Spontaneous Recovery in Idiopathic Unilateral Diaphragmatic Paralysis

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Idiopathic unilateral diaphragmatic paralysis is a rare condition that typically causes minimal symptoms, especially during exercise. Several reports indicated progressive improvement or even complete recovery to normal function of diaphragmatic paralysis that complicated various thoracic and extrathoracic conditions. In this case we describe a 57-year-old male with spontaneous recovery of idiopathic right hemidiaphragm paralysis and review reported cases of reversible diaphragmatic dysfunction. *Key words: diaphragm, idiopathic, phrenic nerve, plication, recovery, spontaneous, unilateral.* [Respir Care 2008;53(3):351–354. © 2008 Daedalus Enterprises]

Introduction

Diaphragmatic paralysis can occur after disruption of the phrenic nerve integrity. Idiopathic unilateral paralysis accounts for the majority of cases,[1] followed by malignancy and surgical trauma. Outcome and prognosis differ among affected subjects, from persistent disease to complete resolution, and appear to be directly related to the underlying etiology and whether muscle dysfunction is unilateral or bilateral.[2] In this report we describe a patient with spontaneous resolution of idiopathic unilateral diaphragmatic paralysis.

Case Summary

A 57-year-old male presented with a history of mild dyspnea on exertion and orthopnea of several months duration. His medical history was unremarkable, and he did not smoke. There was no history of diabetes, viral illness, trauma to the neck or the chest, recent surgery, skin rash, or neurologic disorders or symptoms. Physical examination was noncontributory except for dullness and decreased breath sounds at the base of the right lung. Chest radiograph revealed an elevated right hemidiaphragm (Fig. 1). Laboratory studies, including fasting blood glucose, antinuclear antibody, creatinine kinase, and thyroid stimulating hormone, were normal; fluoroscopy with sniff test revealed paradoxical motion of the right hemidiaphragm. Computed tomography of the chest, flexible bronchoscopy, and magnetic resonance imaging of the cervical spine revealed no additional findings. A moderate restrictive ventilatory defect (total lung capacity and vital capacity of 68% and 61% of predicted, respectively) and normal carbon monoxide diffusion capacity were present on lung function testing. A diagnosis of idiopathic unilateral diaphragmatic paralysis was made, and a conservative approach with regular follow-up was offered, in view of the benign nature of the patient's symptoms. The patient had no change in his clinical, physiological, or radiographic findings at 9 months of follow-up. Complete resolution of symptoms and normalization of spirometry (Table 1) was observed 21 months later. A repeat chest radiograph is shown in Figure 2.

Discussion

Diaphragmatic paralysis can involve either the whole diaphragm (bilateral) or only one leaflet (unilateral). The etiology remains unidentified in more than two thirds of cases.[3] Bilateral diaphragmatic paralysis is characterized by profound symptoms and abnormalities of pulmonary and respiratory muscle function, whereas unilateral dysfunction may present with very subtle symptoms and is

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The authors report no conflicts of interest related to the content of this paper.

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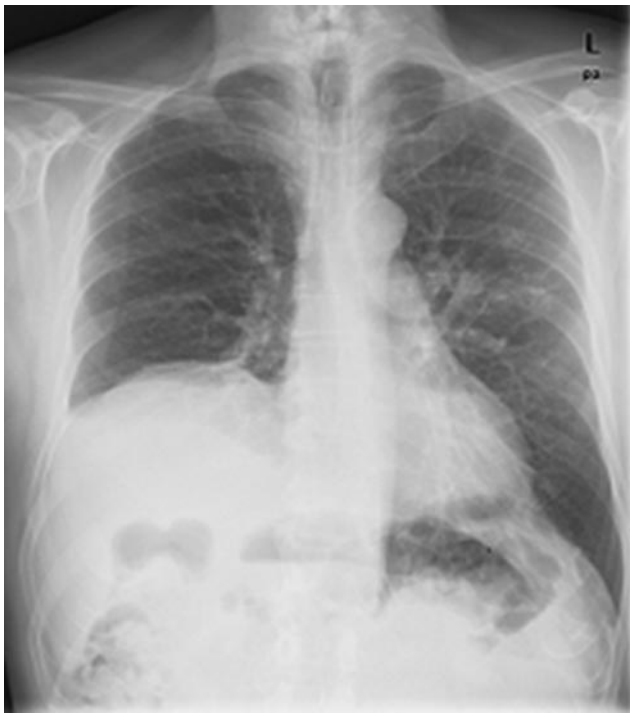

Fig. 1. Chest radiograph shows an elevated right hemidiaphragm.

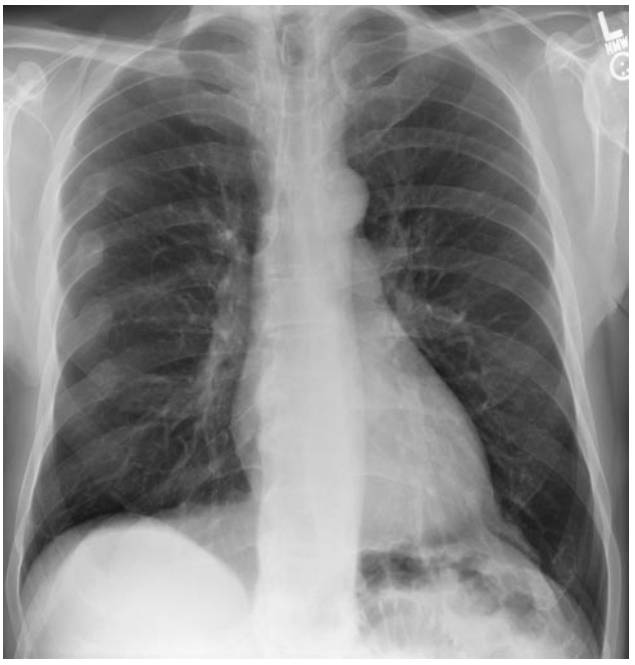

Fig. 2. Repeat chest radiograph shows resolution.

Table 1. Pulmonary Function Test Results*

	Baseline	9 Months	21 Months
Vital capacity (L, % predicted)	2.89 (62)	2.92 (64)	4.36 (87)
Total lung capacity (L, % predicted)	4.48 (66)	4.54 (68)	6.45 (90)

*The results show complete resolution of the restrictive ventilatory defect at 21 months of follow-up.

often discovered incidentally in patients undergoing chest radiography. Hemidiaphragmatic paralysis results in a vital capacity decrement of 10–30%, with the more substantial decrements seen in the supine position.[4] Measurement of the transdiaphragmatic pressure remains the accepted standard diagnostic test for bilateral paralysis, whereas fluoroscopy with sniff test reliably confirms the diagnosis of unilateral diaphragmatic paralysis. The M mode ultrasonography has been also introduced as an accurate method to evaluate the paralyzed diaphragm.[5] Findings on transthoracic ultrasound of the chest include paradoxical cephalad displacement of the hemidiaphragm, muscle atrophy, and evidence of decreased contraction and shortening during inspiration, compared to the normal diaphragm.

Treatment must be individualized and depends mainly on the severity of symptoms. It is now generally accepted that surgical treatment should be reserved for symptomatic unilateral paralysis that causes important derangement of lung function and/or gas exchange. Results from unilateral diaphragmatic plication performed on a series of 17 patients showed subjective and objective improvement that was maintained 5–7 years after surgery.[6] Conservative management for unilateral diaphragmatic paralysis is indicated if the patient is asymptomatic or if impairment is mild or moderate, and may be considered if waiting for spontaneous recovery could be justified in patients with non-neoplastic diaphragmatic paralysis.

Several reports have indicated the potential of recovery of the paralyzed diaphragm. Douglas and Calgett reported 40 patients with idiopathic unilateral diaphragmatic paralysis more than 40 years ago.[7] Six patients exhibited spontaneous return to normal function, as evidenced by chest fluoroscopy, after intervals that ranged from 2 to 19 years. Valls-Sole et al[8] described progressive clinical and electrophysiological progressive improvement in diaphragmatic function in a 41-year-old male with idiopathic bilateral phrenic nerve neuritis over a 2-year period from symptom onset. Spontaneous recovery of diaphragmatic paralysis complicating lobar pneumonia has been described, with recovery time extending up to one year after diagnosis.[9] Progressive improvement of muscle function was also illustrated in the literature by several case series and case reports, for example, after right internal mammary artery harvesting during open heart surgery,[10] obstetrical trauma,[11] liver transplantation,[12] following mediastinal radiotherapy,[13] pulmonary embolism,[14] trauma,[15] neck manipulation,[16] cervical herpes zoster,[17] and spider bite (*Loxosceles* poisoning).[18] Paraneoplastic bilateral

Table 2. Reported Recovery Times in Diaphragmatic Paralysis

First Author	Associated Condition	*n*	Number Who Recovered	Unilateral or Bilateral	Time to Return of Function
Escande[3]	Breech delivery	1	1	Unilateral	2.5 mo
Roy[4]	Liver transplantation	43	8	Unilateral	3 to 9 mo
Riley[5]	Lobar pneumonia	27	15	Unilateral	24 d to 12 mo
De Vito[6]	Mediastinal radiotherapy	1	1	Bilateral	48 mo
Pils[7]	Pulmonary embolism	1	1	Unilateral	2 mo
Iverson[8]	Trauma	10	2	Unilateral and bilateral	6–12 mo
Pandit[9]	Neck manipulation	1	1	Bilateral	36 mo
Stowasser[10]	Zoster	1	1	Bilateral	6 mo
de Rezende[11]	*Loxosceles* poisoning	1	1	Unilateral	1 mo
Rijnders[12]	Renal cell carcinoma	1	1	Bilateral	24 mo
Valls-Sol[13]	Idiopathic	1	1	Bilateral	24 mo
Douglas[14]	Idiopathic	40	6	Unilateral	2–19 y
Present case	Idiopathic	1	1	Unilateral	21 mo

diaphragmatic paralysis with recovery after tumor therapy was observed after nephrectomy in a patient with renal cell carcinoma.[19] The recovery time was highly variable and ranged from less than one month to several years, as shown in Table 2.

The mechanism that leads to phrenic nerve injury and its potential to recover remain elusive. The recovery time of phrenic nerve function may be related to the underlying insult. Injuries that involve a focal demyelination of the phrenic nerve may show complete recovery in a few weeks, via nerve remyelination, whereas longer recovery time is required for axonal regrowth in cases where axonal damage is present.[20] Other mechanisms, such as relative preservation of neuromuscular junction after injury and reflex mechanisms mediated by the vagal nerve, were suggested in experimental animal models to affect the compensatory response of the phrenic nerve electrical activity.[21,22] A retrospective review[23] of ventilated patients with high cervical spine injury and diaphragmatic paralysis showed that 21% of subjects were ultimately able to breathe independently after 4–14 months (mean 246 d), and an additional 15% showed signs of phrenic nerve recovery on electromyography testing. Although the location of the lesion (diaphragm vs phrenic nerve) and transdiaphragmatic pressure gradient could have been further established by more rigorous testing, including electromyogram and insertion of a transesophageal catheter, the absence of atypical features and bilateral involvement of the diaphragm in our patient indicates that the sniff test was probably adequate for a presumptive diagnosis of idiopathic unilateral diaphragmatic paralysis.

In summary, phrenic nerve function recovery depends on the underlying process and may be mediated by local or reflex neurotrophic factors. This report supports the potential spontaneous resolution of unilateral diaphragmatic paralysis, even if the paralysis is idiopathic. Avoiding invasive surgical intervention and considering a conservative approach with close follow-up may be reasonable in these patients, especially if clinical symptoms are minimal or tolerable and an important physiologic derangement is not present.

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