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Primary tracheal microcystic reticular schwannoma – Case report of a rare neurogenic tumor treated by segmental tracheal resection

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Diagnosis and treatment of rare diseases are challenging because experience and evidence are limited. Primary tracheal tumors have a low prevalence but awareness of these is important to avoid misdiagnoses. We present a first case of a tracheal microcystic reticular schwannoma in which diagnosis and treatment recommendations were revised several times resulting in months of anxious uncertainty for the patient before complete resection and extensive histopathologic examination were performed.

CASE PRESENTATION: A 65-year old woman complained about coughing and cervical pain. Tomographies revealed a tumor of the dorsolateral trachea. After repeated biopsies pathologists and tumorboards in different hospitals could not agree on diagnosis or treatment recommendation, so the impatient patient opted for a complete resection for definite treatment.

CLINICAL DISCUSSION: Neoplasms of the posterior mediastinum are mainly neurogenic. In rare cases they may originate in the trachea and can be difficult to differentiate from other tumor entities if only biopsies are available. Although the preoperative diagnosis was unclear, tracheal resection was performed successfully. The differential diagnoses of soft tissue sarcoma or myoepithelioma were discarded and a benign, microcystic, reticular schwannoma was confirmed by international reference pathologists.

CONCLUSION: Microcystic, reticular schwannoma is a rare variant and may occur in the trachea. Diagnosis of such rare neoplasms can be difficult, resulting in delayed or suboptimal treatment. Often biopsies are not sufficient to reach a definite diagnosis. But even without preoperative histology, tracheal resection is a safe and feasible option for definitive treatment with very low recurrence rates.

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1. Introduction

Pathology may be easy at times, but in some cases, it can be difficult to reach a diagnosis, especially when rare tumors are concerned. Due to low incidences, experience with these entities is limited or diagnostic criteria may not be well defined. In addition, these neoplasms can show overlapping features with other diseases.

This case report, prepared in line with the updated SCARE-criteria [1], presents a first case of a rare variant of a neurogenic, primary tracheal tumor, where the patient experienced a long course of different diagnostic procedures and was unsettled by contradicting treatment recommendations.

2. Patient information

A woman of 65 years, non-smoker, with hypertension and a uterine carcinoma treated by hysterectomy 20 years before, had consulted her orthopedist because of cervical pain, slowly progressing over a period of 7 months. Her family history included adenocarcinoma of mamma and colon. Antihypertensive medication and non-steroidal anti-inflammatory drugs had been prescribed. She had been working in a company office for 35 years and retired one year before.

3. Clinical findings

In the weeks before the consultation she complained about coughing, hoarseness under physical strain and intermittent dysphagia. At this point neurological symptoms were absent, but shortly later she developed paresthesia and weakness of her right arm. There was neither weight loss, nor fatigue or other limitation.

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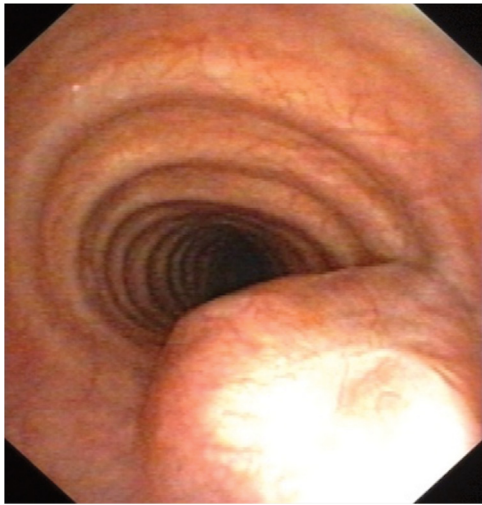


Fig. 1. Bronchoscopic view on the tumor of the dorsolateral tracheal wall.

4. Diagnostic assessment

Besides some cervical spondylosis and disc protrusions magnetic resonance imaging (MRI) showed no affections of vertebrae or spine, but a smooth, paravertebral, tracheal mass with a size of $30 \times 12 \times 10$ mm and a high T2-, but low T1-intensity. Additional imaging of the cerebrum did not reveal pathological findings.

Computed tomography (CT) described a tracheal neoplasm in front of C7/Th1 with smooth extratracheal margins. There was an enlarged lymph node on the contralateral cervical side (max. 17 mm) next to the parotid gland. Pulmonary parenchyma and vascular structures were inconspicuous, there was no mediastinal lymph node enlargement.

F^{18} -flourdesoxyglucose-positrone-emission tomography in combination with low dose CT (F^{18} -FDG-PET/CT) showed a singular tracheal mass with increased metabolic activity (maximal standardized uptake value 6,6).

Esophagoscopy showed no esophageal infiltration or extraluminal compression.

Bronchoscopy revealed a tracheal mass beginning 5 cm below the vocal folds, with intramucosal growth over 3 cm in length, obturating the lumen by 50%. Kryobiopsies were repeatedly taken (Figs. 1 and 2).

Spirometry showed no impairment of lung function.

5. Therapeutic intervention and histopathology

After months of diagnostic interventions in different institutes, with varying histological diagnoses from “myxoid extraskeletal chondrosarcoma” to “benign myoepithelioma” and treatment recommendations ranging from “watch and wait” to definite chemoradiation, the unsettled patient presented herself for a reevaluation. Follow up-tomographies had shown no tumor growth, symptoms and complaints were significant but had not progressed. Flexible Bronchoscopy confirmed a shallow tumor of the dorsal tracheal wall with a size of maximal 3 cm in the middle tracheal third, so tracheal segmental resection with end-to-end-anastomosis was feasible. The resection was performed by experienced specialists of our department of thoracic surgery without complications 12 months after the first symptoms. To prevent sudden traction on the tracheal anastomosis, the neck was stabilized by a neck collar in addition to a fixation by “chin stitch” for one week. There were no adverse events during the operation or the clinical course. The patient left hospital two weeks after the operation.

The resected material was sent to two different, specialized, international histopathological institutes, who described a primary tracheal tumor with slightly eosinophilic, spindle-shaped or ovoid cells arranged around either myxoid matrix or microcystic spaces. Absence of atypia or pleomorphism was confirmed. A diffuse positivity for S100 and focal positivity for CD34 was seen. Staining for epithelial membrane antigen (EMA) showed an incomplete perineural capsule. The reference pathologists agreed on the diagnosis of a benign, microcystic/reticular schwannoma (MRS) (Figs. 3 and 4).

6. Follow up and outcome

The patient recovered well, the follow up over 8 years showed no recurrence or tracheal stenosis. She showed no dyspnea under moderate physical exercise, but a slight dysphonia, caused by unilateral, partial laryngeal paralysis.

7. Discussion

Benign Schwannoma of the head, neck, peripheral nerves or the gastrointestinal tract are well known. Sporadically arising from intercostal or sympathetic nerves they also account for 95% of posterior mediastinal neoplasms [2]. In contrast, the incidence of primary tracheal malignancies is low, but epidemiological data can be derived from national registries. The Netherlands Cancer Registry showed an annual incidence of only 0,142/100.000 [3], the NCI Surveillance, Epidemiology and End Results Register (SEER) reported 0,26/100.000 [4]. In adults 90% of primary tracheal tumors are malignant, therefore the incidence of benign tracheal neoplasms is much lower and no epidemiological data are available for these lesions. The WHO-classification of tumors lists tracheal tumors together with head-/neck tumors rather than tumors of the bronchi, reflecting the position of the trachea as an “interface” where neoplasms of different regions may overlap, making a correct diagnosis quite challenging [5]. This applies even more, when only limited material is available for histopathologic examination, as it was in the presented case.

A few cases of primary tracheal schwannoma have been reported and these publications describe classical schwannoma showing specific features like alternating Antoni A and B regions, hyalinized vessels, Verocay bodies, a capsule and positivity for S-100 and EMA. Only in one case a melanotic psammotous variant was identified. The incidence of these primary schwannoma is higher in the lungs or the peripheral bronchopulmonary tree, but decreases with central location. Clinical symptoms are usually caused by obstruction of the upper airways. They include dyspnoea, coughing or hemoptysis. Even with these classical schwannoma a substantial delay of 10–17 months was seen until definite diagnosis [6].

A variety of schwannoma subforms with distinct histologies and different predilections have been defined in which the typical features of classical schwannoma may not or only partly be present and which may show overlapping features with non-neurogenic neoplasms like sarcoma, gastrointestinal stroma or salivary gland tumors [7]. In 2008 Liegl et al. described a new schwannoma variant with a predilection for the gastrointestinal tract but sporadic occurrences in other organs. This subform lacks the typical features of classical schwannoma but shows a predominant or exclusive reticular pattern of intersecting spindle cell strands with eosinophilic plasma creating microcystic spaces of myxoid material or fibrillary collagen. Atypia is absent, but the mostly unencapsulated boundary may show a pushing growth pattern invading between neighbouring cells in a pseudoinfiltrating way. A strong immunohistochemical S-100-positivity, negative cytokeratins, a



Fig. 2. F18-FDG-PET/CT with a singular tracheal tumor with enhanced glucose metabolism (SUV max 6,6) and MRI showing a paravertebral mass in T2-pictures with homogenous hyperintensity and no capsular enhancement, T1 showed a low intensity (yellow allows).

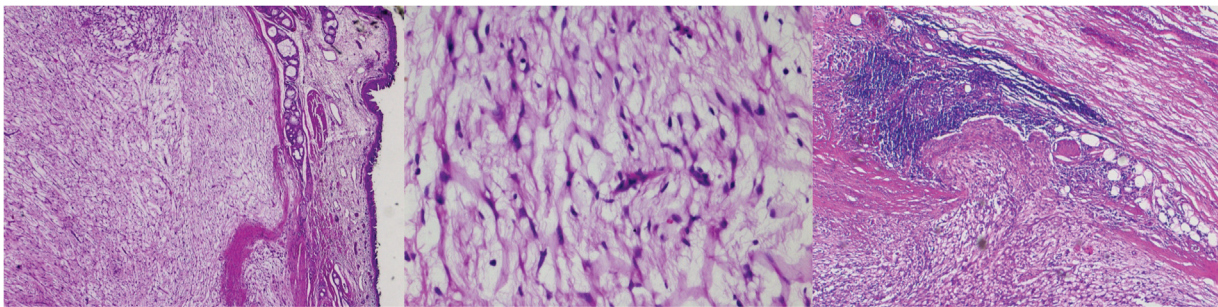


Fig. 3. Histopathology showing normal tracheal mucosa and underneath it a mass of loosely scattered blastoid cells (left) with eosinophilic plasma, spindle-shaped or ovoid nuclei, interspaced myxoid material in microcystic spaces (middle) and occasional bud-like protrusions and infiltrations into the mucosa (right).



Fig. 4. Tracheal anastomosis after segmental resection intraoperative (left), after 1 week (middle) and after 2 weeks (right).

variable positivity for glial fibrillary acidic protein (GFAP) and minor areas with classical features or remnants of an EMA-positive perineural capsule led to the classification as a benign microcystic/reticular variant of schwannoma [8].

In a review Liu et al. [9] collected 36 cases of MRS. These included 14 (39%) in the gastrointestinal tract, 8 (22%) in the subcutaneous or soft tissue, 7 (19%) in the head/neck region, 5 (14%) in endocrine glands (pancreatic, adrenal), 1 (3%) in the lumbal spinal canal and a single bronchial case published in the original case series [8]. Therefore, to our knowledge this is the first report of a tracheal MRS.

Tracheal obstruction is an incriminating condition, requiring short term desobliterating therapy. Pedunculated or small tracheal tumors may be treated by a variety of endoscopic methods with

good results, but sessile tumors with intra- or transmural growth showed recurrence rates as high as 21% after endoscopic resection whereas it was 0% for surgically resected schwannoma [6,10,11]. Radiotherapy plays a role in the adjuvant treatment of tracheal malignancies, but not in the management of benign tracheal diseases [12,13]. In these cases, tracheal resection is the treatment of choice.

In the Netherlands only 12% of malignant tracheal tumors were resected, although more than 50% of the patients were candidates for surgical treatment [14]. The same findings of surgically undertreated tracheal malignancies were reported in other cohorts [4,15]. There is a strong probability, that the same applies for benign tracheal neoplasms, as illustrated in this reported case, where the first treatment recommendations did not include surgery.

About 50% of the trachea can be resected in adults with good neck flexibility via a collar incision with optional manubriectomy or sternotomy. In elderly patients with kyphosis the length of possible resection may be considerably less. Long term results are excellent, although when carinal resections are included, operative mortality may be as high as 10% in patients with tracheal carcinoma depending on the length of resected trachea, tissue infiltration or preoperative high dose irradiation [12,13,16,17].

In summary, schwannoma variants like MRS lacking the features of classical schwannoma may occur in the trachea and make a correct diagnosis extremely challenging. As a result, delay of diagnosis is frequent, incorrect diagnosis and suboptimal treatment recommendations are common. Upper airway obstruction demands short term desobliterating treatment, so repeated diagnostic interventions causing unnecessary delays are not indicated. Although it seems to be undervalued, tracheal resection offers definitive treatment and sufficient material for a histopathological diagnosis with good results and low recurrence rates.

8. Patient perspective

When contacted for written consent, the patient expressed her satisfaction with the decision for a resection to end months of anxious discomfort and uncertainty. Although logopedic training had not improved her dysphonia, she did not experience relevant impairment in everyday life.

Declaration of Competing Interest

The authors have no conflicts of interest.

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Ethical approval

This clinical case report is exempt from the need of ethical approval in our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of this journal on request.

Author contribution

ME: The corresponding author contributed the collection of case details and images, literature work, obtainment of patient consent, writing of the paper.

MT: advise, revision and final approval.

Registration of research studies

Not Applicable.

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