A 48-year-old man with neurofibromatosis 1 presented with progressive shortness of breath over the course of 4 years. There was prominent kyphoscoliosis. Imaging revealed a large thoracic meningocele compressing the left lung (figure). He then developed progressive lower extremity weakness and a decline in pulmonary function. He underwent drainage, resection, and closure of his thoracic meningocele. In a second stage, he underwent spinal cord decompression and fusion. He is no longer requiring supplemental oxygen and is gaining strength. Thoracic meningoceles may occur in neurofibromatosis secondary to congenital mesodermal dysplasia and hypoplastic bone changes.1,2

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