Case Report

Failed Percutaneous Mitral Commisurotomy due to congenital anomaly of inferior vena cava

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Abstract

Since its inception in 1982, percutaneous transvenous mitral commissurotomy (PTMC) has gained increasingly wide use internationally in patients with Rheumatic Mitral stenosis. PTMC offers an alternative to surgery in patients who have pliable mitral valve.Very few reasons were reported to abort the procedure when patient lies on the table. This study presents two case reports in which congenital venous anomalies were one of the reasons to terminate the procedure.

Keywords: Percutaneous transvenous mitral commissurotomy, Rheumatic mitral stenosis, Pliable mitral valve.

Introduction

Percutaneous mitral commissurotomy (PTMC) has been approved as a standard treatment in patients with severe rheumatic mitral stenosis with suitable valve score.[1,2] The procedure is technically successful in over 90% patients, and the long-term durability of commissurotomy is excellent in those with pliable mitral valve leaflets and minimally deformed submitral apparatus.The results are comparable to surgical commissurotomy in relieving the obstructed valve and maintaining a favourable outcome.[2-7] Majority of cases undergo the procedure successfully using traditional transfemoral approach. However, in some rare cases, venous drainage abnormalities or cardiac anatomic disorientation may cause impediments to successful completion of the procedure. Recently, we came across cases which lead to termination of the procedure due to a congenital anomaly of IVC. Although advances in imaging techniques have facilitated early diagnosis of various systemic venous anomalies but most of these patients remain asymptomatic. A persistent left superior vena cava (SVC) with a normal right SVC is most commonly encountered. Anomalies of the inferior vena cava (IVC) and its tributaries are also well known.[8] Mazzucco and colleagues.[9] reviewed the association of IVC anomalies with congenital cardiac anomalies. This study describes two cases of the rarest form of IVC anomaly associated with a valvular lesion.

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Case Report-1:

A 25-year-old pregnant woman, G1P1+0, without any known co morbid was admitted with complaints of progressive shortness of breath (New York Heart Association functional class II-III) for last two months. Cardiovascular examination revealed sinus rhythm, a loud 1st heart sound, and a mid-diastolic rumbling murmur at the apex. On series of investigation, the patient was diagnosed as having acquired rheumatic mitral stenosis. Echocardiography showed an enlarged left atrium and mitral stenosis with a valve area of 1 cm^2. There was associated tricuspid regurgitation with moderate pulmonary artery hypertension with an estimated mean pulmonary artery pressure of 70mm Hg. Chest radiography indicated mild cardiomegaly. Initially she was treated conservatively with diuretics, digitalis, and penicillin prophylaxis for endocarditis and later scheduled for balloon mitral valvuloplasty. After premedication with intravenous alprazolam 1 mg, the procedure was performed under subcutaneous local anaesthetic infiltration with lidocaine, and monitored by electrocardiogram, O2 saturation by spirometry, and invasive blood pressure recordings via the right femoral artery. The right femoral vein was punctured, but the guidewire did not pass through. A radiopaque dye injected through the cannula showed a narrow and tortuous course of the right IVC. Dye injected at this level and at the femoral level, it was seen to travel to the RA bilaterally on both sides of the spine. The procedure was abandoned, and the patient was advised to have a computed tomography (CT) venogram followed by open mitral commissurotomy.

Case Report-2:

A 21 years old male was admitted in NICVD with history of palpitation and shortness of breath on exertion for two years. On clinical examination, he had features consistent with Mitral stenosis. Echocardiographic evaluation showed that he had' Severe Mitral stenosis with moderate Pulmonary Hypertension'. Wilkins score was six. We

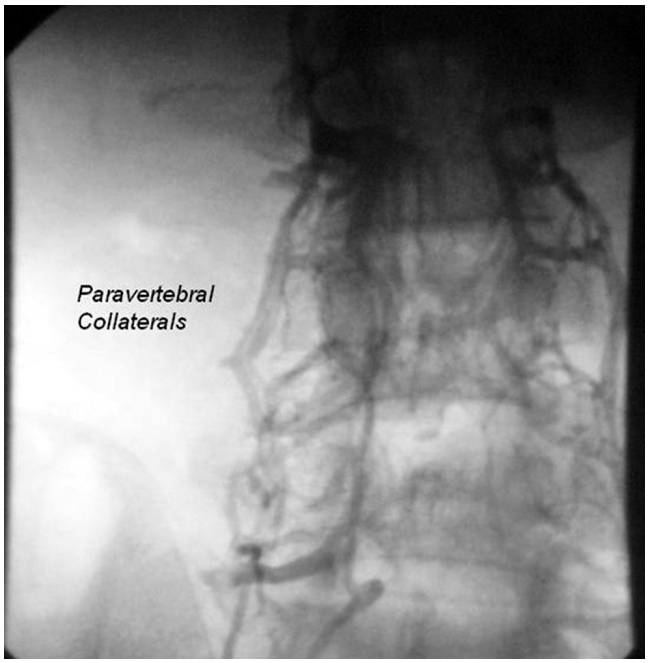

Figure-1: Fluoroscopic Image of IVC Anomaly.

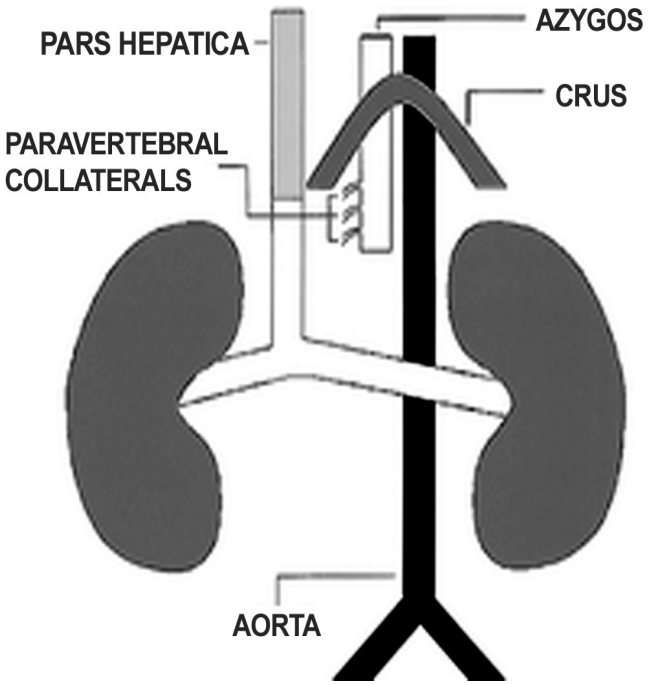

Figure-2: Diagrammatic Illustration of Anomaly of IVC.

decided to treat the patient with PTMC. PTMC via traditional femoral approach was scheduled. Cannulation of right femoral vein disclosed an incidental finding of anomalous venous drainage (Figure-1).

Discussion

The embryogenesis of the IVC is a complex process involving the formation of several anastomoses between three paired embryonic veins. The result is numerous variations in the basic venous plan of the abdomen and pelvis. Briefly, the normal IVC is composed of 4 segments: hepatic, suprarenal, renal, and infrarenal. The hepatic segment is derived from the vitelline vein. The right subcardinal vein develops into the suprarenal segment by subcardinal-hepatic anastomoses. The renal segment develops from right supra-subcardinal and post-subcardinal anastomoses. In the thoracic region, the supracardinal veins give rise to the azygos and hemiazygos veins. Progressive changes in the abdomen lead to replacement of the postcardinal veins by the subcardinal and supracardinal veins. In the pelvic region, the postcardinal veins persist as the common iliac veins.[10] This complex process of IVC formation results in several variations in systemic venous return.[11-13]

In few cases there is an absence of the hepatic segment of the IVC along with the liver; below this level, the IVC and left renal vein were draining into the dilated collateral paravertebral veins and the hepatic veins were draining into an enlarged suprahepatic segment of the IVC. Dilated collateral paravertebral veins were draining into the dilated azygos vein that joins the SVC at the normal location. Absence of the hepatic segment of the IVC with azygos continuation has also been called the azygos continuation of the IVC. The prevalence of this entity was reported as 0.6%.[14] However in our patients interrupted IVC was joined with both side collateral paravertebral veins at the level of the renal veins and the collateral paravertebral veins were draining into the azygos vein (Figure-2). The association of such anomalies with a symptomatic disorder leads to incidental diagnosis, as in these cases.

Radiological diagnosis by CT has led to early recognition of such anomalies, which has been helpful in early treatment. Although vascular structures can usually be readily identified on contrast-enhanced CTscans, identification of unusual venous arrangements may be difficult in those cases in which intravenous contrast material is contraindicated. Among such patients, MR imaging may be used to distinguish aberrant vessels from masses by demonstrating flow voids or flow-related enhancement. A working knowledge of IVC and renal vein anomalies is essential to avoid diagnostic pitfalls.

Limitations:

Non-invasive procedures as CT scan could not be

performed on both cases due to financial constraints.

References

1. No author listed. Multicenter experience with balloon mitral commissurotomy: NHLBI Balloon Valvuloplasty Registry Report on immediate and 30-day follow-up results. The National Heart, Lung, and Blood Institute Balloon Valvuloplasty Registry Participants. Circulation 1992; 85: 448-61.
2. Reyes VP, Raju BS, Wynne J, Stephenson LW, Raju R, Fromm BS, et al. Percutaneous balloon valvuloplasty compared with open surgical commissurotomy for mitral stenosis. N Engl J Med 1994; 331: 961-7.
3. Turi ZG, Reyes VP, Raju BS, Raju AR, Kumar DN, Rajagopal P, et al. Percutaneous balloon versus surgical closed commissurotomy for mitral stenosis. A prospective, randomized trial. Circulation 1991; 83: 1179-85.
4. Patel JJ, Shama D, Mitha AS, Blyth D, Hassen F, Le Roux BT, et al. Balloon valvuloplasty versus closed commissurotomy for pliable mitral stenosis: a prospective hemodynamic study. J Am Coll Cardiol 1991; 18: 1318-22.
5. Arora R, Nair M, Kalra GS, Nigam M, Khalilullah M. Immediate and long-term results of balloon and surgical closed mitral valvotomy: a randomized comparative study. Am Heart J 1993; 125: 1091-4.
6. Bueno R, Andrade P, Nercolini D. Percutaneous balloon mitral valvuloplasty vs open mitral valve commissurotomy. A randomized clinical trial (abstract). J Am Coll Cardiol 1993; 21: 429.
7. Ben Farhat M, Ayari M, Maatouk F, Betbout F, Gamra H, Jarra M, et al. Percutaneous balloon versus surgical closed and open mitral commissurotomy: seven-year follow-up results of a randomized trial. Circulation 1998; 97: 245-50.
8. CAMPBELL M, DEUCHAR DC. The left-sided superior vena cava. Br Heart J 1954; 16: 423-39.
9. Mazzucco A, Bortolotti U, Stellin G, Gallucci V. Anomalies of the systemic venous return: a review. J Card Surg 1990; 5: 122-33.
10. Philips E. Embryology, normal anatomy and anomalies. In: Ferris EJ, Hipona FA, Kahn PC, Phillips E, Shapiro JH, editors. Venography of the inferior vena cava and its branches. Baltimore, MD: Williams & Wilkins, 1969: 1-32.
11. Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. Radiographics 2000; 20: 639-52.
12. Bass JE, Redwine MD, Kramer LA, Harris JH Jr. Absence of the infrarenal inferior vena cava with preservation of the suprarenal segment as revealed by CT and MR venography. AJR Am J Roentgenol 1999; 172: 1610-2.
13. Nagashima T, Lee J, Andoh K, Itoh T, Tanohata K, Arai M, et al. Right double inferior vena cava: Report of 5 cases and literature review. J Comput Assist Tomogr 2006; 30: 642-5.
14. Ginaldi S, Chuang VP, Wallace S. Absence of hepatic segment of inferior vena cava with azygous continuation. J Comput Assit Tomogr 1980; 4: 112-4.