brachial plexus ("wedge-sickle sign"). It was associated with the significant enlargement ($p < 0.0001$) and hypo-echogenicity of the lower trunk. This novel and distinctive ultrasonographic sign allows pre-surgical identification of anomalous fibromuscular bands causing TOS. It is especially useful in patients without neurologic deficit, in whom the diagnosis may not be as straightforward. (E-mail: aranyi.zsuzsanna@med.semmelweis-univ.hu) © **2016 World Federation for Ultrasound in Medicine & Biology.**

Key Words: **Thoracic outlet syndrome, High-resolution ultrasound, Fibromuscular bands, Wedge-sickle sign.**

INTRODUCTION

The term *thoracic outlet syndrome* (TOS) was coined for a group of disorders characterized by compression of the brachial plexus or the subclavian vessels at any point in the thoracic outlet region (Peete et al. 1956). According to the classification presently in use, it comprises five distinct clinical syndromes: arterial vascular TOS, venous vascular TOS, traumatic neurovascular TOS, true neurologic (neurogenic) TOS and non-specific TOS (Ferrante 2012; Wilbourn 1999). In neurogenic TOS, the brachial plexus is typically compressed in the scalenic triangle at the level of the lower trunk or the distal portion of its constituents, the C8 and Th1 anterior primary rami (roots). This gives rise to a characteristic clinical syndrome with selective wasting of the thenar and the first dorsal interosseous muscle (Gilliatt et al. 1970) and sensory disturbance on the medial aspect of the forearm, with or without pain in the affected arm. The electrophysiologic hallmark of neurogenic TOS is the demonstration of post-ganglionic sensorimotor C8–Th1 axon loss, with Th1 being more affected and earlier (Tsao et al. 2014). The category "non-specific TOS," also called "disputed TOS" (Wilbourn 1999), is a controversial category with a lack of consensus on its etiology, pathomechanism and treatment. It is characterized by subjective symptoms such as pain and paresthesia in the arm and the feeling of fatigue of the arm, especially when lifted overhead, with no clinical deficit.

Congenital anomalies or anatomic variations of the thoracic outlet region, particularly the supernumerary cervical rib attached to the seventh cervical vertebra, have been historically implicated in TOS (Roos 1976). However, given that the estimated prevalence of cervical ribs in the general population is 0.5%–2% (Ferrante 2012; Viertel et al. 2012) and that of neurogenic TOS is 1 per million (Gilliatt et al. 1970), statistically the presence of a cervical rib is in itself not diagnostic for neurogenic TOS (Ferrante 2012; Weber and Criado 2014). Its relevance appears to be higher for arterial vascular TOS (Weber and Criado 2014). Roos, with extensive surgical experience in TOS, was the first to focus attention on

Address correspondence to: Zsuzsanna Arányi, Department of Neurology, Semmelweis University, Balassa u. 6, Budapest-1083, Hungary. E-mail: aranyi.zsuzsanna@med.semmelweis-univ.hu

anomalous fibromuscular bands with or without a cervical rib in the thoracic outlet region as the real culprit in neurogenic TOS (Brantigan and Roos 2004; Roos 1976, 1980). He described 10 types of these bands affecting the lower trunk and seven affecting the upper or middle trunks of the brachial plexus (Brantigan and Roos 2004; Roos 1976). These "Roos ligaments" were originally identified based on surgical and cadaveric studies, but today, modern imaging techniques such as magnetic resonance imaging (MRI) and high-resolution ultrasound (HRUS) are available for their possible pre-surgical detection and the facilitation of diagnosis. Some MRI data are already available (Aralasmak et al. 2012; Baumer et al. 2014; Luigetti et al. 2012; Magill et al. 2015; Matur et al. 2013; Poretti et al. 2015; Singh et al. 2014; Yildizgören et al. 2014). However, data in the literature regarding ultrasound are limited to a single case report (Simon et al. 2013), despite the ease and accessibility and recent advent of HRUS in the diagnosis of peripheral nerve disorders (Hobson-Webb et al. 2012). We present here a consecutive case series of patients with neurogenic and non-specific TOS assessed by HRUS.

METHODS

Approval for the retrospective analysis of patient data was obtained from both institutional ethics committees. Twenty consecutive patients, assessed at two tertiary referral centres for neuromuscular disorders between 2014 and 2016, were included in the analysis (Table 1). Criteria for inclusion of patients in the study were clinical symptoms and signs suggestive of TOS and the exclusion of other disorders, such as carpal tunnel syndrome, ulnar nerve lesion and C8–Th1 radiculopathy. All patients gave informed consent for the examinations, and retrospective analysis was performed using anonymized patient data. Healthy controls were examined prospectively with informed consent.

All patients underwent clinical, electrophysiologic and ultrasound assessments and radiographic examination of the cervical spine to look for a cervical rib or elongated transverse process of the seventh cervical vertebra. Additional examinations (*e.g.,* MRI of the cervical spine) were also carried out if deemed necessary for differential diagnosis. Neurogenic TOS was diagnosed if unequivocal clinical *and* electrophysiologic signs of postganglionic sensorimotor C8–Th1 axon loss were observed, unexplained by any other cause. Non-specific TOS was diagnosed when subjective complaints suggesting TOS were present without neurologic deficit (clinical signs of C8–Th1 lesion), with or without electrophysiologic alterations typical of TOS.

Subjective complaints suggesting TOS included pain and paresthesia in the arm, especially when lifted overhead, the feeling of fatigability of the arm and the Tinel sign at the supraclavicular fossa. The paresthesia typically involves the medial side of the forearm and hand, but some patients may not be able to localize it and complain of paresthesia of the whole arm. Provocative maneuvers, such as the Roos test (elevated arm stress test), were not used as a diagnostic element, as they were deemed unreliable (Plewa and Delinger 1998). Eight patients underwent surgery for TOS.

Table 1. Patient characteristics and findings

Case no.	Age (y)	Duration (y)	Side (L/R)	Neurologic deficit	Pain	EDX (C8–Th1 axon loss)	CSA of lower trunk (mm^2)	Radiography (cervical rib/elongated C7)	Surgery
1	64	16	R	Th1 > C8	−	Th1 > C8	29	C7	−
2	27	1	L	Th1 > C8	−	Th1 > C8	47	—	−
3	38	1	L	Th1 > C8	+	Th1 > C8	40	Rib	+
4	36	<1	R	Th1 > C8	+	Th1 > C8	40	—	−
5	37	3	R	Th1 > C8	−	Th1 > C8	20	Rib	−
6	28	5	L	Th1 > C8	−	Th1 > C8	50	Rib	−
7	27	3	R	—	+	Th1 (sens)	45	—	−
8	46	10	R	—	+	—	20	Rib	−
9	40	2	R	—	+	C8-Th1 (sens)	25	C7	−
10	19	2	R	—	+	—	22	—	−
11	74	5	R	Th1 > C8	−	Th1 > C8	29	—	+
12	43	2	R	Th1 > C8	+	Th1 > C8	34	—	+
13	54	5	R	Th1 > C8	+	Th1 > C8	30	Rib	+
14	49	15	R	Th1 > C8	+	Th1 > C8	36	C7	+
15	53	3	R	Th1-C8	+	C8-Th1	34	—	−
16	43	2	R	Th1 > C8	+	Th1 > C8	22	—	−
17	57	13	R	C8-Th1	−	C8-Th1	30	C7	+
18	21	2	R	C8-Th1	−	C8-Th1	37	C7	+
19	24	2	R	C8-Th1	+	C8-Th1	32	Rib	+
20	28	14	R	—	+	—	30	Rib	−

CSA = cross-sectional area; sens = only sensory; EDX = electrophysiologic examination.