Clinicopathologic Conference

A rare primary tracheal tumor

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ABSTRACT

A 20-year-old male without any prior comorbidities presented with 6-month history of hemoptysis as the sole symptom. Clinico-radiological profile and bronchoscopy were suggestive of a tracheal mass. This clinicopathologic conference discusses the differential diagnoses of primary tumors of the trachea and their management options.

KEY WORDS: Bronchoscopy, mass, trachea, tumor

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PRESENTATION OF THE CASE

Dr. Hariharan Iyer

A 20-year-old male presented to the Pulmonary Department with symptoms of hemoptysis for the past 6 months. The hemoptysis was intermittent, fresh red with the maximum amount being 20 ml per episode. The patient denied a history of fever, cough, dyspnea, voice change, facial puffiness, or loss of appetite; however, he reported a 2 kg weight loss over the past 6 months. He had no prior comorbidities, was a lifetime nonsmoker with no significant family history of any major illnesses.

Before presentation to our department, he was evaluated at an outside hospital, where a computed tomography (CT) scan of the chest was done as a part of the evaluation of hemoptysis [Figure 1]. He then consulted our hospital. On examination, he was hemodynamically stable with a room air saturation of 98%. General physical and systemic examination was normal. His routine blood investigations were unremarkable. His CT scan was suggestive of a mass arising from the mid tracheal right postero-lateral wall. For a definite diagnosis, a fiber optic bronchoscopy was done which confirmed the presence of a globular growth in the mid trachea [Figure 2] at a distance of 5 cm above the carina without the presence of a definite stalk. The rest of the tracheobronchial tree was normal. An endobronchial biopsy was obtained from the growth.

SALIENT FEATURES OF THE CASE AND DIFFERENTIAL DIAGNOSIS

Dr. Anant Mohan

This patient is a young male without any comorbidities and addictions. The hemoptysis was originating from the tracheal mass. Important points to be considered while formulating a differential diagnosis are the relatively subacute/chronic course of illness, reflecting a slow-growing mass in the trachea without any features of central airway obstruction or involvement of great vessels such as superior vena cava. Taking into consideration the clinico-radiological profile of this patient, the causes of
primary tracheal tumors (PTT-benign and malignant) need to be excluded from this study. Overall, PTTs constitute around 0.2% of all tumors, with approximately 90% being malignant in adults, as opposed to 30% in children.

Histologically, PTT can arise from the respiratory epithelium, salivary glands, and mesenchymal structures of the trachea. Among malignant PTT, squamous cell carcinoma (SCC) and adenoid cystic carcinoma (ACC) are the two major variants. Other malignant variants include mucocoeplidermoid, neuroendocrine tumors (carcinoid), adenocarcinoma, soft tissue sarcoma, chondrosarcoma, and malignant lymphoma. Among the benign causes, papillomas, pleomorphic adenomas, chondromas, fibrous histiocytoma, leiomyoma, schwannoma, and paraganglioma are some of the possible causes. Tracheal involvement can also be seen due to metastasis from a primary lung malignancy. However, no lung mass was seen on radiology, neither was any lesion visible endobronchially on flexible bronchoscopy. Hence, this possibility is excluded.

DR. HARIHARAN IYER

Squamous cell carcinoma and adenoid cystic carcinoma
SCC accounts for the majority of PTT, followed by ACC. SCC is usually associated with smoking and regional lymph nodal metastasis, while ACC usually occurs in the fourth to fifth decades.\(^1\)\(^2\) Given our patient profile, both these etiologies are less likely in the current case.

Squamous papillomas
These are rare benign tumors composed of stratified squamous epithelium. However, in contrast to a solitary mass in our case, papillomas mostly manifest as multiple exophytic growths into the trachea and are often associated with laryngeal and bronchial involvement.\(^3\) Hence, its possibility is less likely in the case under discussion.

Pleomorphic adenoma
This is a common tumor of major salivary gland and is known to arise from the tracheal and bronchial seromucinous glands. The reported age group of occurrence is 35–74 years with equal incidence in both genders.\(^4\) This is a slow-growing tumor and patients have symptoms such as cough, wheezing which can masquerade as asthma. Although our patient was relatively younger, this entity can be considered in our differential diagnosis.

Leiomyoma
Tracheal leiomyoma is a rare benign tumor; hence, the exact incidence, age, and gender distribution are not well defined. It can involve both the lung parenchyma and the tracheobronchial tree. In the trachea, leiomyoma usually manifests as a smooth solitary submucosal tumor.\(^5\)

Chondroma
This is a benign tumor arising from the cartilaginous tissue. Endobronchial chondromas are found in the larynx, trachea, or major bronchi; however, they are seen more commonly in the trachea than in the bronchi.\(^6\) It usually presents in the fourth decade and has a male preponderance.\(^6\) This also may be considered a possibility in the current patient.

Hamartoma
Tracheal hamartomas are extremely rare group of PTT. A hamartoma usually consists of cells probably derived from primitive connective tissue such as cartilage, fat, bone, and smooth muscle cells. Radiologically, a CT scan may reveal the presence of bone and fat tissue. However, no such finding was seen in our case, hence this possibility was unlikely.\(^7\)

Schwannoma
This is a tumor arising from the nerve sheath. Around 51 cases of primary tracheal schwannoma are reported in the literature.\(^8\) This tumor is more commonly seen in adults and has a female preponderance. The lesion is located most commonly in the distal third of the trachea, followed by proximal, and then the middle third of trachea.\(^9\) These features make this entity less likely as the cause of PTT in our case.

Neuroendocrine tumors
These include typical carcinoid, atypical carcinoid, and paragangliomas. Typical carcinoids are more common than atypical carcinoid. Carcinoid tumors arise more commonly from the bronchi; only 5% of cases arise from trachea.\(^10\) If within the trachea, these tumors most commonly arise from the distal one-third, and from the posterior noncartilaginous wall. Endobronchial carcinoids affect both genders equally and are frequently seen in the young population and have no predisposing factors such as smoking.\(^11\) With this profile, typical carcinoid remains a possible differential in our case.

Paragangliomas are benign neoplasms arising from extra-adrenal chromaffin cells derived from the neural

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**Figure 1:** Computed tomography chest showing a mass in the upper trachea arising from the right postero-lateral wall
They are mostly reported to arise from the membranous part of the upper and mid trachea and are usually sessile. Since our patient is young and has a slow-growing tumor, the possibility of benign variant of PTT is higher than a malignant tumor. Possible etiologies include carcinoid, paraganglioma, leiomyoma, chondroma, or pleomorphic adenoma.

Clinical diagnosis
PTT, possibly of benign etiology.

WITH THESE CLINICAL POSSIBILITIES, CAN WE HAVE THE HISTOPATHOLOGICAL EVALUATION OF THE SPECIMEN?

Dr. Adarsh Barwad
Histopathological examination of the endobronchial biopsy revealed crushed bronchial mucosa with subepithelial infiltration by tumor arranged in nests with prominent vascular network. The cells have abundant eosinophilic cytoplasm with round-to-oval nuclei. These cells were positive for synaptophysin, chromogranin, and negative for pancytokeratin, overall suggestive of paraganglioma [Figure 3].

ATHOLOGICAL DIAGNOSIS
Tracheal paraganglioma.

FINAL DIAGNOSIS

Dr. Anant Mohan
Primary tracheal tumor-paraganglioma
Paraganglioma of the trachea is a rare neoplasm belonging to the category of neuroendocrine tumors. They arise from...
extraadrenal chromaffin cells derived from the neural crest and are typically found in the neck, mediastinum and abdomen, and most commonly in association with the cranial nerves.[12]

Tracheal paraganglioma was first described in 1956.[13] Within the trachea, it has a propensity to arise from its membranous part, which was also the case in our patient. They are highly vascular tumors with hemoptysis as the usual presentation.[14-16] Other presentations include dyspnea, wheeze, and stridor depending on the degree of major airway obstruction.[17-19] Even rarely a mediastinal paraganglioma can mimic as tracheobronchial invasion.[20]

HOW ARE THESE PATIENTS MANAGED AND WHAT IS THEIR PROGNOSIS?

Dr. Sourabh Pahuja

A complete evaluation of patients with paraganglioma includes whole-body imaging with CT and/or magnetic resonance imaging (MRI) with possible vascular evaluation and magnetic resonance angiography of the site of the lesion. Paragangliomas have typical characteristics on CT and MRI. These tumors usually have homogeneous enhancement but may have areas of internal necrosis. They have intermediate signal intensity on T1-weighted MRI images and high signal intensity on T2-weighted images.[15]

Diagnosis is usually made easily by flexible bronchoscopy; however, since biopsy of these tumors may be associated with risk of bleeding due to high vascularity a backup of a rigid bronchoscope and a thoracic surgeon is highly desirable; the definitive management of tracheal paraganglioma is by surgical tracheal resection and anastomosis. A preoperative embolization may be done to reduce the risk of bleeding. Other treatment modalities include radiotherapy and stereotactic radiosurgery, but these are not reported in tracheal paragangliomas.

Our patient was not willing for immediate surgery; hence, we performed an electrosurgical snaring of the mass with a rigid bronchoscope [Figure 4] followed by argon plasma coagulation [Figure 5] of the tumor base to achieve hemostasis control. The patient was kept on follow-up with surveillance bronchoscopy at periodic intervals and he has remained recurrence-free and asymptomatic after 2 years.

SUMMARY

This case describes the various causes of PTTs which are relatively rare in occurrence. A young patient with hemoptysis should be evaluated at the earliest with a CT scan and bronchoscopy to rule out parenchymal and endobronchial abnormalities. Undue delay may lead to critical central airway obstruction. In the event of surgery being not being possible, these tumors may successfully be managed by a combination of interventional bronchoscopic techniques.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed..

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Conflicts of interest

There are no conflicts of interest.

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