Acquired restrictive thoracic dystrophy (ARTD), a rare iatrogenic disease, is characterized by an underdeveloped thoracic cage resulting from an inappropriate surgical correction of pectus excavatum, the most common type of chest wall deformity. We describe a 29-year-old young man with ARTD, stemming from pectus excavatum correction surgery in his toddler age, who developed cor pulmonale with chronic respiratory acidosis, hypoxemia, and moderate pulmonary hypertension. His symptoms were partially ameliorated after medical treatment. Surgical correction of the chest wall was suggested, but declined because of high mortality and morbidity rates incurred by the operation.

Key Words: Acquired restrictive thoracic dystrophy • Cor pulmonale • Heart failure • Pectus excavatum

INTRODUCTION

Chest wall deformities may cause variable cardiopulmonary dysfunction, either symptomatic or asymptomatic. Pectus excavatum is the most common chest wall deformity, for which a variety of correction procedures have been developed. However, inappropriate procedures of surgical correction for pectus excavatum may lead to acquired restrictive thoracic dystrophy (ARTD), which is characterized by an underdeveloped chest wall and hypoplastic lungs. We describe a 29-year-old young man who had severe chronic cor pulmonale. Through a careful review of the medical history, physical examination, as well as the image and hemodynamic studies, we diagnosed this rare iatrogenic disease. To our knowledge, this is the first case report showing severe chronic cor pulmonale induced by ARTD at this late age.

CASE REPORT

A 29-year-old young man, a non-smoking barber, came to our hospital with complaint of progressive shortness of breath, severe dyspnea on exertion (~50 m by walking) and leg edema for at least 3 months. In addition, he also had progressive orthopnea, paroxysmal nocturnal dyspnea, and cough with blood-tinged sputum. Tracing back his history, we found that mild dyspnea on exertion off and on had been noticed for more than 10 years.

On admission, the patient had a body temperature of 37.5 °C, a pulse rate of 100 beats/min, a respiratory rate of 20/minute, and a blood pressure of 129/87 mmHg. His heart sound yielded grade 3 holo-systolic murmur at the left lower sternal border. Both jugular veins were found engorged, with obvious pitting edema on both lower legs. On his lower chest, there was a surgical scar caused by the surgery for correcting pectus excavatum when he was a toddler. Additionally, his chest cage seemed obviously small and underdeveloped (Figure 1A).
Chest film (Figure 1B) revealed cardiomegaly and pulmonary congestion. The twelve-lead ECG (Figure 1C) showed normal sinus rhythm with right ventricular (RV) hypertrophy and a right-deviated cardiac axis. Echocardiography demonstrated RV enlargement and moderate pulmonary hypertension (68-75 mmHg) with paradoxical interventricular septal wall motion. However, the ventilation-perfusion scan excluded the presence of unmatched defect.

To elucidate the cause of right heart failure and pulmonary hypertension, cardiac catheterization was arranged. Left ventriculography exhibited normal left ventricular (LV) systolic function (LV ejection fraction: 79%) with a normal LV end-diastolic pressure (14 mmHg). LV and aortic blood O2 saturation (SaO2) was only 78%, and the patient’s arterial blood gas in room air showed severe hypoxemia (PaO2 39 mmHg), and hypercapnia (PaCO2 59 mmHg) with metabolic compensation (HCO3 34 mmol/l). His hemoglobin level was up to 17 g/dL, implying that the hypoxemia was a chronic process. Data of right-side cardiac catheterization demonstrated a significantly elevated main pulmonary arterial pressure [60/38, 45 mmHg (systolic/diastolic, mean pressure)] with a prominent reduction of oxygen saturation (48.5%). All other data were consistent with features of cor pulmonale, and no obvious intra-cardiac or extra-cardiac shunt was noticed. There was also a decrease of cardiac index (1.63 L/min/m2) with prominently elevated pulmonary vascular resistance (12.8 Wood units). No evidence of proximal pulmonary embolism, pulmonary artery stenosis, pulmonary vein stenosis or stenosis could be found by pulmonary angiography.

Suspecting cor pulmonale from cardiac catheterization, we further looked into whether there was lung parenchymal disease. High-resolution chest computed tomography demonstrated a small and flattened thoracic cage with dilated RV and pulmonary trunks. There was neither lung fibrosis nor other lung parenchymal diseases (Figure 2A). Also, the heart was significantly compressed by the narrowed chest wall, with a Haller severity index ratio (= transverse thoracic distance/sternovertebral distance) up to 3.97 (normal value: < 2.5) (Figure 2B). Pulmonary function test revealed a striking reduction of functional vital capacity (FVC) (0.71 L, 17%
predicted), forced expiratory volume in the first second (FEV1) (0.41 L, 12% predicted), FEV1/FVC (58%), forced expiratory flow, mid-expiratory phase (FEF 25-75%) (0.17 L/S, 4% predicted), total lung capacity (1.88 L, 33% predicted and functional residual capacity (1.31 L, 39%). This data indicated a significant reduction of lung volume with combined severe obstructive and restrictive lung disease. Taken together, all evidence pointed to the diagnosis of acquired restrictive thoracic dystrophy (ARTD) with severe chronic cor pulmonale.

The patient was treated with water and salt restriction, diuretics and bronchodilators during admission. Clinical condition was partially improved afterwards. Although surgical correction was one option for the further treatment, there was no strong evidence supporting that the surgery would be the treatment of choice. On the contrary, there was report showing rapid deterioration of cardiopulmonary function after surgery for pectus excavatum and surgery per se also has high mortality and morbidity rates because of the poor cardiopulmonary functions. Our patient declined any surgical intervention. He was discharged after his clinical condition was stabilized, and was regularly followed up in our outpatient clinic.

**DISCUSSION**

Pectus excavatum results in variable cardiopulmonary function limitation. It is one of the most common congenital chest wall anomalies, occurring in approximately 1 in every 700 births. The laboratory and clinical features of our case demonstrated similar findings. However, this patient had a unique feature: that the chest wall deformity resulted from previous correction surgery for pectus excavatum in his early age. On chest film, this chest wall deformity has been described as “like the rib-cage of a dog” (see Figure 1B). This acquired chest wall deformity, which was previously named “acquired Jeune’s syndrome” or “restrictive lung disease”, is sometimes not easily discernible from other congenital chest wall deformities. Recent studies described the disease using a more comprehensive term “acquired restrictive thoracic dystrophy”, or “ARTD” for short.

ARTD is a rare iatrogenic disease resulting from faulty surgical correction techniques that removes the growth plates from the rib cartilage, and thus, freezes the subsequent chest wall development. Although some authors had also pointed out that the age of surgery was crucial for the development of ARTD, a more recent report suggests that the cause of ARTD is not that the operation is performed at an early age, but that inappropriate surgical technique was performed.

The Ravitch technique or its modifications for pectus deformities correction had been generally performed by different surgeons for decades. One of the major concepts of Ravitch technique is deformed cartilages resection. However, too-large segment of cartilage resection often damages perichondrial sheaths and even interfere with rib growth plates, which further causes ei-
ther unstable chest or depressed chest wall development (such as ARTD). Recently developed minimally invasive repair for pectus excavatum, the Nuss technique, has become more widely accepted and provides excellent outcomes. There is no need to resect rib cartilage or perform sternal osteotomy in the Nuss technique, theoretically minimizing the occurrence of ARTD. To prevent occurrence of ARTD, any surgical procedures should avoid radical subperichondrial resection of the cartilage, extirpation of growth centers, and suturing together of the perichondrial strips retrosternally, as described by Robicsek et al.

Some ARTD patients may also have recurrence of the pectus excavatum. In this case report, a prominently increased Haller index (internal transverse thoracic distance/sternovertebral distance) (Figure 2B), which reached the range (3.97; normal value: <2.5) for the diagnosis of pectus excavatum, was also found, in addition to an underdeveloped chest cage.

Serial studies were needed to figure out the final diagnosis in our case. Firstly, marked cardiomegaly and desaturated arterial blood with a widened alveolar-arterial oxygen gradient (A-a DO2=29.85 mmHg) may suggest the existence of congestive heart failure. A normal LV systolic function with a relatively clear lung field excluded the possibility of LV failure related dyspnea. Secondly, an enlarged RV with moderate pulmonary hypertension and paradoxical movement of interventricular septum, raised the possibility of lung parenchyma, pulmonary artery, or shunting disorder related RV failure. Pulmonary embolism and shunting were excluded by ventilation-perfusion scan of lung and cardiac catheterization, respectively. Other studies for collagen disease related pulmonary hypertension, such as antinuclear antibody, rheumatoid factor and anti-neutrophil cytoplasmic antibody, all turned out to be normal. High resolution chest CT also showed absence of lung parenchymal abnormality. Lastly, the bizarrely small chest cage from the appearance and the chest CT, the unique scar suggestive of a previous extensive resection of rib cartilage, together with an evidence of severe restrictive lung disease from the pulmonary function test, lead us to the final diagnosis of ARTD.

Although ARTD or pectus excavatum basically results in a restrictive lung disease, it may be sometimes complicated with an obstructive lung disease, perhaps caused by repeated lower airway infections or inflammation. Our case was also reported to have an obstructive lung disease, in addition to prominent features of restrictive lung disease. This was the reason why bronchodilators seemed helpful in the treatment of our patient.

Although chest wall deformity is not uncommon, severe cor pulmonale is still a very rare consequence. Symptomatic treatments may include water/salt restriction and diuretics. Whether surgical treatment at this late stage would result in a better clinical outcome is still an issue of debate from currently available evidence.

In summary, we have reported a rare iatrogenic disease, ARTD, which induced severe chronic cor pulmonale and hypoxemia. Even though patient’s symptoms were temporarily ameliorated after treatment, the long-term prognosis was still very poor. This case report alerts us that a more prudent and careful evaluation before the surgical correction for pectus excavatum is essential to avoid this iatrogenic disaster.

REFERENCES