Primary Tracheal Neurilemoma: 
A Case Report and Review of the Literature

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Abstract—A rare case of primary tracheal neurilemoma which was treated successfully by tracheal resection and anastomosis is reported. This case is a 51-year old Korean woman, who had 9 months history of cough, exertional dyspnea and recent aggravation of respiratory distress since 2 months prior to admission. Conventional symptomatic drug treatment and radiation therapy failed to give clinical relief and resection and anastomosis was done without difficulty. Postoperatively, pulmonary edema developed with unknown cause but was recovered on the third postoperative day without clinical problems.

This is the first case in Korea and added as the 4th case of tracheal neurilemoma in yellow race population (3 cases are from Japan). So far 13 cases are reported in Caucasian population in the world literature (Horovitz et al. 1983).

Key Words: Neurilemoma(Primary), Trachea

Benign tracheal tumors, although rare lesions, encompass a wide variety of pathologic types.

In 1978, Grillo published a collected series of 63 patients with primary tracheal tumors. In this series there were only nine cases of benign tumors; chondroma, chondroblastoma, squamous papilloma, granular cell tumor, hemangioma, and fibroma. Other types of tumor can also occur in the trachea. They are hamartoma(Engelking 1959), amyloid tumors(Holinger 1950), mixed tumor of salivary gland type (Ma et al. 1979), fibrous histiocytoma (Hakimi et al. 1975), osteoma(Karlan et al. 1973), intratracheal goiter(D’aunoy et al. 1931) lipoma (Plachta et al. 1962), lymphangioma (Redo et al. 1965), neurofibroma (Meredith et al. 1978), and neurilemoma (Straus et al. 1951; Silverman et al. 1976; Nass et al. 1979; Ma et al. 1981). Only a limited number of tracheal neurilemoma has been reported since the first case report made by Straus and Guckien in 1951. Although Horvitz et al. (1983) reported 13 cases occurred in all white persons, we could find three cases from Japanese literature (Takahashi et al. 1963; Makino et al. 1979; Katagiri et al. 1982). We report a seventeenth case in the world literature and the fourth case in a yellow person.

Case report: The patient was a 51-year old yellow Korean woman who, for nine months, had complained of cough, exertional dyspnea, wheeze on exertion, and of a more recently developed respiratory distress in the two months preceding admission. Treatment with bronchodilators, performed under the diagnosis of bronchial asthma at other clinic, did not give relief. She was transferred to Seoul National University Hospital on January 5, 1984. The patient lost 13 kg of weight during nine months. She had had appendectomy 20 years ago, and nephrectomy 15 years ago for unknown reasons. On physical examination, she looked chronically ill, with wheezing and coarse breath sounds over both lung fields. Arterial blood values were pH 7.46, PO₂ 66, PCO₂ 56, bicarbonate 40 on room air. Vital signs were BP 100/80, pulse rate 100 per minute, body temperature 36.2°C and respiratory...
rate 20 per minute. Laboratory examination: WBC 14,700, hemoglobin 12.0 gm. Urinalysis: trace of glucose. Chest X-ray was normal.

She did not respond to bronchodilators and steroids. On the 15th hospital day a sudden dyspneic attack occasioned with a finding of a tumor mass in the computed tomography of the chest (Fig. 1). This showed an intratracheal mass with protrusion of the tumor beyond the tracheal wall 4 cm above the carina. Radiation therapy without biopsy was recommended. She received 5000 rad in five weeks. The dyspnea was relieved a little, but tomograms after radiation showed a persistent intratracheal mass (Fig. 2). On March 20, 1984 the patient was operated upon. The trachea was exposed through a low cervical and median sternotomy incision. The tumor arose from the posterior wall of the trachea 4 cm above the carina and filled approximately 90% of the lumen.

The surface of the tumor was smooth with well defined margins and prominent vessels. 3 cm of trachea was resected and end to end suture was done by standard anesthetic and surgical techni-
Fig. 5. Fascicles of spindle cells (straight arrows) alternate with edematous areas containing (curved arrows) lymphocytes. (hematoxylin–eosin, ×64)

Fig. 6. Nuclear palisades (arrows) (hematoxylin–eosin, ×160). The nuclei are regimented near areas of fibrillar stroma without cells (F).

Pathology: The length of the resected tracheal specimen was 2.7 cm. The lumen of the trachea was markedly compromised by a 2.7×3.0×2.0 cm encapsulated, pink polypoid tumor attached to the posterior wall. It extended posterolaterally beyond the trachea as a dumbbell shape. The tumor which is composed of spindle cells which form fascicles are separated by edematous stroma with lymphocytes (Fig. 5). The tumor was well delineated but not encapsulated from surrounding connective tissue. It elevated the mucosa, occupied the wall of the trachea, and expanded the peritracheal adventitia. The nuclei of the spindle cells had pointed ends, moderate chromatin, and no mitoses. Throughout the tumor there were palisades of nuclei. These palisades abutted poorly cellular regions of eosinophilic fibrillar tissue to create verocay bodies (Fig. 6).

Comment: Primary tracheal neurilemoma is a rare tumor. Seventeen cases of primary tracheal neurilemoma including our case were found in the literature from 1950 to 1984. They occurred in an age range from 6 to 71 years in both sexes with a slight predilection for women. Nine cases were female and 7 cases were male. Sex was not specified in one (Table 1). Barty and Arean in 1965, commented that neurilemoma of the tracheobronchial tree rarely invades the bronchus, but traverses the intercartilaginous fibrous septa to adopt a dumbbell shape as in our case.

Histologically two types of neurilemoma were described by Antoni. Type A has an orderly arrangement of elongated cells with blunted ends, anastomosing cell bodies and, most characteristically, palisading nuclei. Type B lacks this orientation and consists of haphazardly arranged cells with intercellular edema imparting a loose-textured appearance.

These two configurations often coexist in a given tumor (Nass et al. 1979). Because tracheal tumors
<table>
<thead>
<tr>
<th>Author</th>
<th>Date</th>
<th>Age (yr)</th>
<th>Race</th>
<th>Sex</th>
<th>Site</th>
<th>Symptom</th>
<th>Therapy</th>
<th>Follow-up</th>
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<tr>
<td>Straus</td>
<td>1951</td>
<td>28</td>
<td>C</td>
<td>M</td>
<td>L</td>
<td>Cough, fever, pneumonia</td>
<td>Two-stage endoscopy, silver nitrate cauter y</td>
<td>Well, 6mo</td>
</tr>
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<td>1953</td>
<td>21</td>
<td>C</td>
<td>M</td>
<td>L</td>
<td>Difficulty breathing cough, bronchitis</td>
<td>Endoscopic excision cauter y</td>
<td>No Sx, 5yr</td>
</tr>
<tr>
<td>Ivanov</td>
<td>1953</td>
<td>35</td>
<td>C</td>
<td>F</td>
<td>M</td>
<td>Cough, difficulty breathing</td>
<td>Surgical resection</td>
<td>Well, 1mo</td>
</tr>
<tr>
<td>Kittenger</td>
<td>1961</td>
<td>23</td>
<td>C</td>
<td>F</td>
<td>L</td>
<td>Difficulty breathing stridor on exertion</td>
<td>Endoscopic excision repeated after 2wk</td>
<td>Well, 21/2yr</td>
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<tr>
<td>Takahashi</td>
<td>1963</td>
<td>12</td>
<td>Y</td>
<td>M</td>
<td>U</td>
<td>Difficulty breathing</td>
<td>Tumor excision through tracheotomy</td>
<td>Well, 11/2yr</td>
</tr>
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<td>Gerashchenkol</td>
<td>1964</td>
<td>6</td>
<td>C</td>
<td>F</td>
<td>L</td>
<td>Difficulty breathing cough, weakness</td>
<td>Surgical transection postop.</td>
<td>Pneumonia, died</td>
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<td>1970</td>
<td>43</td>
<td>C</td>
<td>M</td>
<td>U</td>
<td>Difficulty breathing cough, hemoptysis</td>
<td>Tracheal sleeve resection</td>
<td>Well, 21/2yr</td>
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<tr>
<td>Karlan</td>
<td>1973</td>
<td>21</td>
<td>C</td>
<td>F</td>
<td>M</td>
<td>Difficulty breathing cough, pneumonia</td>
<td>Tracheal fissure resection</td>
<td>Well, 1yr</td>
</tr>
<tr>
<td>Conley</td>
<td>1975</td>
<td>28</td>
<td>C</td>
<td>M</td>
<td>U</td>
<td>Cough, difficulty breathing</td>
<td>Surgical resection</td>
<td>Lost to follow-up</td>
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<td>Conley</td>
<td>1975</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Surgical resection</td>
<td>Lost to follow-up</td>
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<tr>
<td>Gouin</td>
<td>1977</td>
<td>71</td>
<td>C</td>
<td>F</td>
<td>M</td>
<td>Recent onset of asthma &amp; cough</td>
<td>Endoscopy aborted, tracheal sleeve resection</td>
<td>Renal insuff., mediastinitis, died postop.</td>
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<td>36</td>
<td>C</td>
<td>M</td>
<td>L</td>
<td>Difficulty breathing hemoptysis, wheezing</td>
<td>Endoscopy with electrocaagulation</td>
<td>Well, 4yr</td>
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<td>Makino</td>
<td>1979</td>
<td>65</td>
<td>Y</td>
<td>F</td>
<td>U</td>
<td>Recent onset of asthma</td>
<td>Tumor excision through tracheotomy</td>
<td>No Sx, 1yr</td>
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<tr>
<td>Ma</td>
<td>1981</td>
<td>23</td>
<td>C</td>
<td>F</td>
<td>L</td>
<td>Difficulty breathing wheezing</td>
<td>Tracheal sleeve resection</td>
<td></td>
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<tr>
<td>Horovitz</td>
<td>1982</td>
<td>38</td>
<td>C</td>
<td>F</td>
<td>L</td>
<td>Difficulty breathing cough, shortness of breathing</td>
<td>1. Endoscopic removal  2. Tracheal resection</td>
<td>Well, 11/2yr</td>
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<tr>
<td>Katagiri</td>
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<td>65</td>
<td>Y</td>
<td>M</td>
<td>U</td>
<td>Difficulty breathing asthma, cough</td>
<td>Tracheal resection</td>
<td></td>
</tr>
<tr>
<td>Current case</td>
<td>1985</td>
<td>51</td>
<td>Y</td>
<td>F</td>
<td>M</td>
<td>Asthma, difficulty</td>
<td>Tracheal resection</td>
<td>Well, 1yr</td>
</tr>
</tbody>
</table>

Legend: C; Caucasian, Y; yellow, L, M, and U; Lower, middle and upper trachea
are rare, they are often not included in the diagnostic consideration of the patient with airway diseases. These patients are given the diagnosis of more common entities such as chronic bronchitis, asthma, and heart failure (Karian et al. 1973).

Rigid or flexible bronchoscopy, the definite diagnostic procedure should have been carried in the present case not only for diagnosis but also for planning the surgical resection. In light of the fact that this tumor has recurred after excision by endoscopy (Horovitz et al. 1983) the treatment of choice of tracheal neurelioma is resection with end to end suture.

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= 국문초록 =

원발성 간절 사염 신경세포증 治験 一例

서울대학교 의과대학 홍부외과학교실, 방대학교실。* 아취과학교실** 및 내과학교실

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저자들은 호흡곤란을 주소로 응급실을 통해 입원하여, 홍부 전산단층촬영 등으로 기관중앙을 확인한 51세 남 여자환자를 경관절체 및 단단문란으로 시행, 완쾌시키고, 조직검사상 일반성 기관 신경섬유조직에 확인된 바 이는 현재까지 세계문헌상 백인에서 13예, 황인(일본인)에서 3예 만이 보고된 희귀한 예로 저자들의 지식으로는 국내 초유의 증례로 사료되어 문헌고찰과 더불어 보고하는 바이다.