Primary Tracheal Neurilemoma (A Case Report)

Yeong Jee Kim, Han Cha, Young Yull Koh, Joo Hyun Kim*, Je G. Chi**
and Kyung Mo Yeon***

Department of Pediatrics, Thoracic Surgery*, Pathology** and Radiology***
Seoul National University Children’s Hospital and College of Medicine, Seoul National University, Seoul 110, Korea

Abstract: Primary tracheal neurilemoma is a very rare neoplasm. Only 17 cases were reported in the literature during 1950–1985. We report a case in a 15-year-old Korean boy who presented with persistent productive cough and dyspnea. An endobronchial lobulated mass was demonstrated in the left main bronchus. He was treated successfully by left bronchial resection and carinal reconstruction. The tumor was diagnosed as primary tracheal neurilemoma. This is the second case of primary tracheal neurilemoma diagnosed in Korea.

Key words: Neurilemoma, Trachea

INTRODUCTION

Primary neurogenic tumors are rare in the lower respiratory tract, being more frequently reported in the lung and bronchi than the trachea (Straus and Guckien 1951; Miller 1969; Silverman et al. 1976; Ma et al. 1981).

Of the neurogenic tumors, neurilemomas form approximately one third of the group (Bartley and Arean 1965). The most common locations are the flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots, and cerebellopontine angle (Oberman and Suller 1967).

To our knowledge, the total number of tracheal neurilemomas reported in the literature from 1950 to 1985 is seventeen (Kim et al. 1985). We report a new case along with a brief review of the previous reports of tracheal neurilemoma.

CASE REPORT

A 15-year-old Korean boy was admitted to Seoul National University Children’s Hospital on July 14, 1986, because of a one month history of difficult respiration that didn’t respond to therapy which included bronchodilators and steroids, and was aggravated 3 days prior to admission. He had a history of persistent productive cough and intermittent hemoptysis of 5 months’ duration, without other symptoms of systemic illness.

On physical examination, respiration rate was 28 per minute. Alae nasii flaring, suprasternal and substernal retractions were detected, with wheezing and moderate aeration over both lung fields. Arterial blood gas values in room air were pH 7.48, pO₂ 41 mmHg, bicarbonate 32 mmol/L. A chest roentgenogram showed right lung herniation with left lower lobe collapse. Bronchogram revealed a round lobu-
lated mass arising from the left tracheobronchial angle which completely obstructed the proximal left main bronchus (Fig. 1). At bronchoscopy, an erythematous movable mass was found covered with whitish necrotic material.

On August 1, 1986, left bronchial resection and carinal reconstruction were performed. At postoperative bronchoscopy, complete removal of tumor and patency of both main stem bronchi were confirmed. He was discharged on the 12th postoperative day, and the patient is doing fine as of the day of this report.

Pathology: The polypoid tumor was a reddish round smooth-surfaced friable mass, 1.5 cm in diameter, covered with whitish brown exudate. Microscopically, the moderately cellular tumor mass consists of spindle cells, arranged mostly in haphazard pattern with many areas of microcystic change, hemosiderin deposits and collagen fibrosis. Vessels were prominent. In a few areas, however, the tumor cells were arranged in a palisading fashion or in an organoid pattern. Occasional large hyperchromatic nuclei were present. However, mitotic figures were absent (Fig. 2, Fig. 3).

DISCUSSION

The first report of a primary tracheal neurilemoma was in 1951(Straus and Guckien). Until 1985, a total of 17 cases were reported. They occurred in a wide age range from 6 to 71 years in both sexes, at all levels of the trachea(Kim et al. 1985). The symptoms may be insidious in onset and the tumor may occlude 3/4 of the lumen before symptoms are reported. The most common symptom is wheezing. Cough, dyspnea and hemoptysis are frequent. Pain, weight loss and malaise are unusual (Cleveland et al. 1977; Parrish et al. 1983). With progressive growth and more complete bronchial obstruction, secondary pulmonary changes occur (Noya et al. 1970; Nass and Cohen 1979; Horovitz et al. 1983).

Because primary tracheal tumors are rare, the inclusion of tumors in the differential diagnosis of adult-onset dyspnea and wheeze would be non-practical. However, if the asthmatic patient’s progress is less than satisfactory, the physician should consider obstructing lesions of the major airways (Karlan et al. 1973; Horovitz et al. 1983; Parrish et al. 1983). High-penetration chest radiography, fluoroscopy, tomograms and computerized tomography of the trachea(Horovitz et al. 1983; Parrish et al. 1983), pulmonary function tests(Parrish et al. 1983), and bronchogram(Noya et al. 1970) are helpful adjuncts for this. Bronchoscopy should be carried not only for diagnosis but also for planning the surgical resection(Kim et al. 1985).

Because of the possibility of severe hemorrhage and incomplete excision with early recurrence and due to technical difficulties in removal, endoscopic removal of tumors is a controversial approach, and tracheal resection with primary anastomosis, lobectomy, or pneumonectomy have been used (Abdulio et al. 1976; Nass and Cohen 1979; Ma et al. 1981; Horovitz et al. 1983). The prognosis is excellent following surgical excision, and postoperative recurrence is rare(Harkin and Reed 1969). However, malignant von Recklinghausen’s disease was reported(Hanada et al. 1982). Because the tumor is characterized by slow growth and later recurr-
ence is possible, annual follow-up physical examinations, including chest X-ray films are recommended (Nass and Cohen 1979).

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=국문초록=
원발성 기관 신경섬유초종 1례

서울대학교 의과대학교 소아과학교실・통부외과학교실**・병원학교실***・방사선과학학교실***

김영자・차 한・김영주・김주현**・김재근**・연경모***

저자들은, 6개월간의 기관신경초종의 확장성 및 부신림절 호르몬 등의 치료에 반응없는, 1개월간의 진행성 호흡곤란을 주소로 내원한 15세 남아에서 기관절은을 발견하고, 기관절은이 성장 속도 및 단단함을 심화하였던 내의 증상의 조직검사상 원발성 기관신경섬유초종을 나타내었 다. 이는 1951년부터 1985년까지 전체적으로 17례만이 보고된 매우 드문 증상이고 국내에서 는 두번째 증례로 사료되어 보고하는 바이다.