A 28-year-old woman was referred to our hospital after successful resuscitation from out-of-hospital cardiac arrest due to pulseless ventricular tachycardia. The patient’s past echocardiographic record documented normal cardiac function and structure when she had received a dual-chamber pacemaker for complete atrioventricular block at 21 years of age. On this admission, however, echocardiography showed left ventricular systolic dysfunction and dilatation with thinning of the basal septal wall (Panel A). Additionally, a mobile nodule (16 × 17 mm) mimicking a thrombus in the right atrium adjacent to the coronary sinus outlet (Panels B and C, arrows) was detected. Although we decided to upgrade the pacemaker system to cardiac resynchronization therapy with a defibrillator (CRT-D), the nodule seemed to obstruct left ventricular lead cannulation due to its location over the coronary sinus. Anticoagulant therapy was initially attempted, but this did not reduce the nodule’s size. We decided to excise the nodule and surgically implant a left ventricular lead. Intraoperatively, the smooth-surfaced light red nodule was found attached to the top of the oval fossa of the right atrium (Panel D, arrows) and was completely excised. Pathological examination of the nodule revealed that it contained numerous non-caseating epithelioid granulomas (Panel E) with multinucleated giant cells (Panel F, arrows). Finally, we implanted the defibrillation lead transvenously and successfully completed the CRT-D system. To the best of our knowledge, this is the first case report of a sarcoid nodule located on the endocardium.