Discovering persistent left superior vena cava (PLSVC) during Pacemaker implantation

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Abstract: Persistent left superior vena cava (PLSVC) is rare but important congenital vascular anomaly. It results when the left superior cardinal vein caudal to the innominate vein fails to regress. It is most commonly observed in isolation but can be associated with other cardiovascular abnormalities including atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus and ostial atresia. The presence of PLSVC can render access to the right side of heart challenging via the left subclavian approach, which is a common site of access utilized when placing pacemakers. We report on a rare case of persistent left superior vena cava (PLSVC) with absent right superior vena cava (RSVC). This venous malformation was identified incidentally in a 25-year-old male during Permanent pacemaker. Isolated PLSVC is usually asymptomatic but it can pose difficulties for establishing central venous access, pacemaker implantation. This condition is also associated with an increased incidence of congenital heart disease, arrhythmias and conduction disturbances. A wide spectrum of clinicians should be aware of this anomaly, its variations and possible complications.

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1.Introduction

Persistent left superior vena cava (PLSVC) is the most common congenital malformation of the thoracic venous system and it affects about 0.3 to 0.5% of the general population^{1,2}. PLSVC is usually discovered incidentally. PLSVC is generally coexistent with right superior vena cava (RSVC), but rarely associated with absent RSVC. Isolated PLSCV is very uncommon, occurring in 0.07 to 0.13% of patients who have congenital heart defects .Nearly half of the patients with isolated PLSVC have other cardiac malformations, such as atrial septal defect, endocardial cushion defect or tetralogy of fallot³. Its prevalence is in fact much higher in patients with congenital cardiac abnormalities compared to general population, ranging from 2.8% to 4.3% ⁵ in addition, about 10% of these subjects with congenital cardiac abnormalities do not have a right superior vena cava (RSVC)⁴⁻⁷.

Precise anatomical knowledge of the great vessels of the neck is essential for safe anesthesia, pacemaker implantation and cardiac surgery. A wide spectrum of clinicians should be aware of this anomaly, its variations and possible complications. In this case report we present a patient with isolated PLSCV who had undergone an implantation of a dual chamber pacemaker system.

2.Case Presentation

A 25-year-old male patient complained of shortness of breath (SOB), dizziness, fatigue and chest pain was presented to the cardiology clinic. The patient has history of closure of ventricular septal defect (VSD) before 5 years. His electrocardiograms (ECG) revealed a sinus rhythm of 51 beats/minute. A normal chest X-ray and Echocardiogram was obtained however the Holter monitor recording clearly indicated sinus bradycardia. Thus, considering the state of sinus bradycardia and as the patient was symptomatic, it was planned to implant a dual chamber permanent pacemaker. Usually left-sided approach is considered during the procedure several attempts was carried out to position the ventricular lead through left subclavian vein but failed.

During pacing, the high threshold of >10mV of ventricular lead raised the suspicion of persistent left superior vena cava. A right bundle branch block morphology also indicated that the lead is towards left ventricle. In case of LSVC persistence, the lead enters the subclavian vein and does not cross the column, but lays parallel to it before entering the coronary sinus posteriorly to the right ventricle. A venogram was done, which confirmed the presence of isolated PLSVC i.e. absence of RSVC. We observed a large PLSVC draining into the coronary sinus. It showed a bridging vein draining the right jugular and right subclavian veins; it joined the left brachiocephalic vein and formed the PLSVC, which descended at the left side of the mediastinum leftward of the pulmonary artery and left atrium (LA) before draining into the right atrium (RA) via a dilated coronary sinus. The RSVC was absent and the PLSVC carried all venous blood from the head, neck and upper extremities.

A permanent dual chamber pacemaker was planned. The guide wire was introduced into the left subclavian vein by a puncture and fluoroscopy revealed their unusual left-sided downward course; venography confirmed the presence of PLSVC draining into a large coronary sinus. A Medtronic model, Sensia SESR01 (Medtronic, Inc., Minneapolis, MN, USA) was used. The guidance of the ventricular lead was achieved after manually forming the shape of a stylet with a proximal bend and a distal rounded curve. Several attempts was carried out but failed. Vein. Eventually, the ventricular lead was positioned at the apex of the right ventricle through a puncture of the right subclavian vein. The lead presented optimal measured parameters intraoperatively (R wave > 12mV, impedance of 524 ohms and threshold of 0.4 mv). These values were maintained in the system interrogation which was performed during the one month follow-up visit. Several attempts were done to position the atrial lead but were failed.

3.Discussion

In early embryonic development the venous blood of the upper part of the body drains into the right atrium via two bilateral and symmetrically running veins (left and right anterior cardinal veins).^{2,4} At approximately eight week gestation, the left brachiocephalic vein develops a bridge between the left and right cardinal veins. The of the left anterior cardinal vein below this anastomosis usually collapse and degenerates, leaving only the right anterior cardinal vein which becomes anastomosed to the superior vena cava. A subtype of PLSVC is isolated PLSVC where the RSVC is absent resulting in drainage of venous blood from the head and both arms through the left brachiocephalic vein, the PLSVC and coronary sinus into the right atrium.

Isolated cases of LSVC persistence in undergoing permanent pacemaker patients implantation have reported in literature; the most common problems related to the unusual anatomic access to the heart were reaching a convenient pacing site and ensuring stable lead placement. When, isolated LSVC persistence is usually not recognized until a left superior approach to the heart is required, when it becomes a relevant anatomic finding. The left sided approach could complicate the positioning of left side-pacemaker leads. The physician's defined different steps to overcome implant related difficulties: hand shaping the stylet in predefined fashion, using active fixation leads, and moving to the right-sided implant after right peripheral venography.

In patients with poor handling through the coronary sinus, a right approach is recommended after visualization of a right vena cava entering the right atrium by contrast venography or echocardiography; in absence of RSVC an epicardial implantation is suggested. The right-sided approach requires a longer procedure time; contrast venography compared to leftsided approach and also may cause greater discomfort to the patients.

Positioning a pacemaker lead through the PLSVC and coronary sinus in the right ventricle may be difficult in these patients⁵. After we had demonstrated the anatomical relations in our patient we decided to introduce the pacemaker leads via the PLSVC through the left brachiocephalic vein .The presence of PLSVC complicates the implantation of pacemaker while it not feasible to attempt right approach due to concomitant absence of the right SVC. The recognition of a markedly dilated coronary sinus (CS) without right atrial dilatation in a standard echocardiography may raise the suspicion of PLSCV. Radiographic features or clinical examination findings that may also suggest the presence of a PLSCV include a shadowy configuration on chest X-ray and on ECG.

The practical outcome of the different policies was assessed by lead performance, total fluoroscopy time and total procedure time. Technical difficulties associated with PLSCV-related anatomic peculiarities need to be addressed during pacemaker implantation. The major barrier that has to be overcome is the steering of the ventricular lead through the PLSVC, the coronary sinus and then through the tricuspid annulus in the right ventricle. When the lead is advanced out of the ostium of the CS it points away from the tricuspid orifice and towards the opposite right atrial wall. Therefore, the use of a pre-shaped stylet is needed in order to redirect its course around the tricuspid annulus and through the valve. Another obstacle to device implantation in patients with PLSCV is the length of the leads used. Taking into consideration the circuitous, elongated course of the ventricular lead along the PLSVC, CS and right ventricle, the length of conventional leads may be inadequate and therefore longer leads may be necessary. In addition, it is a common practice to favor the use of the active fixation leads in order to minimize the risk of subsequent lead dislodgement.

Attempts to define the anatomy of the superior caval system before pacemaker implantation are not routine because anomalies are rare and because the most commonly encountered anomaly, a persistent left superior vena cava, does not preclude successful lead placement. When the pacemaker lead enters the right atrium through the persistent left superior vena cava and the coronary sinus it often forms a loop in the right atrium and then enters the right ventricle. Rarely a lead may reach the right ventricle without forming a loop or enter a branch of the coronary sinus; these two courses are sometimes difficult to distinguish in the anterior-posterior view. Fortunately, ventricular pacing (usually from a branch of the coronary sinus) is safe long term procedures.

Appropriate curving of the stylet, without complete "closing" of a loop, as in our cases, facilitated insertion of a lead into the tricuspid orifice. In conclusion, we have presented a case report of the patient where the isolated PLSVC was discovered during the implantation of PM, had to consecutively undergo successive observations to diagnose the anomaly and opt for different approach. In right approach, the ventricular lead was positioned at the apex of the right ventricle .The post-operative and follow-up observations of the patient was satisfactory.

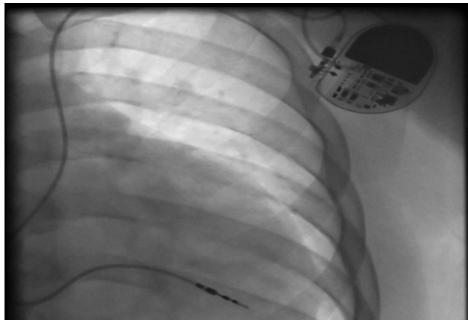


Fig 1. Left-sided approach: Figure reveals the course taken by the ventricular lead.

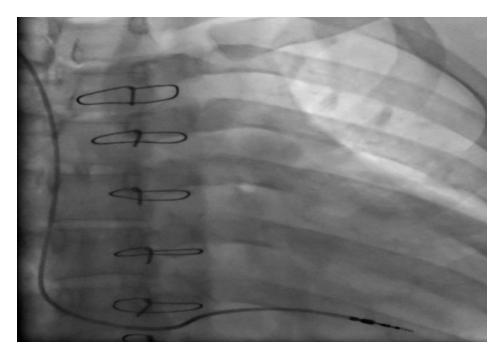


Fig. 2. Right- sided approach: Figure reveals the course taken by the ventricular lead and successful placing of the ventricular lead at the apex of right ventricle.

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