Recurrent Spontaneous Pneumothoraces Associated With Juvenile Polymyositis*

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A 17-year-old man, who had received a diagnosis of juvenile polymyositis (PM) at the age of 1 year, developed recurrent spontaneous pneumothoraces and underwent surgical treatment by means of video-assisted thoracic surgery. Intraoperative observation and microscopic studies demonstrated numerous bleb-like lesions below the visceral pleura. To our knowledge, this is the first article that describes a case of spontaneous pneumothorax associated with juvenile polymyositis.

PM. Our observation should lead to broadening of the spectrum of pleuropulmonary manifestations of PM.

Key words: cyst-like lesions; juvenile polymyositis; spontaneous pneumothorax

Abbreviations: DM = dermatomyositis; PM = polymyositis

Polymyositis (PM) and dermatomyositis (DM) are related idiopathic inflammatory myopathies. Although spontaneous pneumothorax has been reported as a rare complication of DM,1–4 to our knowledge, no case of PM associated with spontaneous pneumothorax has been reported. In this article, we describe a case of spontaneous pneumothorax in a 17-year-old patient with juvenile PM. Intraoperative and microscopic findings were unique, suggesting that pleural changes due to PM contributed to recurrent spontaneous pneumothoraces.

Case Report

A 17-year-old man had received a diagnosis of juvenile PM when he was 1 year old, based on the following signs: his proximal muscle weakness (Gower’s sign, +); elevated myogenic enzymes, including creatinine phosphokinase, 626 IU/L (normal level, 30 to 200 IU/L) and aldolase 10.3 IU/L (normal level, 1.7 to 5.7 IU/L); myogenic pattern of electromyogram; and histology. The myogenic enzymes were normalized during high-dose prednisolone therapy (2 mg/kg/d). He received prednisolone until age 9 years. From age 13 to 16 years, he was treated with growth hormone for his growth retardation.

In May 1997, right spontaneous pneumothorax was diagnosed for the first time. During the next 2 years, he was conservatively treated for recurrent pneumothoraces, four times on the left side and three times on the right side. In June 1999, he had a right pneumothorax and was referred to our hospital for surgical intervention. There had been no previous attempts for prevention of recurrence, such as chest tube with sclerosis.

On admission, he was 155 cm in height and 27 kg in weight. In addition to systemic proximal muscular atrophy, he had prominent thoracic deformity and scoliosis. This skeletal abnormality was attributed to his muscular disease and to steroid therapy. Other musculoskeletal, collagen, and metabolic diseases that could cause skeletal abnormalities were ruled out based on his symptoms and clinical data. Chest radiograph showed right-sided pneumothorax. Chest CT, which had been performed before this pneumothorax occurrence, demonstrated bullae at the apex of both lungs (Fig 1, top). No evidence of pulmonary fibrosis was found (Fig 1, bottom). Pulmonary function testing showed severe restrictive impairment (vital capacity, 1.5 L; percent of predicted vital capacity, 38.2%; FEV1, 1.37 L; percent of FEV1, 90.1%). Blood gas analysis while breathing room air demonstrated CO2 retention (PAO2, 72 mm Hg; PACO2, 60 mm Hg; pH, 7.34).

On June 21, 1999, the patient underwent a right bullectomy under video-assisted thoracic surgery. Only one cystic lesion (about 5 × 10 mm) was identified at the apex of the right lung; however, almost all the surface of the visceral pleura was covered with numerous cystic lesions, about 0.5 to 1.0 mm in diameter. The larger cystic lesion was resected together with its surrounding tissue. Microscopic studies revealed nonspecific cystic lesion.

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and multiple bleb-like lesions just below the visceral pleura (Fig 2). No evidence of interstitial fibrosis was found. The patient was extubated after 18 h of postoperative respiratory support and discharged 7 days after surgery.

**Discussion**

Juvenile PM/DM is an acquired inflammatory disease of unknown etiology that normally affects children < 15 years old. The disease is manifested as severe proximal muscle weakness and, in juvenile DM, cutaneous eruption is characteristic.5 Muscle biopsies characteristically show patchy mononuclear infiltrates with perifascicular muscle fiber degeneration.7 Juvenile PM/DM is considered as a part of the spectrum of PM/DM and is distinguished from the adult form in having a number of different clinical and pathologic features.7 Development of calcinosis—subcutaneous and/or muscular calcification—is a more frequently encountered complication in juvenile PM/DM than in the adult form. Juvenile PM/DM is regarded as a syndrome rather than an entity because of its various clinical pictures.6,8 We consider that our patient has a type of juvenile PM that can be complicated with spontaneous pneumothorax.

The mechanism of recurrent pneumothoraces in our patient is not clear. The multiple bleb-like lesions played some role in the development of pneumothorax. Other collagen diseases, such as progressive systemic sclerosis and systemic lupus erythematosus, are associated with increased incidence of pneumothorax.9 Interstitial fibrosis is believed to degenerate alveolar walls, which leads to cyst formation, rupture of the cyst, and spontaneous pneumothorax.9 A few cases of DM associated with spontaneous pneumothorax have been reported.1–4,10 Besides interstitial fibrosis, widespread vascular inflammation and infarction are other possible mechanisms of pneumothorax associated with DM.1 We found no interstitial fibrosis or vascular inflammation in the parenchyma of the lung in our patient.

In general, pleural changes in PM/DM occur primarily in association with interstitial lung disease. In this regard, PM/DM differs from other connective tissue diseases.8 However, no interstitial changes were found in our patient. This case demonstrates that pleural changes can occur without interstitial fibrosis in a type of PM.

In conclusion, we report a case of recurrent spontaneous pneumothoraces associated with PM. This case presents a different type of pleuropulmonary manifestation of PM.

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**References**

Coronary Angiodysplasia of Epicardial and Intramural Vessels*

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A case of coronary angiodysplasia combining large aneurysms of epicardial arteries with diffuse malformation of intramural vessels is reported. Clinical presentation may mimic a vascularized cardiac tumor. Although leaking of the aneurysms in the pericardial space may occur, this entity seems to have a benign prognosis not requiring surgical repair. (CHEST 2000; 118:1511–1513)

Key words: angiodysplasia; coronary aneurysms; myocardial vessels

Abbreviations: LAD = left anterior descending artery; LV = left ventricle; RV = right ventricle

Coronary artery aneurysms and ectasia are uncommon abnormalities that have been found in up to 4.9% of angiographic1 and in about 1.4% of autopsy cases2; they may be of congenital or acquired origin, and their most frequent complications are rupture or thrombosis with distal embolization causing myocardial infarction and sudden death. Such disorders are usually confined to the epicardial coronaries, sparing the intramural vessels and the ventricular myocardium.

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We describe an unusual case of coronary angiodysplasia where large aneurysms of epicardial vessels were associated with diffuse dysplastic lesions of the intramural vessels giving rise to a spongy malformation of the ventricular myocardium.

CASE STUDY

A 30-year-old man was admitted because of prolonged chest pain. No fever, chest trauma, and systemic symptoms or diseases were referred. Physical examination was normal except for the presence of a continuous heart murmur graded 3/6 heard at the apex. Heart rate was 75 beats/min, and BP was 130/80 mm Hg. Chest radiography showed a mild prominence of the left ventricular (LV) border. The ECG showed a sinus rhythm with signs of left LV hypertrophy and deep negative T waves in the precordial leads and in D1-aVL. The two-dimensional echocardiogram revealed a moderate echogenic, diffuse pericardial effusion without signs of cardiac tamponade and a right ventricular (RV) and LV hypertrophy (interventricular septum, 22 mm; LV free wall, 20 mm) mainly localized in the apical region. At this level, there was an epicardial mass that was difficult to differentiate from the underlying myocardium, protruding in the pericardial space, with an inside cavity that showed a systolic-diastolic pulsatility. RV and LV dimensions and contractility were normal. The left atrium was mildly dilated (44 mm). Doppler echocardiographic analysis of the transmittal flow revealed a diastolic dysfunction (ratio of peak flow during early diastolic filling to peak flow during atrial contraction < 1).

The presence of a cardiac mass and of a pericardial effusion suggested, in the first instance, a cardiac tumor infiltrating the myocardium; for further evaluation, cardiac nuclear magnetic resonance was performed, showing clear thickening of the middle anterior and inferior septum, and apical and anterolateral walls of both ventricles with inhomogeneous signal intensity; furthermore, there was evidence of huge intramural and epicardial high-flow vessels. Intermediate signal pericardial effusion was also present, suggesting a hemopericardium (Fig 1); on flow images, the left anterior descending artery (LAD) was clearly demonstrated as a very large vessel (diameter = 8.5 mm) with

![Figure 1. Cardiac nuclear magnetic resonance in horizontal long-axis spin-echo showing septal and apical wall thickening with inhomogeneous signal intensity and large intramural and epicardial vessel (black blood round structures). Pericardial effusion with intermediate signal intensity was seen on T1-weighted images, suggesting a hemopericardium is also present.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21954/ on 04/05/2017)