

Tracheal Paraganglioma: Unusual Cause of Hemoptysis. A Case Report and Review of the Literature

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Abstract

Background: Paragangliomas are neoplasms arising from extraadrenal chromaffin cells. In the trachea, these tumors are exceedingly rare and only a few cases have been reported in the medical literature. Tracheal paragangliomas usually present with hemoptysis.

Case report: This report describes a 32 year old Moroccan man suffering from progressive dyspnea and hemoptysis due to tracheal tumor. Histopathology of tumor after surgery showed paraganglioma. A circular resection of the trachea carrying the first 4 rings allowed for resecting the entire tumor. No recurrence of tumor was detected at bronchoscopy six months later and it demonstrated good healing of the anastomosis.

Conclusion: Paraganglioma is an unusual histology for tracheal neoplasms. The originality of this case lies in the rarity of this type of tumor, tracheal location that is even more rare and differential diagnostic problem which arises. Clinical features, histological appearance and differential diagnosis as well as treatment are discussed.

Introduction

Paraganglioma of the trachea is a rare neoplasm, with a few cases reported in the literature. They are members of the extensive and diffuse category of neuroendocrine tumors [1]. Hemoptysis is the main symptom, and patients are at high risk for bleeding with airway manipulation. We recently encountered a new presentation of this rare entity and share our experience in its diagnosis and treatment.

Case Report

We report a 28 year old Moroccan man, no smoking. He suffering from progressive dyspnea for 6 months associated with recurrent hemoptysis low abundance. The chest radiograph was normal. Bronchoscopy was performed and showed a hemorrhagic tumor appearance, at the expense of the posterior wall of the trachea. A chest CT confirmed the existence of a posterior tracheal tumor 2.5 cm long axis without extra-tracheal extension and without significant lung injury (Figure 1). One biopsy was performed, it was very hemorrhagic and inconclusive and it was not renewed. A circular resection of the trachea carrying the first 4 rings allowed for resecting the entire tumor. Continuity



Figure 1: Unenhanced CT images show polypoid mass in posterior wall of trachea.

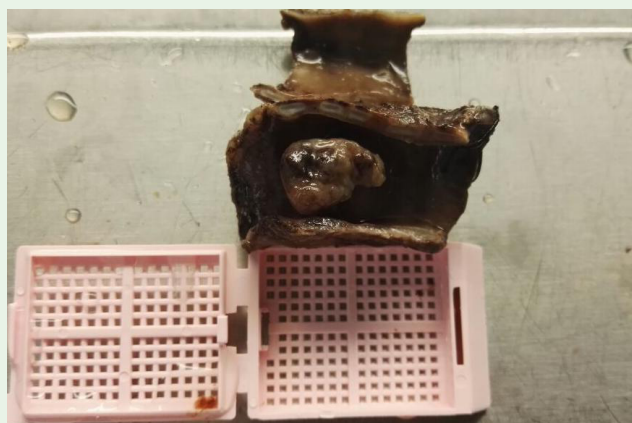


Figure 2: Photograph of the gross specimen obtained at tracheal resection shows a small polypoid mass arising from the posterior tracheal wall.

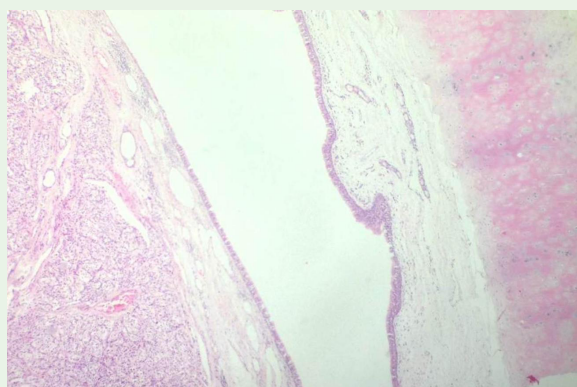


Figure 3: Tracheal wall shows a tumor proliferation with zellballen pattern surrounded by respiratory mucous. Note the cartilage on the right of the picture (Low magnification, HE).

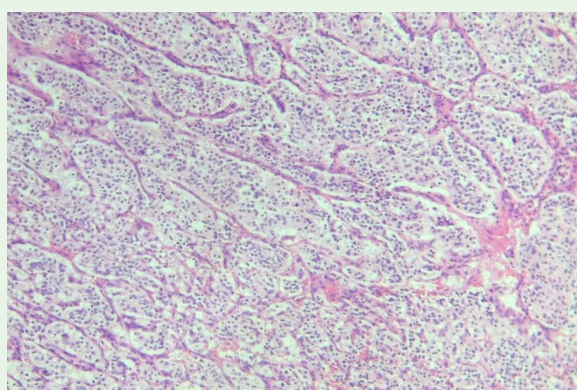


Figure 4: Medium-power photomicrograph of paraganglioma showing the typical nested, or zellballen, pattern. Surrounded by fibrovascular septa.

tracheal was reconstructed with end-to-end anastomosis. The postoperative course was unremarkable. Macroscopic examination of the piece shows an exophytic protruding tumor with a smooth surface, hemorrhagic appearance and soft consistency and pendant to the posterior wall of the trachea. Surgical margins were negative (Figure 2).

Histology of the lesion revealed tumor cells with abundant eosinophilic cytoplasm arranged in clusters (zellballen pattern) interspersed with fibrovascular stroma in keeping with a paraganglioma (Figure 3 and 4). The patient was kept on follow-up. No recurrence of tumor was detected at bronchoscopy six months later and it demonstrated good healing of the anastomosis.

Discussion

Paragangliomas are neuroendocrine neoplasms which were first described in 1880. Paraganglioma arises from multicentric system of extra adrenal paraganglia cells derived from the neural crest. Paraganglionic tissues have been found in more than 20 anatomic sites [2].

Tracheal paraganglioma was first described in 1956 [3]. Paragangliomas in the trachea are exceedingly rare and only a few cases have been reported in the medical literature. They usually present between the fourth to sixth decades of life and are three to four times more common in women [4]. These tumors can be either functional with secretion of active neuropeptides or nonfunctional. Tumor cells of the head, neck, and thorax are usually nonfunctional whereas tumor cells of the abdomen tend to be functional [5-7].

Up to 50% of patients with extra-tracheal paragangliomas are asymptomatic and the diagnosis is incidental [8,9]. Patients with tracheal paraganglioma often present with intermittent hemoptysis and symptoms relating to airway narrowing [10]. The highly vascular nature of paragangliomas results in hemoptysis and biopsy may be fraught with difficulty because of bleeding, as in our case.

Paragangliomas have typical characteristics on CT and MRI. They are usually show intense homogeneous enhancement except in necrotic areas, which enhance poorly. These tumors show intermediate signal intensity on T1- weighted MR images and high signal intensity on T2-weighted images [3,9].

The correct diagnosis of paraganglioma can only be made by histological examination following surgical biopsy or resection. Microscopically, paragangliomas are composed of organoid nests of chief cells (type I) surrounded by sustentacular cells (type II). This growth pattern is commonly referred to as a “zellballen” pattern. A prominent capillary network, which outlines the organoid nests, can be demonstrated histochemically with a reticulin stain or immunohistochemically with CD34. The chief cells are relatively uniform polygonal cells with centrally placed nuclei and eosinophilic granular cytoplasm. The chief cells typically demonstrate the presence of at least one neuroendocrine marker such as neuron-specific enolase, synaptophysin, or chromogranin. The type I cells are typically negative for epithelial markers. The type II, or sustentacular cells, are almost always positive with S-100 protein [11]. Immunohistochemical study is important to rule out other diagnostic namely: Parathyroid and thyroid neoplasms, carcinoid tumors, alveolar soft-part sarcoma, granular cell tumor, malignant melanoma, and metastatic renal cell carcinoma.

Histological parameter for charactering malignancy is presence of local invasion of primary tumor and distant metastasis. Metastatic involvements of lung, skull, vertebral bodies and liver have been described [12].

Tracheal paragangliomas are rare. Therefore, there are rare therapeutic strategies for these tumors [13,14]. Treatment is surgical excision, and many methods have been described, including tracheal resection, pre-operative embolization with subsequent resection. Preoperative embolization can be used to devascularize the lesion before removal because paraganglioma are very hemorrhagic [15].

Simple excision with minimal adequate margin and primary reconstruction was the most commonly used and most successful surgical technique in the reported cases and can be recommended as optimal therapy [10]. In the literature, only one case was died intraoperative resulting from marked hemoptysis [3]. Good surgical excision reduces recurrences and complications [10].

Conclusion

Paraganglioma is an unusual histology for tracheal neoplasms. It was first described in 1956.

Paragangliomas have tendency to be multicentric and unusual locations. Most of paragangliomas are benign and can be treated and cured by only surgical excision.

Availability of data and materials: Relevant data and supporting material will be available on request.

Authors' contributions: All authors contributed and approved the final version of the manuscript.

Consent for publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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