

CASE REPORT

Acute respiratory failure as the first sign of Arnold-Chiari malformation associated with syringomyelia

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Acute respiratory failure as the first sign of Arnold-Chiari malformation associated with syringomyelia. D. Alvarez, I. Requena, M. Arias, L. Valdés, I. Pereiro, R. De la Torre. ©ERS Journals Ltd 1995.

ABSTRACT: We report a rare case of acute respiratory failure in a previously asymptomatic patient showing clinical signs of inferior cranial nerve palsy together with weakness and muscular atrophy of the upper limbs.

Magnetic resonance imaging revealed Arnold-Chiari malformation associated with platybasia, basilar impression, syringomyelia and Klippel-Feil syndrome. Episodes of apnoea required tracheostomy and recurred upon tentative closure of the tracheostome, but remitted upon decompression of the posterior fossa.

This case involved both obstructive mechanisms and dysfunction of the respiratory centre. Patients with respiratory failure not explained by pulmonary pathology should be checked for underlying neurological disease.

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Arnold-Chiari malformation is a dysraphic congenital disorder, frequently associated with other malformations of the same kind, including syringomyelia, Klippel-Feil syndrome and tethered cord [1]. Its clinical signs can be very diverse, but only exceptionally consist of paroxysmal episodes (fainting, choking or acute respiratory failure) [2–6]. We report a case of Arnold-Chiari malformation (associated with syringomyelia, platybasia, Klippel-Feil syndrome and single kidney) that was detected upon investigation of untractable repeated respiratory failure and episodes of apnoea originally precipitated by respiratory infection.

Case report

The patient, a 38 year old seaman with facial dysmorphism, cleft palate, low hairline and a short neck with limited mobility, presented to the Casualty Service with predominantly nocturnal respiratory difficulty that had arisen following several days of coughing and purulent expectoration. Arterial blood gas analysis showed hypoxaemia (arterial oxygen tension (P_{aO_2}) 4.3 kPa (32 mmHg)), and hypercapnia (arterial carbon dioxide tension (P_{aCO_2}) 10 kPa (75 mmHg)); bicarbonate concentration was high (45 mmol·l⁻¹), and pH was 7.29. Chest radiography revealed left basal laminar opacity. Acute respiratory failure whilst in the Casualty Service prompted endotracheal intubation (which proved troublesome) and the application of ventilatory assistance.

Upon transfer to the Intensive Care Unit, the patient received broad-spectrum antibiotic treatment. Three

attempts at extubation were each followed by respiratory failure. In view of this and the difficulty of intubation, tracheostomy was performed. No further respiratory failure occurred until tentative closure of the tracheostome.

Upon transfer to the Pneumology Service, the patient underwent the following diagnostic procedures: blood cell counts and biochemical analyses (including a proteinogram and determination of immunoglobulins, alpha₁-antitrypsin and thyroid hormones), which were all normal. Arterial basal blood gas analysis: P_{aO_2} 8 kPa (60 mmHg), P_{aCO_2} 7.7 kPa (58 mmHg), pH 7.38, arterial oxygen saturation (S_{aO_2}) 90%, bicarbonate 35 mmol·l⁻¹, and alveolar to arterial oxygen difference ($A-aDo_2$) 19.2. Sputum and blood cultures were negative. Spirometry: forced vital capacity (FVC) 2.8 l (70% predicted), forced expiratory volume in one second (FEV_1) 2.1 l (67% pred), $FEV_1\%$ 75 (78% pred), single breath transfer factor $TLCO$ 15.5 ml·min⁻¹·mmHg⁻¹ (68%), and diffusion/alveolar volume (D/VA) 4.97 (83%). Fibreoptic bronchoscopy showed abundant purulent secretion in the bronchus of the left inferior lobe, with no other abnormalities. Spinal radiography showed cervical scoliosis and fusion of C5 and C6 vertebrae. Abdominal ultrasonography and urography revealed a single hypertrophic lobulate kidney with a single ureter. Chest radiography and radioscopy showed elevation of the left hemidiaphragm, with conserved mobility. Holter-electrocardiography (ECG) revealed sinus arrhythmia with frequent episodes of tachycardia and supraventricular extrasystole, predominantly at night.

Neurological examination revealed a left Bernard-Horner syndrome; paralysis of the 9th, 10th and 12th cranial



Fig. 1. – Sagittal craniocervical magnetic resonance imaging (MRI) section (SE-T1 mode), showing herniated cerebellar tonsils, basilar impression and Kippel-Feil syndrome, with an extensive syringomyelia extended from the lower medulla to the level of the T10 vertebra (not seen in the figure).

nerves; distal paresia, muscular atrophy and areflexia of the upper limbs; abolition of left abdominal cutaneous reflexes; hyperreflexia in the lower limbs, with left Chaddock's sign; and preservation of all modalities of sensibility.

Magnetic resonance imaging (MRI) (fig. 1) showed a large basilar impression (the whole of the C2 vertebra was above the Chamberlain's line); cervical Klippel-Feil syndrome; Arnold-Chiari malformation and bulbar deformation, with a syringomyelic cavity extending from the lower medulla to the T10 vertebra.

Covering the patient's tracheostomy tube provoked further episodes of respiratory failure, which only remitted if the tube was uncovered.

Following occipitovertebral decompression (suboccipital craniectomy, C1 and C2 laminectomy), the tracheostome was closed without inducing further diurnal episodes of apnoea. Subsequent polysomnography revealed numerous episodes of sleep apnoea, some central but most often obstructive.

Discussion

Breathing is an essentially automatic process governed by the respiratory centres of the brainstem, which receives information on metabolic status from the peripheral

chemoreceptors *via* afferent nerve fibres, and projects efferent fibres to the motor nuclei innervating the respiratory muscles. This automatic respiratory mechanism is subject to continual voluntary intervention in the waking state, but not during sleep, when the anatomical and functional integrity of the circuits is crucial [7]. Arnold-Chiari malformation, whether alone or in combination with syringomyelia, can give rise to a variety of sudden or progressive respiratory disorders, including central alveolar hypoventilation [3, 8, 9], sleep apnoea [8, 10–14], and acute respiratory insufficiency brought on by aspiration in dysphagic patients [4, 15, 16].

Our patient led a normal life until a respiratory infection led to respiratory failure requiring intubation and, eventually, tracheostomy. His arterial blood gas characteristics suggested alveolar hypoventilation, and the risk of apnoea derived from both obstructive causes (short neck, limited mobility of soft palate and tongue) and central causes (medullary compression and/or vascular lesion because of the ectopic cerebellar tonsils and/or central cavitation). Pure obstructive apnoea was unlikely, since it ought to have been totally prevented by tracheostomy, whereas, in fact, it did not cease completely until surgical decompression of the posterior fossa was carried out. The occurrence of both central and obstructive apnoea was confirmed by polysomnography, which showed numerous episodes of sleep apnoea, most of them obstructive.

In other cases in which the first clinical manifestation of Arnold-Chiari malformation has been acute respiratory failure, the patient's neurological performance has been normal [9]. The clinical presentation of Arnold-Chiari malformation with syringomyelia as acute respiratory failure initiated by a limited respiratory infection is exceptional. When reported in a previous case, it was attributed to dysfunction of afferents from chemoreceptors, which was assumed to be responsible for the absence of a respiratory centre response to pneumonia-induced hypoxia [3]. In addition to this mechanism, a restrictive one must also be considered in the case of our patient, who showed signs of the 9th, 10th and 12th cranial nerves being affected, and muscular respiratory weakness due to syringomyelia.

In conclusion, we suggest that in cases in which acute respiratory failure is not explicable in terms of the patient's pulmonary pathology, the possibility of underlying neurological disease should be considered. Conversely, all patients with diagnosed dysraphic malformation should undergo thorough investigation of their respiratory function, including polysomnography, with a view to detection of subclinical functional disorder.

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