A 33-year-old male patient was admitted to our hospital with shortness of breath and fever. The patient’s past medical history was remarkable for Ewing’s sarcoma (ES) in the left kidney, for which the patient had received therapy 2 years previously. On physical examination, body temperature was 38.5°C, there were rales in the lungs, and a palpable supraclavicular mass was present. Echocardiography revealed a mobile, smooth shaped, hyperechoic mass in the left atrium (Figure A and Video*). At first glance, the lesion appeared to be a myxoma. Mitral valve flow was normal, and there was no gradient with positional change. Furthermore, examination exhibited local pericardial effusion adjacent to the left ventricle and a large mass lesion in the lung on the left posterior side of the left atrium (Figure B). Echocardiographic assessment of the patient 1 year prior to current admission had shown normal findings. Computed tomography scan of the thorax confirmed a large mass lesion that completely occupied the inferior lobe of the left lung. The mass lesion located in the left atrium appeared to arise from the pulmonary vein (Figure C). Histopathological findings of supraclavicular and lung mass lesion were all consisted with ES. Due to expectation of poor prognosis with surgery in patients with recurrent ES, the patient elected to undergo chemotherapy. ES is a malignant primary neoplasm of bone, which usually affects children and adolescents. ES without involvement of bone rarely develops from the soft tissues of the lower extremities, paravertebral region, gastrointestinal tract, kidney, uterus, and other areas of the body. Cardiac metastasis of ES is extremely rare. To our knowledge, the present case is the first example of extraskeletal ES metastasized to the left atrium and pulmonary structures, as indicated by echocardiographic examination. It must be considered that mass lesions may represent not only a primary cardiac tumor, thrombus formation, vegetative lesions, and foreign bodies, but also metastasis from a malignant tumor located in other organs.