Acquired Paralysis of the Diaphragm

Michael Augustine Ko, MD, PhD, Gail Elizabeth Darling, MD*

KEYWORDS

• Diaphragmatic paralysis • Plication
• Phrenic nerve palsy • Dyspnea • Diagnosis

Diaphragmatic paralysis (DP) occurs when traumatic injury, systemic disease, or neurologic process results in the loss of control of the hemidiaphragms. Symptomatology is dependent on whether or not one or both hemidiaphragms are affected, the onset of paralysis, and the presence or absence of underlying pulmonary disease. Patients with severe respiratory derangement may require intermittent continuous positive airway pressure (CPAP) or bilevel positive airway pressure (BiPAP) or even prolonged mechanical ventilation. Less severely symptomatic patients can be managed through a program of respiratory rehabilitation or by operative diaphragmatic plication. Video-assisted techniques for plication are described and may be a less invasive alternative to open surgery. This review describes the etiology, clinical presentation, diagnosis, and treatment of acquired DP in adults.

ETIOLOGY

The diaphragm is a dome-shaped aponeurotic muscular structure that is enervated by the phrenic nerve, which arises from the third to the fifth cervical nerve roots. From the origin at the level of the scalenus anterior, the phrenic nerve follows a circuitous route in the posterior neck and mediastinum before entering the central tendon of the diaphragm. Because the only enervation of the diaphragm is via the phrenic nerve, total DP can be achieved by compression or damage to the phrenic nerve anywhere along its length (**Fig. 1**). The differential diagnosis of acquired DP is, therefore, broad and includes traumatic, compressive, neurogenic, myopathic, and inflammatory conditions (**Box 1**).

Unilateral DP (UDP) is commonly caused by ipsilateral phrenic nerve palsy. The most common cause of UDP is currently open heart surgery, occurring at an incidence of 2% to 20%.[1,2] The left nerve is more commonly affected than the right, perhaps due to injury during mobilization of the left internal mammary artery.[2] Another theory is that cold cardioplegia induces a direct thermal injury to the phrenic nerve during cardiac arrest.[1,2] In support of this, animal studies have demonstrated that local hypothermia induced by cardioplegia induces acute demylenation and axonal degeneration of the phrenic nerve.[3] In contrast, phrenic nerve injury is relatively uncommon in other noncardiac thoracic surgery, such as lung resections for carcinoma.[4] One exception is during thymectomy for thymoma, where the tumor is often invading or in close proximity to the superior extent of the nerve. The phrenic nerve was intentionally sacrificed for oncologic purposes in approximately 7% of a series of 183 patients.[5] Other rare causes of iatrogenic phrenic nerve palsy include injury after head and neck surgery,[6] chiropractic manipulation,[7] anesthetic blockade,[8] and central venous catheter placement.[9]

Unilateral phrenic nerve palsy can be induced by direct invasion or compression by space-occupying lesions at any point along its length. These include primary neoplasms, most commonly bronchogenic carcinoma or invasive thymoma (see **Box 1**). Less frequently, secondary neoplasms, such as metastatic pulmonary nodules or bulky mediastinal lymphadenopathy, may also lead to unilateral phrenic nerve palsy. Fortunately, most patients with incidentally found UDP do not have an occult intrathoracic

Division of Thoracic Surgery, Department of Surgery, University of Toronto, Toronto General Hospital, 200 Elizabeth Street, Room 9N-955, Toronto, ON, Canada M5G 2C4
* Corresponding author.
E-mail address: Gail.Darling@uhn.on.ca (G.E. Darling).

Thorac Surg Clin 19 (2009) 501–510
doi:10.1016/j.thorsurg.2009.08.011
1547-4127/09/$ – see front matter

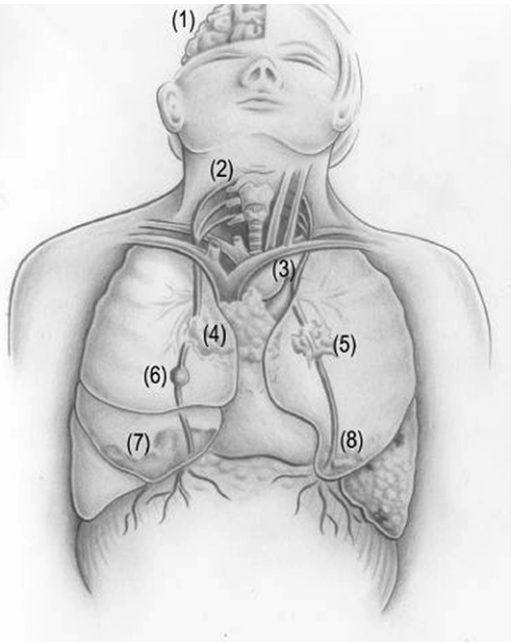

Fig. 1. Etiology of phrenic nerve palsy. DP can be caused by phrenic nerve damage anywhere along its length. **(1)** ALS. **(2)** Cervical osteoarthritis. **(3)** Aortic aneurysm. **(4)** Thymoma. **(5)** Lung carcinoma. **(6)** Peripheral nerve sheath tumor. **(7)** Pneumonia. **(8)** Tuberculosis.

malignancy. In a series of 142 patients with incidental diaphragmatic paresis, only 3.5% (n = 5) had an undiagnosed malignancy accounting for their symptoms.[10] Rarely, benign neoplasms, such as intrathoracic goiter[11] or phrenic nerve schwannoma,[12] may also present with unilateral phrenic nerve palsy. Non-neoplastic examples of compressive phrenic neuropathy include cervical osteoarthritis at the C3 to C5 nerve roots,[13] von Recklinghausen's disease,[14] and an expanding aortic aneurysm.[15]

Any peripheral neuropathy may affect the phrenic nerve, causing UDP. These include acute processes, such as MS[16] and chronic inflammatory demyelinating neuropathy.[17] For unknown reasons, diabetic neuropathy has a special predilection for phrenic nerve involvement.[18] Inflammatory conditions may adversely affect phrenic nerve function by causing local injury to the nerve (ie, pneumonia or tuberculosis) or by direct nerve damage, such as in herpes zoster or poliomyelitis.[19] Also, there have been several case reports of radiation therapy causing delayed phrenic nerve palsy several months to years after its administration.[20] If no obvious cause of DP is identified, it is labeled idiopathic. A recent report has suggested that such cases of idiopathic DP may be an acute viral infection of the nerve with herpes zoster; so-called Bell's palsy of the diaphragm.[21] Treatment of these

Box 1
Differential diagnosis of acquired diaphragmatic paralysis

1. Traumatic
    a. Penetrating injury
    b. Traction injury
        i. Motor vehicle collision
        ii. Cervical manipulation
    c. Iatrogenic
        i. Head and neck surgery
        ii. Cardiothoracic surgery
        iii. Anesthetic blockade
        iv. Central venous catheter placement
2. External phrenic nerve compression
    a. Neoplastic
        i. Pulmonary tumors
            1. Primary
                a. Non–small cell lung cancer
                b. Small cell lung cancer
                c. Carcinoid
            2. Secondary
        ii. Mediastinal tumors
            1. Thymoma
            2. Lymphoma
            3. Germ cell
            4. Intrathoracic goiter
            5. Phrenic nerve stimulation test
    b. Non-neoplastic
        i. Aortic aneurysm
        ii. von Recklinghausen's disease
        iii. Cervical osteoarthritis
3. Neurogenic
    a. Neuropathy
        i. Multiple sclerosis (MS)
        ii. Chronic inflammatory demylenating neuropathy
        iii. Diabetic neuropathy
        iv. Amyotrophic lateral sclerosis (ALS)
    b. Brainstem injury
        i. Ischemia
        ii. Compression
4. Inflammatory
    a. Pneumonia
    b. Polio
    c. Inclusion body myositis
    d. Herpes zoster
    e. Vasculitis
    f. Tuberculosis
    g. Radiotherapy
5. Idiopathic