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(Brief Communication)

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, PO_2 66, PCO_2 56, bicarbonate 40 on room air. Vital signs were BP 100/80, pulse rate 100 per minute, body temperature 36.2°C and respiratory

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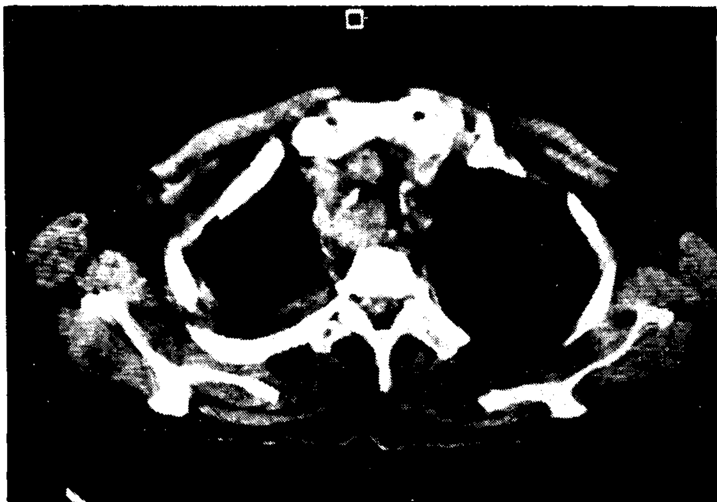

Fig. 1. Computed tomography of the chest. 4 cm above carina the tumor occupies 90% of the tracheal lumen and extends extratracheally.

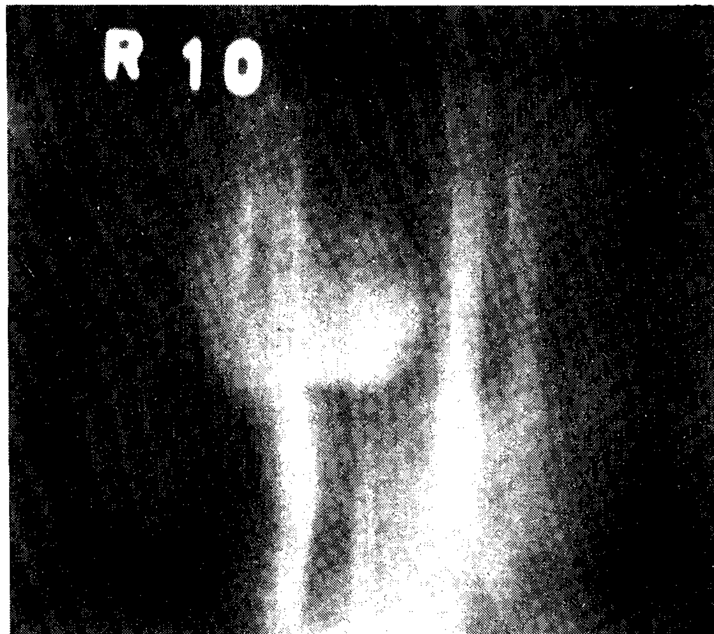

Fig. 2. Tomography of the trachea after 500 rad of radiation therapy. The size of the tumor has not changed.

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 tho trachea 4 cm above the carina and filled approximately 90% of the lumen.

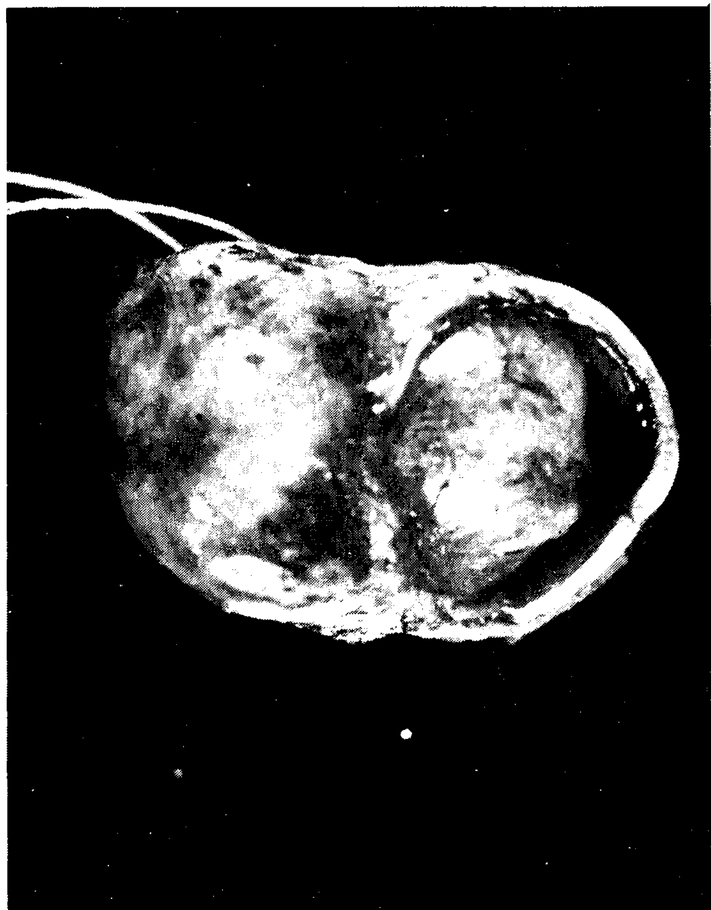

Fig. 3. The resected tracheal segment shows a dumbbell shaped tumor.

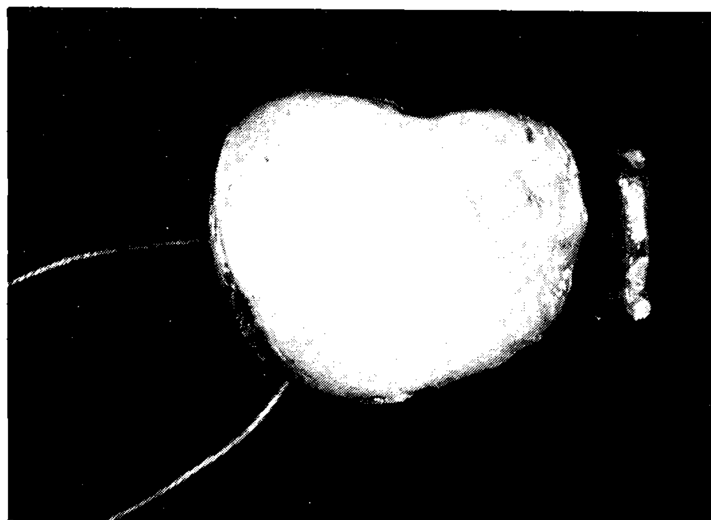

Fig. 4. Cut surface of the tumor

The surface of the tumor was smooth with well defined margins and prominent vessels. 3cm of trachea was resected and end to end suture was done by standard anesthetic and surgical techni-

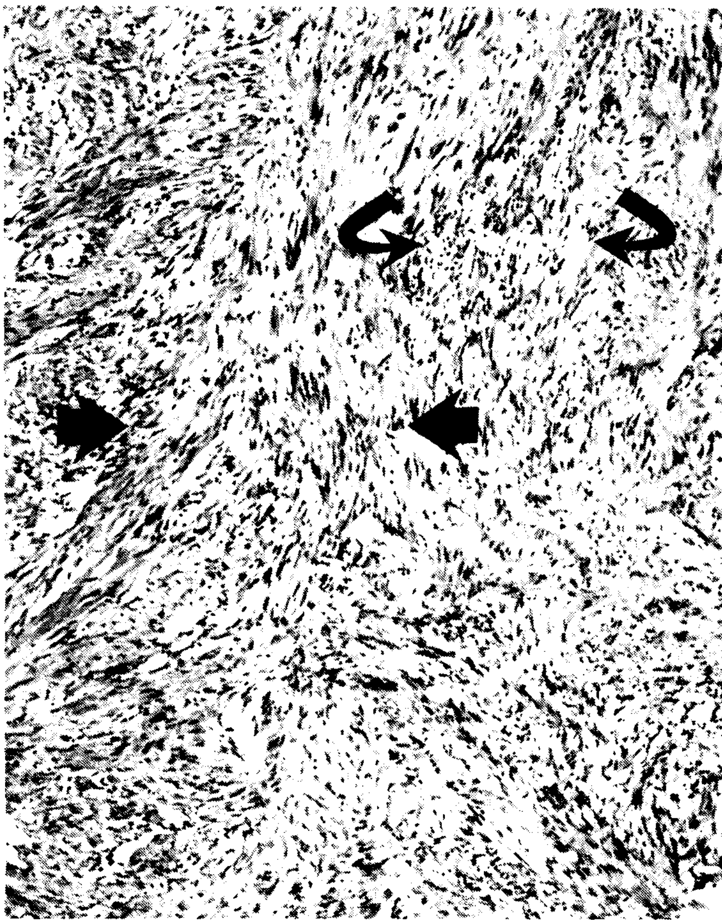

Fig. 5. Fascicles of spindle cells (straight arrows) alternate with edematous areas containing (curved arrows) lymphocytes. (hematoxylin-eosin, ×64)

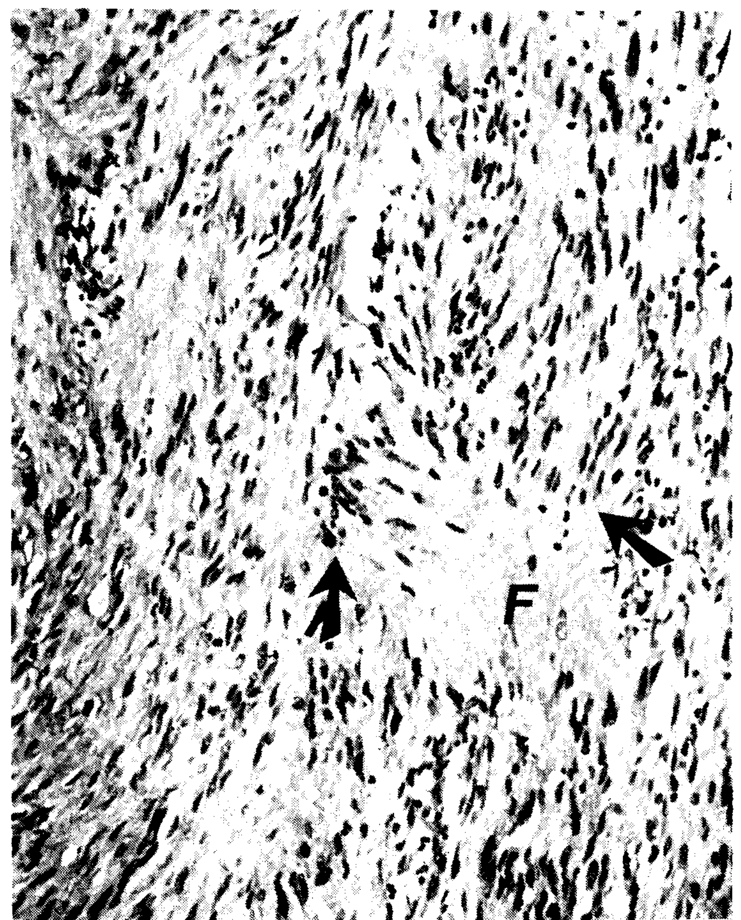

Fig. 6. Nuclear palisades (arrows) (hematoxylin-eosin, × 160) The nuclei are regimented near areas of fibrillar stroma without cells(F).

ques(Grillo, 1978; Grillo, 1982). The tumor was dumbbell shaped. The main portion lay in the tracheal lumen and a smaller portion in the extratracheal space(Fig. 3, Fig. 4). The patient had pulmonary edema immediately after operation with unknown cause, but recovered on the third postoperative day. This patient was discharged on the 15th postoperative day in good condition.

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 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 veroccay 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). Bartly 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.

Histologicaly 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 charcteristically, 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 1. Primary tracheal neurilemoma

Author	Date	Age(yr)	Race	Sex	Site	Symptom	Therapy	Follow-up
Straus	1951	28	C	M	L	Cough, fever, pneumonia	Two-stage endoscopy, silver nitrate cautery	Well, 6mo
Secretan	1953	21	C	M	L	Difficulty breathing cough, bronchitis	Endoscopic excision cautery	No Sx, 5yr
Ivanov	1953	35	C	F	M	Cough, difficulty breathing	Surgical resection	Well, 1mo
Kittinger	1961	23	C	F	L	Difficulty breathing stridor on exertion	Endoscopic excision repeated after 2wk	Well, 21/2yr
Takahashi	1963	12	Y	M	U	Difficulty breathing	Tumor excision through tracheotomy	Well, 11/2yr
Gerashchenkol	1964	6	C	F	L	Difficulty breathing cough, weakness	Surgical transection postop.	Pneumonia, died
Kim	1970	43	C	F	L	Difficulty breathing asthma, hemoptysis	Tracheal sleeve resection	Well, 21/2yr
Karlan	1973	21	C	M	U	Difficulty breathing cough, pneumonia	Tracheal fissure resection	Well, 1yr
Conley	1975	28	C	M	U	Cough, difficulty breathing	Surgical resection	Lost to follow-up
Conley	1975	—	—	—	—	—	Surgical resection	Lost to follow-up
Gouin	1977	71	C	F	M	Recent onset of asthma & cough	Endoscopy aborted, tracheal sleeve resection	Renal insuff., mediast-initis, died postop.
Nass	1979	36	C	M	L	Difficulty breathing hemoptysis, wheezing	Endoscopy with electrocoagulation	Well, 4yr
Makino	1979	65	Y	F	U	Recent onset of asthma	Tumor excision through tracheotomy	Well, 6mo
Ma	1981	23	C	F	U	Difficulty breathing wheezing	Tracheal sleeve resection	No Sx, 1yr
Horovitz	1982	38	C	F	L	Difficulty breathing cough, shortness of breathing	1. Endoscopic removal 2. Tracheal resection	1. Recurrence, 12y 2. No Sx
Katagiri	1982	65	Y	M	U	Difficulty breathing asthma, cough	Tracheal resection	Well, 11/2yr
Current case	1985	51	Y	F	M	Asthma, difficulty	Tracheal resection	Well, 1yr

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(Karlan *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 neurilemoma is resection with end to end suture.

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= 국문초록 =

原發性 氣管 神經纖維梢腫 治驗 一例

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김건열***, 한용철***

저자들은 호흡곤란을 주소로 응급실을 통해 입원하여, 흉부 전산단층촬영 등으로 기관종양을 확인한 51세된 여자환자를 기관절제 및 단단문합으로 시술, 완쾌시키고, 조직검사상 원발성 기관 신경섬유초종이 확인된 바 이는 현재까지 세계문헌상 백인에서 13예, 황인(일본인)에서 3예 만이 보고된 희귀한 예로 저자들의 지식으로는 국내 초유의 증예로 사료되어 문헌고찰과 더불어 보고하는 바이다.