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A rare cause of upper extremity deep venous thrombosis: Paget Schroetter syndrome

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The authors stated that written consent was taken from the patient for the procedure and publication of data including clinical photographs

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Abstract

Paget-Schroetter syndrome is a rare clinical entity, which is characterized by the thrombosis of the axillary and/or subclavian vein resulting from the activation of the coagulation cascade due to vigorous and repetitive activity of the arm and the external compression related microtrauma to the venous intima. The first imaging modality for diagnosis should be color Doppler ultrasonography. Thrombolysis and surgical decompression of the thoracic outlet are focused on relieving the external compression of the veins. In this case report, we aimed to present the greyscale ultrasonography and dynamic Doppler ultrasonography findings of a patient with upper extremity deep venous thrombosis.

Keywords: Paget-Schroetter Syndrome, Effort thrombosis, Color Doppler ultrasonography, Upper extremity deep venous thrombosis

Introduction

Paget-Schroetter syndrome (PSS) or ‘effort’ thrombosis is the thrombosis of the subclavian vein due to its compression at the thoracic outlet level by variation or deformities of adjacent anatomic structures with excessive physical activity in otherwise healthy individuals [1]. PSS corresponds to approximately 10-20% of all upper extremity deep venous thrombosis cases [2]. It is suggested that coagulation cascade activation, which is triggered by intimal microtrauma in the vein, is responsible for the pathogenesis of PSS [3]. Plain radiography, ultrasonography (US), color Doppler US (CDUS), computerized tomography (CT)/magnetic resonance imaging (MRI) venography, and catheter venography can be used for diagnosis [4].

In this case report, we aimed to present the US and dynamic CDUS findings of a 46-year-old female patient who was admitted to our emergency room with discoloration and swelling in the right arm and diagnosed with PSS.

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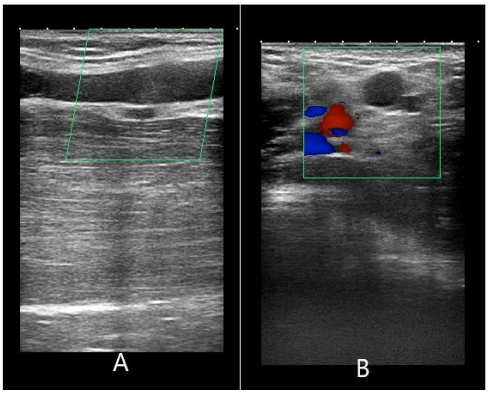
Case presentation

A 46-year-old female patient with discoloration, swelling, and increasing pain in the right arm for 1 week was admitted to our emergency department. In her medical history, there were no related traumas, chronic diseases, no regular drug use such as oral contraceptives, and no history of insect bites. She had been working with repetitive and excessive arm movements in a chicken processing factory. In physical examination, slight blue-purple discoloration and edema were present in her dominant arm, forearm, and hand, compared to the contralateral one. The range of motion in her joints was normal. The peripheric pulses were normal in the neutral position. Her lab test results and posteroanterior chest radiography showed no clinical significance. Together with all those findings, CDUS was requested for the right arm to confirm the preliminary diagnosis of right upper extremity deep venous thrombosis. US and CDUS examinations were performed with a 6-12 MHz band range, linear, high-frequency transducer (Toshiba Aplio MX SSA-780A, Tokyo, Japan). In greyscale US examination, hypoechoic thrombus material filling the whole lumen of the right subclavian vein was observed and the vein was non-compressible. In color mode and spectral mode, no flow signal or recanalization finding was observed (Figure 1 a-b).

Figure 1 a-b: Color Doppler Ultrasonography (CDUS) examination showed hypoechoic, uncompressed thrombus and an absence of flow in the right subclavian vein lumen in the a-long axis and b-transverse plane.



More distal veins showed mild dilatation and slow flow findings. The findings were consistent with acute/subacute thrombosis of the right subclavian vein. The patient was given anticoagulant medication and called for control evaluation 2 weeks later. Control examinations revealed that the blue-purple discoloration and edema in her right arm had regressed. Also, thrombus material had resolved and recanalized flow was observed in the neutral position in greyscale US and CDUS imaging. The recanalized flow ceased by dynamic CDUS examination, which was combined with Adson's test (the arm was abducted to the level of the head and the head was rotated to the contralateral side during deep inspiration). Because all those findings pointed to the external compression to the subclavian vein at the thoracic outlet, the diagnosis was venous thoracic outlet syndrome (TOS) (Figure 2 a-b). When the patient was questioned about the activities, such as hanging clothes or curtains, she stated that she had difficulties when performing activities that required raising her arm above her head. During the Adson maneuver, normal flow and the waveform of the right subclavian artery were gradually dampened then completely ceased (Figure 3 a-b). Hence, the patient also had arterial TOS apart from PSS. After anticoagulant treatment, the patient did not have any further examinations or surgical treatment during the pandemic. Written informed consent was obtained from the patient.

Figure 2 a-b: In the dynamic control examination of CDUS performed two weeks later, a-prominent recanalized flow in the subclavian vein was seen in the neutral position, especially in the posterior part of the lumen, b- cessation of the recanalized flow during the provocation test.

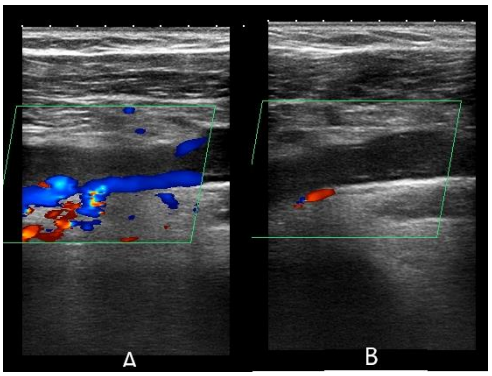
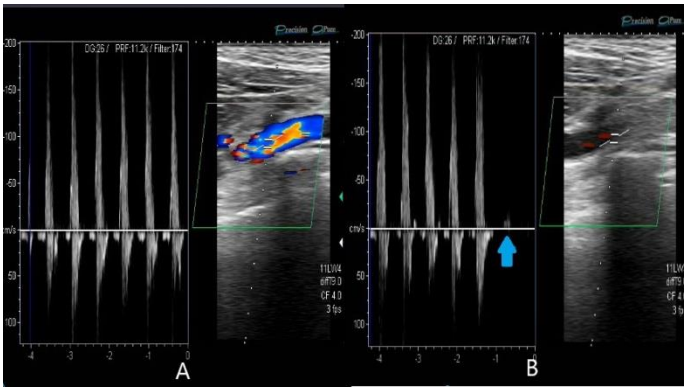


Figure 3 a-b: In dynamic CDUS examination, a- color mode and spectral mode in the neutral position showed normal flow velocity and waveform in the subclavian artery; b- color mode and spectral mode during the provocation test showed complete cessation of flow (blue arrow).



Discussion

This entity was first described by Paget in 1875 and Von Schroetter in 1884, and Hughes named it "PSS" in 1949 [1]. PSS is seen in approximately 2-11/100,000 people and constitutes 1-4% of all venous thrombosis [5,6]. In PSS, there is primary thrombus development without any traumatic injury that may cause secondary thrombosis in the subclavian vein or an iatrogenic cause such as a pacemaker, or central venous catheter insertion [3, 4]. Approximately 60-80% of PSS cases are due to excessive physical activity and are seen in the dominant arm. It is often seen in males, athletes, and in the 3rd decade of life. The entity, which is named "strain thrombosis", was thought to be related to dominant use; however, external compression can also contribute. For this reason, compression of vascular structures due to narrowing in the thoracic outlet region may also be one of the causes of PSS [4]. Microintimal trauma in the subclavian vein caused by excessive shoulder-arm movement and the external compression at the thoracic outlet level causes intimal hyperplasia and fibrosis in the connective tissue around the vein [7]. In recent years, studies reported a thrombophilia incidence in PSS patients, but there has been no consensus to screen these patients for hypercoagulability disorders [4,8]. In our case, because the patient had no family history or a similar history of thrombosis, no further investigation was conducted.

Vascular compressions may be caused by abnormal subclavius or anterior scalene muscles, cervical vertebrae with long transverse processes, cervical ribs, congenital fibromuscular bands, 1st rib deformations due to trauma, osteodegeneration, or narrow costoclavicular space [3, 4, 9]. External compression of the subclavian vein (venous thoracic outlet syndrome - venous TOS) in the thoracic outlet that may cause PSS is a rare condition with an annual incidence of 1/100,000 [3]. The clinical picture in which symptoms related to venous TOS is seen without evidence of thrombosis is called intermittent obstruction or McCleery's syndrome [4]. In this process, diagnosis and treatment with 1st rib resection or scalenectomy may prevent the formation of PSS. Clinically, it may be difficult to distinguish between McCleerly's syndrome and neurogenic TOS due to compression on the brachial plexus. Pain, numbness, and tingling are common symptoms in both entities [4]. CDUS and electromyography can be used to distinguish between the two. Our patient repeatedly used her arm due to her job, which may have triggered thrombus formation. Chest radiography of the patient allowed us to exclude possible bone pathologies. However, etiological factors related to soft tissues such as anomalies or hypertrophic changes in the muscle and ligament structures in this region might have triggered thrombus formation. Due to the pandemic period, this suspicion could not be confirmed by cross-sectional imaging methods, such as a CT or an MRI.

Clinical symptoms, blood tests, and radiological imaging methods should be evaluated together when diagnosing PSS. The clinical findings alone have a specificity of less than 50% [12, 13]. Since partial venous drainage will be provided, especially due to the collateral network that develops after a while, clinical findings regress, and diagnosis may be difficult [4]. Although the Plasma D dimer test is relatively highly sensitive, its specificity is very low as it can be increased in many inflammatory processes [14]. Bone anomalies or pathologies can be evaluated radiologically on cervical vertebrae or chest radiographs. With dynamic CDUS, CT venography, MRI venography, and catheter venography examinations combined with dynamic maneuvers, the exact location and degree of vascular compression can be revealed. Among them, CDUS stands out as the first choice because it is non-invasive, non-ionizing, inexpensive, and ubiquitous [6, 9]. Besides, as an advantage against CT and MRI, high false-negative results that may occur in the supine position can be avoided by performing the CDUS in a sitting position [10]. With dynamic CDUS examination, both the subclavian vein and subclavian artery can be examined in the same session. There are publications about both arterial and venous TOS association in the literature, reporting a rate of 13.7% [10, 11]. Different provocation tests can be performed during the examination. The most common one is the Adson test. A dynamic examination can also be performed with EAST (elevated arm stress test) or ULTT (upper limb tension test) [15]. With the dynamic CDUS examination performed during the test, the venous flow, which normally changes with respiration, is disrupted. Infraclavicular enlargement can be observed in the vein together with loss of respiratory changes, increased velocity due to narrowing of the vein, or complete cessation of flow [4, 10]. In detecting thrombosis in the subclavian vein, CDUS has a sensitivity of 78-100% and a specificity of 82-100% [6]. Although CT and MRI venography are expensive compared to CDUS examinations, they are alternative radiological imaging methods because they can give better anatomical details about the surrounding structures for the etiology of thrombosis. However, since this detail does not affect the initial treatment management for thrombosis, they are not the first choice of radiological imaging [14]. Catheter venography examination, on the other hand, is preferred in catheter thrombolytic treatment rather than in making the diagnosis because it is invasive and ionizing [10].

Conclusion

PSS is a rare, primary deep vein thrombosis of the upper extremity, which is more common in the young population and is caused by repetitive excessive arm activity and vascular compression. The diagnosis of PSS should be considered especially when there is no trauma to the subclavian vein or any iatrogenic procedures in the patient's history. CDUS examination is the imaging method that can detect thrombosis with high accuracy and should be the first choice in diagnosis. This method also gives an idea about the etiological factors because it is easily suitable for dynamic examination.

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