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Pulmonary puzzle

Orthopnoea and arm weakness

PULMONARY PUZZLE

A 70-year-old Caucasian man was transferred back to New Zealand from an Italian hospital having been admitted there 6 weeks earlier with acute dyspnoea. There was no associated cough or fever and, despite antibiotic treatment, he remained dyspnoeic at rest with persistent orthopnoea. His left shoulder had been weak for over 10 years but he had recently noticed weakness in the right shoulder and arm. He had moderate chronic obstructive lung disease secondary to smoking. On examination he had a short neck, with limited range of movement in all directions, and a body mass index of 32. His respiratory rate was 24/min, oxygen saturation 80% on air. The chest was mildly hyperinflated but expansion was decreased and lung bases were dull to percussion with decreased breath sounds. There were no signs of pulmonary hypertension. Bilaterally, there was severe weakness in the shoulders (power grade 2/5 on the right, 1/5 on the left) and very mild weakness of arm flexion (power grade 5–/5). Tone was increased and reflexes brisk in all limbs. He was in asymptomatic urinary retention. An arterial blood gas on air was consistent with chronic type II respiratory failure (pH 7.41, PCO_2 66 mm Hg, PO_2 42 mm Hg, HCO_3^- 41 mmol/l); spirometric tests showed a 52% reduction in his vital capacity in the lying position compared with sitting (0.71 l to 1.46 l) and the chest radiograph showed bilateral loss of lung volume with elevated hemidiaphragms. A sniff test showed bilateral diaphragmatic paralysis without paradoxical movement and serum creatine kinase levels were normal. A CT scan showed moderate bibasal atelectasis only. MRI of the neck demonstrated extensive vertebral abnormalities in the cervical spine (fig 1).

QUESTION

What is the diagnosis and what further investigations are required?

See page 1069 for answers

H K Chen,¹ D Jardine,² L Beckett¹

¹ Department of Respiratory Medicine, Christchurch Hospital, Christchurch, New Zealand;

² Department of General Medicine, Christchurch Hospital, Christchurch, New Zealand

Figure 1 T2 MRI of sagittal cervical spine showing congenital synostosis of the C3/C4 and C5/C6/C7 vertebrae consistent with Klippel-Feil syndrome. The C3/4 disc-osteophyte complex is causing severe compression to the cord (arrow) and myelomalacia above this level. The relevant vertebral bodies are numbered.

Correspondence to: D L Jardine General Medicine, Christchurch Hospital, Christchurch 8011, New Zealand; david.jardine@cdhb.govt.nz

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Pulmonary puzzle

ANSWER

From the question on page 1031

The scan showed synostoses of the C3/C4 and C5/C6/C7 vertebrae, consistent with Klippel-Feil syndrome. At C3/C4 the disc-osteophyte complex was compressing the adjacent cord, causing severe myelomalacia well above this level. Phrenic nerve studies, including amplitudes and conduction velocities, were normal bilaterally, reliably excluding lower motor neurone pathology.¹ Bilateral diaphragmatic paralysis, demonstrated by fluoroscopic monitoring during the sniff test, was therefore secondary to interruption of the descending corticospinal pathways supplying the phrenic nerve cell bodies, most of which lie in the C4 neuromere adjacent to the C2/C3 disc. This would account for his respiratory failure and we suspect that the interruption was included in the extensive area of myelomalacia above the cord lesion. This may initially have been unilateral and asymptomatic, progressing only to respiratory failure when the right side became involved.² His ability to compensate for this would have been decreased by his underlying chronic obstructive airways disease and high body mass index.

Klippel-Feil syndrome is a congenital condition (incidence 1:40 000 births) of uncertain inheritance characterised by synostosis of the cervical spine at one or multiple levels,

involving the vertebral bodies, facet joints and posterior arches. The neck may be short and webbed with restricted movement and the hairline low. Associated conditions include other musculoskeletal deformities, deafness, heart defects and genitourinary abnormalities.³ Neurological complications secondary to spinal cord and nerve root compression may occur in childhood or later. The differential diagnosis of bilateral diaphragmatic paralysis includes motor neurone disease, multiple sclerosis, cervical myelopathy secondary to degenerative spondylosis and spinal tumours.^{4,5} The rapid onset of his dyspnoea and absence of lower motor neurone pathology is against motor neurone disease. His dyspnoea has subsequently improved with conservative treatment.

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