Venous thoracic outlet syndrome caused by a congenital rib malformation

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Abstract

Summary: Venous thoracic outlet syndrome (VTOS) represents a rare disorder. Hypertrophy of the anterior scalene musculature is the cause of the compression syndrome in most cases. To our knowledge, we describe the first reported case worldwide of a venous compression syndrome caused by a congenital malformation of the 1st and 2nd ribs. Treatment by transaxillary partial rib resection was necessary and a very good postoperative result was achieved.

Keywords: Venous thoracic outlet • Subclavian venous thrombosis • Rib malformation

INTRODUCTION

Venous thoracic outlet syndromes (VTOSs) are unusual. They occur in one of 100 000 people [1]. Generally, chronic damage to the subclavian vein occurs because of the hypertrophy of the scalene musculature and the subclavian muscle, or because of the constriction of the costoclavicular space. According to our research, the case presented here is the only case reported worldwide in which a congenital malformation of the 1st and 2nd ribs caused a VTOS.

CASE

A 23-year old male presented with an external deformity of the right clavicle (see Fig. 1) and recurrent swelling and blue discoloration of the right arm which had been noticed a couple of weeks earlier. He was referred to us with the presumptive diagnosis of a sarcoma of the upper thoracic aperture. He is a trained machinist and currently works with a computer-guided machine. His work frequently entails lifting heavy objects overhead. There were no other disorders. An initial physical examination revealed a diffusely swollen right arm. The circulation, motion and sensation of the right arm were intact. There was a deformity of the right clavicle. Phlebography of the right arm revealed an abrupt stop of the contrast medium column at the level of the subclavian vein’s entrance into the thorax. Thoracic computed tomography (CT) and 3D-CT reconstruction revealed that a malformation of the 1st and 2nd ribs with a pseudoarthrotic connection was the cause of the thrombosis (see Fig. 2A–C). A surgical decompression was indicated because of the presence of VTOS. A partial resection of the 1st and 2nd ribs and also an anterior scalenotomy were performed using a transaxillary approach.

The postoperative clinical course was uneventful. A Doppler study of the veins prior to discharge showed a partial restoration of blood flow in the subclavian vein. The patient was prescribed a compression stocking for the affected arm for the next three months and continued oral anticoagulation over this time period.

DISCUSSION


The VTOS must fundamentally be differentiated from neurogenic and arterial outlet syndromes. VTOS is further subdivided into three types: intermittent venous constriction, secondary thrombosis of the subclavian vein caused by a central venous catheter or pacemaker cables and thrombosis resulting from external compression because of muscle contraction.

The incidence of this latter type is approximately one in 100 000 people annually [1]. Men are affected twice as often as women. Symptoms occur on the right side in 60–80% of the cases [1]. VTOS is most often caused by the hypertrophy of the anterior scalene muscle or the subclavian muscle or by a constriction of the space between the clavicle and the 1st rib. An accessory cervical rib is not a risk factor for VTOS.

In our patient, VTOS was caused by a rib malformation present since birth. There was no history of trauma in this patient. Clinically, this patient exhibited the typical signs of intermittent
Posture-dependent venous compression: episodes of swelling of the right arm with blue discolouration and pain. Externally, a deformity of the right clavicle and prominence of the superficial venous plexus were evident. These are the typical signs of the formation of venous collaterals as often described in the literature [5, 6]. Phlebography confirmed the clinically suspected thrombosis of the subclavian vein and also suggested a possible aetiology. A CT study of the upper thoracic aperture also confirmed the finding of a costal malformation with a neoarticulation of the 1st and 2nd ribs. We performed a 3D thoracic reconstruction for the purposes of visualization and planning.

The thrombosis was initially treated using a compression stocking and beginning a weight-adjusted therapy using a low molecular weight heparin. A transaxillary partial resection of the 1st and 2nd ribs as well as an anterior scalenotomy were necessary to treat the problem of chronic compression. The approach described by Roos [7] offers the best exposure to the anterior aspect of the 1st and 2nd ribs along with an excellent cosmetic result. A surgical dissection, however, is elaborate and injuries to the axillary vessels are very complex to repair using this approach. The long-term results using this approach are excellent [8]. Supra-, trans- and infraclavicular approaches are also possible, but at the expense of increased perioperative morbidity [9]. Nowadays, 1st rib resection using a videothoracoscopic technique is also possible, but this new method has to be evaluated in more studies before becoming a first-choice approach in most of the patients with a VTOS [10].

Oral anticoagulation with coumarine for three months is recommended and the patients are followed up at postoperative months one and six. A postoperative recurrence of VTOS appears extremely rare.

To our knowledge, the case we presented here is the first case worldwide of VTOS caused by a rib malformation requiring a surgical correction.

Conflict of interest: None declared.

REFERENCES