A Case of Tracheal Neurilemmoma Treated as Bronchial Asthma

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Abstract

A fifteen-year-old female was referred to our hospital with a 3-year-history of dyspnea and dry cough. Stridor was noted on physical examination. Fiberoptic bronchoscopy revealed a mass in the upper third of the trachea. The mass had a smooth surface and occupied approximately 85% of the tracheal lumen. Tracheotomy was done and the mass was excised. The histopathology of the lesion was evaluated as neurilemmoma.

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Key words: *Neurilemmoma, trachea.*

Introduction

Tracheobronchial tumours of neurogenic origin are extremely rare (1). Only 23 intratracheal neurilemmoma cases have been described in the literature and half of these cases were reported from Japan (2,3). Neurilemmomas take origin from Schwann cells of the neural sheath (4).

In this report we describe a case of intratracheal neurilemmoma who had been treated and followed as an asthma patient from the onset of the symptoms.

Case Report

A 15-year-old female patient was referred to our hospital with a three-year-history of dyspnea, cough and night sweats. She had previously been diagnosed as an asthma patient and had received palliative medical treatment.

On physical examination, stridor and minimal suprasternal retractions were noted. The remainder of the general physical examination was unremarkable. The results of the routine laboratory tests, including complete blood cell count, blood chemistry and urinalysis were also within the reference limits. A

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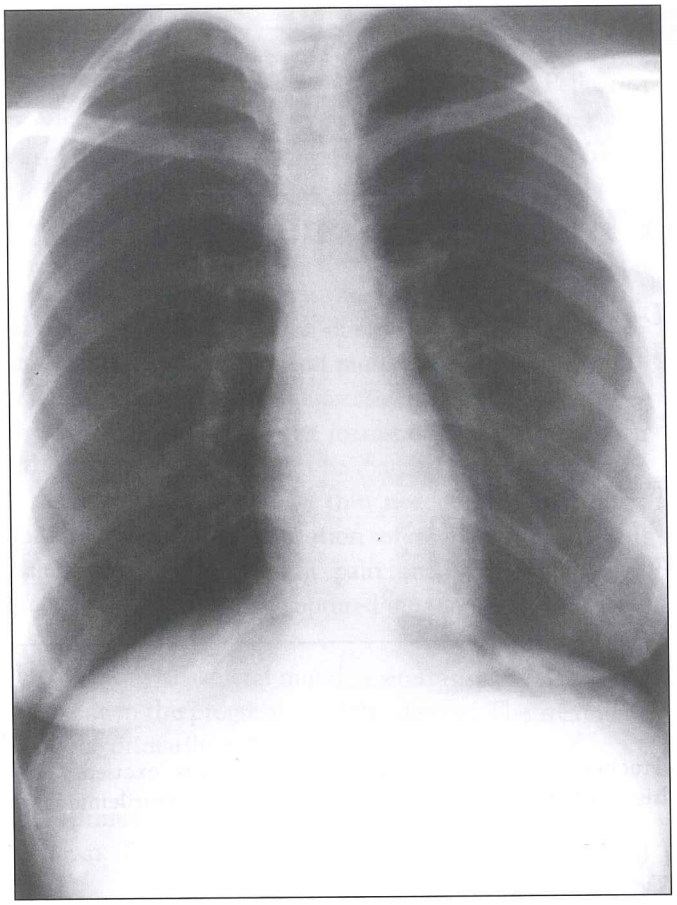

Figure 1. *Postero-anterior chest X-ray*

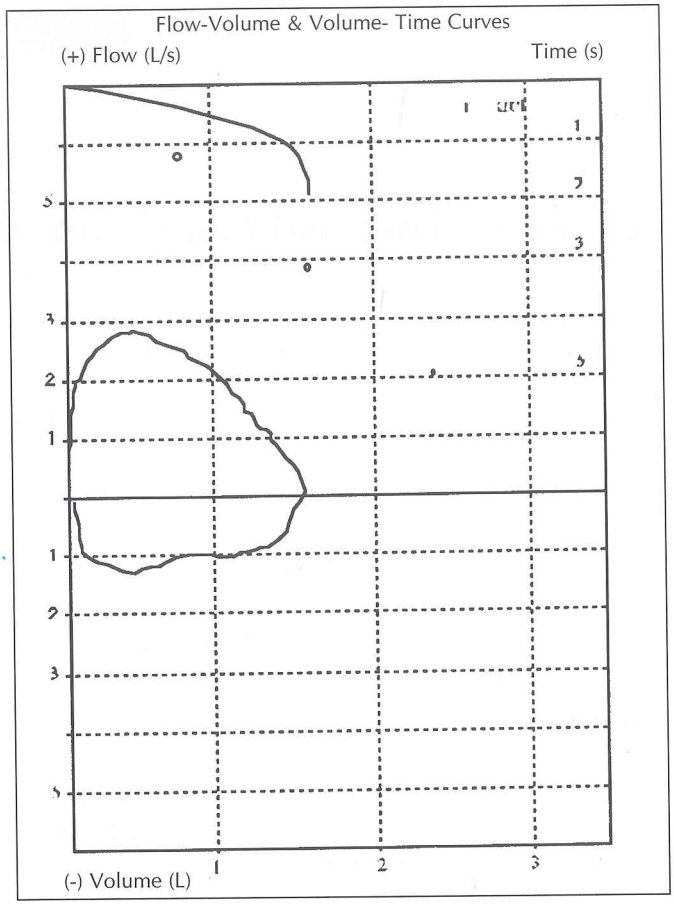

Figure 2. *The flow volume curve, showing remarkably reduced inspiratory flow rates*

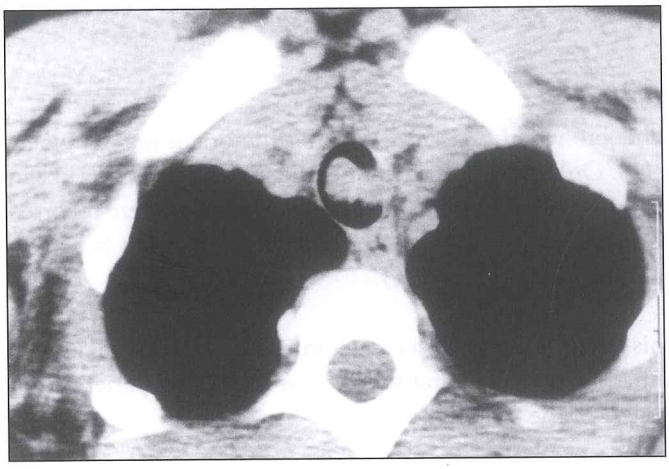

Figure 3. *Computerised chest tomography showing an intratracheal solid and heterogeneous mass lesion occupying almost all of lumen.*

postero-anterior (P-A) chest X-ray was evaluated as normal (Figure 1). In spirometric analyses at the flow volume curve, inspiratory flows were found to be markedly reduced (Figure 2).

Computerised chest tomography revealed an intratracheal solid, lobulated and heterogeneous mass lesion which was attached to the left anterior and lateral walls of the proximal trachea, occupying approximately 85% of the tracheal lumen (Figure 3).

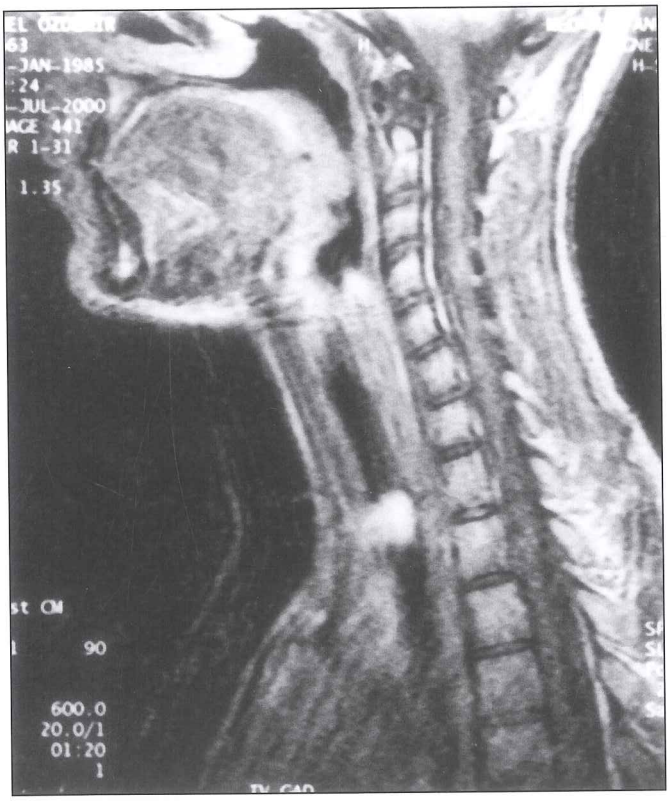

Figure 4. *MRI of the neck showing the intraluminal mass in the middle third of the trachea.*

Figure 5. *Tumor cells are strongly positive for S-100 protein (S-100 protein x100).*

To clarify the relationship between the tumoral lesion and the mediastinal structures, magnetic resonance imaging (MRI) of the neck was performed. MRI showed an intraluminal mass in the middle third of the trachea, with no invasion to any mediastinal structures (Figure 4).

Fiberoptic bronchoscopy showed a lobulated, smooth surfaced, mildly vascularized lesion, yellow in colour and located 1-2 cm distal to the vocal cords, on the left lateral wall of the trachea. The lesion had caused a narrowing of the lumen by about 85 %.

The patient was referred to the Thoracic Surgery Department of our hospital. To assess the nature and the location of the lesion, rigid bronchoscopy under general anesthesia was applied to the patient prior to the operation. The tumour was found to originate from the left anterior wall of the trachea, 2 cm down from the vocal cords. It measured almost 2 cm in diameter. After the patient was placed in the supine position, a straight endotracheal tube with an internal diameter of 5 mm was inserted via fiberoptic bronchoscopy (Olympus LF-DD). Coller incision was made and pretracheal structures were dissected. To have a better exposure, an upright traction to the thyroid tissue was made. The isthmus of the thyroid gland was divided and the trachea was freed from adjacent structures. After palpation of the most rigid part of the trachea, a dissection was performed from the distal part of the lesion. The tumour was seen on the left anterolateral wall of the trachea. To maintain ventilation during excision, a sterile spiral endotracheal tube was inserted from the dissected part of trachea The mass was excised by circumferencial resection of the involved trachea. Both the proximal and the distal sides were resected segmentally about 3 mm away from the tumor macroscopically, a distance equal to 4 tracheal rings. Then an end-to-end anastomosis of the resected trachea was made with 3-0 monocryls for the membranous part of trachea.4-0 vicryl sutures were used for the cartilaginous part. The operation was completed with closure of the anatomical structures.

The surgical material from the intratracheal mass was sent for pathological evaluation. On gross examination, the material was a smooth surfaced mass; 2x1.5x1 cm in size, with a nodulated and yellow appearance at cross section. Microscopically a tumoral structure was observed under the bronchial epithelium. It was surrounded by a thin fibrous capsule, separating it from the bronchial cartilage and glandular structures. This tumoral structure was mainly composed of the 'verrocay bodies' that is characterised by the fusiform cells with ovoid nuclei forming palisades around a fibrillary material. No mitotic figures could be seen in any area of the tumoral structure (Figure 5). Squamous metaplasia was observed in a part of the bronchial epithelium of the tumour. Lymphocyte and plasma cell infiltration were detected in an area under the epithelium. Immuno-histochemically, the tumour cells showed high immunoreactivity with S-100 protein and vimentin, while no immunoreactivity was detected with desmin and actin.

The postoperative course was uneventful and no granulation tissue, strictures or recurrence were encountered at the end of two years of follow-up.

Discussion

Benign tracheobronchial tumours are extremely rare and they are usually operable and curable (4). Most of the benign tracheobrochial tumours occur in infants and children. The more common benign tumours encountered in adults are adenoma, fibroma, papilloma, chondroma, lipoma and hemangioma (5). Benign tumours are most frequently seen in the upper third of the trachea in children and in the lower third in adults. They usually originate from the membranous part of the trachea. Most of the reported intrapulmonary neurilemmoma cases were shown to originate from the region of terminal segmental bronchi. Some present as an intrabronchial polypoid mass (2). In our patient the lesion was located in the trachea, 1-2 cm below the vocal cords.

Although there is no sex predilection for endobronchial tumours, bronchial or intrapulmonary neurilemmomas have a slight female predominance. The age range is wide, the youngest patient in the literature being 5 years old, and the oldest is 78 years (2,4). The clinical features of the neurilemmomas differ depending on their location. In most of the reported cases, the first symptom was a dry, constant, irritating cough that was not relieved by sedatives or expectorants. Stridor was another important symptom. Since intratracheal tumours are generally not included in the differential diagnosis of wheezing, there is frequently a

long delay from the time the patient seeks medical attention until the diagnosis is established. In reviewing the literature, the average delay was found to be 15 months (6). In two endotracheal neurilemmoma cases reported by Pang (4), the delay in diagnosis was 8 years in one case and 2 years in another case. This delay was 3 years in our patient.

On chest X-rays, the tumour appears as a round or ovoid, lobulated homogenous intraluminal mass (2,4). Computerised thorax tomography gives the exact localisation of the polypoid mass lesion and the attachment of its stalk in the tracheal lumen. Schraegen et al., reported that magnetic resonance imaging is superior to computerised tomography in determinig the exact location and extension of the tumour in the trachea (7). Viewing the tracheobronchial tree through the bronchoscope directly is the most reliable method in diagnosis of endotracheal lesions (4).

In our patient, the postero-anterior chest X-ray was evaluated as normal. Both CT and MRI revealed an endotracheal lesion originating from the left wall of the trachea with no invasion of other mediastinal structures.

As to the treatment, the first priority is usually the establishment of the airway. This frequently is an emergency procedure consisting of the forceps removal of the obstructive lesion with bronchoscopy (4). Since recurrence due to malignant nature can rarely occur, the most dependable procedure in the treatment is segmental tracheal resection (5). As was also the case in our patient, postoperative prognosis is usually excellent (4,5).

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