A 64-year-old female who complaint shortness of breath and coughing was admitted to our emergency department. On her physical examination the blood pressure was 100/60 mmHg, and heart rate was 110/min. Her neck veins were distended, there was crepitations in basal of the lungs and grade 1 to 2 pretibial edema. A grade 2 pansystolic murmur were present all precordial areas. Electrocardiography showed sinus rytm and giant p waves. The angiographic X-ray revealed marked cardiomegaly of the right heart and pulmonary artery dilatation (Fig. 1). Transthoracic echocardiography showed right chambers dilatation (Fig.2), a giant right atrium measuring 13.5x9.4 cm in the apical four-chamber view (Fig.3) and severe tricuspid regurgitation (Fig.4). The patient’s condition was stabilized after medical therapy. The right heart catheterization revealed severe pulmonary artery hypertension (systolic pressure was 83 mmHg, mean pressure was 50 mmHg). We thought that the patient’s primary problem is severe primary pulmonary hypertension. After that, she was discharged on medical therapy including warfarin, digoxin, a diuretic, a beta-blocker and bosentan as an endotelin reseptor antagonist.