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# Surgical treatment of primary Tracheal Carcinoma in a Patient with Recklinghausen's disease (Neurofibromatosis type 1)

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## Abstract

This is the case of a 56-year-old man with significant history of neurofibromatosis and smoking who pre-sented with respiratory distress and workup revealed intra-tracheal mass. Even though the expectation was the tracheal tumor to be benign the pathology revealed malignancy. Notably, pulmonary nodules were at first thought to be benign tumors, yet workup revealed these were metastatic. Presence of two rare conditions can coexist and it is important for the clinicians to keep an open mind while managing patients with newly found mass who have history of benign tumors. This case also represents good learning opportunity for surgical management of intratracheal tumor when intubation is a challenge

**KEYWORDS: tracheal Carcinoma; Recklinghausen's disease; neurofibromatosis type; tracheostomy**

# Introduction

Primary Tracheal Carcinoma is a very rare tumor, accounting about 0.2% of malignant neoplasms of the respiratory tract. The most common primary tracheal neoplasms are squamous cell carcinomas which is related to smoking. Clinically, tracheal tumors present with cough, dyspnea, hemoptysis, stridor or symptoms of invasion to contiguous structures (such as laryngeal nerve, esophagus or thyroid gland). Tracheal stenosis is most common radiologic and endoscopic finding. Stage of presentation is advanced with approximately 50% of cases. Five-year survival for all tracheal neoplasms is 40% but falls 15% for those with stage IV disease. Neurofibromatosis is a rear genetic disorder as well, that cause tumors to form on nerve tissue. These tumors can develop anywhere in the nervous system, including the brain, spinal cord and nerves. There are three types of neurofibromatosis: neurofibromatosis 1 (Recklinghausen's disease, NF1). Neurofibromatosis 2 (NF2) and schwannomatosis. The tumors in these disorders are usually benign. NF1 is usually diagnosed during childhood. Signs are often noticeable at birth or shortly afterward and almost always by age 10. Signs and symptoms are often moderate, but can vary in severity. It depend on site and size of tumor. We reviewed literature and were not able to find a case of tracheal carcinoma in a patient with Recklinghausen's disease. It is the reason of presentation of this case report.

## Case description

A 56 year old smoker male was admitted to our clinic with complaints of cough, dyspnea and hemoptysis for about a year. He was not able to lay on his back, due to choking sensation. He had history of bronchial asthma for which he was on treatment. He also had history of neurofibromatosis (van Recklinghausen's disease) with multiple neurofibromas with maximum diameter of 5 cm on the chest wall, neck and upper limbs. Also, volumetric formations (tumors) of C1 and C2 intervertebral foramina with propagation in the spinal canal and cord compression at the same level. He had limited movement in the right upper extremity.

In the past he had undergone total surgical resection of big size neurofibromas of anterior surface of neck (1978 year), left upper limb, left axillary fossa (1991 year) and in the area of the right sternoclavicular joints (2013 year) with local anesthesia. Physical examination revealed multiple neurofibromas of different size (from 3 mm to 5 cm) on upper limbs, chest wall, and neck. No enlarged peripheral lymph nodes were detected. At anterior surface of neck, left elbow, right axillary fossa postoperative scars were seen. Thoracic CT

revealed sharp narrowing of the tracheal lumen 4 cm distal from the vocal cords. There was a solid oval-shaped tissue density nodular lesion of 20 X 22 mm with irregular edges. It was causing narrowing of tracheal lumen in this area up to 6 mm. The tumor expended to the pretracheal location, close to both carotid arteries. Fibrous changes were seen in the apices of lungs bilaterally. In the 8-th and 10-th segment of the right lung, the round formations with size 4-6 mm. were seen in the 10-th segment of the left lung, a round–shaped formation with a size 10 mm was detected. In summary, primary assessment consistent of tracheal tumor with focal lesions in both lungs. Fibertracheobronchoscopy revealed paralyzed right wall of the larynx and right vocal cord, the larynx shifted frontally, free subglottic space, and a large exophytic formation in the upper part of trachea, which occupied a large part of the lumen, originated from the anterior wall with the proximal border at the level of the 2-nd cartilaginous ring (Figure 1). Lumen was preserved along the posterior wall which limited further evaluation of the deeper structures. The surface of the tumor was irregular, hyperemic and bled easily. Biopsy was not done due to risk of serious bleeding and asphyxiation. Endoscopic findings: A large exophytic tumor of the upper part of trachea (neck part), paralysis of the right part of the larynx.

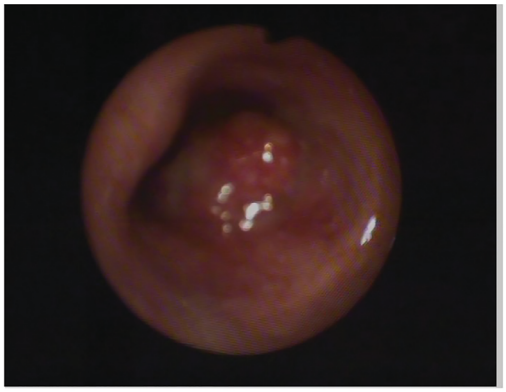

*Fig. 1*

Magnetic Resonance imaging of the cervical spine revealed volumetric formations of C1 and C2 intervertebral foramina with spread in the spinal canal and cord compression at the same level. Swelling of the retropharyngeal soft tissues and C6 – C7 locking plates (Fig. 2).

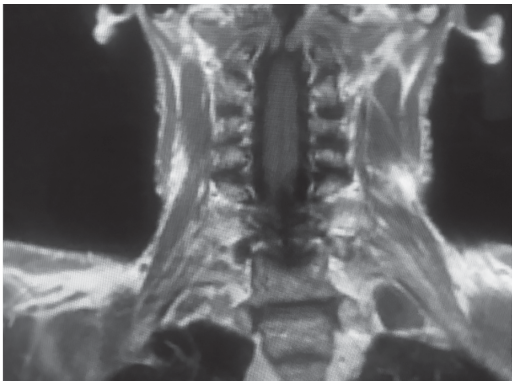

*Fig. 2*

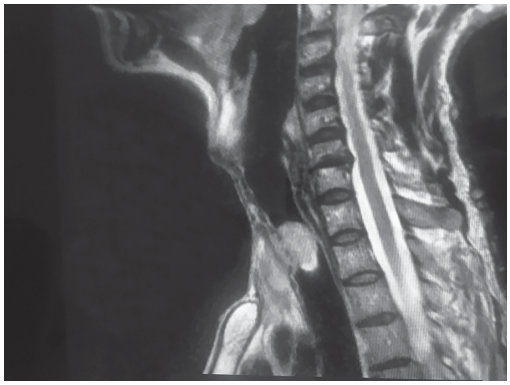

*Fig. 3*

MRI also revealed tracheal tumor which originated from anterior and lateral walls of the trachea narrowing its lumen (Fig. 3). Anterior contrasted X-ray examination of the esophagus and stomach and ultrasound of the abdominal organs did not reveal any pathology.

Given prior history of neurofibromatosis, and absence of malignant tumor after previous operations (excision of tumors of different localizations) expectation was that the tumor in the trachea would also be benign. We took into account the expected tracheal obstruction and the risk of suffocation of the patient and decided to perform the operation: A circular resection of the 2-nd, 3-rd and 4-th cartilaginous rings of the trachea along with the tumor in healthy margins and primary anastomosis.

The operation was started under local anesthesia with tracheostomy. The patient was not able to lay on his back because of respiratory compromise. So we started operation in semi-sitting position. A lower tracheotomy was performed at the level of the 4-th cartilaginous ring. During the insertion of the intubation tube into the trachea from the tracheotomy, the patient began to asphyxiate. Fiberbronchoscopy from tracheotomy incision was performed immediately and 2 pieces of tumor were removed from the bifurcation area of the trachea. Then, the trachea was intubated from the tracheotomy incision. Tumor's spread was checked by oral fiberbroncoscopy. The tumor extend from the upper edge of the 2-nd cartilaginous ring to the lower edge of the 4-th cartilaginous ring, affecting the anterior and lateral walls of the trachea. The posterior membranous wall was intact. Taking into account spread of the tumor, the 2-nd, 3-rd and 4-th cartilaginous rings were cut in a con-like manner along with the tumor. We preserved the membranous part in order to keep the tracheal blood supply and improve healing.

Intraoperative revision gave the impression that the tumor was spreading extratracheally in the right lobe of the thyroid gland, so resection of right lobe of the thyroid was done as well. The integrity of the lateral walls of the trachea was restored with 3/0 absorbable suture (Vicril, Ethicon). The intubation tube was removed from the tracheotomy incision and pulmonary ventilation was continued with oral intubation. The integrity of the anterior tracheal wall was restored with knotted absorbable suture. The integrity of the midsection of the neck was restored in layers with knotted sutures. The chin was temporarily (for 7 days) attached to the sternum. The specimen was sent to the histomorphological laboratory: two pieces of the tracheal tumor (which broke at the beginning of the operation and fell in to the trachea), two pieces of extratracheal tumor, the lateral and anterior walls of the trachea in form three cartilaginous rings together with tumor and piece of resected thyroid gland.

The postoperative period was uneventful. The sutures of the trachea were healed, neck wound healed with primary intention.

Morphological report of the specimen was consistent with low-differentiated squamous cell carcinoma of the trachea with free margins. Fibrous changes were found

in the resected part of the thyroid tissue with no malignancy. In extratracheal tissues fibrous changes without tumor (scar tissue after previous operation) was seen. The patient was discharged from the clinic on the 12-th day.

In control fibrobronchoscopy performed at postoperative second month, the right half of larynx and right vocal cord remained paralyzed. The anastomosis was solid, the tracheal lumen was open. There was minimal fibrosis of the anastomosis line. Distally, tracheal walls were unchanged with visible bifurcation.

After the operation, the patient underwent 3 courses of chemotherapy. During this time, patient's condition was satisfactory. There was no cough, difficulty breathing or shortness of breath. He had only neurological symptoms associated with neurofibromas of the cervical spine and intervertebral foramen extending into the spinal canal which were unchanged from prior. 1 year after the operation, the patient underwent a control CT scan examination. He was not complaining of cough and shortness of breath. He only mentioned neurological symptoms. According to chest CT scan, the tracheal lumen is free, although it is slightly deformed in the upper third. Fibrous changes are seen in the apices of both lungs. The lymphatic nodes in the mediastinum are not enlarged. Small (5 to 12 mm) round nodules can be seen in both lungs (metastasis). There is no effusion in the pleural cavity and pericardium. CT result is unchanged from post tracheal resection changes and focal (metastatic) lung damage.

The last follow-up examination he had 3 years after the operation. He complained of cough, loss of appetite, weight loos, and limitation of movement of limbs. He used wheelchair due to generalized weakness and difficulty with ambulation. Patient refused further examination and chemotherapy treatment. We placed him in the palliative care unit.

## Discussion (review)

This is the case of a 56-year-old man with significant history of neurofibromatosis and smoking who presented with respiratory distress and workup revealed intra-tracheal mass. Even though the expectation was the tracheal tumor to be benign the pathology revealed malignancy. Notably, pulmonary nodules were at first thought to be benign tumors, yet workup revealed these were metastatic. Presence of two rare conditions can coexist and it is important for the clinicians to keep an open mind while managing patients with newly found mass who have history of benign tumors. This case also represents good learning opportunity for surgical management of intratracheal tumor

when intubation is a challenge. Starting the operation with tracheostomy under local anesthesia was the likely cause the part of the tumor to break off and drop down into the trachea which caused asphyxiation and changed the course of the surgery. This could have been prevented if the surgery was begun under general anesthesia with a small-caliber rigid bronchoscopy ventilation. This can be achieved with carefully passing the tube behind the tumor adjacent to the undamaged membranous wall of the trachea under the visual control. Membranous wall maintenance proven to be effective. The trachea was healed without narrowing the lumen.

## Conclusion

In conclusion we want to present our summary. Starting the operation with tracheostomy under local anesthesia was the likely cause the part of the tumor to break off and drop down into the trachea which caused asphyxiation and changed the course of the surgery.

This could have been pre-vented if the surgery was begun under general anesthesia with a small-caliber rigid bronchoscopy ventilation. This can be achieved with carefully passing the tube behind the tumor adjacent to the undamaged membranous wall of the trachea under the visual control. Membranous wall maintenance proven to be effective. The trachea was healed without narrowing the lumen.

## References

1. Bhattacharyya N. Contemporary staging and prognosis for primary tracheal malignancies: a population-based analysis. Otolaryngol Head Neck Surg. 2004;131:639-642.
2. Gassert HA, Grillo HC, Shadmehr MB, et al. Uncommon primary tracheal tumors. Ann Thorac Surg. 2006;82(1):268-272.
3. Regnard JF, Fourquier P, Levasseur P. Results and prognostic factors in resections of primary tracheal tumors: a multicenter retrospective study. The French Society of Cardiovascular Surgery. J Thorac Cardiovasc Surg. 1996;111:808-813. discussion 813.
4. Willmann JK, Weishaupt D, Kestenholz PB, von Smekal A,

Marincek B. Endotracheal neurofibroma in neurofibromatosis type 1: an unusual manifestation. Eur Radiol. 2002;12:192-194. PMID: 11868097. DOI: 10.1007/s 003300100892.

5. Ghalayani P, Saberi Z, Sardari F. Neurofibromatosis type I (von Recklinghausen's disease): A family case report and literature review. Dent Res J (Isfahan). 2012;9(4):483-8. PMCID: PMC3491339. PMID: 23162593.