Schwannoma of the accessory phrenic nerve

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Received 22 August 2006

Abstract

Otolaryngologists are frequently confronted with the management of cervical tumors. Neurogenic tumors concern, especially, the cranial, sympathetic, or peripheral nerve sheaths. These tumors are benign and grow slowly. The involvement of the cervical part of the phrenic nerve is exceptional, and only 2 cases are reported in the literature. We describe the first case of a cervical schwannoma involving the accessory phrenic nerve. The anatomy and function of the accessory phrenic nerve are reviewed.

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1. Case report

A 45-year-old woman was referred to the ear, nose, and throat department of the Réseau Hospitalier de Médecine Sociale with a 6-year history of left cervical mass. In fact, this mass was first diagnosed by her general practitioner in 1999. At that time, a computed tomography (CT) revealed a tumor measuring 2.3 cm and involving the cervicothoracic inlet (Fig. 1A). The patient was asymptomatic and refused the surgery. Five years later, she consulted for pain in the supraclavicular area. The solid mass was palpable in the supraclavicular region, but the rest of the standard ear, nose, and throat examination was normal. The CT demonstrated a very small increase in the size (0.4 cm) of the mass involving the cervicothoracic inlet.

We performed the excision of this tumor using a cervical approach (Fig. 1B). The tumor was firm, adherent to the jugular internal vein, and located close to the phrenic nerve. A second nerve was visualized adjacent to the phrenic nerve and going through the tumor (Fig. 1B). Caudally, this nerve merged into the phrenic nerve and was therefore considered as an accessory phrenic nerve. A radical excision of this mass was performed, including the accessory phrenic nerve. Histologic examination revealed a benign schwannoma. There were no postoperative complications and no respiratory complaints. The pulmonary function tests were within normal limits. The vital capacity and total lung capacity were, respectively, 2.87 L (109% of predicted value) and 5.04 L (115% of predicted value), and the inspiratory capacity was 2.26 L. A functional magnetic resonance imaging demonstrated a partial left diaphragmatic palsy. On frontal views (Fig. 2), the caudal displacement of the diaphragmatic dome during a deep inspiration was 40.6 mm on the right side but only 3.2 mm on the left. In addition, on the right side, the displacement of the anterior portion of the diaphragm was less than half of the posterior, whereas on the left side, this anterior portion was the only moving portion. Six months after surgery, inspiratory capacity had increased by 270 mL.

2. Discussion

Our review of the literature shows that schwannomas involving the phrenic nerve are exceptional (21 reported cases) and are most commonly observed as intrathoracic masses [1]. Mevio et al [1] reported 2 cases of schwannoma...
involving the cervical part of the phrenic nerve. We describe the first case of schwannoma of the accessory phrenic nerve. Management of these neurogenic tumors is primarily surgical excision, which is curative. No recurrence has been described so far. The parent nerve should be preserved, if possible, by using intracapsular enucleation. Repair using graft or end-to-end anastomosis have been proposed but never carried out [1].

The section of the accessory phrenic nerve led us to reinvestigate its origin and its functional contribution to the diaphragmatic innervation. In humans, the main trunk of the phrenic nerve commonly originates from the cervical nerve roots C-3, C-4, and C-5 [2]. In an autopsy study, Cailot et al [2] observed accessory phrenic nerves accounting for only 4% of nerves in humans. On the other hand, rabbit and rat cervical dissections demonstrated a higher percentage of accessory phrenic nerves [3]. In those species, the accessory phrenic nerve originates from C-6 and leaves the brachial plexus far lateral from the position of the phrenic nerve. Studies in different species have demonstrated that the diaphragm is innervated somatotopically after an anteroposterior distribution [3]. Specifically, the ventral portions of the costal and crural regions are innervated by the upper ventral roots, and the dorsal portions of both regions are primarily innervated by the lower roots. In animals, the distribution of the accessory nerve into the diaphragm corresponds to the

Fig. 1. (A) A CT showing the relationship between the tumor, the subclavicular vessels, and the pulmonary apex. (B) Operating view revealing the entrance of the accessory phrenic nerve into the tumor.

Fig. 2. On frontal views (magnetic resonance imaging), the caudal displacement of the diaphragmatic dome during a deep inspiration (B) was 40.6 mm on the right side and only 3.2 mm on the left [panel (A) shows diaphragmatic dome during expiration].
somatotopical distribution of phrenic nerve fibers originating from C-6 ventral root [3]. In experimental conditions, the preservation of this accessory nerve during phrenicotomy is sufficient to maintain significant diaphragmatic function [4]. In our case, no respiratory consequences of the selective resection of the accessory phrenic nerve have been experienced. The reduction of displacement was predominant in the posterior region, confirming in man the somatotopical distribution of the accessory phrenic nerve observed experimentally in animals. In spite of this huge reduction in displacement, vital capacity as well as total lung capacity were in reference range. Several factors explain this observation. First, even in the presence of a complete unilateral paralysis of the diaphragm, the functional consequence is limited. In the literature, the reported reduction in vital capacity in that case amounts to only 20% to 25% in the sitting position [5]. This is in relation with the nonlinearity of the respiratory system compliance. At high lung volume, a lowering in pleural pressure only induces a small change in lung volume. Thus, a reduction in the ability to generate a high level of pleural depression is associated with only a small reduction in change in lung volume. In addition, there is interdependence between the 2 hemidiaphragms, and during the contraction of the intact hemidiaphragm, there is a passive tension generated in the paralyzed hemidiaphragm that contributes to the change in lung volume. Moreover, in the case of a partial paralysis, such as that observed here, the residual activity has its own inspiratory effect but also impedes excessive shortening of the contralateral hemidiaphragm that would otherwise reduce the aptitude to lower pleural pressure. Finally, 6 months after surgery, the vital capacity of our patient was above reference range. We postulate that the improvement of 12% corresponds to the initial loss and was due to collateral reinnervation of diaphragmatic muscle fibers.

References