A 39-year-old woman with a known history of Gardner syndrome was referred for management of a chest wall desmoid tumor with marked intrathoracic extension (Fig. 1). Through an open thoracotomy, tumor resection and chest wall reconstruction was done (Fig. 2). Severe dyspnea and lower extremity edema were soon resolved.

Fig. 1. The tumor, originating from the right posterior chest wall and involving the 8th, 9th and 10th ribs, had invaded the right thoracic cavity and was causing compression of the right lung and leftward displacement of the mediastinum. The patient had no history of pregnancy or surgical trauma in her chest wall. Preoperatively, the patient’s vital capacity was 940 ml (34% of normal) and echocardiogram revealed pulmonary hypertension (estimated right ventricular pressure: 62 mmHg).

Fig. 2. A right posterolateral skin incision was made and the involved 8th, 9th and 10th ribs were resected. Through the chest wall defect, the intrathoracic part of the tumor could be pulled out. The tumor had not invaded the lung parenchyma or the mediastinal structures. The resected tumor was a mostly intrathoracic mass, measuring 27 cm × 17 cm × 7 cm and weighing 3232 g. The surgical margins were pathologically positive, and anti-estrogen therapy was conducted. Desmoid tumors of the chest wall with life-threatening intrathoracic extensions, as well as with maximum diameters greater than 20 cm, are extremely rare. CW: chest wall.