AZJCVS

SURGICAL REPAIR OF CONGENITAL STENOSIS OF THE INFERIOR VENA CAVA

Seymur MUSAYEV
: https://orcid.org/0000-0001-9699-3614
Baku Health Centre, Baku, Azerbaijan

Emin GURBANOV
: https://orcid.org/0000-0002-2919-7922
Baku Health Centre, Baku, Azerbaijan

Namiq CABBAROV
: https://orcid.org/0000-0001-5471-9414
Baku Health Centre, Baku, Azerbaijan

Natik ALISHOV
: https://orcid.org/0000-0003-0106-042X
Baku Health Centre, Baku, Azerbaijan

Anar HASANOV
: https://orcid.org/0000-0001-9978-6397
Baku Health Centre, Baku, Azerbaijan

ARTICLE INFO

Article type:
Case report

Article history

Received:
18 September, 2020

Accepted:
29 November , 2020

Published:
29 January, 2021

ABSTRACT

The congenital stenosis of the inferior vena cava (IVC) is a rare anomaly causing numerous clinical manifestations, depending on the variant of drainage patterns or collaterals. This case presents a five-year-old Azerbaijani male, who suffered with massive ascites and a leg edema for three months. IVC stenosis was detected on echocardiography, due to a high velocity flow and visualisation of a focal narrowing between the IVC orifice and the proximal end of the hepatic vein entry. The IVC stenosis was confirmed by computed tomography. Surgical dilatation of the IVC using xeno-pericardial patchplasty was performed under the cardiopulmonary bypass. This successful surgical correction of IVC stenosis is the only case ever reported in Azerbaijan.

Key words: surgical repair, congenital stenosis, inferior vena cava, computed tomography , IVC stenosis

Volume 2, Issue 1, 2021. 66-73
DOI: 10.30546/azjcvs.2021.2.1.66

INTRODUCTION

Congenital variations and anomalies of the IVC are rare in clinical practice. They can lead to life-threatening complications if not identified, resulting in abdominal and renal surgery or interventional procedures (1).

A five-year-old Azerbaijani male was presented. He had been suffering with massive ascites, back pain and a leg edema for three months. Examination indicated obvious clinical evidence of an IVC obstruction. An echocardiogram assessment indicated that the IVC orifice was significantly narrowed (Figure 1).

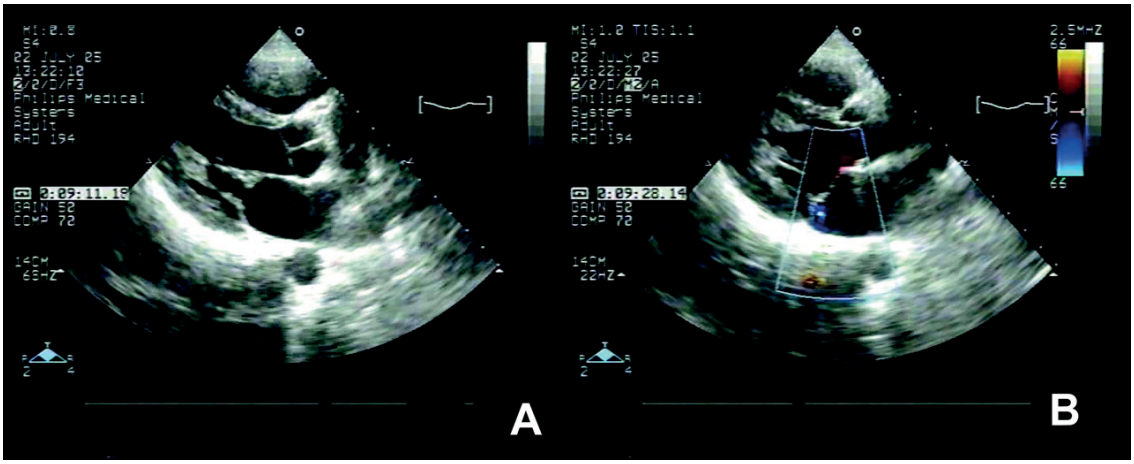

Figure.1.

Furthermore, a narrowing of the IVC proximal to the hepatic vein entry was noticed on sub-costal views measuring 4.3mm, when compared to a proximal segment of 14.7 mm and distal segment of 12.5 mm. Doppler studies of the IVC revealed a high velocity turbulent flow with a peak velocity of 1.24 m/s, and a peak gradient of 6 mmHg with a fusion of S/D waves on a parasternal RV inflow. Such findings were highly suggestive of IVC stenosis.

Additional imaging modalities were requested for further evaluation. Evaluation with the abdominal ultrasound detected 1200 ml of free intra-peritoneal fluid. This finding was highly suggestive of IVC stenosis. Computed tomography of the abdomen confirmed the presence of IVC stenosis proximal to the hepatic vein entry.

Surgical dilatation of the IVC was performed. A sternotomy was performed under general anesthesia. The right femoral vein and superior vena cava were cannulated and antegrade cold cardioplegia was given through the aorta. The superior vena cava was clamped. An incision on the diaphragm enabled around 1100 ml of IVC ascites to be flushed away. The body temperature of the patient was decreased to 29°C. The cannula was inserted in the IVC to aspirate the blood and keep the surgical field clear. The IVC was incised for about 1.5 cm up to the hepatic segment, and dilation of the IVC using xeno-pericardial patchplasty was performed. Drainage was placed near the diaphragm area. After rewarming, the patient was weaned from cardiopulmonary bypass (CPB). The sternotomy was closed in the classic manner and the patient was transferred to the intensive care unit (ICU). Subsequently, the patient was extubated after hours and after 20 hours was transferred to the ordinary cardiac ward. The drainage was removed on the third postoperative day, his total period in hospital being five days. The belly of the patient drastically reduced to its normal appearance. The postoperative

echocardiography undertaken prior to discharge showed no residual stenosis of the IVC.

DISCUSSION

Congenital stenotic lesions of the IVC are rare vascular defects and result from aberrant development of the IVC segments during embryogenesis (2,3). IVC stenosis is characterised by narrowing, mostly at the diaphragmatic level or hepatic segment of the IVC (3–6). The infrahepatic IVC may continue as the azygos (7) or hemiazygos vein, intrathoracic veins (5), or anomalous intrahepatic veins (8). The hepatic segment of the IVC then drains directly into the right atrium (3). To present the prevalence of congenital interruption or stenosis of the IVC, Koc *et al* analysed 7972 patients who had undergone consecutive routine abdominal multidetector row computed tomography. The existence of interruption (n=8) or congenital stenosis (n=4) of the IVC occurred in 12 (0.15%) of 7972 patients (1).

In many cases, IVC stenosis can be asymptomatic-and, in other cases, can lead to fluid accumulation and cardiopulmonary collapse (3,4). Adolescent males with the condition have shown to present such symptoms as spontaneous lower extremity deep venous thrombosis (DVT), leg swelling, leg pain, varices in the lower extremities, hepatic thrombosis and hematochezia (5). Chylothorax_has been occasionally observed as a symptom of congenital stenosis of the superior vena cava in infant patients (6). Ruggeri *et al*. (7) found an anomalous IVC in four of 75 young patients who were suffering with their first experience of DVT. Those authors estimated the prevalence of an anomalous IVC in that group of patients to be around 5.3%. Other symptoms which may become present after adolescence include venous hypertension, post-necrotic hepatic cirrhosis and portal hypertension from the manifestation of the condition (8).

Embryologic development of the IVC is a complex process that includes formation, regression, and fusion of the three longitudinal pairs of veins: postcardinal, subcardinal, and supracardinal (9,10). These processes occur between the fifth and seventh weeks of gestation, and five embryologic segments form the final structure of the IVC (11,12,13). In caudal-cranial order, these segments are: posterior cardinal veins (iliac segment), right supracardinal vein (subrenal segment), anastomosis between the right supra- and subcardinal veins (renal segment), right subcardinal vein (suprarenal segment), and the hepatocardiac canal (hepatic segment) (14).

Noninvasive imaging modalities, such as computed tomography (MDCT) and magnetic resonance imaging (MRI), are the most reliable methods for identifying these anomalies (10,15).

Interruption or stenotic lesion of the IVC can be diagnosed as absent or atretic segment/ segments of the IVC on contrast-enhanced computerised tomography (CT) or magnetic resonance (MR) angiography (5,7). There may also be associated findings, such as venous collaterals between upper and lower segments of the interruption or stenosis, azygos/hemiazygos continuation, DVT below the obstructed segment, and varices. The identification of dilated azygos/hemiazygos veins on CT or MRI are supplementary findings in the diagnosis. In our case, computed tomography indicated severe ascites, significant collateral veins around the stoma, and severe stenosis of the IVC.

Knowledge of the presence of variant venous anatomy also is important when undertaking surgical and interventional procedures. If well-developed azygos/hemiazygos continuation is present, the patient presumably will be asymptomatic, whereas the converse is true if such continuation is not present. The presence of acute or recurrent DVT, diffused varices, varicocele, hemorrhoids, venous

aneurysms, or venous collaterals (including the abdominal wall) in a relatively younger patient can be indicative of interruption or congenital stenosis of the IVC (16).

There is sparse data relating to surgical IVC reconstruction, and the clinical outcome is unclear. At present, the standard of treatment strategy includes endovascular IVC reconstruction for treatment of the chronic ilio-caval compression uncovered by thrombus removal and in patients at risk from venous leg ulcers (17). Data on endovascular IVC reconstruction has mainly been published for self-expanding steel alloy stents, predominantly from Wallstents (18–24). From the prospective Bern Venous Stent Registry, Sebastian *et al.* investigated the clinical outcome of 62 patients suffering with IVC obstruction who received endovascular reconstruction using dedicated venous nitinol stents. Both the Villalta score and revised venous clinical severity score (rVCSS) improved substantially, with 43% being free from any symptoms and signs of venous hypertension. Overall, freedom from postthrombotic syndrome (PTS) (Villalta score <5 points) was achieved in approximately two-thirds of the patients, only 10% of whom experienced no clinical improvement. Approximately one-third of the patients required endovascular reintervention to maintain stent patency. The cause of loss of patency was most likely stent thrombosis, due to impaired venous return through post-thrombotic leg inflow veins (25).

Finally, the three-month follow-up indicated no restenosis of IVC on echocardiography. The patient proved asymptomatic with good exercise capacity. Surgical treatment of the IVC stenosis was found to be safe, with acceptable short-term results. The limitation of the study is its status as a case report, with a longer follow-up being warranted to elucidate long-term results of the technique.

CONCLUSION

IVC stenosis is a rare disorder. The presence of DVT; diffused venous collaterals, including those in the abdominal wall; varices; varicocele; hemorrhoids; or venous aneurysms in relatively young patients are frequently seen in those suffering from interruption or congenital stenosis of the IVC and can be diagnosed via CT or MRI. Interrupted IVC is commonly asymptomatic, if associated with well-developed azygos/hemiazygos continuation, whereas commonly symptomatic if this is not present. The case study indicates that the surgical repair of congenital IVC stenosis can be successfully performed.

ABBREVIATIONS

IVC: Inferior vena cava

MRI: Magnetic resonance imaging

CPB: Cardiopulmonary bypass

ICU: Intensive care unit

DVT: Deep venous thrombosis

MDCT: Multi-detector computed tomography

CTA: Computed tomographic angiography

PTS: Postthrombotic syndrome

CONFLICT OF INTERESTS

The authors declare that there are no conflict of interests.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

FUNDING

None.

STUDY ASSOCIATION

This study is not associated with any thesis or dissertation work.

REFERENCES:

1. Koc, Z; Oguzkurt, L. *Interruption or Congenital Stenosis of the Inferior Vena Cava: Prevalence, Imaging, and Clinical Findings*. Eur J Radiol. 2006; 62 (2): pp. 257–66.

2. Siragusa S, Anastasio R, Falaschi F, Bonalumi G, and Bressan MA. *Congenital Absence of Inferior Vena Cava.* (letter) Lancet 2001; 357:1711

3. Harrison, D, Sullivan, P, Christman, G, and Takao, C. *Intravascular Stent Implantation for Refractory Chylothorax Secondary to Congenital Superior Vena Cava Stenosis in an Infant,* Structural Heart Disease. 2017; 3 (2): pp. 49–54

4. Halparin, J, Mongale P, and Newall, F. *Congenital Abnormalities of the Inferior Vena Cava Presented Clinically in Adolescent Males*. Thrombosis Research. 2015; 135 (4): pp. 648–651

5. *Congenital Heart Defects,* Medline Plus. U.S. Department of Health and Human Services, 2016

6. Bass, J; Redwine, M; Kramer, L; and Harris, J. *Absence of the Infrarenal Inferior Vena Cava with Preservation of the Suprarenal Segment as revealed by CT and MR Venography.* AJR 1999; 172: pp. 1610–1612

7. Ruggeri, M; Tosetto, A; Castaman, G; and Rodeghiero, F. *Congenital Absence of the Inferior Vena Cava: A Rare Risk Factor for Idiopathic Deep-Vein Thrombosis*. (letter) Lancet 2001; pp. 357–441

8. Smith, L. *A Case of Congenital Stenosis of the Inferior Vena Cava with Portal Hypertension.* British Journal of Surgery. 2018: 52 (11): pp. 913–916

9. Mathews, R; Smith, P; Fishman, E; and Marshall, F. *Anomalies of the Inferior Vena Cava and Renal Veins: Embryologic and Surgical Considerations.* Urology 1999; 53 (5): pp. 873–880

10. Bass, J; Redwine, M; Kramer, L; Huynh, P; and Harris Jr., J. *Spectrum of Congenital Anomalies of the Inferior Vena Cava: Cross-Sectional Imaging Fndings.* Radiographics 2000; 20 (3): pp. 639–652

11. Vijayaraghavan, S; Raja, V; and Chitra, T. *Interrupted Inferior Vena Cava and Left-Sided Subrenal Inferior Vena Cava: Prenatal Diagnosis.* J Ultrasound Med 2003; 22 (7): pp. 747–752

12. Ramanathan, T; Hughes, T; and Richardson, A. *Perinatal Inferior Vena Cava Thrombosis and Absence of the Infrarenal Inferior Vena Cava.* J Vasc Surg 2001; 33 (5): pp.1097–9

13. Drouillard, J; Bruneton, J; Elie, G, *et al*. *Congenital Malformations of the Inferior Vena Cava: Embryological, Anatomical and Radiological Study*. J Radiol Electrol 1978; 59 (12): pp. 669–677

14. Minniti, S; Visentini, S; and Procacci, C. *Congenital Anomalies of the Venae Cavae: Embryological Origin, Imaging Features and Report of Three New Variants.* Eur Radiol. 2002; 12 (8): pp. 2040–2055

15. Fishman, E. *From The RSNA Refresher Courses: CT Angiography: Clinical Applications in the Abdomen.* Radiographics 2001; 21: pp. 3–16

16. Gayer, G; Luboshitz, J; Hertz, M. *et al*. *Congenital Anomalies of the Inferior Vena Cava Revealed on CT in Patients with Deep Vein Thrombosis*. AJR Am J Roentgenol 2003;180 (3): pp. 729–732

17. O'Donnell Jr., T; Passman M. *Clinical Practice Guidelines of the Society for Vascular Surgery (SVS) and the American Venous Forum (AVF) and Management of Venous Leg Ulcers. Introduction.* J Vasc Surg 2014; p. 60 (Suppl): 1S-2S

18. Murphy, E; Johns, B; Varney, E; and Raju, S. *Endovascular Management of Chronic Total Occlusions of the Inferior Vena Cava and Iliac Veins*. J Vasc Surg Venous Lymphat Disord 2017; 5: pp. 47–59

19. Chick, J; Jo, A; Meadows, J; Abramowitz, S; Khaja, M; Cooper, K, *et al. Endovascular Iliocaval Stent Reconstruction for Inferior Vena Cava Filter-Associated Iliocaval Thrombosis: Approach, Technical Success, Safety and Two-Year Outcomes in 120 Patients.* J Vasc Interv Radiol 2017; 28: pp. 933–939

20. Delis, K; Bjarnason, H; Wennberg, P; Rooke, T; and Gloviczki; P. *Successful Iliac Vein and Inferior Vena Cava Stenting Ameliorates Venous Claudication and Improves Venous Outflow, Calf Muscle Pump Function and Clinical Status in Postthrombotic Syndrome.* Ann Surg 2007; 245: pp. 130–139

21. Partovi, S; Kalva, S; Walker, T; Taj, S; and Ganguli, S. *Long-Term Follow-Up of Endo-Vascular Recanalisation of Chronic Inferior Vena Cava Occlusion Secondary to Inferior Vena Cava Filters.* Vasa 2017; 46: pp. 121–126

22. Grotta, O; Enden, T; Sandbaek, G; Gjerdalen, G; Slagsvold, C; and Bay, D, *et al. Patency and Clinical Outcome after Stent Placement for Chronic Obstruction of the Inferior Vena Cava.* Eur J Vasc Endovasc Surg 2017; 54: pp. 620–628

23. Ye, K; Lu, X; Li, W; Yin, M; Liu, X; and Qin J, *et al. Outcomes of Stent Placement for Chronic Occlusion of a Filter-Bearing Inferior Vena Cava in patients with Severe Post-Thrombotic Syndrome*. Eur J Vasc Endovasc Surg 2016; 52: pp. 839–846

24. Meltzer, A; Connolly, P; Kabutey, N; Jones, D; and Schneider, D. *Endovascular Recanalisation of Iliocaval and Inferior Vena Cava Filter Chronic Total Occlusions*. J Vasc Surg Venous Lymphat Disord 2015; 3: pp. 86–89

25. Sebastian, T; Dopheide, J; Engelberger, R; Spirk, D. and Kucher, N. *Outcomes of Endovascular Reconstruction of the Inferior Vena Cava with Self-Expanding Nitinol Stents. Journal of Vascular Surgery: Venous and Lymphatic Disorders.* 2017; 11.012

Cite this article as: MUSAYEV Seymur, GURBANOV Emin, CABBAROV Namiq, ALISHOV Natik, HASANOV Anar. Case Report: Surgical repair of Congenital stenosis of the Inferior vena cava. *Azerbaijan Journal of Cardiovascular Surgery,* Volume 2, Issue 1, 2021, pp. 66-73

XÜLASƏ

Klinik hal:
Aşağı boş venanın anadangəlmə stenozunun cərrahi müalicəsi

Seymur MUSAYEV
Bakı Sağlamlıq Mərkəzi, Bakı, Azərbaycan

Emin QURBANOV
Bakı Sağlamlıq Mərkəzi, Bakı, Azərbaycan

Namiq CABBAROV
Bakı Sağlamlıq Mərkəzi, Bakı, Azərbaycan

Natik ALIŞOV
Bakı Sağlamlıq Mərkəzi, Bakı, Azərbaycan

Anar HƏSƏNOV
Bakı Sağlamlıq Mərkəzi, Bakı, Azərbaycan

Aşağı boş venanın (IVC) anadangəlmə stenozu, drenaj şəbəkəsinin və yaxud kollateralların versiyasından asılı olaraq, çoxsaylı klinik təzahürlərə səbəb olan nadir anomaliyadır. Hazırkı halda söhbət üç ay ərzində massiv assitdən və ayaq şişməsindən əziyyət çəkən 5 yaşlı Azərbaycanlı oğlandan gedir. Exokardioqrafiya zamanı IVC dəliyi ilə hepatik vena girişinin proksimal ucu arasında yüksək sürətli axın və fokal daralma ilə şərtlənən IVC stenozu aşkar olunmuşdur. IVC stenozu kompüter tomoqrafiyası ilə təsdiqləndi. Ksenoperikardial plastikadan istifadə etməklə IVC-nin cərrahi diletasiyası süni qan dövranı şəraitində həyata keçirilmişdir. IVC stenozunun bu uğurlu cərrahi korreksiyası Azərbaycanda indiyədək qeydə alınmış yeganə haldır.

Açar sözlər: Cərrahi müalicə, anadangəlmə stenoz, aşağı boş vena, kompüter tomoqrafiyası, IVC stenoz

РЕЗЮМЕ

Клинический случай:
Хирургическое лечение врожденного стеноза нижней полой вены

Сеймур МУСАЕВ
Бакинский Центр Здоровья, Баку, Азербайджан

Эмин ГУРБАНОВ
Бакинский Центр Здоровья, Баку, Азербайджан

Намик ДЖАББАРОВ
Бакинский Центр Здоровья, Баку, Азербайджан

Натиг АЛЫШОВ
Бакинский Центр Здоровья, Баку, Азербайджан

Анар ГАСАНОВ
Бакинский Центр Здоровья, Баку, Азербайджан

Врожденный стеноз нижней полой вены (НПВ) - редкая аномалия, вызывающая многочисленные клинические проявления в зависимости от версии дренажной сети или коллатералей. В данном случае речь идет о 5-летнем азербайджанском мальчике, который в течение трех месяцев страдает массивным асцитом и отеком ног. Эхокардиография выявила стеноз НПВ, вызванный высокоскоростным потоком и очаговым сужением между отверстием НПВ и проксимальным концом входа в печеночную вену. Стеноз НПВ подтвержден компьютерной томографией. Хирургическая дилатация НПВ с помощью ксеноперикардиальной пластики выполнена в условиях искусственного кровообращения. Эта успешная хирургическая коррекция стеноза НПВ – является пока единственным зарегистрированным случаем в Азербайджане.

Ключевые слова: Хирургическое лечение, врожденный стеноз, нижняя полая вена, компьютерная томография, стеноз НПВ