A 50-year-old woman with highly symptomatic, drug refractory paroxysmal atrial fibrillation (AF) was referred to our institution for transcatheter pulmonary vein isolation. The pre-operative magnetic resonance imaging (MRI) scan showed a congenital vascular anomaly: right superior vena cava was absent and both brachiocephalic veins drained in a persistent left superior vena cava (LSVC) that ended up in a huge coronary sinus (Panel A). Before performing the transseptal puncture, a self-terminating AF episode triggered by focal activity from LSVC was observed. Planned pulmonary vein isolation was then abandoned, and the electrical isolation of the LSVC from distal coronary sinus was attempted. A three-dimensional reconstruction of the right atrium and the giant coronary sinus together with the proximal persistent LSVC was obtained merging the MRI scan and the Carto 3 map. Circumferential ablation at the junction between LSVC and distal coronary sinus was performed (Panel B, red dots: ablation sites). Complete isolation was documented by the presence of focal activity in the vein, highlighted by asterisk in Panel C, dissociated from the atrium.

The patient was discharged without antiarrhythmic drugs and experienced no clinical recurrence during 6-month follow-up. Persistent LSVC is the most common congenital thoracic venous anomaly. The association between LSVC and AF has already been documented in small series of cases, in which both pulmonary veins and LSVC isolation were performed. This is the first reported case in which LSVC disconnection alone was effective in the cure of paroxysmal focal AF.