Occult Respiratory Dysfunction in a Craniovertebral Anomaly*

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Craniovertebral anomalies are developmental disorders that affect the skeleton and the enclosed neuraxis at the junction of the cranium and cervical spine. The two fundamental types of malformations result from faulty metameric segmentation or from dysraphic anomalies in the median sagittal plane. The associated clinical syndromes may be caused by any one of several factors, among which are bony deformities that produce compression or traction upon the neuraxis, intrinsic malformations of the nervous system, and disturbed circulation of cerebrospinal fluid.

The effect of these lesions on respiratory function has received scant attention in the literature. The hazards accompanying operative intervention were first appreciated by List and were described secondarily by Mullan and Raimondi. The flexed position of the head, which is of utmost importance for proper surgical exposure, may stretch already compromised neural tissues and so produce changes in respiration which, if not corrected, may result in apnea and death. Pathological confirmation of respiratory peril was described by Bharucha and Dastur who, in 20 operations, reported four operative mortalities from ventilatory failure. Necropsy in two patients demonstrated areas of softening and hemorrhage in the cervicomedullary region. In a later communication, a similar danger was associated with decompression of atlanto-axial dislocations.

Case Report

A 50-year-old woman was admitted with a complaint of occipital pain of 2 years duration. The pain was increased by coughing and sneezing; and, associated with these maneuvers, she experienced momentary periods of apnea and tetraparesis. She noted dizziness when she extended the neck. She had increasing difficulty in gait, as well as clumsy movements and numbness of her arms and hands. She denied bowel or bladder dysfunction.

Examination. She was a short, dysmorphic, woman with a short neck; otherwise, the general systemic examination was unremarkable. She was alert and oriented, the cranial nerves were intact, except for absent gag reflexes. Motor examination demonstrated a spastic, ataxic gait, worse on the right, but no loss in strength on either side. Coordination was impaired in all four extremities. Deep tendon reflexes were moderately hyperactive bilaterally, as was the jaw jerk. Hoffman and Babinski signs were present bilaterally. On sensory testing, vibration and position sense were impaired in both legs, but temperature, pinprick, and light touch were appreciated normally. Routine laboratory data were unremarkable.

Roentgenologic Findings. The chest x-ray was normal. Plain films (Fig. 1) of the cervical spine demonstrated abnormalities of the neural arches of C-1 and C-2. The anterior element of the atlas was fused with the base of the skull. In addition, platybasia was present with basilar invagination. There was minimal mobility of the dens in relation to the anterior arch of the atlas on flexion and extension. The diameter of the foramen magnum, from dens to posterior lip, measured 18 mm. Pantopaque myelography (Figs. 2 and 3) demonstrated widening of the spinal cord shadow in both the anteroposterior and lateral views beginning at the third cervical vertebra. The cisterna magna was small, and the fourth ventricle was in normal position. Further examination of the upper cervical spinal cord was carried out with air (Fig. 4). After the introduction of

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Fig. 1. X-ray films of the lateral cervical spine. *Left:* Fusion of the arches of C-1 and C-2 is seen (*white arrow*) with minimal basilar invagination (*black arrow*). *Right:* Fusion of the anterior arch of the atlas with the base of the skull is seen (*white arrow*). The distance from the posterior lip of the foramen magnum to the dens is only 18 mm.

Fig. 2. Myelograms with the patient in the prone position. *Left:* The spinal cord is abnormally wide above the C-3 level. *Right:* There is encroachment on the anterior subarachnoid space.
Complication of Craniovertebral Anomaly

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10 ml of air, the cord shadow was found to be enlarged in its anteroposterior diameter; after the introduction of more air, however, the cord appeared to collapse and its diameter became abnormally small. The ventricular system was symmetrical and moderately dilated.

Hospital Course. Our experience with percutaneous cervical cordotomy and the involvement of this area of the neuraxis in the control of pulmonary function stimulated the investigation of this patient, despite the absence of clinical evidence of pulmonary disease. Pulmonary function testing revealed a reduction of vital capacity to 40% predicted, a reduction of maximum breathing capacity to 47% predicted, and a reduction of lung compliance to 50% predicted. No underlying pulmonary disease could be detected to account for these changes. The low maximal intrathoracic pressure of 14 cm H2O in the presence of a reduced lung compliance supported the impression that the reduced vital capacity and maximum breathing capacity were due to extrapulmonary disease probably impaired motor function. The arterial pCO2 of 48 mm Hg and the pH of 7.38 indicated compensated alveolar hypoventilation, and the ventilatory response to inhaled CO2 was severely impaired. During testing, an irregular respiratory pattern was noted with the head in the neutral position; this was exaggerated by flexion or extension (Fig. 5).

The respiratory and neurological dysfunction was considered to be the result of repeated trauma to the cervicomedullary region by the pincer action of the bony anomalies. It, therefore, was decided to immobilize the upper cervical spine. This was accomplished with a collar, as surgery was thought to be contraindicated at this time.

Follow-Up. When the patient was re-examined at 1 and 3-month intervals, she
showed remarkable improvement in walking and less numbness of her arms and hands. The pulmonary status had improved significantly (Table 1). The vital capacity rose to 67% predicted and the resting ventilation was now adequate. The ventilatory response to inhaled CO₂, although still impaired, showed a significant increase, probably due to both a decreased threshold and an increased sensitivity.

Comment. The diameter of the foramen magnum was so very narrow that neurologic symptoms could have been predicted on this basis alone. The presence of bony anomalies did suggest the possibility of an associated neural lesion, such as hydromyelia or tumor; and, although splaying of the spinal cord and a small cisterna magna may be seen with the Arnold-Chiari malformation, the normal position of the fourth ventricle argued against this likelihood. There was demonstrated a change in spinal cord size, from abnormally large to abnormally small, suggesting the presence of hydromyelia, but the Pantopaque did not enter the central canal (presumably because of a small ostium connecting it to the fourth ventricle), as has been demonstrated in other reported cases of hydromyelia. We could find only one other report of a change in the size of the spinal cord during pneumography.

Discussion

The potentially serious changes in pulmonary function associated with craniovertebral anomalies are worth noting. A recent report of respiratory dysfunction in an attempt to relieve compression in the cervicomedullary region resulting from atlantoaxial dislocation emphasizes this problem. The patient had had respiratory insufficiency before surgery and was made worse by attempted reduction.
with skeletal traction. A transoral resection of the dens resulted in improvement, but intermittent respiratory assistance was still necessary 11 months after surgery.5

Similarly, three patients reported by Severinghaus and Mitchell11 became apneic while undergoing surgery involving the high cervical spinal cord or brain stem. Postoperatively, the patients had long periods of apnea associated with an irregular respiratory pattern. They were not able to maintain adequate ventilation while asleep and required assistance. Their ventilatory response to CO₂ was reduced, but the irregular respiratory pattern became regular at a high Pa CO₂.

It must be emphasized that chronic alveolar hypoventilation may be occult and difficult to recognize. Inadequate ventilation may not be suspected, because the patient may not have overt clinical evidence of pulmonary disease. Even when hypoventilation is considered, the physician’s observation and concern about the patient’s breathing may evoke sufficient hyperventilation in the patient to restore the arterial gas tensions to normal but the diagnosis may not be confirmed and an abnormally high serum bicarbonate may be the only tangible evidence of the carbon dioxide retention from chronic alveolar hypoventilation. The observation of alveolar hypoventilation is vital, furthermore, since it may be responsible for serious ventilatory depression or apnea which may follow the administration of sedatives or general anesthesia.

The surgeon’s index of suspicion must remain high, since occult respiratory dysfunction can be a serious threat to life in the treatment of otherwise benign and correctable diseases of the cervicomедullary junction.

Summary

A case of occult respiratory dysfunction in a patient with a craniovertebral anomaly has been reported. The ventilatory defect was, in part, reversed by immobilization of the head and neck.

It is clear that lesions in this region may result in chronic alveolar hypoventilation. The signs of respiratory insufficiency must be sought, despite the absence of clinical symptoms of pulmonary distress. Occult respiratory dysfunction may be responsible for ventilatory depression or apnea following the

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TABLE 1

Pulmonary function data before and after immobilization

<table>
<thead>
<tr>
<th></th>
<th>Room Air</th>
<th>5% CO₂</th>
<th>7.5% CO₂</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Initial</td>
<td>2 wks 3½ mos</td>
<td>Initial</td>
</tr>
<tr>
<td>Vital Capacity (% predicted)</td>
<td>40</td>
<td>67 66</td>
<td>5.0</td>
</tr>
<tr>
<td>FEV 1 sec/VC (%)</td>
<td>19</td>
<td>7.9</td>
<td>6.5</td>
</tr>
<tr>
<td>MBC (% predicted)</td>
<td>100</td>
<td>37    37</td>
<td>18</td>
</tr>
<tr>
<td>Lung Compliance (L/cmH₂O)</td>
<td>.08</td>
<td>.08</td>
<td>.08</td>
</tr>
</tbody>
</table>

Minute Ventilation (liter/min) 4.0 5.0
Rate 19 17 21 18 18 18 20
Tidal Volume (cc) 210 294 377 361 583 472 510 957
PACO₂ (mm Hg) 44 39 37 60 50 53 63 57
PaCO₂ (mm Hg) 48 39 37 60 50 53 63 57
pH 7.38 7.33 7.30
Oxygen Saturation (%) 97 99

FEV₁ seco/VC = forced expired volume in 1 second/vital capacity
MBC = Maximal breathing capacity
PACO₂ = Alveolar carbon dioxide tension
PaCO₂ = Arterial carbon dioxide tension
All volumes corrected to BTPS.
administration of sedatives or anesthesia, it may also become a serious threat to life in the surgical treatment of otherwise benign and correctable diseases of the cervicomedullary junction.

References