Nine-year Follow-up of a Patient with Thoracic Outlet Syndrome and Paget-Schroetter syndrome

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ABSTRACT

Thoracic outlet syndrome (TOS) is a challenging clinical condition with regards to its diagnosis and treatment. Its management may turn out to be challenging a case when an adverse complication such as Paget-Schroetter syndrome (PSS) also co-exists. Herein, we report a nine-year follow-up of a patient with TOS (a 41-year old cardiovascular surgeon) who had suffered multiple PSS episodes and repeat TOS surgeries. In essence, we want to highlight the diverse clinical findings of TOS and discuss its management in light of the relevant literature.

Keywords: Paget-Schroetter syndrome, subclavian thrombosis, surgery, thoracic outlet syndrome

Nueve años de seguimiento de un paciente con síndrome de salida torácica y síndrome de Paget-Schroetter

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RESUMEN

El síndrome de salida torácica (SST) es una condición clínica que constituye un desafío con respecto a su diagnóstico y tratamiento. Su tratamiento puede llegar a convertirse en un verdadero atolladero ser de añadirse una complicación adversa como el síndrome de Paget-Schroetter (SPS) Reportamos aquí nueve años de seguimiento de un paciente con SST (un cirujano cardiovascular 41 años de edad) que había sufrido varios episodios de SPS y repetidas cirugías por SST. En esencia, queremos destacar los diversos resultados clínicos del SST y discutir su tratamiento a la luz de la literatura pertinente.

Palabras claves: síndrome de Paget-Schroetter, cirugía, trombosis de la vena subclavia, síndrome de salida torácica

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INTRODUCTION

Thoracic outlet sydrome (TOS) is a constellation of findings that ensue due to compression of the neurovascular structures in the cervicoaxillary region. Having a wide range of clinical findings, the diagnosis and treatment of TOS may be challenging in clinical practice. Concerning the pertinent venous complications, Paget-Schroetter syndrome (PSS) – otherwise called primary thrombosis of the axillary/subclavian vein– would be one of the worst clinical scenarios; prompt management is paramount (1, 2).

In this report, we present a nine-year follow-up of one of our TOS patients who had suffered multiple PSS episodes and three TOS surgeries bilaterally.

CASE REPORT

In 2003, a 41-year old female patient (a cardiovascular surgeon) was seen for discoloration and swelling on her right upper extremity. She reported that her complaints had ensued after performing a coronary bypass surgery. She also described previous similar but less severe complaints including mild pain, numbness and fatigue of both arms. She added that they usually occurred after playing tennis and after coronary artery bypass grafting operations, especially during left internal thoracic artery take down during which the surgeon's arms

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are hyperabducted. Her medical/family history was otherwise non-contributory.

Physical examination revealed right arm swelling and positive Adson and Roos tests (fatigue and pain). Immediate upper limb Doppler ultrasonography showed thrombosis of the right subclavian vein. Blood tests for thrombophilia were all normal. As magnetic resonance imaging (MRI) also confirmedthe diagnosis of PSS (Fig.1A, B), she was treated with early tissue plasminogen activator (TPA) with partial thrombolysis resulting in partial residual stenosis of the subclavian vein.

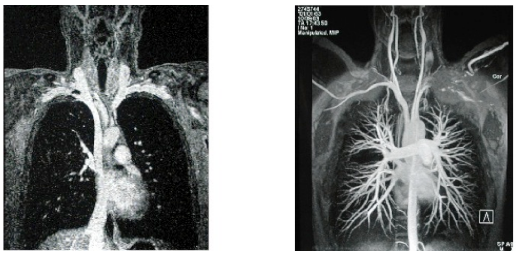

Figure: A) T1 vibe fat-sat postcontrast magnetic resonance imaging (coronal view) demonstrating the hypointense thrombus partially occluding the right-subclavian vein. B) Postcontrast flash three-dimension magnetic resonance angiography depicting mild narrowing in the right subclavian artery during hyperabduction. The left-sided "pseudoocclusion" is due to the technique (coronal oblique sequence adjusted for prompt right-sided imaging).

Thereafter, her treatment was maintained with pneumatic compression for the right arm swelling. Multidetector computed tomography, which was performed for TOS, uncovered compression of the subclavian vessels between the clavicle and the 1st rib bilaterally. Accordingly, the patient was also diagnosed with bilateral TOS. She preferred not to undergo a corrective TOS surgery at that time and, therefore, was followed with a conservative treatment which comprised home-based exercises for strengthening the shoulder elevators and avoidance of provocative activities.

One year later, in 2004, she suffered from swelling, this time, of her left upper extremity again after a long-lasting surgery. Magnetic resonance angiography (MRA) demonstrated thrombosis in the left subclavian vein, and along with early TPA treatment, this time corrective surgery for the left-sided TOS was also carried out. Unfortunately, she had to be operated twice on the same day as the postoperative X-ray unmasked the wrong resection of the 2nd rib after the initial surgery (transaxillary approach). Paget-Schroetter syndrome recurred on the left limb during the night of the operation and she received low molecular weight heparin, followed with warfarin therapy. Her complaints (swelling and heaviness) persisted due to partial obstruction of the left subclavian vein with multiple collateral venous circulation.

During follow-up with conservative treatment, her TOS symptoms relatively improved on the left-side but did not change much on the right-side. Therefore, one year later in 2005, corrective TOS surgery was applied for the right side as well. Only incomplete resection of the first rib could have been performed due to very difficult accessibility of the right axillary region. Postoperative hospital stay was uneventful.

Currently (2012), she is free of any PSS attacks since 2004; however, her complaints of pain and numbness (especially during hyperabduction) in the right arm, and occasional heaviness and minimal swelling in the left arm still exist.

DISCUSSION

Thoracic outlet syndrome encompasses a variety of symptoms and signs due to compression of the brachial plexus, subclavian vessels and the sympathetic ganglia in the cervicoaxillary region. The common clinical scenario consists of pain, paraesthesia, weakness, fatigue, swelling and temperature changes. On the other hand, its recognition – as well as its management – may be confounded by its other less common findings like tachycardia, dyspnoea, dysphagia, angina-like chest pain, occipital headache, vertigo, dizziness, tinnitus, complex regional pain syndrome, PSS and Raynaud-like vasomotor changes (1–3). Although the diagnosis of TOS is clinical, depending on the extent of involvement or for those patients who are to undergo surgery, the underlying aetiology needs to be delineated precisely with radiological assessment [*ie* with Doppler ultrasound, computed tomography, MR imaging or angiography] (4). The treatment is either conservative exercise therapy (strengthening shoulder elevators, postural restoration and avoiding provocative activities) or corrective surgery that can be as challenging as the diagnosis. Indisputably, the whole scenario may turn out to be an impasse in case an adverse complication (*eg* PSS) does also coexists.

Paget-Schroetter syndrome – thrombosis of the subclavian vein – is a serious clinical condition with significant morbidity/mortality (5). The syndrome is not commonplace and may be present in 5% of TOS patients (6). In addition to external causes (anatomical factors or overuse of the upper limbs), certain haematologic risk factors for thrombophilia (factor V Leiden, prothrombin G20210A, protein S deficiency, hyperhomocysteinaemia) or microtraumatizations of the venous intima (due to retroversion or hyperabduction of the arm) may all predispose to PSS (7). Its management embraces mainly arm elevation and routine anticoagulation, thrombolysis, and early or subsequent surgical decompression *ie* first rib resection, scalenectomy and venous reconstruction (4, 8). On the other hand, while non-surgical treatment may be sufficient in most cases, decompression surgery is generally warranted especially if venous obstruction persists during the follow-up. Yet, the risk of recurrence is high without correction of the underlying anatomical cause (4, 8).

In our case, TOS (due to the aforementioned congenital factors), long-lasting isometric contractions of the upper limbs and dehydration with hyperabduction of the arms (2) during her every-day surgery practice seemed to be the main contributors for the eventual clinical scenario. In accordance with the literature, whereby early anatomical correction in addition to anticoagulation and thrombolytic treatment are

suggested (4, 8), she was operated on (though inconveniently) for her persistent complaints. Herein, it is noteworthy that timing of surgery in PSS is really a matter of debate; while it is effective for returning to activity (4, 8), there is the possibility of overtreating the patient with early surgery as well. Moreover, in a highly active patient (like ours) with a known previous history, contralateral surgery may even be a reasonable early therapeutic approach if relevant signs and symptoms ensue in the presence of similar predisposing factors. Yet, contralateral recurrence is rare but does occur – 7/103 – in a retrospective case series (6).

Overall, presenting this case of ours – a cardiovascular surgeon with TOS and PSS – we have once again drawn attention to the challenges of TOS management. It is not only the prompt diagnosis out of a wide range of findings, but also the challenges pertaining to surgery.

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