Glomus Tumor of the Trachea Managed by Spiral Tracheoplasty

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Conflict of interest: None declared

Patient: Female, 58
Final Diagnosis: Glomus tumor of trachea
Symptoms: Hemoptysis
Medication: —
Clinical Procedure: —
Specialty: Otolaryngology

Objective: Rare disease
Background: Glomus tumors are usually found over the dermis of the extremities, particularly over the subungal region of the fingers, and occurrence in the trachea is an extremely rare event. To date, only 29 cases of tracheal and 2 main bronchus glomus tumors have been reported in the English literature. Our patient is the first ever reported case in Taiwan that was managed by spiral tracheoplasty.

Case Report: A 58-year-old woman was admitted to our hospital because of hemoptysis. Computed tomographic (CT) scan revealed a mass over the posterior wall of the trachea. Surgical resection with spiral tracheoplasty was performed due to uncontrolled bleeding and airway compromise. Histopathology and immunostaining confirmed a glomus tumor. Postoperative course was unremarkable and she was discharged in improved condition after 9 days of hospital stay.

Conclusions: Although chronic symptom presentation is the rule for tracheal glomus tumors, airway obstruction and bleeding are life-threatening presentations. Histopathological examination and staining are important to differentiate it from hemangiopericytoma or carcinoid tumors. Spiral tracheoplasty after tangential resection may be tried, as this preserves more tracheal tissue, decreases tension, and prevents postoperative leakage at the anastomotic site.

MeSH Keywords: Glomus Tumor • Thoracic Surgery • Tracheal Diseases

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Background

Glomus tumors are usually found over the subungual region of the fingers of the hand and the lower extremities. Its occurrence in the trachea is extremely rare. To date, there are 31 reported cases of tracheal and proximal main bronchus glomus tumors in the English literature [1–28] since it was first reported by Mason in 1924 [29]. Sleeve resection with primary reconstruction of the trachea is usually the treatment of choice for tracheal glomus tumors. In patients with tangential tracheal lesions, spiral tracheoplasty has never previously been performed, but can be attempted after tangential resection. Our patient is the first reported case to undergo successful spiral tracheoplasty, as pioneered by one of our authors, after resection of this tumor that occurred in a very rare location.

Case Report

A 58-year-old Taiwanese woman was admitted to our hospital due to hemoptysis. She had no known systemic illnesses in the past and did not smoke cigarettes or consume alcoholic beverages. She denied any weight loss, fever, cough, dyspnea, or epistaxis prior to this incident. A chest CT scan was performed, showing a 2.2×2.2 cm polypoid lesion over the posterior wall of the lower third of the trachea (Figure 1A, 1B).

On examination, the patient was afebrile and breath sounds were clear. Laboratory test results were unremarkable. A bronchoscopic examination was performed, showing a polypoid tumor over the posterior wall of the trachea (Figure 2A). During admission, the patient had another episode of massive hemoptysis, which resulted in hypotension, respiratory distress, and anemia that necessitated a blood transfusion. Our thoracic surgeon was consulted and surgery was performed. Excision of the tracheal tumor via sternotomy was performed (Figure 2B) followed by reconstruction of the trachea with spiral anastomosis. Afterwards, hemostasis was achieved, hemodynamics stabilized, and intraoperative fiberoptic bronchoscopy showed tracheal patency. Hemangiopericytoma was reported on fast-frozen section. However, histopathology showed a hypervascular tumor composed of branching, dilated, thick-walled vascular channels and thin-walled capillary-like vascular spaces, surrounded by lobular arrangements of oval-to-spindle cells, with abundant eosinophilic cytoplasm and centrally-located rounded nuclei. A focal hemangiopericytoma-like pattern interspersed with thin-to-coarse collagenous bundles was also noted (Figure 3A–3B). Immunohistochemical staining was positive for smooth muscle actin (Figure 3C), focally reactive with synaptophysin and negative for cytokeratin, chromogranin A, S-100 protein, and HMB-45. This was later diagnosed as a glomus tumor. Seven days after surgery, fiberoptic bronchoscopy was repeated, showing a patent trachea with mild granulation tissue over the oblique anastomotic line and no stenosis (Figure 2C). The patient was discharged the next day. She is currently asymptomatic and no complication or tumor recurrence has occurred in approximately 2 years of follow-up.

Discussion

Glomus tumors are usually benign and rarely exhibit malignant potential. However, they can cause airway obstruction and bleeding. In 2001, the World Health Organization reclassified these tumors with atypical and malignant features into 4 categories [30]. The diagnosis of malignant glomus tumor is reserved for tumors having: a size of >2 cm and subfascial or visceral location, atypical mitotic figures or marked nuclear atypia, and any level of mitotic activity. Glomus tumors not fulfilling the criteria for malignancy, but having at least 1 atypical
Figure 3. (A) Representative low-power magnification of a cut section of the glomus tumor showing a hypervascular tumor composed of branching, dilated, thick-walled, vascular channels and thin-walled, capillary-like vascular spaces (hematoxylin and eosin, 40×). (B) High-power magnification of the tumor showing lobular arrangements of oval- to-spindle-shaped cells, with abundant eosinophilic cytoplasm and centrally-located rounded nuclei (hematoxylin and eosin, 400×). (C) Immuno-staining showing tumor cells strongly reactive to actin.

Figure 2. (A) Preoperative bronchoscopic image of the tumor almost completely obstructing the trachea. (B) Gross appearance of the excised tumor. (C) Bronchoscopic image performed 7 days postoperatively showing mild granulation tissue over the oblique anastomotic site. The trachea is patent and the axis of the distal anastomotic site is not on the same plane as the proximal segment.
| No. | Author Reference | Year | Age | Sex | Symptoms | Tumor site | Size (cm) | Treatment | Outcome |
|-----|------------------|------|-----|-----|----------|------------|-----------|-----------|---------|
| 1   | Hussarek [1]     | 1950 | 43  | F   | Dyspnea, stridor | Upper 3rd post. wall | Bean size | Tracheal resection | Not stated |
| 2   | Fabich [2]       | 1980 | 63  | M   | Cough     | Lower 3rd post. wall | 2.5×2.0×1.0 | Sleeve resection | Died of complications on 10th post-op day |
| 3   | Warter [3]       | 1980 | 69  | M   | Dyspnea, hemoptysis | Mid-trachea post. wall | 2.3×1.5×1.5 | Segmental resection | Unremarkable |
| 4   | Heard [4]        | 1982 | 50  | M   | Dyspnea, asthma-like symptoms | Lower 3rd post. wall | 2.5×1.5×1.0 | Sleeve resection | Died of sepsis on 15th post-op day |
| 5   | Ito [5]          | 1988 | 51  | M   | Respiratory infection, hemoptysis | Upper 3rd post. wall | 1.5×1.2×1.0 | Segmental resection | No recurrence at 2 years |
| 6   | Sheffield [6]    | 1988 | 74  | M   | Dyspnea, cough | Lower 3rd post. wall | 2.2 | Endoscopic removal | Unremarkable |
| 7   | Kim [7]          | 1989 | 54  | F   | Dyspnea, cough, hemoptysis | Mid-trachea post. wall | 1.5×1.2 | Segmental resection | No recurrence at 13 months |
| 8   | Shin [8]         | 1990 | 47  | F   | Cough, hemoptysis | Lower 3rd post. wall | 1.5×1.0×1.0 | Wedge resection | Not stated |
| 9   | Garcia-Prats [9] | 1991 | 58  | M   | Dyspnea, cough, hemoptysis | Mid-trachea post. wall | 2.5×1.8 | Segmental resection | No recurrence at 8 months |
| 10  | Haraguchi [10]   | 1991 | 61  | M   | Asymptomatic | Mid-trachea post. wall | 1.2 | Sleeve resection | Not stated |
| 11  | Arapantoni [11]  | 1995 | 65  | M   | Dyspnea, hemoptysis | Lower 3rd post. wall | 4.5×3.0 | Bronchoscopy with Nd-Yag laser excision | No recurrence at 1 year |
| 12  | Koskinen [12]    | 1998 | 66  | M   | Asymptomatic | Lower 3rd post. wall | 2.0×3.0 | Multiple endoscopic laser ablations and external radiotherapy | Not stated |
| 13  | Watanabe [13]    | 1998 | 43  | M   | Hoarseness | Lower 3rd post. wall | 2.0×1.6×1.4 | Sleeve resection | No recurrence at 20 months |
| 14  | Menaissy [14]    | 2000 | 34  | M   | Hemoptysis | Mid-trachea post. wall | 2.4×2.1×1.6 | Tracheal resection | No recurrence at 4 months |
| 15  | Lange [15]       | 2000 | 20  | M   | Dyspnea | Left main bronchus | 1.4×1.3×0.6 | Bronchial sleeve resection | No recurrence at 9 months |
| 16  | Gowan [16]       | 2001 | 73  | M   | Cough, chest pain, dyspnea, hemoptysis | Mid-trachea post. wall | 1.6×0.3×0.6 | Segmental resection | No recurrence at 6 years |
| 17  | Chien [17]       | 2003 | 50  | F   | Cough, dyspnea, hemoptysis | Lower 3rd post. wall | 2.5×2.5×2.0 | Segmental resection | No recurrence at 1 year |
| 18  | Nadrous [18]     | 2004 | 39  | M   | Hemoptysis | Upper 3rd post. wall | 2.0×1.5×1.5 | Sleeve resection | No recurrence at 3 months |
| 19  | Altinok [19]     | 2006 | 83  | F   | Dyspnea, hemoptysis | Upper 3rd post. wall | 2.0×1.5×1.2 | Partial sleeve resection | No recurrence at 1 year |
| 20  | Haver [20]       | 2008 | 70  | F   | Dyspnea | Mid-lower trachea post. wall | 1.8×1.3×1.3 | Tracheal resection | No recurrence at 2 years |
feature other than nuclear pleomorphism, as in this case, are classified as glomus tumor of uncertain malignant potential.

From 1950 to 2014, 29 cases of tracheal and 2 cases of proximal main bronchus glomus tumors have been reported in the English literature. Characteristics of these patients are summarized in Table 1. There were 21 males and 10 females with a mean age of 52±17 years. The youngest was a 10-year-old girl and the oldest was an 83-year-old woman. These tumors occurred anywhere along the length of the trachea and proximal main bronchus, and were all located over the posterior or posterolateral tracheal wall.

Tracheal glomus tumor is sometimes mistaken for a carcinoid tumor, or, as in our case, a hemangiopericytoma. Therefore, immunostaining and careful histopathologic examination should be performed to avoid misdiagnosis. Some would argue for conservative treatment or watchful waiting for treating this disease since it is benign in 95% of cases. However, complete resection was performed in this case due to recurrent bleeding, acute airway obstruction, and a classification of “uncertain malignant potential”.

Sleeve resection with primary reconstruction of the trachea is the treatment of choice for tracheal glomus tumor. Complete surgical resection is usually curative, but endoscopic intervention may be performed for lesions that are confined to the airway lumen without extension into the airway wall. This can also be done when the tumor is benign, in patients with high surgical risks, or when the patient refuses surgical intervention. Of the 31 cases described in the literature, most underwent surgical resection followed by reconstruction, with only 9 patients receiving endoscopic resection combined with laser ablation.

| No. | Author Reference | Year | Age | Sex | Symptoms | Tumor site | Size (cm) | Treatment | Outcome |
|-----|------------------|------|-----|-----|----------|------------|-----------|-----------|---------|
| 21  | Colaut [21]      | 2008 | 70  | M   | Dyspnea  | Mid-trachea post. wall | 2.0×1.0×1.0 | Endoscopic resection and Nd-YAG | No recurrence at 2 years |
| 22  | Shang [22]       | 2010 | 59  | M   | Chest pain, dyspnea | Lower 3rd post. wall | 2.0×1.0×0.5 | Endoscopic removal | No recurrence at 1 year |
| 23  | Sakr [23]        | 2011 | 66  | M   | Stridor, cough, dyspnea | Upper 3rd post. wall | 1.2×0.8×2.0 | Sleeve resection | No recurrence at 21 months |
| 24  | Mogi [24]        | 2011 | 56  | F   | Dyspnea, cough | Lower 3rd post. wall | 1.3×1.2×1.1 | Sleeve resection | No recurrence at 9 months |
| 25  | Okereke [25]     | 2011 | 58  | M   | Stridor, dyspnea | Lower 3rd post. wall | 1.1 | Tracheal resection | No recurrence at 6 months |
| 26  | Fan [26]         | 2013 | 15  | M   | Cough, dyspnea, hemoptysis | Mid-trachea post. wall | 2.0×2.5 | Tracheal resection | No recurrence at 1 year |
| 27  | Choi [27]        | 2014 | 64  | M   | Asymptomatic | Mid-trachea post. wall | 2.6 | Tracheal resection | No recurrence at 2 years |
| 28  | Xiong [28]       | 2014 | 55  | M   | Hemoptysis, cough, chest pain | Lower 3rd post. wall | 0.5×0.3×0.3 | Bronchoscopic cryoablation with brachytherapy | No recurrence at 6 months |
| 29  | Wu [current case]| 2014 | 58  | F   | Hemoptysis | Lower 3rd post. wall | 1.2×1.0×0.8 | Bronchoscopic cryoablation and argon plasma coagulation | No recurrence at 6 months |

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In 2009, we introduced a technique called spiral tracheoplasty to preserve tracheal tissue while reducing tension at the anastomotic site of the trachea after tangential wall resection (Figure 4). Briefly, the procedure for spiral tracheoplasty consists of 5 steps. 1) After identifying the tumor, the trachea is separated from the esophagus by about 2 cm to allow for manipulation and exposure of the operative field. 2) This is followed by tangential resection of the tracheal lesion instead of a circumferential resection. For example, a 2-cm area of tracheal tissue and tumor was resected in this picture. 3) Both transected tracheal ends are further separated from the esophagus and are rotated 90 degrees in opposite directions. The proximal end is rotated clockwise and the distal end is rotated counter clockwise. 4) The tracheal ends are then trimmed for irregularities to obtain good apposition. In this example, the 1-cm lengths at the proximal and distal ends are positioned next to each other, to obtain a mirror image and are tested for exact fit. 5) Then, the anastomosis is performed using running sutures [31]. Intra- and post-operative fiberoptic bronchoscopy should be performed to check for tracheal patency, bleeding, and optimal anastomosis.

In performing tracheal surgery, it is of utmost importance that the safety of the anastomosis takes precedence over the completeness of the resection. In spiral tracheoplasty, a tangential tracheal wall excision instead of a circumferential resection with end-to-end anastomosis is more advantageous since the resected tracheal tissue is only half of the length removed or lost compared with circumferential resection, preserving and sparing more of the trachea in the process. The tension created by twisting both ends in opposite directions is acceptable, since a 2-cm or more separation from the esophagus was created prior to the twisting and anastomosis. This important step also frees the esophagus and allows it to shorten or contract a little to accommodate and match the already shortened trachea. This procedure is most suitable for surgery of tangential lesions of the cervicothoracic trachea, and laryngeal release or other types of release of the proximal trachea is unnecessary.

This is the first case wherein a tracheal glomus tumor was managed with this technique, and this is the first time that a bronchoscopic image is available post-procedure. As seen in Figure 2C, the anastomotic line is tangential and oriented to the left, and the axis of the distal anastomotic segment is not in the same plane as the proximal segment.

**Conclusions**

Although glomus tumors are mostly benign, tumor location is important because airway compromise and bleeding can be life-threatening. Histopathologic staining is critical to avoid mistaking it for a hemangiopericytoma or a carcinoid tumor. Spiral tracheoplasty may be performed after tangential resection, as it preserves more tracheal tissue, decreases tension, and prevents leakage at the anastomotic site, which is almost always fatal when it occurs.

**Statement**

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References:

1. Hussarek M, Reider W: Glomus tumor the air tubes. Krebsarzt, 1950; 5: 208–12
2. Fabich DR, Hafez GR: Glomangiom of the trachea. Cancer, 1980; 45: 2337–41
3. Warter A, Vetter JM, Morand G, Philippe E: Tracheal glomus tumor. Arch Anat Cytol Pathol, 1980; 28: 184–90
4. Heard BE, Dewar A, Firmin RK, Lennox SC: One very rare and one new tracheal tumor found by electron microscopy: glomus tumor and acinic cell tumor resembling carcinoid tumors by light microscopy. Thorax, 1982; 37: 97–103
5. Ito H, Motohiro K, Nomura S, Tahara E: Glomus tumor of the trachea: immunohistochemical and electron microscopic studies. Pathol Res Pract, 1988; 183: 778–84
6. Sheffield E, Dewar A, Corrin B et al: Glomus tumour of the trachea. Histopathology, 1988; 13: 234–36
7. Kim YJ, Kim JH, Suh JS et al: Glomus tumor of the trachea: report of a case with ultrastructural observation. Cancer, 1989; 64: 881–86
8. Shin DH, Park SS, Lee JH et al: Oncocytic glomus tumor of the trachea. Chest, 1990; 98: 1021–23
9. García-Prats MD, Sotelo-Rodríguez MT, Ballestín C et al: Glomus tumor of the trachea: report of a case with microscopic, ultrastructural and immunohistochemical examination and review of the literature. Histopathology, 1991; 19: 459–64
10. Haraguchi S, Yamamoto M, Nishimura H: A glomus tumor of the trachea: a case report. Nippon Kyobu Geka Gakkai Zasshi, 1991; 39: 214–18
11. Arapantoni-Dadioti P, Panayiotides J, Fatsis M, Antypas G: Tracheal glomus tumor. Respiration, 1995; 62: 160–62
12. Koskinen SK, Niemi PT, Ekfors TO et al: Glomus tumor of the trachea. Eur J Radiol, 1998; 6: 364–66
13. Watanabe M, Takagi K, Ono K et al: Successful resection of a glomus tumor arising from the lower trachea: report of a case. Surg Today, 1998; 28: 332–34
14. Menaissey YM, Gal AA, Mansour KA: Glomus tumor of the trachea. Ann Thorac Surg, 2000; 70: 295–97
15. Lange TH, Magee MJ, Boley TM et al: Tracheobronchial glomus tumor. Ann Thorac Surg, 2000; 70: 292–95
16. Gowan RT, Shani JF, Perkins DG, Maziak DE: Glomus tumor of the trachea. Ann Thorac Surg, 2001; 72: 598–600
17. Chien ST, Lee TM, Hsu JY et al: Glomus tumor of the trachea. J Chin Med Assoc, 2003; 66: 551–54
18. Nadrous HF, Allen MS, Bartholmai BJ et al: Glomus tumor of the trachea: value of multidetector computed tomographic virtual bronchoscopy. Mayo Clin Proc, 2004; 79: 237–40
19. Altnik T, Cakir E, Guhan E, Tastee I: Tracheal glomus tumor. J Thorac Cardiovasc Surg, 2006; 132: 201–2
20. Haver KE, Hartnick CJ, Ryan DP et al: Case 10-2006: a 10-year-old girl with dyspnea on exertion. N Engl J Med, 2008; 358: 1382–90
21. Colaut F, Toniolo L, Scapinello A, Pozzobon M: Tracheal glomus tumor successfully resected with rigid bronchoscopy: a case report. J Thorac Oncol, 2008; 3: 1065–67
22. Shang Y, Huang Y, Huang HD et al: Removal of glomus tumor in the lower tracheal segment with a flexible bronchoscope: report of two cases. Int Med, 2010; 49: 865–69
23. Sakr L, Palaniappan R, Payan MJ et al: Tracheal glomus tumor: a multidisciplinary approach to management. Respir Care, 2011; 56: 342–46
24. Mogi A, Kosaka T, Yamaki E et al: Successful resection of a glomus tumor of the trachea. Gen Thorac Cardiovasc Surg, 2011; 59: 815–18
25. Okereke IC, Sheski FD, Cummings OW: Glomus tumor of the trachea. J Thorac Oncol, 2011; 6: 1290–91
26. Fan M, Liu C, Mei J et al: A rare large tracheal glomus tumor with postoperative haematemeses. J Thorac Dis, 2013; 5: E185–88
27. Choi IH, Song DH, Kim J, Han J: Two cases of glomus tumor arising in large airway: well organized radiologic, macroscopic and microscopic findings. Tuberc Respir Dis, 2014; 76: 34–37
28. Xiong W, Cai CL, Zhou YZ et al: Tracheal glomus tumor: two cases with bronchoscopic intervention. Chin Med J, 2014; 127: 189–90
29. Masson P: Le [The glomangiomyoma of the tactile regions and their tumors]. Lyon Chir, 1924; 21: 257–80 [in French]
30. Folpe AL: Glomus tumors. In: Fletcher CDM, Unni KK, Mertens F (eds.), World Health Organization classification of tumors: pathology and genetics of tumours of soft tissue and bone. Lyon (France): IARC Press, 2002; 136–37
31. Wu MH: Spiral tracheoplasty after tangential resection of the trachea. Ann Thorac Surg, 2009; 88: 2042–43

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