Wolff-Parkinson-White Syndrome and a Left Atrial “Mass” – Is There a Link? –

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Figure 1. CMR imaging still frames showing left-sided SVC (LSVC). (a) Transverse T1-weighted image showing right-sided SVC, LSVC and no bridging veins. (b) Still frame of SSFP: 4-chamber view, and intra-atrial course of LSVC. (c) CMR-SSFP systolic frame. (d) CMR-SSFP diastolic frame. (e) CMR aortogram-phase reconstruction showing a “bovine arch” arrangement. CMR, cardiac magnetic resonance; CS, coronary sinus; SSFP, steady-state free precession; SVC, superior vena cava.

Left-sided superior vena cava (LSVC) a common incidental finding during imaging, invasive procedures and/or surgery. It may be associated with other congenital abnormalities of the heart and great thoracic vessels. We report a case of a young man who, during the work-up for and treatment of a Wolff-Parkinson-White (WPW) syndrome, was found to have LSVC with a particularly unusual course, initially raising the suspicion of a space-occupying lesion in the left atrium (LA).

A 19-year-old male with history of palpitations, presyncope

and ECG suggestive of a posteroseptal accessory pathway (AP) was referred for electrophysiological studies (EPS). A prior transthoracic echocardiogram suggested a LA “mass”. During the EPS, the coronary sinus (CS) mapping electrode was placed from the right femoral vein and had an unusual “vertical” appearance. Hence, a CS angiogram was performed, which suggested a possible LSVC.

Consequently, cardiac magnetic resonance (CMR) imaging was performed to further investigate the possible LA mass and delineate the course of the assumed LSVC.

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Figure 2. Transoesophageal echocardiography images. (a) Pedunculated “mass-like” appearance of the left-sided SVC (LSVC) in the LA. (b) Agitated saline contrast study showing drainage of LSVC into the CS. (c) Color comparison showing LSVC flow independent of LA flow. CS, coronary sinus; LA, left atrium; SVC, superior vena cava.

Cardiovascular morphology was studied using diaphragmatic navigated T1-weighted TurboSpin Echo (HASTE) imaging, as well as steady-state free precession cine imaging along the left ventricular long axis as described before.¹

In addition, contrast-enhanced aortography was also performed to rule out significant thoracic aortic abnormalities. CMR imaging confirmed the presence of a LSVC, which displayed a posteromedial displacement with respect to the atrioventricular groove, posterior mitral annulus and the LA wall. Rather than the usual course of running outside the LA, sharing a small area of LA wall as part of its circumference, the LSVC formed an integral part of the LA wall between the left lower pulmonary vein and mitral annulus. In the cranial (superior) part of the LA, more than 70% of the LSVC circumference was located intra-atrially (“atrialised”), while from the mid LA to the floor, the LSVC appeared almost com-

pletely “atrialised” via a small stalk attachment to the LA wall (Figures 1a–d). The aortogram confirmed a “bovine arch” arrangement with the 3DVR late-phase images further confirming the presence of a LSVC (Figure 1e).

Transoesophageal echocardiography was performed and conventional views² were obtained, which showed a pedunculated mass-like appearance of the LSVC within the LA (Figure 2a). Agitated saline contrast injection via the left arm showed this to be a LSVC with a vertical invaginated course within the LA wall, draining into a roofed CS (Figure 2b). Color Doppler assessment confirmed that there was no admixture of blood from the “intra-atrial” LSVC and LA (Figure 2c).

The patient underwent successful radiofrequency ablation of the AP, which was mapped close to the CS OS. Catheter ablation of the AP was difficult because of poor catheter stability in the location of the AP. A series of linear lesions were

made using an irrigated radiofrequency energy ablation catheter (Biosense Webster, F curve, irrigated catheter at 40 W output max temp 50°C), held in position with the help of a support sheath (Agilis, St Jude). Eventually, restoration of a normal His–ventricle interval was achieved, which changed from –18 ms pre-ablation to +46 ms post-ablation with loss of pre-excitation in the surface ECG compared with the ECG at presentation and there was evidence of underlying right bundle branch block.

LSVC is a congenital malformation that occurs in approximately 0.4% of individuals,³ and represents the persistence of the embryological left anterior cardinal vein, which usually involutes to form the left brachiocephalic vein.⁴ The most common type of this venous anomaly, comprising 90% of persistent LSVC, is the presence of both right and left SVC. A bridging innominate vein is observed in less than 30% of cases. A rare variation is a complete absence of the RSVC in the presence of persistent LSVC.

In 80–90% of individuals, the persistent LSVC drains via the distal CS into the right atrium (RA). Of the remaining 10–20%, drainage into the LA, resulting in a right to left shunt, and into the hepatic vein or inferior vena cava have been described.⁵

The usual course of the LSVC is along the posterolateral aspect of the LA, lateral to the left-sided pulmonary veins draining into the CS near the OS. In our case, the course of the LSVC was unusual because most of its circumference was “atrialised”, giving rise to the suspicion of a space-occupying lesion. Rather than running outside the LA with minor wall sharing of the posterolateral LA wall, it traversed cranio-

caudally through the cavity of the LA and drained into the distal aspect of the CS close to the junction of the RA and CS OS, while having no direct flow communication with the LA cavity.

With respect to the clinical presentation of the WPW syndrome, it is worthwhile noting that LSVC may carry muscle sleeves, resulting in anatomical and electrical communication with the atria, which may promote the initiation and maintenance of atrial fibrillation and in rare situations even lead to sudden death.⁶ Although the presence of an AP in this patient could have been incidental, the possibility of the LSVC carrying a muscle sleeve resulting in an atrioventricular communication (AP) could not be ruled out. This may be the reason for the requirement of multiple linear lesions to abolish conduction through the AP.

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