A 64-year old woman with a 2 year history of a slow-growing mass in the left infero-posterior mediastinum was admitted for increasing dyspnea on exertion and severe left-sided chest pain. Thorax computed tomography (Fig. 1) showed a large mass above the left hemidiaphragm surrounding the distal esophagus. Bronchoscopy and esophagoscopy were normal, and transthoracic biopsy was inconclusive. There was no evidence of extrathoracic disease. The patient underwent a left-thoracoabdominal approach and a well-circumscribed lobulated heterogeneous multicystic tumor with a hemorrhagic area of 150 × 145 × 50 mm was resected along with a 95 mm long esophagus segment (Fig. 2), and reconstruction was made with the stomach. Microscopically, the tumor showed numerous large venous, cavernous and capillary-sized vessels infiltrating the wall of the entrapped esophagus segment whose submucosa and muscularis propria showed ectatic vascular spaces. No cellular atypia or mitoses were observed. The patient is asymptomatic and disease-free 2 years following operation.