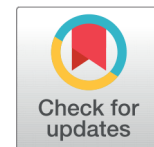


SIMULTANEOUS IVC/SVC ENDOVASCULAR SHARP RECANALIZATION IN A PATIENT WITH BUDD-CHIARI SYNDROME, SYSTEMIC LUPUS ERYTHEMATOSUS AND ANTIPHOSPHOLIPID SYNDROME, A CASE REPORT



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ABSTRACT

Background: One of the challenging life-threatening conditions is Budd-Chiari syndrome (BCS). This is a case report of BCS patient presented with inferior vena cava (IVC) and superior vena cava (SVC) stenosis and treated by a simultaneous IVC/SVC sharp recanalization.

Case presentation: A 40-year-old female patient with a history of systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS) presented with distended abdomen. A triphasic computed tomography (CT) scan revealed attenuation of hepatic veins and hypertrophy of the caudate. There was a severe dilatation of the azygos and hemiazygos veins with multiple posterior mediastinal and retrocrural tortuous collaterals recanalizing the IVC distally. Chest CT with contrast revealed left brachiocephalic vein occlusion. Although, the SVC was patent, multiple calcified foci of the wall were noted. An inferior venacavogram demonstrated a complete suprahepatic IVC occlusion. Another venogram illustrated a complete right brachiocephalic vein blockage with extensive collaterals. Sharp recanalization of the brachiocephalic vein to the SVC was done, followed by covered stent placement. Then Multiple balloon angioplasties were made at the level of suprahepatic IVC with a placement of non-covered stent.

Conclusion: It was a successful recanalization for both completely occluded suprahepatic IVC and right brachiocephalic vein with placement of stents.

المخلص

الخلفية: تعد متلازمة بود خياري (BCS) من أكثر الأمراض التي تشكل خطورة على الحياة. هذا تقرير حالة لمريض تم تقديمه مع تضيق الوريد الأجوف السفلي (IVC) وتضيق الوريد الأجوف العلوي (SVC) وتم علاجه عن طريق إعادة الاستقراء الحاد للوريد الأجوف العلوي والسفلي في وقت واحد.

Received 22 May 2022

Revised 26 July 2022

Accepted 2 August 2022

Published 27 August 2022

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eISSN: 1658-8959



عرض الحالة: مريضة تبلغ من العمر 40 عامًا ولديها تاريخ من الذئبة الحمامية الجهازية (SLE) ومتلازمة أضداد الشحوم الفوسفورية (APS) مصحوبة بانتفاخ في البطن. كشف التصوير الأشعة المقطعية عن توهين الأوردة الكبدية وتضخم في الفص الكبدية المذنب. كان هناك توسع شديد في الأوردة الوريد المفرد (azygos) الوريد الفرد الصغير السفلي (hemiazygos). أظهر التصوير المقطعي المحوسب للصدر مع الصبغة انسداد الوريد العضدي الرأسي الأيسر. على الرغم من أن الوريد الأجويف السفلي كان مفتوح، فقد لوحظت بؤر متكلسة متعددة داخل جدار الوريد. أظهر تصوير الأوردة انسداد كامل للوريد الأجويف السفلي فوق الكبد وانسدادًا كاملاً للوريد العضدي الرأسي الأيمن. تم إجراء استقراء للوريد العضدي الرأسي إلى الوريد الأجويف العلوي، متبوعًا بوضع دعامة مغطاة. تم عمل عدة قساطر بالبالون على مستوى الوريد الأجويف السفلي فوق الكبد مع وضع دعامة غير مغطاة.

الخلاصة: لقد كانت إعادة استقراء لكل من الوريد الأجويف السفلي فوق الكبد المغلق تمامًا والوريد العضدي الرأسي الأيمن مع وضع الدعامات المناسبة للمريض.

Keywords: BuddChiari syndrome, Systemic lupus erythematosus, Antiphospholipid syndrome, Sharp recanalization 2

1. INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare life-threatening condition and is caused by an obstruction to the hepatic venous outflow. The obstruction is often located in the level of the main hepatic veins, in the suprahepatic part of the inferior vena cava (IVC) or both [1]. BCS is classified into a primary or secondary depending on the underlying etiology behind the hepatic venous outflow obstruction. BCS is considered a primary if the source of obstruction is inside the vein and is usually caused by a prothrombotic condition, and a secondary if it was caused by a compression or a tumor outside the veins [2]. Regardless of the etiology, hepatic venous outflow obstruction leads to an increase in hepatic sinusoidal pressure and subsequently portal hypertension [3]. To avoid complications such as hepatic fibrosis and cirrhosis, the obstruction must be relieved by therapeutic interventions such as thrombolysis, percutaneous transluminal angioplasty, ± stent placement [4]. We report a case of a patient with BCS, systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS) with IVC/SVC stenosis that was treated by a simultaneous IVC/SVC sharp recanalization.

2. CASE REPORT

A 40-year-old female patient presented with distended abdomen. She has a history of systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS). The patient was on Warfarin and the dose upon admission was 7mg 4 days per week and 6mg 3 days per week. She had a history of right atrial mass resection, Mechanical Valve Replacement (MVR), thrombophilia with recurrent thrombotic events including pulmonary embolism,

for which she underwent thrombectomy, and IVC & SVC stenosis. Physical examination revealed ascites without jaundice or organomegaly. Laboratory investigations showed high Alkaline phosphatase and Gamma-glutamyl transferase, leukocytosis, hypochromia, and anisocytosis. The coagulation profile indicates prolonged Prothrombin time (PT), International Normalization Ratio (INR), and Partial Thromboplastin Time (PTT). A triphasic computed tomography (CT) scan revealed a heterogeneous, nutmeg appearing liver. In addition to, attenuation of hepatic veins and hypertrophy of the caudate with splenomegaly. Initially the ascitic fluid was drained under ultrasound (US) guidance. Under general anaesthesia two accesses were made. The right femoral vein and the right internal jugular vein. An inferior venacavogram through the right femoral vein demonstrated a complete occlusion of the suprahepatic IVC, extensive paravertebral collateral networks, and patent hepatic veins (Figure 1).

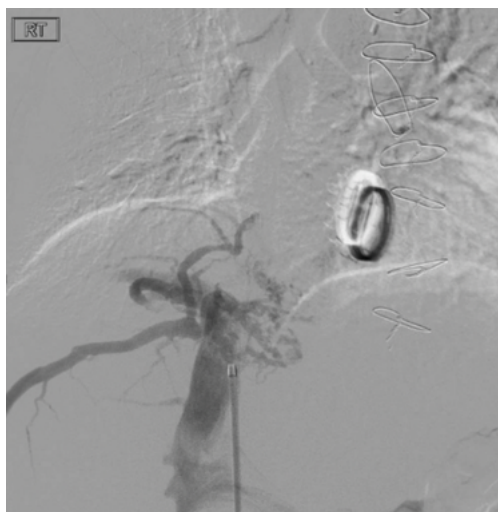


Figure 1 Suprahepatic IVC complete occlusion, extensive paravertebral collateral networks, and patent hepatic veins.

Another venogram through an access of the right internal jugular vein illustrated a complete blockage of the right brachiocephalic vein with extensive collaterals (Figure 2).

Multiple non-successful attempts were made to bypass the occluded segments at the IVC from the right femoral access and at brachio-cephalic vein through internal jugular access. A venogram through the right brachial vein access demonstrated similar findings to the venogram through the internal jugular vein. Infrarenal IVC venogram was obtained and showed a huge hypertrophied dilated azygos. Decision was made to advance a wire into the right atrium through the azygos vein. Through the jugular access, sharp recanalization of the brachiocephalic vein to the SVC was done to target the balloon within the SVC, which was advanced from the azygos vein. Then, an 8mmx4cm ARMADA balloon (Abbott Vascular, Santa Clara, CA, USA) was inflated in the SVC to right atrium to serve

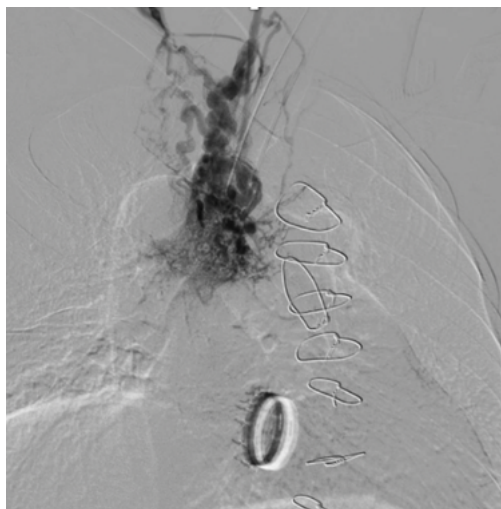


Figure 2 Right internal jugular vein showed complete obstruction at the right brachio-cephalic vein with extensive collaterals.

as a target for puncture for sharp recanalization, which was successfully achieved with a Chiba needle. Then a vascular sheath was advanced into the right atrium. Simultaneous venogram was made through the right femoral and right internal jugular access which revealed an occluded segment between the right atrium and the IVC with multiple collaterals. After multiple attempts of sharp recanalization, Through-and-through access between the vascular sheaths was obtained. Multiple balloon angioplasties were made at the level of suprahepatic IVC utilizing 2mmx4cm PACIFIC (Medtronic, Minneapolis, MN, USA), 10x4cm ARMADA (Abbott Vascular, Santa Clara, CA, USA), 12mmx4cm MUSTANG (Boston Scientific, Natick, MA, USA) and 16mmx6cm ATLAS (BD Interventional, Covington, GA, USA) balloons. These were followed by placing a non-covered 20mmx6cm VENOVO stent (BD Interventional, Covington, GA, USA). Post stent deployment showed significant improvement of contrast flow from the IVC to the right atrium (Figure 3).

Then the brachial cephalic to SVC stenosis was targeted with balloon angioplasty utilizing 8mmx4cm ARMADA balloon (Abbott Vascular, Santa Clara, CA, USA). This is followed by placing a covered stent graft 14mmx49mm BENTLEY BeGraft (Bentley InnoMed GmbH, Hechingen, Germany). Significant improvement of contrast flow is noted as well from the right jugular to the SVC (Figure 4).

The intraprocedural medication was a Total of 10,000Unit IV-Heparin every 30 min. The next day after the procedure the patient was feeling well and clinically stable. US showed the stent within the IVC with a patent IVC and hepatic veins without any sign of thrombosis. The latest US follow-up which showed a stable heterogeneous liver parenchyma due to BCS-related parenchymal changes without focal lesions. Also, the left

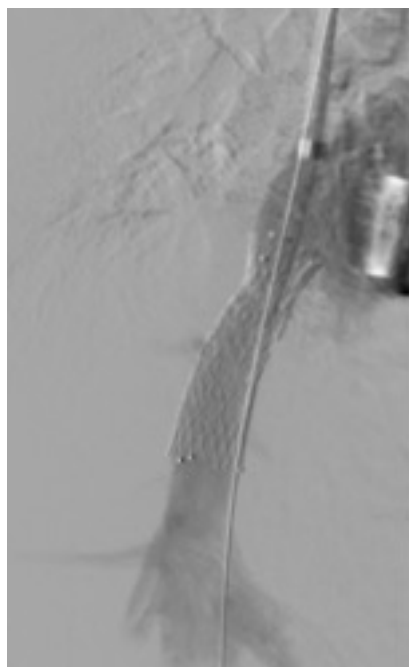


Figure 3 Successful recanalization for both BC/ SVC and suprahepatic IVC respectively.

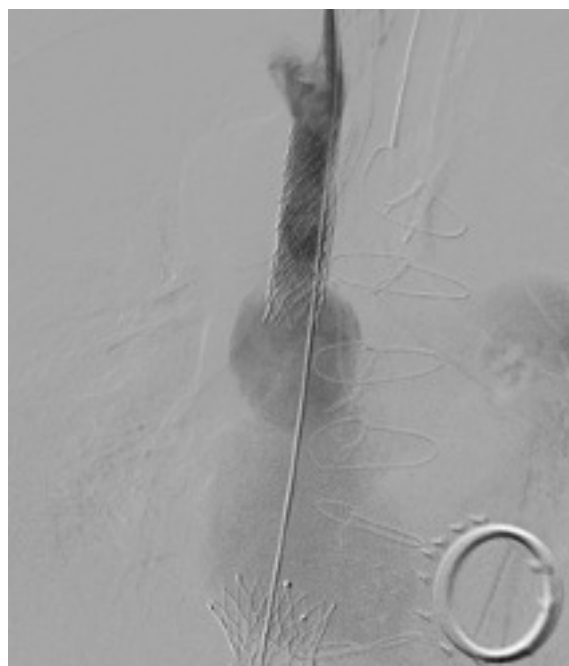


Figure 4 Successful recanalization for both BC/ SVC and suprahepatic IVC respectively.

hepatic vein was stable with a small non-occlusive focal thrombosis or web, otherwise patent IVC stent and hepatic vasculature. In the latest follow-up clinic, the patient is doing well with no active issue.

3. DISCUSSION

In this case, the patient was diagnosed with primary BCS that was caused by chronic thrombosis. BCS that are caused by thrombosis are usually treated with thrombolysis, TIPS, or liver transplant [5]. However, in our case, the selected approach was angioplasty and thrombolysis. Although this approach is preferred for a different aetiology of BCS, which is membranous occlusion, it was chosen for our case. Moreover, a study that was conducted by researchers from china supported this approach for patients with chronic thrombus [6]. In their study, out of their 39 chronic thrombus patients who were treated with a large balloon dilation and stent placement, only one of them failed due to extensive calcification of the thrombus. As for the risk of re-intervention for this approach, a case report that was conducted in India stated that recurrence and re-intervention rates are increased with balloon angioplasty alone. As a result, angioplasty with stent placement is preferred for recurrent cases or as long-term treatment for better results of occlusions associated with BCS [4]. Four months later, a post procedure ultrasound was done, which showed small non-occlusive thrombus in the left hepatic vein with patent IVC stent and vasculature. Clinically, the patient is doing well. However, long-term outcomes of our case could not be fully assessed after only four months of follow-ups.

4. CONCLUSION

It was a successful recanalization for the completely occluded suprahepatic IVC and placement of IVC stent and a successful recanalization for the completely occluded right brachiocephalic vein and placement of a stent. Patient was followed up and found clinically improved as well as significant improvement of contrast flow is noted post stenting and no complication were encountered.

CONFLICT OF INTEREST

The authors declare that the research was conducted without any commercial or financial relationships that could be construed as a potential conflict of interest.

ACKNOWLEDGEMENTS

N/A

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