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Primary Tracheal Schwannoma

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ABSTRACT

Primary neurogenic tumors of trachea are extremely uncommon and account for only about 9% of all neoplasms of trachea. Tracheal schwannoma is among the rarest of them and there is no unanimity of opinion regarding its treatment.

We report a 30 year-old woman with symptoms of airway obstruction due to primary tracheal schwannoma. She was suffering from cough and exertional dyspnea. She was admitted to the thoracic surgery ward with stridor and hemoptysis and underwent rigid bronchoscopy and biopsy. The patient then underwent primary tracheal resection and anastomosis through a cervicomediastinal approach. The diagnosis was made through histopathological examination as schwannoma.

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Key words: Schwannoma, Trachea, Surgical treatment

INTRODUCTION

Primary tracheal tumors account for about 1% of all neoplasms. (1) Two-thirds of these neoplasms are squamous cell carcinoma and adenoid cystic carcinoma. These tumors usually affect the adult population. The remaining third is a widely heterogeneous group of benign lesions that occur more frequently in pediatric group except for neurogenic tumors. Squamous papiloma, leiomyoma, and hemangiomas are most frequently seen. Benign neurogenic tumors are extremely rare and present at any age. This type of tumor arises from nerves located inside the tracheal wall and is composed of Schwann cells of the nerve sheath (2).

CASE SUMMARIES

A 30-year-old woman presented with dry cough since 7 years ago, that had intensified during the last 2 months prior to admission. Hemoptysis was added to symptoms one week before the admission. She also complained of exertional dyspnea. There were no exacerbating or attenuating factors. In her past medical history she had no pulmonary or other type of disease. She was non-smoker. On physical examination, the blood pressure was normal; she had normal general appearance and no symptom of respiratory distress. She only had stridor on physical examination. Her skin was normal. She had no findings correlating with von-Recklinghausen syndrome. Laboratory tests and chest x-ray were normal. Pulmonary function test revealed restrictive pattern related to tracheal outlet obstruction. CT-scan was performed and a mass with intra and extra tracheal extension was seen; there was no evidence of lung collapse (Figure 1). The patient was a

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candidate for rigid bronchoscopy because of hemoptysis and stridor. A bulging was seen 4cm under the vocal cords which was pale and tense; the length of tumor was 2cm and a biopsy was taken (Figure 2).



Figure 1. Intra and extraluminal extension of the tumor

Histopathological study revealed a benign tumor without definitive diagnosis.

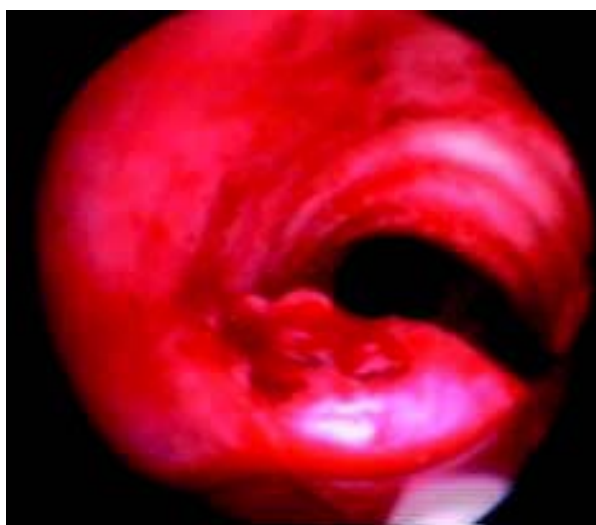


Figure 2. Bronchoscopic view of the tumor

She underwent surgery for tumor resection. A cervicomedial incision was made. The tumor was located 4 cm below the vocal cords. It was tense, capsulated and had no adhesion to the adjacent

tissue. The tumor had extra-luminal extension and had involved 4 tracheal rings. The tumor was to proximity of innominate artery and esophagus (Figure 3).

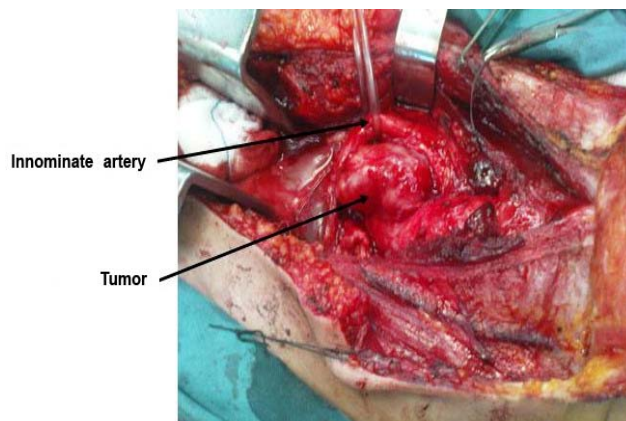


Figure 3. Tumor appearance with extra luminal extension adjacent to innominate artery

The tumor was resected with small safe margin and end to end anastomosis was performed with absorbable vicryl sutures.

On gross pathology, the tumor was encapsulated, dumbbell shape with yellow-tan color, and homogenous appearance with firm consistency growing outward from the ring space (Figure 4).



Figure 4. Macroscopic view of the tumor

On microscopic examination, two different patterns were identified. Most areas were cellular

(Antoni type A) and revealed proliferation of spindle cells with wavy nuclei arranged in palisade fashion or in organoid arrangement (Verocay bodies), with extremely scanty mitotic figures (Figures 5 and 6). Some areas were hypocellular with abundant edematous stroma (Antoni type B).

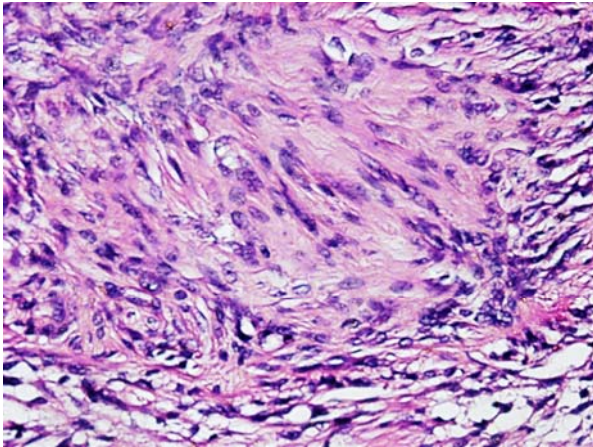


Figure 5. Spindle cells with wavy nuclei arranged in a palisading fashion.

On immunohistochemical IHC study, the tumor cells showed nuclear immunoreactivity for S-100 protein (Figure 7). IHC study was negative for desmin, smooth muscle actin, cytokeratin, and EMA.

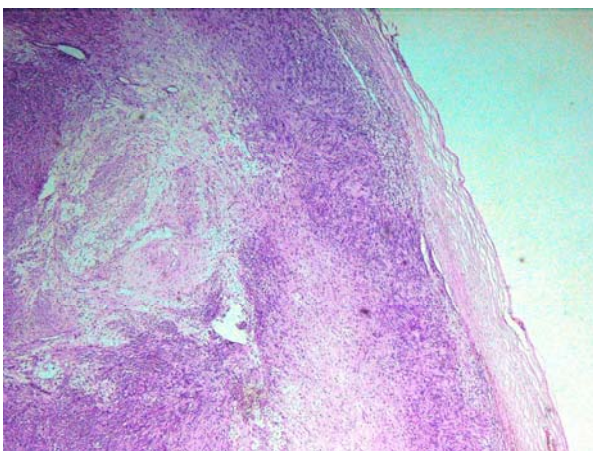


Figure 6. Encapsulated neoplasm with palisade fashion in Antoni type A area ($\times 40$).

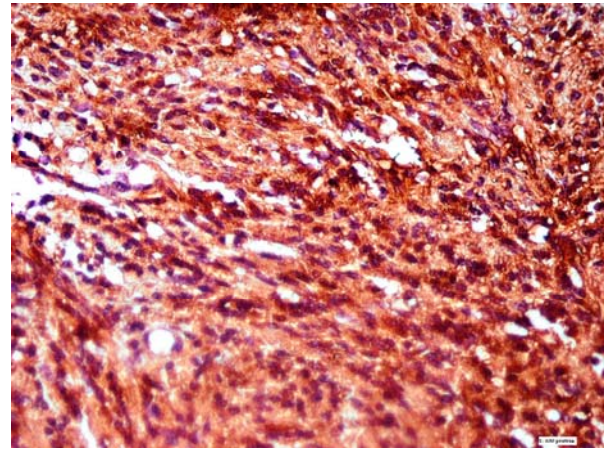


Figure 7. S-100 immunoreactivity in tumoral cells

DISCUSSION

Benign neoplasms of trachea could be classified according to their cell origins including: neural (carcinoid, benign clear cell, neurofibroma, myoblastoma), epithelial (papiloma, chondroma, lipoma, fibroma, fibrous histiocytoma), and epithelial and mesenchymal (hamartoma). Primary neurogenic tumors in the upper respiratory tract are extremely uncommon, being more frequently reported in the lungs and bronchi than trachea. Neurogenic tumors are divided into two groups, schwannoma and neurofibroma (1). To date, neurofibroma has been reported only in men and schwannoma has shown a predilection for women. Most of the cases have occurred in adults. Schwannoma occurs most frequently in the distal of trachea, followed by decreasing order in the proximal and middle thirds. The expanding lesion is compressed by tracheal cartilaginous rings so that the tumor is forced to extrude into tracheal lumen or out into extrinsic soft tissue (3). Schwannoma of the trachea arises from intra-luminal neurogenous tissue (not vagus nerve), to be more precise, Schwann cells of the nerve sheath. (1)

Schwannomas are typically single, circumscribed, encapsulated tumors attached to a nerve but containing no axon (2). Schwannomas are rarely

associated with von-Recklinghausen disease, and rarely undergo malignant changes. Neurofibromas are usually multiple and frequently associated with von-Recklinghausen disease. Neurofibroma shows proliferation of all elements of the nerve including the Schwann cells, perineural cells and nerve axons. They may go under malignant changes. (4)

The diagnosis is usually delayed. The delay in diagnose is due to common and non-specific symptoms. The tumor must generally become large enough to obstruct more than half the diameter of the airway before dyspnea is experienced. Tracheal neoplasm would be treated for months or even years with bronchodilators and steroids until hemoptysis or obstructive pneumonia leads to a diagnostic bronchoscopy (5). Routine CXR is not an efficient radiologic procedure because of the superimposition of soft tissues and bony structures. Sometimes it could demonstrate the tracheal mass and outline its intraluminal extension and secondary complications such as collapse due to pneumonia. Conventional computed tomography (CT) is more sensitive. The CT features of tracheal schwannoma are a well demarcated, low density mass before contrast injection and there is homogenous and excessive contrast enhancement after injection. In magnetic resonance image (MRI), tracheal schwannoma is a well demarcated isotense mass on T1- weighted images and is homogeneously hyperintense on T2- weighted images. There is an excessive homogeneous contrast enhancement on the T1- weighted images after injection of contrast material (gadolinium). MRI images have the advantage of sectioning the trachea into both coronal and sagittal planes, therefore, the location and extension of the tumor can be more precisely defined (3). The best definitive diagnostic procedure is represented by tracheal bronchoscopy. It can be carried out with a flexible or rigid tube and should be done under local or general anesthesia with the patient breathing

spontaneously according to the procedure. If the local appearance is typical of a benign lesion and especially if the tumor appears highly vascular, biopsy should be avoided or deterred until the time of definitive surgical resection (1). Histopathologically, there are regions of high and low cellularity called Antoni type A and B areas, respectively. The tumor cells stained yellow with von Gieson special stain and immunohistochemical stains revealed strong positivity for S100 proteins, especially in Antoni type A areas (7). Stains for muscle specific actin, desmin and cytokeratin were negative and no mitotic activity or necrosis was seen (8).

It is difficult to determine what the optimal treatment is, because schwannomas are very rare and patient's follow up period has often been short (2,6,9). The choice of treatment should probably be influenced by clinical presentation of the tumor which is pedunculated or sessile, the risk of tracheal resection and the presence or absence of extratracheal component. Laser resection with CO₂ or Nd- YAG laser seems to be effective for pedunculated tumors without the extratracheal part. The tumor of extratracheal component should be definitively managed by surgery as tracheal resection and primary anastomosis. Bronchoscopic surveillance should not be ignored (10).

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