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Difficult-to-wean: High index of suspicion

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Female, 57
Final Diagnosis: Syryngomyelia • cervical
Symptoms: Acute respiratory insufficiency
Medication: —
Clinical Procedure: —
Specialty: Pulmonology

Objective: Rare disease

Background: Failed planned extubation occurs in a minority of patients with acute respiratory failure requiring invasive mechanical ventilation. In patients presenting with acute respiratory failure with no identifiable cardiopulmonary causes, uncommon conditions, such as cervical spondylotic myelopathy, should be considered. In very rare instances, when cervical spondylotic myelopathy and syringomyelia present concomitantly, they can be devastating.

Case Report: A 57-year-old woman visited the emergency room (ER) after feeling unwell for several days. She was brought to the ER with acute respiratory distress and obtunded state with GCS of 6/15. She was hypotensive and agonally breathing. Her initial neurologic evaluation was unrevealing. Based on these findings, she was intubated. Over the next several days, she was difficult to wean from the ventilator and had persistent respiratory acidosis. After a short-lived extubation, the patient was again re-intubated. This time the neurologic evaluation showed decreased movements of all muscle groups against gravity and forces, with generalized weakness. An MRI of the brain and cervical spine demonstrated moderate degenerative disc disease and syringomyelia extending from C2 to C7 level. The patient underwent de-compression laminectomy. After failing several weaning trials, she underwent bronchoscopically-assisted tracheotomy.

Conclusions: Acute cardiopulmonary and intensive care unit-acquired neuromuscular conditions have been attributed as a major cause of difficult weaning and extubation. Failure to identify and correct other rare combinations (such as cervical degenerative disc disease and syringomyelia) may cause acute respiratory failure and subsequent failure to wean and extubation, resulting in high rates of mortality and morbidity.

MeSH Keywords: Spinal Cord Compression • Syringomyelia • Difficult-to-wean

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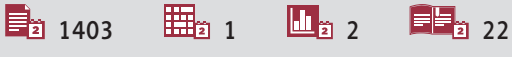

Background

Acute respiratory failure (either hypoxemic or ventilator) requiring mechanical ventilation is one of the most common reasons for ICU admission. ICUs have implemented weaning protocols to extubate patients. Despite meeting all weaning criteria and succeeding in a weaning trial, planned extubation fails in about 10–20% of general ICU cases [1]. Patients who fail extubation have a high mortality rate of approximately 25–50% [2,3]. Repeated unsuccessful attempts at weaning usually signify incomplete resolution of the illness that precipitated mechanical ventilation and/or the development of 1 or more new problems. Some patients remain difficult-to-wean even though their acute illness has resolved and they are otherwise stable. Several reversible causes such as respiratory and/or cardiac load, neuromuscular, and metabolic and endocrine disorders have been attributed as causes of difficult weaning. Besides cardiovascular-related causes [4], neuromuscular weaknesses is also a common cause of failure to wean from the ventilator; the causes include polyneuropathy and myopathy [5]. Rarely, undiagnosed, advanced cervical spondylotic myelopathy and syringomyelia may cause acute respiratory failure and subsequent failure to extubate [6].

We present the case of a 57-year-old female patient admitted with severe sepsis and acute respiratory failure requiring mechanical ventilation. After several attempts, the patient was finally extubated. Six days later, she developed acute onset quadriparesis and worsening of her subacute respiratory insufficiency, which required noninvasive positive-pressure ventilation. An MRI showed spinal syringomyelia with cervical spondylotic myelopathy causing cord compression.

Case Report

A 57-year-old woman visited the emergency room (ER) after feeling unwell for several days with sore throat, subjective fever, neck pain, and stiffness. Her medical history is significant for chronic neck pain, questionable stroke, generalized anxiety, and depression. She is a current smoker. Her vital signs were within normal limits. Her laboratory workup was unremarkable but an x-ray of the cervical spine showed moderate disc space narrowing at C4-C5, C5-C6, and C6-C7 with marginal hypertrophic spurring (Figure 1). Her neck pain improved with morphine and she was discharged home with pain medications. The next day, she returned to the ER complaining of difficulty urinating. She revealed a history of intermittent urinary retention. She denied other urinary symptoms, weakness, or difficult breathing. Otherwise, her exam results were normal. A Foley catheter was inserted and 600 cc of urine was evacuated. Repeat urinalysis was unrevealing.

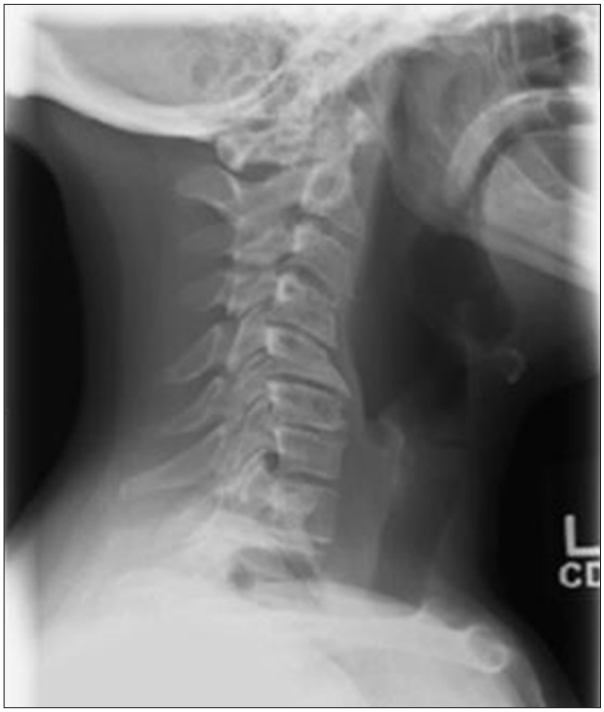

Figure 1. Cervical spine x-ray showed moderate disc space narrowing at C4-C5, C5-C6, and C6-C7 with marginal hypertrophic spurring.

The following day, the patient felt generally weak and became unresponsive. She was brought to the ER with acute respiratory distress and obtunded state with GCS of 6/15. Vitals were heart rate of 121 bpm (beats per minute), systolic blood pressure of 50 mmHg, agonal breathing with respiratory rate of 6 breaths per minute, and oral temperature of 35.2°C. Pupils were 3 cm and reacted bilaterally. No focal neurologic deficit was noted. The chest exam revealed decreased air entry bilaterally. Her cardiovascular, abdominal, and extremities exam results were normal. She was immediately intubated and fluid resuscitation was started. Initial lab results showed white cell count of 13.3 with 82% neutrophils, BUN 40 mg/dl, Cr 2.0 mg/dl, and lactic acid level of 10.5 mmol/dl. ABG (arterial blood gases) showed pH 7.01, PCO_2 58 mmHg, PO_2 489 mmHg, and HCO_3 14.6 on FiO_2 100%. EKG was normal except for sinus tachycardia. A computed tomography (CT) of the head and chest showed no acute processes. Further work-up with urine and serum drug screen, and cerebrospinal fluid analysis were unremarkable. The patient was treated for severe septic shock with multi-organ failure per guideline. After a few days, the patient's level of consciousness had started to improve but she remained hemodynamically unstable. Also, she failed multiple weaning trials and remained ventilator-dependant. Her ABG analysis showed persistent respiratory acidosis (Table 1). After the patient remained on the ventilator for 6 days, she was finally extubated, although she intermittently required noninvasive positive-pressure ventilation. Neurologic re-evaluation showed decreased movements

Table 1. Evolution of arterial blood gases during her hospital stay.

	Admission	Day 5	Day 6	Day 7	Day 8	Day 9	Day 14	Day 17	Day 19	Day 22
pH	7.01	7.252	7.278	7.305	7.291	7.348	7.338	7.302	6.848	7.133
$PaCO_2$, mmHg	58	55.7	51.9	53	53.5	50.6	55.8	62.8	154.7	71.6
PaO_2, mmHg	489	135.4	97.2	115	103.5	80.1	111.2	138.1	138.8	134.2
HCO_3	14.6	24.4	23.4	25.8	25.2	27.2	29.3	30.3	26.3	23.4
FIO_2, %	100	40	40	40	35	35	–	40	–	40
Mode	ETI	CPAP	CPAP	BiPAP	BiPAP	BiPAP	NC	VM	NC	CPAP
PEEP	5	5	5	15/5	PS-15	18/10	–	–	–	5

$PaCO_2$ – partial arterial carbon dioxide tension; PaO_2 – partial arterial oxygen tension; FIO_2 – fractional oxygen concentration of the inspired air; ETI – endotracheal intubation; PEEP – positive end expiratory pressure; CPAP – continuous positive airway pressure; BiPAP – bi-level positive airway pressure; NC – nasal cannula; VM – venturi mask.

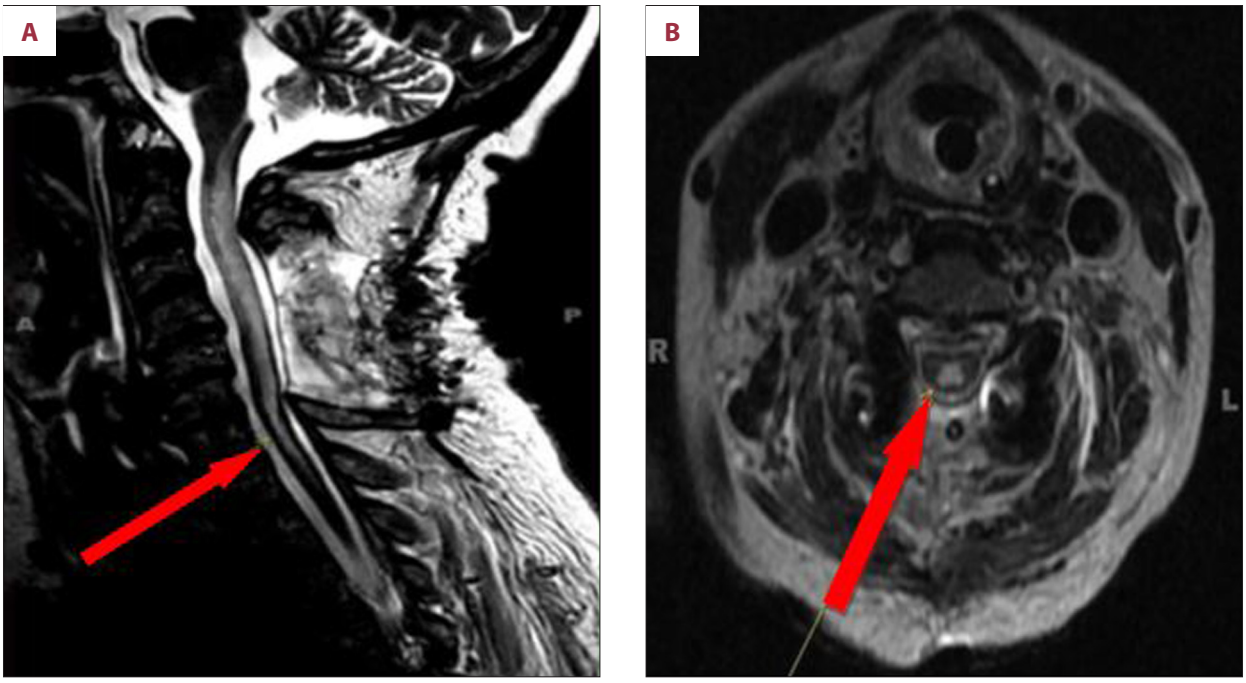

Figure 2. (**A, B**) An MRI of the brain and cervical spine demonstrated moderate degenerative disc disease and syringomyelia extending from C2 to C7 level (red arrows).

of all muscle groups against gravity and forces, with generalized weakness attributed to critical illness myopathy.

A few days later, she had progressive weakness of all extremities, causing quadriparesis and respiratory failure with marked respiratory acidosis. The patient was re-intubated. An MRI of the brain and cervical spine demonstrated moderate degenerative disc disease and syringomyelia extending from C2 to C7 level. No intraspinal tumors were identified (Figure 2A, 2B). Because of profound quadriparesis, she underwent C3-to-C6 decompressive laminectomy with posterior fusion. After failing several weaning trials, she was elected to undergo bronchoscopically-assisted tracheostomy. She was then transferred to a long-term care facility for further care.

Discussion

Difficult weaning is defined as failure of initial weaning and requiring up to 3 spontaneous breathing trials (SBT) or as long as

7 days from the first SBT to achieve successful weaning. This signifies that further weaning trials should be withheld and a thorough and systematic search for potentially reversible pathologies should be conducted. Cardiopulmonary causes account for the majority of difficult-to-wean conditions. Cardiac dysfunction leads to further reduction in pulmonary compliance and increase the work of breathing. It could be detected using bedside echocardiography [7] and high basal levels or an increase in B-type natriuretic peptides measured at the end of a spontaneous breathing trial [8,9]. After respiratory or cardiac causes have been excluded, the contribution of neuromuscular abnormalities should be considered.

Most neuromuscular dysfunction that complicates weaning is acquired during the ICU stay. ICU-acquired paresis due to critical illness myopathy (CIM) or polyneuropathy (CIP) is an independent predictor of difficult and prolonged weaning [10]. Weakness acquired in the ICU is 2–3 times more common than primary neuromuscular disorders such as Guillain-Barré syndrome (GBS), myopathies, or motor neuron diseases [5]. Primary neuromuscular disorders are usually apparent before weaning difficulties are encountered. However, new diagnoses can occasionally be made in the course of investigating the difficult-to-wean patient. Under-diagnosed disorders of the cervical spine spinal cord, including cervical spondylotic myelopathy (CSM), can compromise neuromuscular function of the respiratory system and motor function of the extremities. The diagnosis of CSM, which occurs in 5–10% of patients with symptomatic cervical spondylosis, is often missed because signs and symptoms are usually subtle. The hallmark symptoms associated with CSM are gait abnormalities and weakness or stiffness of the legs, which usually develop insidiously. However, abrupt worsening can occur, often following minor neck injuries [11]. The myelopathy can be attributed to 1 or more of 3 possible mechanisms: direct compression of the spinal cord by bony or fibrocalcific tissues, ischemia caused by compromise of the vascular supply to the cord, and/or repeated trauma secondary to normal flexion and extension of the neck [12].

Syringomyelia, central cavitation of the spinal cord, is rarely a primary disease process. It is generally associated with congenital malformations, such as Arnold-Chiari malformation, spinal arachnoiditis (post infectious or inflammatory), tumors (especially ependymoma and hemangioblastoma), and trauma. Spinal spondylosis has rarely been described as a cause of syringomyelia [13–15]. The pathophysiology of syringomyelia in association with cervical spondylosis is not completely understood. It is thought that craniospinal pressure dissociation from intermittent spinal cord compression might cause syrinx formation within the spinal cord. The clinical features of the syndrome are highly variable, usually with an insidious onset and a long and unpredictable progression. Patients usually present with progressive painless weakness, muscle wasting of the upper extremities, and segmental dissociated sensory loss. Acute deteriorations in syringomyelia could be related to rupture of the syrinx due to increased venous pressure associated with violent coughing or Valsalva maneuvers. Acute progression to paraplegia and/or quadriplegia and respiratory failure are certainly rare occurrences. Rarely, it has been reported to be associated with respiratory insufficiency and failure [6,16,17]. In a report, Fuller and Stanners postulated that the acute respiratory failure was due to sudden dissection of a long-standing syrinx, precipitated by violent coughing [18]. The diagnosis of syringomyelia is best established using MR imaging. The signal differences between watery CSF and neural tissue clearly outline the syrinx cavity on MR images. Surgical decompression with fenestration and/or shunt placement is recommended for patients with neurologic deterioration or intractable central pain. Syringoperitoneal shunting is a simple and effective means of reversing or arresting neurological deterioration in patients with syringomyelia [19,20].

Conclusions

Rare instances when cervical spondylotic myelopathy and syringomyelia present concomitantly can be devastating. In patients presenting with respiratory failure with no identifiable cardiopulmonary causes, syringomyelia should be considered. Health-care professionals should remember the possibility of asymptomatic spondylotic changes and syringomyelia and avoid abrupt or prolonged hyperextension of the cervical spine to minimize the risk of cord compression and respiratory failure.

Conflict of interest

None.

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