Persistent left superior vena cava (PLSVC) may coexist with a variety of cardiovascular disorders, including coarctation of the aorta. Here we describe the rare coexistence of two different forms of narrow-QRS tachycardia in a patient with PLSVC and coarctation of the aorta.

Case presentation

We present the case of a 21-year-old patient who was referred to our department for an electrophysiologic (EP) study because of recurrent palpitations usually associated with exercise. An episode of narrow-QRS, short RP tachycardia with a cycle length (CL) of 370 ms had been documented on Holter monitoring. The patient had a severe long-segment coarctation of the aorta, surgically repaired with a prosthetic tube bypass graft extending from the ascending to the descending aorta. Additionally, he had a patent foramen ovale and a PLSVC connecting to a dilated coronary sinus (CS) (Figure 1).

An EP study was performed with two quadripolar catheters placed at the His bundle and right ventricular (RV) apex and a decapolar catheter in the CS. Basic intervals were in the normal range. Dual atrioventricular nodal physiology was not demonstrable. During RV pacing, retrograde activation was initially concentric, but during pacing at a CL shorter than 330 ms a change in retrograde activation pattern from concentric to eccentric without decremental conduction properties was noted. During programmed atrial stimulation, a narrow QRS tachycardia was induced (CL 370 ms) with a distal to proximal CS activation sequence and a ventriculoatrial (VA) interval of 104 ms. Delivery of a ventricular extra-stimulus during His refractoriness resulted in slight advancement of the atrium with an increase in the VA interval. Ventricular overdrive pacing resulted in cessation of the tachycardia and therefore an A-V versus A-A-V response could not be ascertained. The tachycardia terminated after adenosine administration. Based on these findings we presumed atrioventricular reentry (AVRT)
through a concealed left free wall accessory pathway (AP) as the underlying mechanism.

We selected the transseptal approach for mapping and ablation, in view of the presence of the prosthetic bypass tube and prior documentation of a patent foramen ovale that would allow left atrial access without the need for transseptal puncture. Using a steerable long sheath (Agilis™ Nxt, St Jude Medical) the AP was successfully ablated in the anterolateral region of the mitral annulus using an irrigated tip catheter (Celsius® Catheter, Biosense Webster).

Following AP ablation, ventricular pacing demonstrated a consistently concentric retrograde activation pattern with decremental conduction properties. However, during programmed ventricular and atrial stimulation, a second type of narrow QRS tachycardia (CL 390 ms) with a long RP pattern was induced, with earliest retrograde activation at the proximal CS. Following ventricular overdrive pacing 10 ms faster than the tachycardia CL, tachycardia resumed with a V-A-V response, excluding the possibility of an atrial tachycardia. For further differentiation of AVRT versus atypical atrioventricular nodal tachycardia (AVNRT), we measured the interval between the last pacing stimulus and the last entrained atrial depolarization (S-A interval), as well as the post-pacing interval (PPI) at the right ventricular apex, and compared them with the VA and tachycardia cycle length (TCL) before pacing (Figure 2). The calculated values of S-A minus VA and PPI minus TCL were greater than 85 ms and 115 ms, re-
spectively, and strongly favored atypical AVNRT as the underlying mechanism of the second tachyarrhythmia. Furthermore, the response to parahisian pacing was nodal (shortened VA interval with His capture).

Based on the abovementioned criteria, we proceeded with ablation of the slow AV nodal pathway following CS venography. Ablation was successfully performed at the bottom of the “barrel” CS and subsequently no type of tachycardia was inducible.

Discussion

PLSVC may coexist with a variety of cardiovascular disorders, including coarctation of the aorta, while it has also been associated with several types of supraventricular tachycardias.2-4 PLSVC is an unusual site of accessory pathways, while it can also be the arrhythmogenic source of atrial fibrillation.2,4 To our knowledge this is the first report documenting the coexistence of two different forms of narrow-QRS tachycardias in a patient with coarctation of the aorta and PLSVC.

The combination of tachycardia features and the use of diagnostic maneuvers allows the differentiation of underlying tachycardia mechanisms.5,6 Several diagnostic maneuvers can be implemented for the differentiation of long RP tachycardias (Table 1).1,7,8 The most frequently used pacing maneuvers are parahisian pacing, delivery of ventricular extrastimuli when the His is refractory, ventricular overdrive pacing, atrial overdrive pacing and atrial pacing at the tachycardia cycle length.

Parahisian pacing can be used for differentiation between retrograde conduction over the atrioventricular node or over a septal accessory pathway. Delivery of ventricular premature stimuli when the His is refractory can identify the presence of accessory pathways based on the presence of tachycardia reset or not. Several key points associated with this pacing maneuver should be outlined. The timely delivery of the premature beat when the His is refractory can be confirmed, even when the His is not recorded, in the case of ECG fusion, since then antegrade His depolarization is validated. Advancement of the atrium indicates the presence but not the participation of the accessory connection in the tachycardia. The latter scenario is documented in case of termination of the tachycardia without affecting the atrium.

Ventricular overdrive pacing during tachycardia is a valuable maneuver that can provide insights concerning the mechanism of the tachycardia. A V-A-A-V electrogram sequence after the last ventricular paced complex indicates an atrial tachycardia, while a V-A-V response can be encountered in both an AVRT and an AVNRT. An important caveat during ventricular overdrive pacing is the documentation of consistent retrograde atrial capture, which can be missed owing to the slightly faster ventricular pacing rate in comparison to the tachycardia rate. Measurement of the post-pacing interval and the VA interval can be used, as in our case, in order to differentiate an atypical AVNRT from an orthodromic reciprocating tachycardia using a septal accessory pathway.2 Furthermore, if ventricular

Table 1. Basic pacing maneuvers for differentiation of long RP tachycardias.

Atypical AVNRT versus AT versus AVRT using a slowly retrogradely conducting accessory pathway.

1. Parahisian pacing (nodal versus extranodal response).
2. Ventricular premature stimuli when His is refractory:
   – Advancement of the atrial activation is suggestive of an AVRT.
   – Termination of the tachycardia without atrial depolarization is diagnostic of an AVRT.
3. Ventricular overdrive pacing with consistent retrograde atrial capture:
   – AV dissociation excludes AVRT and disfavors AVNRT
   – Difference between ventricular post-pacing interval and tachycardia cycle length (>115 ms favors AVNRT, <115 ms favors AVRT)1
   – S-A (interval between the last pacing stimulus and the last entrained atrial depolarization) minus VA interval during tachycardia (> 85 ms favors AVNRT, <85 ms favors AVRT)
   – Reproducible termination of tachycardia without atrial depolarization excludes AT
   – Different atrial activation sequence during tachycardia and ventricular pacing favors AT
4. Atrial overdrive pacing:
   – Atrial overdrive pacing and comparison of VA interval of return beat with VA during tachycardia (ΔVA <10 ms disfavors AT)
5. Atrial pacing at the tachycardia cycle length:
   – Comparison of AH during tachycardia and AH during pacing: ΔAH>40 ms favors atypical AVNRT.

AT – atrial tachycardia; AVNRT – atrioventricular nodal reentrant tachycardia; AVRT – atrioventricular reentrant tachycardia.
overdrive pacing results in AV dissociation then an AVRT can be excluded, while if it reproducibly terminates the tachycardia without atrial depolarization an atrial tachycardia can be excluded.

Atrial overdrive pacing is performed on the premise of comparing the VA interval of the return beat, after atrial pacing with 1-to-1 conduction, with the VA interval during tachycardia. If the two intervals are the same then the presence of VA linking is supported, indicating that the ventricular activation is dependent on the atrial activation. The documentation of VA linking disfavors atrial tachycardia and speaks in favor of an AVRT or an AVNRT. However, an atrial tachycardia may rarely be combined with apparent VA linking, while on the other hand VA linking may not be evident in 3% of AVNRTs. Finally, atrial pacing at the tachycardia cycle length can differentiate the mechanism of a long RP tachycardia based on the difference in the AH interval between tachycardia and atrial pacing. If the AH interval during atrial pacing exceeds the tachycardia AH by more than 40 ms then the presence of an atypical AVNRT is indicated.

It should be emphasized that, although additional maneuvers have been reported in the literature, the presented constellation includes those which are routinely used in our institution because they are considered to be useful, easy to perform and to interpret. In conclusion, this case describes the implemented diagnostic approach and successful management of two types of narrow QRS tachycardia in a patient with PLSVC and surgically repaired coarctation of the aorta.

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References