Sternal fracture with fatal outcome in cystic fibrosis

A 16 year old girl with cystic fibrosis (CF), with very severe pulmonary disease and osteoporosis, presented to the outpatient clinic with sudden onset of back pain. A radiograph of the spine revealed multiple vertebral fractures leading to new thoracic kyphosis. She was briefly admitted to hospital for pain control and discharged without respiratory distress. Ten days later she presented again with increasing dyspnoea and difficulties in inflating her chest secondary to a new funnel chest. Despite ongoing respiratory support, appropriate antibiotics, pain control and physiotherapy, the patient died in respiratory failure 2 days after admission. It is likely that the recent vertebral fractures (fig 1, small arrows) generated forces on the sternum that led to the spontaneous fracture of the mid sternum (fig 1, large arrow).

Low bone mineral density (BMD) is common in adults' and adolescents with CF. Spontaneous fractures of the vertebra and ribs may occur in these patients, especially if they have additional risk factors for the development of bone disease. For example, Elkin et al found evidence of vertebral deformity by radiography in 17% of 107 adult CF patients. Although several well designed studies on bisphosphonates as treatment of CF related bone disease exist, none of these trials included children. This case emphasises the importance of screening for BMD and the need for more aggressive treatment, not only in adults but also in children with CF.

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