A 71-year-old asymptomatic man was referred for a cardiac magnetic resonance study to better evaluate a pericardial cyst, for which the patient was on an echocardiographic follow-up for 10 years and in which, during the last echocardiogram, a feeble flow was detected. Upon cardiac magnetic resonance imaging (A: four-chamber view; B: short-axis view), the pericardial ‘cyst’ (arrowheads in A) turned out to be the section of a polyaneurismatic and tortuous vessel (asterisks in B) with an unclear drainage in the superior portion of the right atrium or the superior vena cava. The fistula had systodiastolic blood flow and a maximum diameter of 43 mm. In order to refine the diagnosis, a 64-row computed tomography coronary angiography was performed. Computed tomography coronary angiography images (C: MPR paratransversal view; D: MPR paracoronal view; E: 3D volume rendering upper-anterior view; F: 3D volume rendering right-posterior view) confirmed the polyaneurismatic and tortuous path in the atrio-ventricular root (asterisks in E and F) of an enlarged left circumflex artery with diffuse parietal calcifications (arrow in C). This enlarged artery was connected with the superior vena cava, confirming the presence of a pathological coronary artery fistula whose drainage at the right atrium–superior vena cava passage (arrows in D, E, and F) was compressed by one of the vessel’s many aneurisms. Two months later, the patient underwent cardiac surgery: the diagnosis was confirmed and the native left circumflex was ligated at its end, where it drained into the superior vena cava. Computed tomography coronary angiography allowed a complete non-invasive preoperative evaluation of the complex anatomy of this very uncommon coronary artery fistula. CT coronary angiography with ECG gating is faster than MR angiography and has a higher spatial and temporal resolution that allows us to perform sub-millimetre reconstructions. Owing to these features, it has to be considered the non-invasive diagnostic method of choice in the evaluation of all the coronary anomalies being particularly useful in the preoperative planning.