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Thoracic outlet syndrome in a patient with SAPHO syndrome – A case report

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Thoracic outlet syndrome (TOS) includes disorders caused by compression of the neurovascular structures in the upper thoracic outlet (Roos and Owens, 1996 [1]; Bürger, 2014; Curuk, 2020 [3]). Depending on the compressed structure, it is categorized into neurological, arterial and venous TOS.

SAPHO syndrome (synovitis–acne–pustulosis–hyperostosis–osteitis syndrome) is a rare chronic inflammatory disease of unknown etiology. With its typical involvement of sternoclavicular joint and clavicle, complication due to hyperostosis in this region, leading to thrombosis of the subclavian vein have been reported in some cases of SAPHO syndrome.

Between 2015 and 2019 488 patients, suffering from neurological, vascular or combined TOS presented at our department. Depending on clinical and diagnostic results surgical therapy was performed in 175 cases via the transaxillary approach, including complete first rib and/or cervical rib resection, neurolysis of plexus brachialis, thoracic sympathectomy and vascular reconstruction if indicated (Curuk, 2020). During this period, only one single patient presented with SAPHO syndrome with thrombosis of the subclavian vein and neurovascular TOS.

CASE PRESENTATION: We present a 50-year-old female patient, in line with the SCARE 2020 criteria (Agha et al., 2020 [12]) suffering from extremely rare combination of neurovascular TOS and SAPHO syndrome with thrombosis of the left subclavian vein due to hyperostosis of the left clavicle.

CONCLUSION: Progressive bone changes associated with SAPHO syndrome can lead to narrowing of the thoracic outlet. Pharmacological therapies to avoid the progression of the hyperostosis of the costoclavicular joint and the clavicle do currently not exist. First rib resection is a therapeutic option to widen the space in the upper thoracic region. Surely, it is a rare condition and more long-term follow-up data are required.

1. Background

Thoracic outlet syndrome (TOS) is a rare condition caused by compression of neurovascular structures within the anatomic space posterior to the clavicle, above the first rib and extending to the subcoracoid space [4]. The prevalence is uncertain and varies between 0.1 per million and 1% [5].

The three main components in the thoracic outlet area are the brachial plexus, subclavian vein, and subclavian artery. According to the compressed structure, TOS is classified as neurogenic (nTOS), arterial (aTOS) and venous (vTOS). In some cases, there is a joined compression of neurological and vascular components leading to a neurovascular TOS. The nTOS is the most common form of TOS with a prevalence up to 95%. vTOS is the second most common with 3–5% and affecting mainly young sporty patients. The least common type is the aTOS with a prevalence 1–2% [2].

Due to the complex clinical presentation and the lack of diagnostic criteria, objective laboratory tests and standardized therapy approach, the TOS continuous to be underdiagnosed and insufficiently treated [2,4,6].

SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, and osteitis) was first described by Chamot and colleagues [6] in 1987. SAPHO syndrome covers a spectrum of heterogeneous diseases characterized by osteoarticular and dermatological manifestations [7].

The diagnosis of SAPHO syndrome is based on history, characteristic scintigraphic and radiological results, as well as skin manifestations [8].

Abbreviations: SAPHO, synovitis–acne–pustulosis–hyperostosis–osteitis; TOS, thoracic outlet syndrome; nTOS, neurogenic thoracic outlet syndrome; aTOS, arterial thoracic outlet syndrome; vTOS, venous thoracic outlet syndrome; CT, Computed tomography.

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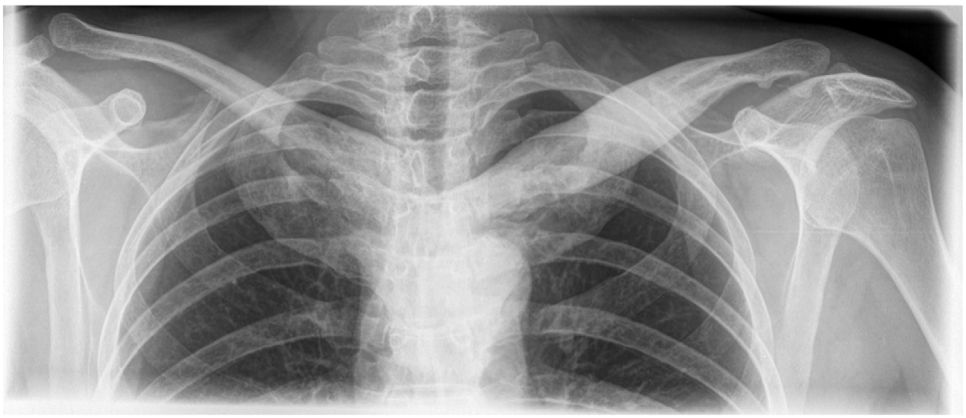

Fig. 1. X-ray of the upper thoracic region showing hyperostosis of the sternoclavicular joints and left clavicle.

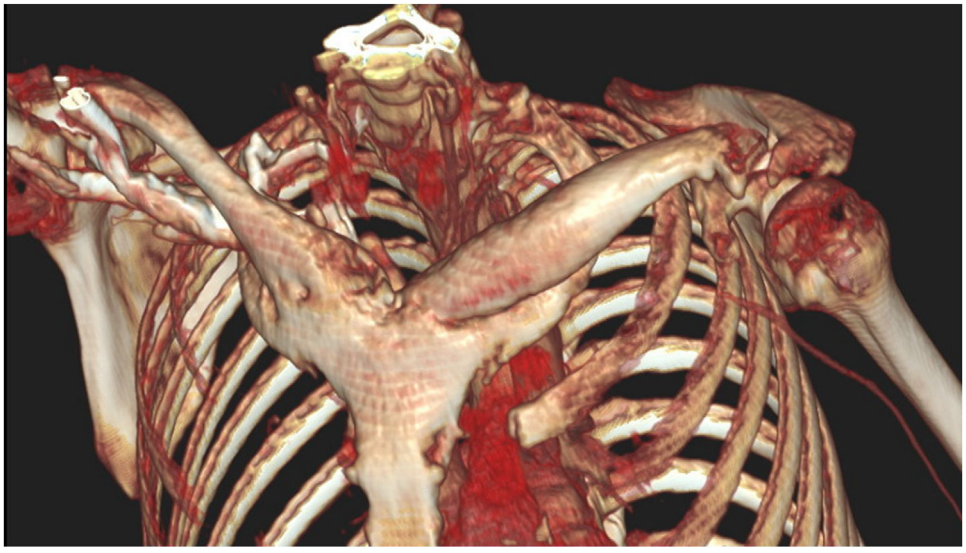

Fig. 2. 3D CT scan reconstruction showing hyperostosis of the left clavicle and narrowing of the left upper thoracic region.

The anterior chest wall involvement because of the hyperostosis of the sternoclavicular joint and clavicle is characteristic of SAPHO syndrome. In cases of advanced disease course, it can lead to complications such as thrombosis of the subclavian vein.

However, venous thrombosis is a rare complication of SAPHO syndrome. In a series of 120 patients with this syndrome, Hayem et al. found only one case (0.8%) reporting thrombosis of the subclavian vein [9].

A literature review of thrombotic manifestation in SAPHO syndrome revealed 15 published cases of venous thrombosis associated with the presence of SAPHO syndrome. In 14 of these cases, thrombosis was located in the subclavian veins [10].

2. Case presentation

In November 2019, a 50 years old female patient was referred to our vascular surgery department. The patient reported to have experienced pain, weakness and paresthesia in the left upper limb accompanied with recurrent swelling and cyanosis of the left arm. She had been diagnosed with SAPHO syndrome several years ago. However, the patient reported symptoms and complaints did differ from the SAPHO related complaints the patient had experienced before.

2.1. Past medical history of SAPHO syndrome

The patient was first diagnosed with SAPHO syndrome in 2013 using bone scintigraphy. At the time of diagnosis, the patient had suffered more than 10 years from pain in the left upper thoracic region. The patient had even gone through multiple surgeries of the clavicle and the left shoulder to ameliorate the symptoms. As early as 2012, the patient began to notice occasional swelling and cyanosis of the left arm. A thrombosis of the subclavian vein was excluded at that time and later in early 2019. The patient presenting with swelling of the upper extremity without thrombosis can be explained by McCleery syndrome, an intermittent compression of the subclavian vein [11]. No further diagnostics or therapies were applied then until the presentation of the patient in our clinic in November 2019.

The initial clinical examination in our department revealed a prominent left clavicle bone, a collateral venous circulation in the left upper chest region and a suppression of the left radial pulse on arm elevation. Neither swelling respectively cyanosis of the left arm, nor signs of peripheral embolization were apparent. Neurophysiological examination showed no evidence of neurological damage.

No family History of thrombosis. Laboratory Diagnostics showed no signs of thrombophilia or genetic disorders that could lead to venous thrombosis.

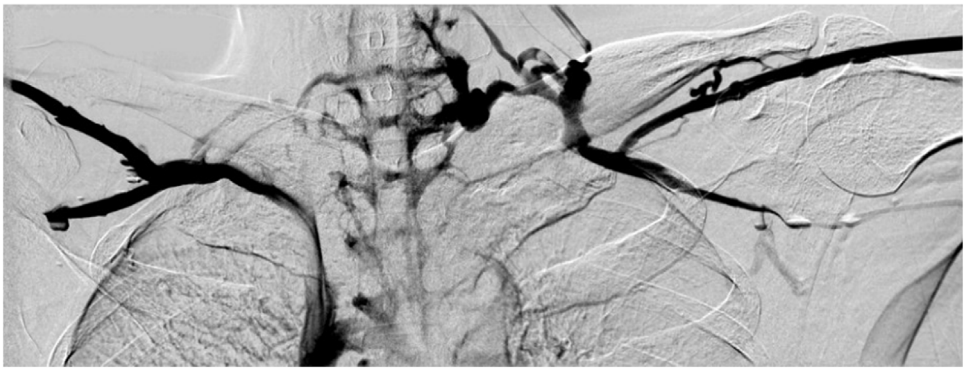

Fig. 3. Central-venous angiography with occlusion of the left subclavian vein and collateral circulation in abduction.

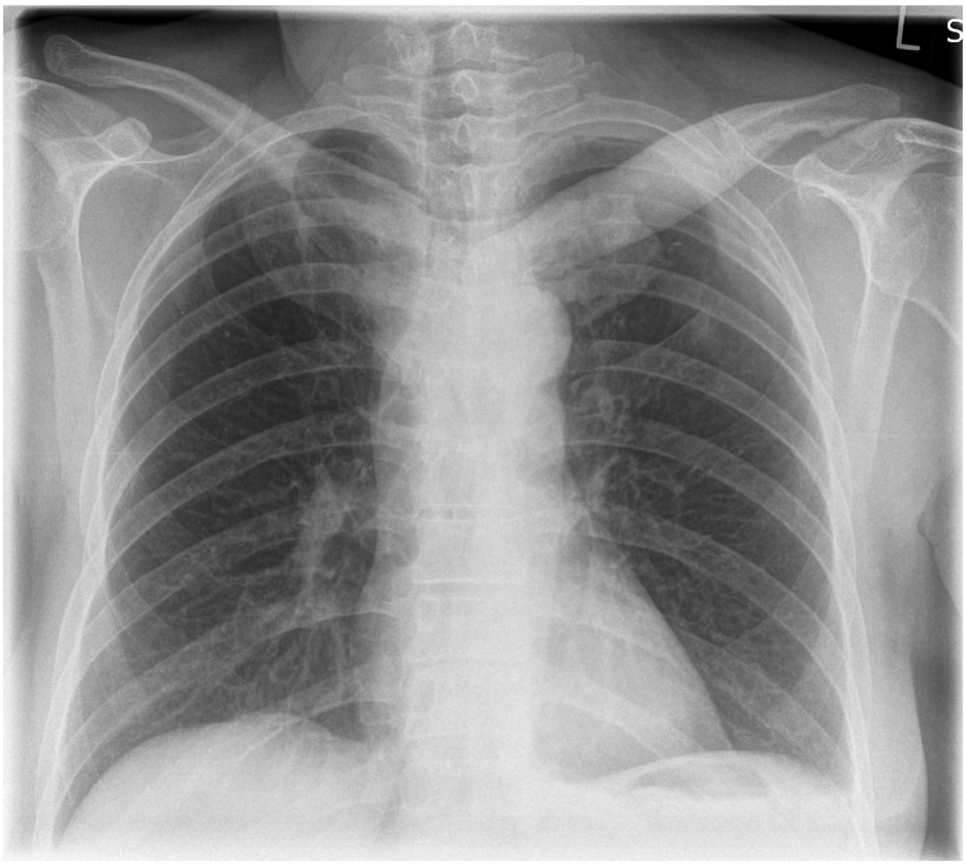

Fig. 4. Postoperative chest X-ray showing partial removal of the left first and second rib.

The x-ray of the upper thoracic region confirmed the hyperostosis of the left clavicle and no evidence of cervical rib (Fig. 1).

The duplex ultrasound and the CT scan showed a thrombotic occlusion of the left subclavian vein, compression of the left subclavian artery during abduction and elevation of the left arm and a massive hyperostosis of the sternoclavicular joint and the left clavicular bone with compression of the neurovascular structures (Fig. 2).

Our standard routine central venous angiography in sitting position showed an occlusion of the left subclavian vein with collateral circulation (Fig. 3) and a compression of the left subclavian artery during arm elevation without any aneurysmatic wall changes.

The clinical and radiological findings all indicate a neurovascular compression in the left thoracic outlet region mainly due to the hyperostosis of the left clavicle.

Our initial recommendation was conservative treatment of the patient with pain management and physiotherapy. Anticoagulation was not prescribed given the total occlusion of the proximal subclavian vein with well-established collaterals.

Six months after the initial presentation, the patient was readmitted to our clinic for operative treatment with progressive complaints in the left arm. We performed atypical resection of the first and second rib because of the specific anatomical situs and the massive tissue adhesions and the hyperostosis of the clavicular bone to achieve a complete decompression of the upper thoracic outlet (Fig. 4). The postoperative care included pain management with intravenous and oral Medications and Physiotherapy. The comparison between pre and postoperative optical pulse oscillography shows a significant improvement of the arterial flow in the left arm (Fig. 5). Two weeks after surgery, we discharged the patient

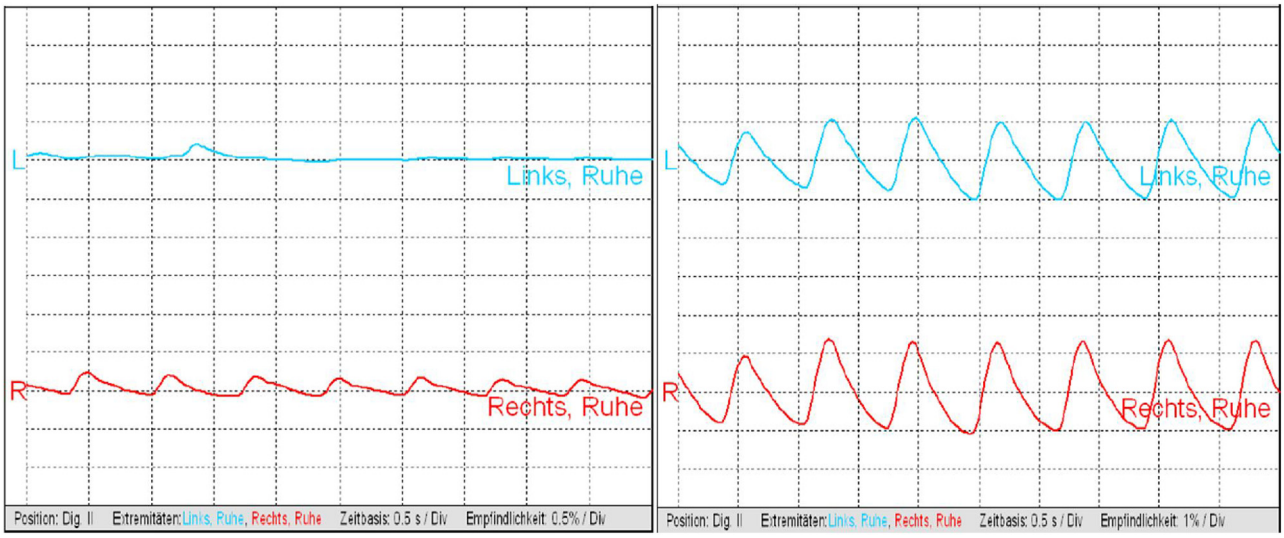

Fig. 5. Optical Pulse Oscillography of 1. Finger in arm elevated position (Left = preoperative, Right = postoperative).

with sufficient symptoms relief. A special TOS Physiotherapy exercise was instructed and recommended until the next follow up.

In the postoperative follow up after 2 months, the Patient showed a complete recovery and was not taking any analgesic after 4 weeks of surgery.

3. Discussion and conclusions

Progressive bone changes in SAPHO syndrome can lead to narrowing of the upper thoracic region and venous compression leading to vessel damage and thromboembolic events.

In TOS, first rib resection widens the space and releases the compression on the neurovascular structures in the upper thoracic region. In the absence of a current therapy to avoid the progression of the hyperostosis of the costoclavicular joint and clavicle, the rib resection offers a surgical therapy. Resection of the first rib in patients with SAPHO syndrome suffering from compression of the subclavian vein can help in avoiding more vascular damage and can relief the compression on the collateral venous circulation. More cases need to be studied to prove the effectiveness of this surgery in patients suffering from this rare disorder.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

The need for approval was waived.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor in-Chief of this journal on request.

Author contribution

Idea and Data collection initially by the author, during writing this paper HO, CC, HP, TB and ES were involved constantly in discussion und improvements of the manuscript.

Surgery was performed by Prescher H.

Registration of research studies

researchregistry6599 available at: https://www.researchregistry.com/browse-the-registry#home/registrationdetails/6032c0dd53d1b7001b53376a/.

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Thomas Burger, M.D.

Provenance and peer review

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Availability of data and material

Data collection done by reviewing literature and health records at our department. Data was collected and stored at our server, to avoid violation access encrypted.

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