Atrioventricular Block in a Patient with Persistent Left Superior Vena Cava and Absent Right Subclavian Vein: A Case Report

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ABSTRACT

Persistent Left Superior Vena Cava (PLSVC) is an uncommon congenital anomaly, with an incidence of 0.5% in the general population. Moreover, the right superior vena cava anomaly with PLSVC is even rarer, with an incidence of 0.09%-0.13% in the general population. Patients with PLSVC and an absent right subclavian vein may present complications when undergoing conventional pacemaker lead implantation due to their vein anomalies. In this case report, a leadless pacemaker proved to be a useful alternative in a patient with the complete atrioventricular block with PLSVC and an absent right subclavian vein defect, the latter being found incidentally during the initial pacemaker implantation. This case highlights the importance of evaluating the presence of vascular malformations before surgical intervention.

INTRODUCTION

Persistent Left Superior Vena Cava (PLSVC) is an uncommon congenital anomaly; it has an incidence of 0.5% in the general population. A concurrent right superior vena cava anomaly with PLSVC is even rarer, with an incidence of 0.09%-0.13% in the general population. Patients with PLSVC are usually asymptomatic, and PLSVC is incidentally detected by Computed Tomography (CT) and catheter angiography, or during complicated surgical procedures, such as pacemaker implantation [1].

In this report, we present a case of an elderly patient with PLSVC and concurrent occurrence of absent right subclavian vein and Complete Atrioventricular Block (CAVB). In clinical settings, both a CAVB and an absent right subclavian vein are rarely encountered in PLSVC. Leadless pacemakers, an emerging technology, is an effective treatment option for patients with such vascular anomalies, including the present case.
CASE PRESENTATION

A 79-year-old man with a history of colon carcinoma presented to a local hospital with complaints of dizziness and staggering while walking. Bradycardia was suspected as CAVB-induced bradyarrhythmia. The same day, he was transferred and admitted to our hospital. At the time of admission, a physical examination revealed a height of 162 cm, body weight of 50 kg, blood pressure of 129/78 mmHg, pulse rate of 25 beats/min, normal lung sounds, and absence of murmurs. Chest radiography showed no abnormalities, while Electrocardiography (ECG) revealed CAVB, complete right bundle branch block, and no ST changes (Figure 1). Transthoracic echocardiography revealed an ejection fraction of 60% without asynergy or valvular disease. Since the ECG suggested symptomatic CAVB, emergency temporary pacemaker (Abbott Medical Japan) implantation was performed from the right jugular vein prior to the permanent pacemaker implantation, which was attempted 3 days later.

The initial treatment plan was to approach the left axillary vein. Preoperative left axillary venography and angiography confirmed the presence of PLSVC (Figure 2A) and the innominate vein and Superior Vena Cava (SVC), respectively. Given that the guidewire (guidewire 0.038 inches, 8451694, Abbot Medical Japan) also passed smoothly through the innominate vein to the SVC, we decided on the left axillary vein approach, as originally planned, to implant leads in the right atrium and ventricle through the innominate vein. However, although the sheath (sheath introducer: 405104, Abbot Medical Japan) could be placed from the left axillary vein to the SVC via the innominate vein, it was difficult to pass the leads (Tendril STS: 2088TC, Abbott Medical Japan) through the sheath because of vein tortuosity. Venography was performed after the patient complained of pain, which confirmed that the lead had perforated the innominate vein (Figure 2B). To investigate the feasibility of approaching from the right axillary vein, right brachial venography was performed; however, the right subclavian vein was absent, and the procedure was suspended (Figure 2C). For further evaluation, plain CT was performed for chronic kidney disease, and tortuosity of the innominate vein passing in front of the heart was confirmed (Figure 3).

Thus, it was assumed that lead insertion would be difficult for both approaches—the left and right axillary veins; therefore, we decided to perform the implantation with a leadless pacemaker. On day 8 of hospitalization, a right femoral vein approach was undertaken and a leadless pacemaker (Micra TPS, Medtronic Japan) was placed in the right ventricle (Figure 4). The postoperative course was good, and the patient was discharged on day 17 of hospitalization.

Written informed consent to publish this case report was obtained from the patient, and the publication procedure was approved by the ethics committee of our hospital (No.2022-08-16-1).

Figure 1: Electrocardiogram on admission showed a complete atrioventricular block.
DISCUSSION

PLSVC usually drains into the right atrium through the coronary sinus. However, in rare cases in which the PLSVC presents an absent right superior vena cava. A right-to-left shunt usually does not occur; occasionally, direct flow into the left atrium may occur [1].

PLSVC makes catheterization and pacemaker lead implantation difficult. Complications such as shock and cardiac arrest may occur as a result of complicated catheter manipulation [2]. Preoperative diagnosis and procedure consideration is important.

The leadless pacemaker was developed to overcome the weaknesses of the conventional pacemaker: infection, tricuspid regurgitation, and lead failure [3-5]. The original indications for leadless pacemaker are chronic atrial fibrillation with atrioventricular block, normal sinus rhythm
with second or third-degree AVB and a low level of physical activity or short expected lifespan, and sinus bradycardia with infrequent pauses or unexplained syncope with abnormal electrophysiological findings such as prolonged HV interval. Congenital vascular malformations were not included [5-7].

Although the efficacy of leadless pacemakers for CAVB associated with congenital SVC deficiency has been recently reported [8], a leadless pacemaker-implanted CAVB case with PSLSV and an absent right subclavian vein defect is sporadically mentioned in literature.

In a report of leadless pacemaker implantation in combination with a subcutaneous implantable cardiac defibrillator, the usefulness of the technique for patients in whom it is difficult to insert a pacemaker lead into the right atrium and ventricle due to venous malformation has been clearly demonstrated [9]. Therefore, leadless pacemakers will be beneficial in device-implantation therapies for venous malformations in which it is difficult to insert leads and implant devices.

In our case, a prior CT scan was not performed as there was no known vascular malformation. Therefore, intraoperative venography of the right axillary vein confirmed, for the first time, the venous anomaly as a complication of PSLSV and its inflow into the coronary sinus.

Initially, lead insertion was determined to be possible by confirming the normality and blood flow of the blood vessels with selective angiography using a sheath and guidewire. However, because the lead could only be delivered till the SVC and could not reach the right atrium and ventricle, the insertion of leads through the left axillary vein was interrupted. We considered implantation from the right axillary vein and performed venography of the right brachial vein that showed hypoplasia of the right subclavian vein. CT revealed severe tortuosity of the innominate vein. These findings led us to abandon conventional pacemaker placement; instead, we decided to perform leadless pacemaker implantation. Thus, we were able to complete the procedure without complications and in a short duration (55 minutes).

Considering that CT showed severe tortuosity of the innominate vein, intraoperative venography and two-dimensional image evaluation using a guidewire alone were not sufficient for the initial evaluation. Therefore, this case highlights the importance of preoperative CT evaluation.

A non-replaceable battery with a short battery life of 4.7 years is the major limitation of leadless pacemakers [3]. In our case, the patient was 79 years old; considering the average male life expectancy of 81 years in Japan, the short battery life was not of concern. On the other hand, even in patients with venous malformation and other conditions in which conventional pacemaker implantation is difficult, it is necessary to consider whether there are any contraindications for leadless pacemakers, including age or complication risks [10].

CONCLUSION

Leadless pacemakers are effective in patients with CAVB and PSLSV with an absent right subclavian vein. This case highlights the importance of confirming a vascular anomaly prior to surgical interventions and consideration of individual adaptation for the leadless pacemaker.

CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest.

References