Hindsight is 20/20: Reflections on an elusive diagnosis

Arman Kilic, MD

In this issue of the Journal, Hamuro and colleagues1 present an interesting report in which a membranous ventricular septal defect (VSD) with an aneurysmal septal leaflet of the tricuspid valve was misdiagnosed as an aneurysm of the membranous septum. This confusion in diagnosis has been reported as early as the 1960s. Chesler and colleagues2 described 5 cases in which anomalies of the tricuspid valve resembled aneurysms of the membranous septum. Moreover, there were 3 cases in which a VSD was associated with a tricuspid leaflet pouch, and 2 cases in which anomalous attachment of the septal leaflet of the tricuspid valve to the edge of a VSD resulted in a pouchlike structure. A subsequent study published in the 1970s reviewed 118 patients with endocardial cushion defects, and tricuspid pouch was diagnosed in 15 of these patients.3 Pouches appeared similar to membranous septal aneurysms, although an important distinction was that most pouches protruded into the inflow portion of the right ventricle, whereas most membranous septal aneurysms protruded to the outflow portion of the right ventricle.

The images provided in the report by Hamuro and colleagues1 are informative, highlighting a large membranous VSD that was missed during the initial operation. This misdiagnosis led to an early reoperation and closure of the VSD on postoperative day 21. The absence of a shunt during the index hospitalization likely clouded the diagnosis. So, what clues can surgeons use in the future to identify this elusive diagnosis? As suggested by Mavroudis and Backer4 in an article focusing on operative techniques, membranous defects are often covered by the tricuspid septal leaflet and may be difficult to visualize. A clue to the VSD may be depression in the tricuspid valve caused by valve tissue being sucked down into the defect, particularly in cases in which the chordal attachments of the tricuspid valve to the edge of the defect are short, or there is aneurysmal or redundant tricuspid leaflet tissue. As was done during the reoperation in the report of Hamuro and colleagues,1 detachment of the tricuspid leaflet often will provide necessary exposure to appreciate the underlying VSD and treat it surgically.

Placement of a rigid tricuspid ring that was abutting a thinned out and aneurysmal septal leaflet of the tricuspid valve that was subject to systemic pressures predictably led to erosion and creation of a shunt in this case. In a way, this was fortunate in that it led Hamuro and colleagues1 to the correct diagnosis with appropriate surgical revision and treatment early after the initial operation. Hindsight is certainly 20/20, and the authors1 provide an important case report detailing how these various diagnostic entities are easily confused, as corroborated by previous reports as well, underscoring the importance of careful preoperative and intraoperative anatomic evaluation in these cases.

References