General Thoracic and Cardiovascular Surgery (2019) 67:991–995
https://doi.org/10.1007/s11748-019-01085-8

CASE REPORT

Behavior of primary tracheal glomus tumor, uncertain malignant potential subtype

Yuxing Jin[1] · Samer AL Sawalhi[1] · Deping Zhao[1] · Haomin Cai[1] · Ahmad M. Odeh[2] · Huikang Xie[3] · Jiong Song[4]

Received: 16 September 2018 / Accepted: 3 February 2019 / Published online: 25 February 2019

Abstract

The behavior of the pathological subtypes of tracheal glomus tumor (benign, uncertain malignant potential UMP, malignant) is vague. In a 51-year-old gentleman, suffering from cough for 4 months, computed tomography scan showed a mass at lower third of the trachea and the bronchoscope revealed exophytic mass at the tracheal lumen. Segmental tracheal resection was done using special modified endotracheal tube for the distal lung ventilation. The tracheal glomus tumor was also diagnosed based on the immunohistochemical staining. The tumor was 2 cm in diameter, deeply located, mitotic phase was difficult to identify, and a diagnosis of UMP subtype was made. There was no recurrence after 2 years follow-up. This is the first reported case of UMP subtype in lower trachea and we studied the treatment options with the clinic-pathological behavior of this tumor and its sequel by regular follow-up.

Keywords Glomus tumor · Trachea · Endotracheal intubation

Introduction

Glomus tumor (GT) is a rare soft tissue neoplasm derived from modified smooth muscle cells of the normal glomus body surrounding arteriovenous anastomosis [1]. Tracheal GT occurs commonly at lower portion of the trachea, because the glomus glands and blood vessels are more abundant in the lower trachea and bronchial mucosa than in the upper and middle bronchial mucosa, therefore, most common location of tracheal GT is the distal trachea (54%), then the middle trachea (22%) [2]. Most of the reported tracheal GT are benign with only 5% displaying malignant histology [3]. Previous authors reviewed the literature on primary broncho-pulmonary glomus tumors. They have found UMPS either in lung parenchyma or in the lobar bronchi [1]. However, this is the first reported case of UMPS in the lower trachea. Furthermore, this case discussed the clinic-pathological differentiation between carcinoid and tracheal GT, because both of them have shared clinical, radiological and histopathological similarity to glomus tumor.

Case report

A 51-year-old gentleman, non-smoker, was complaining for cough for 4 months without other symptoms. The physical findings were unremarkable. Contrast-enhanced computed tomography scan showed an intensely enhanced nodular mass measuring about 1.5 × 2 cm at the lower third of the trachea (Fig. 1a). Bronchoscopy revealed a polypoidal exophytic smooth surface mass, located just above the carina, originating from the membranous portion of the trachea, and almost occluding the distal tracheal lumen (Fig. 1b). Tracheal tumor resection and reconstruction requires multidisciplinary team work and the challenging share issue between the surgeon and anesthesiologist is to maintain the airway through the operation. The tracheal tumor was in the lower part of the trachea; therefore, we chose right posterior-lateral thoracotomy approach at the level of the fourth intercostal space. Endotracheal intubation was done using special modified endotracheal tube for

✉ Deping Zhao
zdp1992@163.com; drsawalhi@yahoo.com

[1] Department of Thoracic Surgery, Shanghai Pulmonary Hospital, Tongji University, Shanghai, China

[2] Department of Surgery, King Faisal University, Al Hufuf, Saudi Arabia

[3] Department of Pathology, Shanghai Pulmonary Hospital, Tongji University, Shanghai, China

[4] Department of Anesthesia, Shanghai Pulmonary Hospital, Shanghai, China

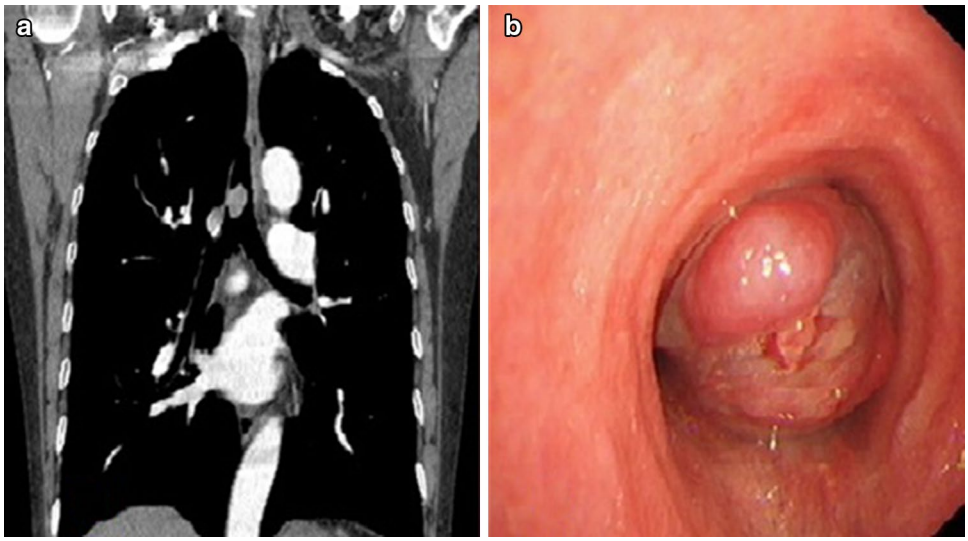

Fig. 1 **a** Chest CT scan, a nodular tumor arises from the right distal tracheal wall. **b** Bronchoscope shows a cauliflower-polypoidal 2-cm mass resulting in a 90% obstruction of distal trachea

Fig. 2 A modified double-lumen tube DLT size 32 F. The distal end of the tracheal cuff of DLT was tied by a silk suture wrapped around distal part of the tracheal cuff to be away from the tracheal tumor

the distal lung ventilation (Fig. 2). A left double-lumen tube (DLT) size 32 F was employed to selectively intubate left main bronchus. The distal end of the tracheal cuff of DLT was tied by a silk suture wrapped around the lower end of the cuff in order to reduce the size of the cuff (Figs. 2, 3), so it will not be in contact with the tracheal tumor and it facilitates suctioning, therefore this modified technique has dual function through one lung ventilation and suctioning in the same time. A fiberoptic scope was used to achieve proper placement of the tracheal cuff as well as the bronchial cuff. After right-thoracotomy and stapling the azygos vein using endovascular cartridge (Medtronic, Minneapolis, MN, USA). Dissection of the anterior and posterior tracheal wall with preservation of its blood supply and the recurrent laryngeal nerve were performed. We used intraoperative a flexible bronchoscope with transillumination as a guide to localize accurately the tumor margins while the assistant surgeon used a needle through the trachea to mark the superior and inferior tumor margins. After that the distal segment of the trachea was transected, Then a second reinforced endotracheal tube size 16 F was used to intubate the left main bronchus (cross-table ventilation) (Fig. 4). Proximal end

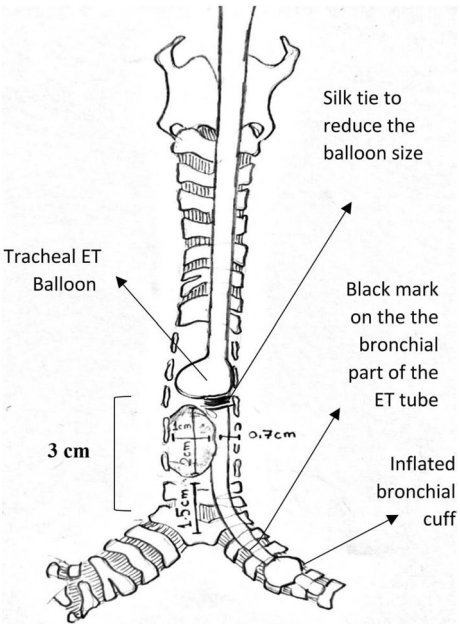

Fig. 3 Left modified double-lumen tube DLT size 32 F was employed to intubate left main bronchus with aid of fiberoptic scope was used to achieve proper position

of tracheal tumor was cut with clear margin circumferentially. Both tracheal ends were approximated after releasing the inferior pulmonary ligament and lung hilum using 4/0 interrupted vicryl sutures with 4 mm apart. Retained blood in the trachea–bronchial airway was sucked before starting the tracheal anastomosis from the posterior membranous part keeping the knots outside. Sometimes the cross-field tube was withdrawn frequently during anastomosis allowing accurate placement of anastomotic sutures and enabling suctioning of blood from the distal airway. After that sutures were used to approximate the

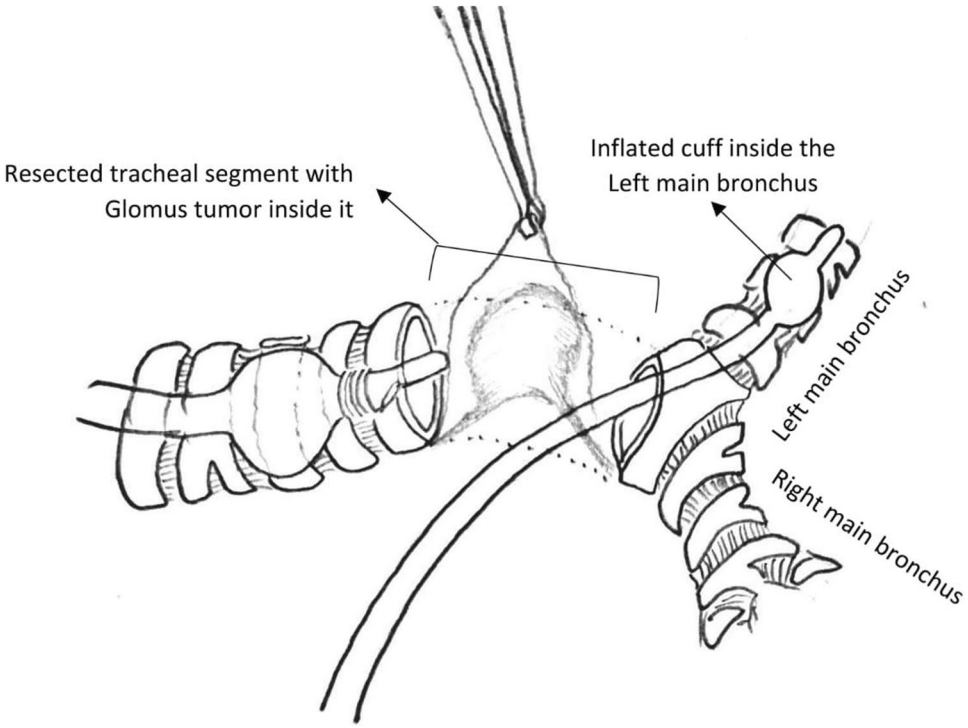

Fig. 4 Distal intraluminal tracheal mass almost resected completely and endotracheal tube (ET) size 16 F was used to intubate the left main bronchus (cross-table ventilation) to keep one lung ventilation through the procedure

cartilaginous wall and the pericardial flap was wrapped around the anastomosis. During last step in the anastomosis, we switched the (cross-field ventilation) to the oral endotracheal tube with its cuff pushed carefully distal to the anastomotic line. The anastomosis was checked under the water with an inflation pressure of 20–40 cm of H_2O. A guardian silk suture was used between the submental and anterior chest wall keeping the neck flexed enough to maintain tension free tracheal anastomosis. The patient was extubated at the end of the operation and he recovered smoothly without post-operative air leak. The histopathology result showed 2×1.5 cm well circumscribed solid lesion. Microscopically, the tumor contains branching vascular channels that are lined with endotheliocytes and interspersed by uniformly round epithelioid cells with eosinophilic cytoplasm, mitotic phase (difficult to see)

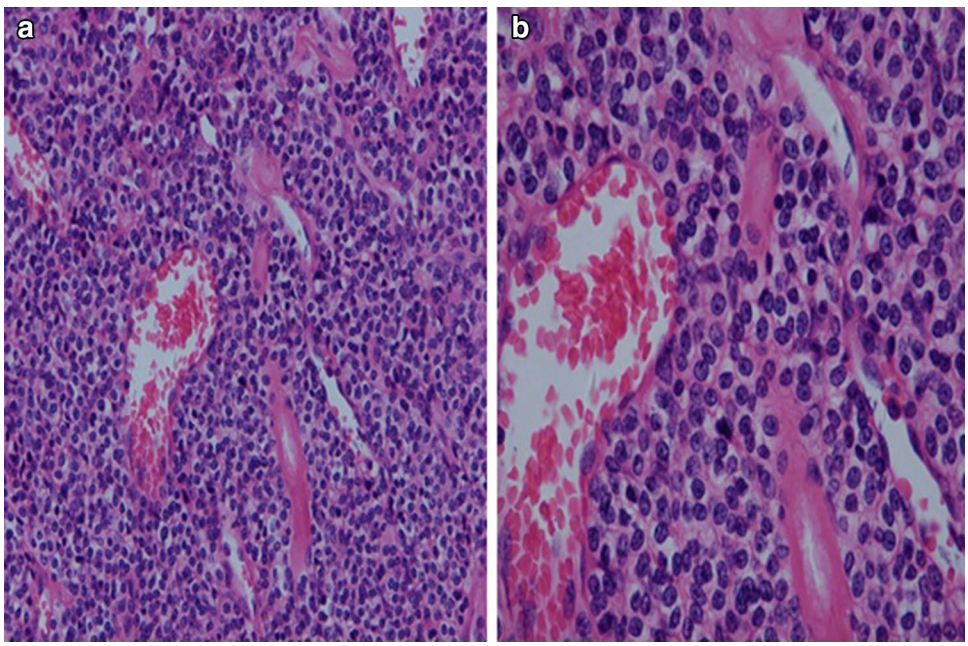

Fig. 5 Pathology and H&E staining findings. **a** A sheet-like growth pattern and clusters of round epithelioid cells in abundant eosinophilic cytoplasm and round nuclei with numerous vascular spaces. H&E stain. Original magnification ×200. **b** Single round cells with a uniformly monotonous with a round nucleus, clear or pale eosinophilic cytoplasm and hyperchromatic nuclei. No necrosis, cellular atypia, vascular invasion or mitotic figures were noticed. H&E stain. Original magnification ×400

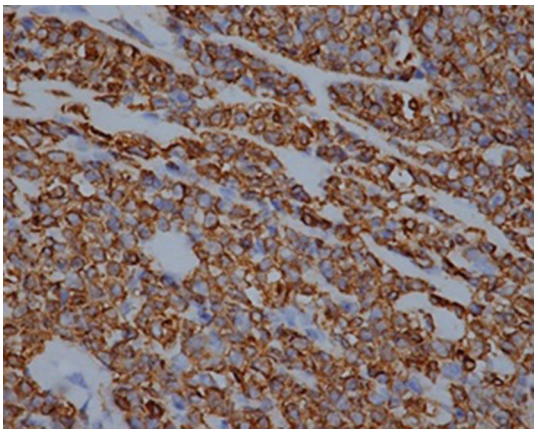

Fig. 6 Positive immunohistochemical staining for anti-α-smooth muscle actin antibody. Original magnification ×400

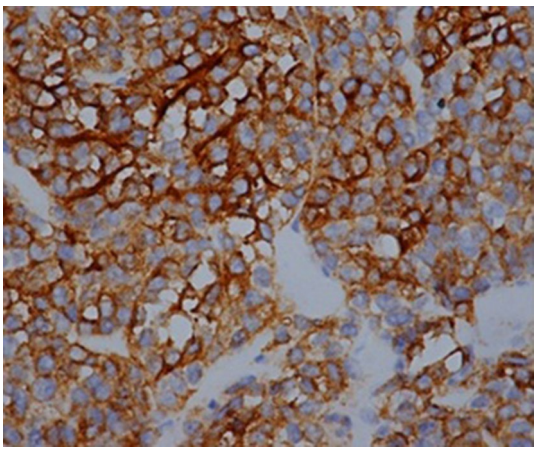

Fig. 7 Positive immunohistochemical staining for CD34. Original magnification ×400

(Fig. 5a, b), the upper and lower margins were free of the tumor. Seven lymph nodes were harvested, all were negative for malignancy. Immunohistochemical staining of the cells was demonstrated positive for SMA (smooth muscle actin antibody) (Fig. 6), CD34 (Fig. 7), SHA, Syn (synaptophysin), K167 FOCAL, whereas negative for Cg-A (chromogranin-A), CD56, CK (casein kinase), TOT, LCA, VIN, and CD31. The tracheal glomus tumor was diagnosed based on immunohistochemical staining and histological characteristics. The diagnostic criteria for malignant glomus tumors include; Large size (> 2 cm) and deep location position, atypical mitotic phase and nucleus with obvious atypia, mitotic phase number ≥ 5/10 HPF [1]. In this case, the tumor was 2 cm in diameter; deeply located, mitotic phase is not easy to see, so we diagnosed it as uncertain malignant potential. Bronchoscopy was performed 1 month later. No incidence of tracheal stenosis was noticed and the patient was followed up by regular bronchoscopy, no recurrence after 2 years.

Discussion

Tracheal GT is a rare type of myoepithelial tumor that may be associated with an autosomal dominant mutation in a gene located on chromosome 1p [4]. No causative relationship has been reported between GTs and smoking. This disease mainly occurs in middle-aged people [5] and is more common among men than women, male-to-female ratio (3:1) [4]. The world Health Organization (2002) defined (GTs) as benign tumors comprising perivascular cells [6]. This kind of the tumor is growing slowly and behaves like benign. Primary pulmonary glomus tumors affect both bronchi and peripheral lung parenchyma. In the bronchus, the tumor looked like a protruding, polypoidal mass, whereas it appears as nodular lesion in the peripheral lung. Clinically, symptoms related to airway irritation are common in bronchial or tracheal GTs, but usually asymptomatic once GT arising in peripheral lung parenchymal. Among primary lung tumors, the carcinoid tumor is a mimic of the glomus tumor, and differentiating these tumors is known to be difficult, especially in small samples. Because of this rare pathological entity, primary broncho-pulmonary GTs were misdiagnosed as carcinoid tumors based on preoperative biopsy or intraoperative frozen section. Diagnosis of tracheal GTs is usually made on the histopathological features and immunohistochemical staining patterns. Glomus tumor cells are round or oval in shape and line up tightly. They showed positive staining for SMA, VIM, and type IV collagen, but do not stain positive for carcinoid tumors, including chromogranin, CK, desmin and S-100 protein. In addition, glomus tumors lack the characteristics (salt and pepper appearance) of nuclear chromatin distribution which is typical for carcinoid tumors, whereas carcinoid tumors have less vascular mass, and their cells are arranged as nests.

Growing evidence has shown that most of the large and deeply located glomus tumors behave in a clinically benign fashion, hence the first diagnostic criterion was modified, and these lesions are now considered glomus tumors of uncertain malignant potential [7]. However, we consider that the degree of nuclear atypia, and mitotic activity could be used to estimate the malignancy of tracheal GTs, but these criteria related to tumor size, and deep location are unsuitable because most previously reported tracheal GTs with diameter > 2 cm were benign. Odie et al. [1] reviewed the literature on primary broncho-pulmonary glomus tumor of UMPS taking consideration of malignant grade estimation. He found that UMPS located in the lung parenchyma, right and left main bronchus, bronchus intermedius and subpleural, but not in the trachea. Till now, no standard

approach for treating patients with tracheal GT. Segmental tracheal resection with adequate safe margins is the preferred method used.

Sakr et al. [8] considered that endoscopic intervention has limited indications including; tumor biopsy, and to restore airway patency in urgent situations. Also, if the lesion is benign and strictly confined to the airway lumen or the patient is not fit for surgery, and those with a tumor located high in the trachea (C7–T1 level), which associated with phonation dysfunction. Completeness of large tracheal tumors resection by endoscopy may be questionable, due to interventional technical problem and the complications rates. Usage of bronchoscopy intervention in such kind of big tracheal tumors is sometimes dangerous, because of bleeding and risk of suffocation. Wang et al. [5] arrested bleeding by using bronchial artery embolization. On the other hand, radical resection is mandatory to prevent tumor recurrence and growth of fibrinous coagulation tissue material. Furthermore, these patients need further bronchoscopy examination follow-up, and bronchoscopy intervention like; electrocautery, high-frequency electrocautery, argon-plasma coagulation, or radiotherapy to deal with tumor recurrence. Masoum et al. [9] reported the case of a 21-year-old man who underwent GT resection via bronchoscopy that recurred after 1 year. Even though tracheal GTs could be resected via bronchoscopy with similar long-term results as compared to open resection [2], we preferred the surgical resection, because the patient was young and the size of the lesion was large enough. Therefore, even bronchoscopy biopsy was avoided to avoid the risk of bleeding and high recurrence rate. Open resection was definitely recommended based on the tumor location and an estimated 5% risk of malignancy. The major cause of death of malignant GT is distant metastasis. The rate of metastasis is 31.2–38.0%, and tumors often recur between 3 and 4 years after surgery [7]. Estimation of the malignant potential of bronchopulmonary glomus tumors should be addressed with close follow-up.

Conclusion

Glomus tumor is a rare benign neoplasm arising in the tracheobronchial respiratory tract. High index of suspicion of tracheal glomus should be based on the location and the pathological immunohistochemical staining. Tracheal GT should be treated with segmental tracheal resection instead of bronchoscope therapy to avoid tumor recurrence and bleeding.

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