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Endobronchial Schwannoma: Literature Review in Account of a Case Report

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Abstract: Background: Schwannomas are benign mesenchymal tumor originated in the Schwan cells of peripheral, spinal, or cranial nerve sheaths. They represent approximately 0.2-2% of benign tumors in the tracheobronchial tree. To date, only sporadic case reports exist in western literature. Case presentation: We present a 35-year-old female, whose initial complaints were dyspnea, persistent cough, and chest pain for two months. In a chest CT-scan, a solid tumor was identified in the juncture of the right main bronchus and the emergence of the ipsilateral superior bronchus. Said tumor had both an intra and extra-luminal component, and its dimensions where 2.9×2.6×2.7cm. Results: A right posterolateral thoracotomy was performed with selective intubation. The possibility of preserving the right upper lobe was identified transoperatively, with the right upper bronchus being sutured to the right main bronchus, without complications. The histopathologic report was of a benign bronchial Schwannoma, with the formation of Verocay bodies in the HE stain, and intense positivity for the S100 protein in the subepithelial lesion. Patient follow-up was performed with a new CT-scan and flexible bronchoscopy 3 months after surgery, with no evidence of recurrence. 23 cases of endobronchial Schwannoma reported in the available literature were subsequently analyzed, with emphasis on clinical presentation, treatment offered and follow-up. Conclusions: Endobronchial Schwannomas are an uncommon type of airway tumor, and are rarely thought of as an initial diagnosis. Total resection of the tumor is of upmost importance to avoid recurrence. Surgical resections should be considered in all patients with high probability of residual tumor through endoscopic resection, most notably those with a known or suspected extraluminal component. More case reports are needed to develop a better understanding of the best diagnostic, treatment, and follow-up options.

Keywords: Endobronchial Schwannomas, Lung Tumors, Endoscopic Resections

1. Introduction

Schwannomas, also known as neurilemmomas, are benign mesenchymal tumor originated in the Schwan cells of peripheral, spinal or cranial nerve sheaths [1, 3, 7]. They may present themselves in any part of the body; however, they are most often found in the extremities, cerebellopontine angle, posterior mediastinum, retroperitoneum, and spinal roots [1, 3]. Neurogenic tumors represent approximately 15-25% of all tumors in the posterior mediastinum, with Schwannomas representing less than half of those [2, 11]. They tend to be associated the type 2 neurofibromatosis, with presentation in patients without said diagnosis also being possible [1]. These endobronchial tumors represent approximately 0.2% [2, 6] of all lung tumors, and 2% of benign tumor of the trachea-bronchial tree [4, 9]. Most present themselves as single tumors, but up to 25% of patients may present themselves with 2 or more endobronchial tumors [7]. To date, only sporadic case reports exist in western literature. The biggest case report series are from Takeda in Japan [2], who first reported on 37 cases in 2004 [11] and later reported on another 12 cases in 2007 [16].

Endobronchial Schwannomas were classified by Kasahara in central when they were in the trachea or proximal bronchus, and peripheral when they could not be located through bronchoscopy [2, 15]. Central Schwannomas were subdivided in those that have only an intraluminal component or mixed when they had both intra and extraluminal components.

Endobronchial Schwannomas are most often found in the third decade of life, with no sex predilection [1, 2]. The clinical presentation and evolution of these tumors depends on their size, location, and degree of bronchial obstruction produced [1, 17]. There is no specific clinical presentation, with most patients developing unspecific symptoms secondary to airway obstruction [2]. They are often asymptomatic, being incidentally discovered in an imaging study performed for other indications [3, 8]. The most often described symptoms are dyspnea and hemoptysis, with post-obstructive pneumonia, repetitive respiratory tract infections, fever, and productive or dry cough frequently portrayed as well [1]. Yoon Yang and cols. identified a greater probability of developing dyspnea in patients with more central tumors, as well as a higher likelihood of the patient being asymptomatic when possessing a peripheral tumor [4].

Radiologic imaging may unveil the presence of a solid, homogenous intrabronchial mass, often rounded or lobulated. Parenchymal consolidations or atelectasis may be present when the tumor produces partial or total endoluminal obstruction [1, 6]. There are no specific radiologic signs capable of differentiating the type of tumor. Among the most often cited differential diagnosis are both benign and malignant tumors, including hamartomas, carcinoid tumors, fibrous histiocytomas, fibrous polyps, papilloma, leiomyomas, neurofibromas, neurosarcomas, adenoid cystic carcinoma, squamous cell carcinoma, and inflammatory pseudotumor [1-4, 16]. There are also reports of patients with multiple endobronchial Schwannomas [5].

Bronchoscopy plays a vital role in this entity, being capable of not only identifying the location, size, and degree of bronchial obstruction of the tumor, but also giving the opportunity to perform proximal tumor biopsies, as well as partial or total resection of the tumor [1].

The confirmatory way of diagnosing endobronchial Schwannoma is through histopathology, with the classic presence of Antoni A type formation, as well as Verocay bodies in the Hematoxylin-Eosin (HE) stain. It is also characteristic to find positivity for protein S100 in immunoperoxidase [1, 4, 15]. Leiomyoma or inflammatory myofibroblastic tumor may be suspected when the typical morphology for Schwannoma is not present; however, the first tends to be negative for protein S100, while the second is usually associated with a dense inflammatory infiltrate [6].

Treatment for all endobronchial Schwannomas is resection. The main objective is the complete excision of the tumor to avoid possible recurrences. This might be achieved through bronchoscopy or surgery (both open or VATS). Endoscopic resection is most feasible for small tumors, and is generally performed with the aid of electroresection, cryoresection, argon-plasma coagulation, aluminum laser resection, among other technologies [2, 5, 7, 20]. Lobectomy is considered the standard surgical procedure in patients who can tolerate it [2]. Sleeve lobectomy and bronchoplasty can also be considered [1]. In peripheral tumors, resection of the surrounding lung parenchyma is recommended to avoid recurrence. It is challenging to compare the results of different resecting procedures due to the limited information of this pathology [2, 4].

There are those, however, who promote conservative management and close monitoring in asymptomatic patients [1, 4]. While malignant transformation of Schwannomas is rare, follow-up with or without resection of the tumors is recommended to detect recurrence in the former and possible growth of the tumor in the latter [1, 15]. There are currently no stablished follow-up protocols, and the risk of malignant transformation is still unknown [5].

2. Case Report

We present a 35-year-old female, whose initial complaint was dyspnea, persistent cough, and chest pain for the past two months. In a computed tomography of the chest, a solid tumor was identified in the juncture of the right main bronchus and the emergence of the ipsilateral superior bronchus. It was identified that said tumor had both an intra and extra-luminal component, and its dimensions where 2.9×2.6×2.7cm.

3. Results

Flexible bronchoscopy with tumor biopsy was performed, with a histopathologic report of a mesenchymal malignant tumor. Later, an attempt to resect the tumor via cryoresection through bronchoscopy was performed, with only partial

success. The excised fragment was sent to pathology, who reported a malignant neurofibroma.

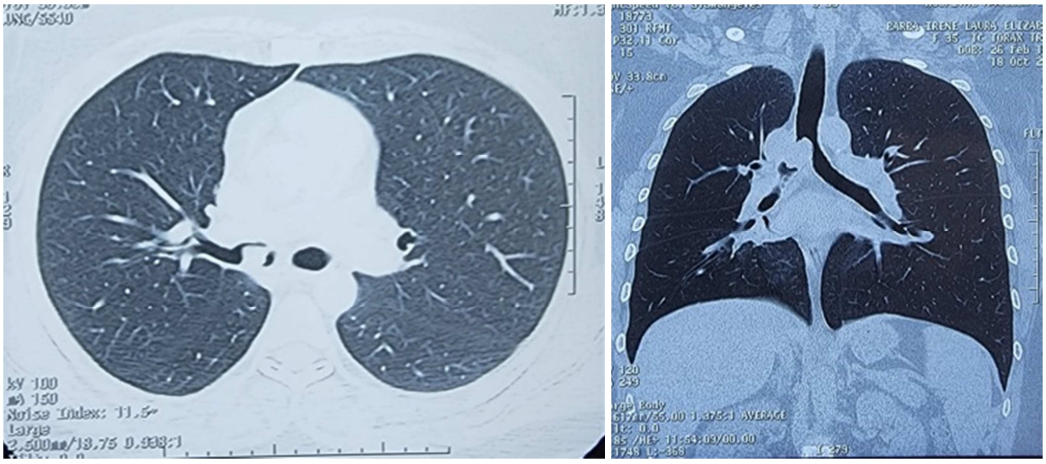

Figure 1. *Axial (a) and coronal (b) CT-scan in lung-parenchyma window, demonstrating a tumor in the right main bronchus.*

Seeking a therapeutic alternative, the patient was sent to the authors for medical assessment, deciding the was a good candidate for surgical tumor resection most likely with a sleeve right upper lobectomy. A right posterolateral thoracotomy was performed with selective intubation. Trans operatively, the extraluminal component was confirmed in the posterior angle of the emergence of the right upper bronchus. After a margin-free bronchial resection of the tumor was performed, the possibility of preserving the right upper lobe was identified. The right upper bronchus was then sutured to the right main bronchus, with no complications. The patient was successfully extubated after surgery without the need of intensive care, and with no evidence of air leakage through the chest tube. The chest drain was removed on the third postoperative day, and the patient was discharged the following day without incidents.

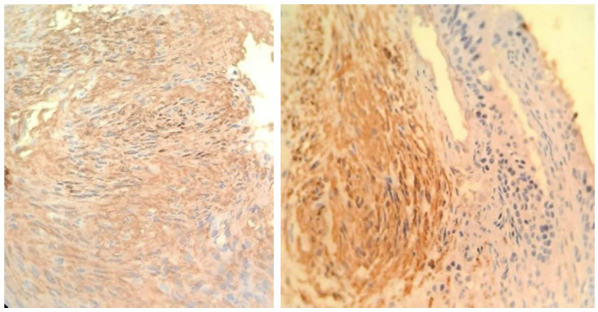

Figure 2. *Hematoxylin-Eosin stain in 40x, demonstrating palisading spindle cells forming Verocay bodies.*

The histopathologic report was of a benign bronchial Schwannoma, with the formation of Verocay bodies in the HE stain, and intense positivity for the S100 protein in the subepithelial lesion. Patient follow-up was performed with a new CT-scan and flexible bronchoscopy 3 months after surgery, with no evidence of recurrence. The only outstanding finding was a shortening of the right main bronchus and an altered angle in the emergence of the right upper bronchus, consistent with the surgical procedure. A one-year follow-up was performed, with the patient being asymptomatic and no evidence of tumor recurrence.

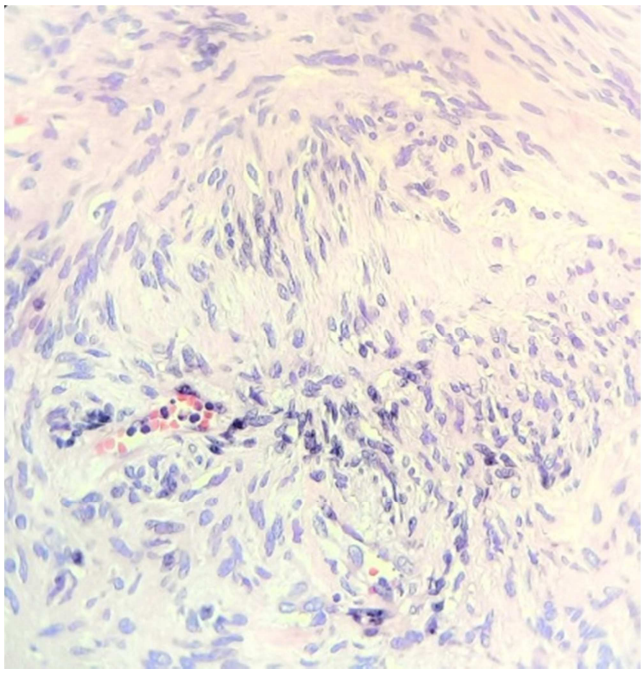

Figure 3. *Intense positivity for S100 protein in the subepithelial lesion.*

4. Discussion

In the cited literature [1-14, 17-19], twenty-three cases of endobronchial Schwannomas were reported between 2010 and 2021. Most where simple case reports, while two [4, 18] where small case series of 7 and 2 endobronchial Schwannomas, respectively. The described ages ranged from 8 to 81 years old, with an average of 48.2. It was found, as well, that most patients (14, 60.8%) were over 45 years of age, while 8 (34.7%) were 60 years old or older. This is in contrast with the previous reports, where it is often cited that most endobronchial Schwannomas present themselves during the third decade of life. Thirteen patients were female (56.52%), and no patient was known to have

neurofibromatosis.

Among comorbidities, three patients with history of extra thoracic malignancy stand out, as in all three cases a metastatic tumor was initially suspected. These previous malignancies where breast cancer, thyroid cancer and colorectal cancer. The span of symptom development at presentation was wide, ranging from 2 days to 2 years. It is also important to note that two of the reported cases where in asymptomatic patients, while in 3 cases no symptom temporality was reported. Eleven patients (47.8%) sought medical attention due to their symptoms within the first 3 months of presentation.

The most often reported symptoms where dyspnea (12, 52.17%), cough (11, 47.83%) and hemoptysis (4, 17.39%). From the total of reported cases, 8 patients (34.78%) had concomitant pneumonia at the time of diagnosis, while only 3 (13.4%) developed a fever. Preoperative flexible bronchoscopy was performed in all patients. Prior to bronchoscopy, the most considered differential diagnosis were metastatic tumor in 3 patients, carcinoid tumor in 3 patients, pneumonia in 2 patients, and inflammatory pseudotumor in 1 patient.

AGE	SEX	EVOLUTION	SYMPTOMS	SIZE IN CM	LOCALIZATION	PROCEDURE
60	M	3 MONTHS	DYSPNEA, CHEST PAIN	6	RMB	RU LOBECTOMY
18	F	5-7 DAYS	DYSPNEA, COUGH PNEUMONIA	2.1	LUB	LU LOBECTOMY
42	F	1 MES	HEMOPTYSIS	4.3	LLB	LL SLEEVE LOBECTOMY
81	F	19 MONTHS	DYSPNEA	4	LMB	RIGID BRONCHOSCOPY
52	F	9 MONTHS	DYSPNEA, HEMOPTYSIS	3	C	RIGID BRONCHOSCOPY
21	F	1 MES	COUGH, FEVER, PNEUMONIA	2.1	LMB	FLEXIBLE BRONCHOSCOPY
50	F	4 MONTHS	COUGH, DYSPNEA	2.8	C, RMB	RIGID BRONCHOSCOPY
58	F	2 MONTHS	CHEST PAIN, PNEUMONIA	1.5	BINT	FLEXIBLE BRONCHOSCOPY
16	M	ND	ASYMPTOMATIC	2.3	LS8	LL LOBECTOMY
36	M	7 MONTHS	DYSPNEA	3	LMB	FLEXIBLE BRONCHOSCOPY
64	M	4 MONTHS	DYSPNEA, PERSISTENT ATELECTASIA, PNEUMONIA	ND	C; MB; RS7	FLEXIBLE BRONCHOSCOPY
66	F	2 MONTHS	COUGH, HEMOPTYSIS	3.7	LMB	LU LOBECTOMY
60	F	2 DAYS	FEVER, COUGH, PNEUMONIA	2	LUB	LU LOBECTOMY
80	M	3 DAYS	FEVER, COUGH, PNEUMONIA	1	LMB; INTB	FLEXIBLE BRONCHOSCOPY
69	M	6 MONTHS	DYSPNEA	3	C	FLEXIBLE BRONCHOSCOPY, CARINECTOMY
51	F	2 MONTHS	COUGH, DYSPNEA	ND	C, RMB	FLEXIBLE BRONCHOSCOPY
44	F	ND	ASYMPTOMATIC	1.9	LMB	RIGID BRONCHOSCOPY
26	M	3 WEEKS	HEMOPTYSIS, COUGH, DYSPNEA	ND	C, LMB	RIGID BRONCHOSCOPY
11	M	3 MONTHS	COUGH, BRONQUITIS, PNEUMONIA	1	LMB	FLEXIBLE BRONCHOSCOPY
64	M	ND	DYSPNEA	2	T	RIGID BRONCHOSCOPY, TRACHEOPLASTY
8	F	2 YEARS	COUGH, PNEUMONIA	2	LMB	PNEUMONECTOMY
67	M	ND	COUGH	ND	LUB	FLEXIBLE BRONCHOSCOPY
66	F	ND	DYSPNEA	ND	LMB	FLEXIBLE BRONCHOSCOPY

Figure 4. *Patient distribution. ND: not disclosed, RMB: right main bronchus, LUB: left upper bronchus, LLB: left lower bronchus, LMB: left main bronchus, C: carina, INTB: intermediary bronchus, LS8: left segment 8, ML: middle lobe, RS7: right segment 7, T: trachea, RU: right upper, LU: left upper, LL: left lower.*

The reported size of endobronchial Schwannomas, hereby described in centimeters by their largest diameter, ranged from 1 to 6cm, with an average of 2.65cm. Two patients had more than one synchronic Schwannoma, in whom their largest tumor was taken into account. It is noteworthy that in 5 cases, no tumor size was reported. Most tumors had a central localization (24, 92.3%). Of all described tumors, 4 were found in the trachea, 7 in the right bronchial tree, and 15 in the left bronchial tree. The most frequently identified location for endobronchial Schwannomas was in the left main bronchus, accounting for 43.48% (10) of all cases.

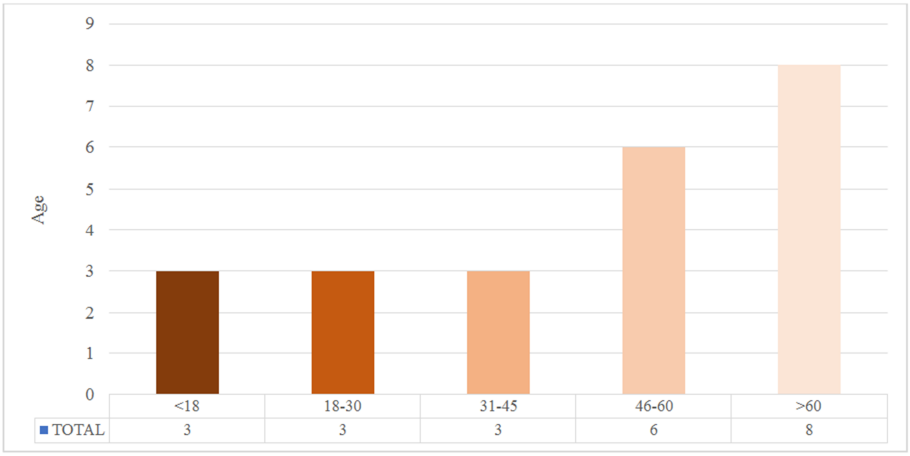

Figure 5. *Age distribution among cited patients with endobronchial Schwannoma.*

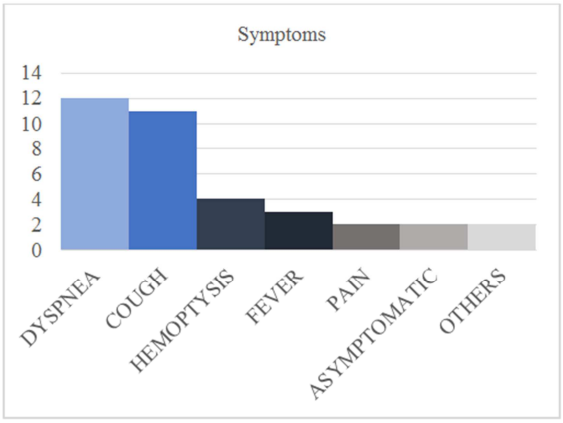

Figure 6. *Most prevalent reported symptoms at presentation.*

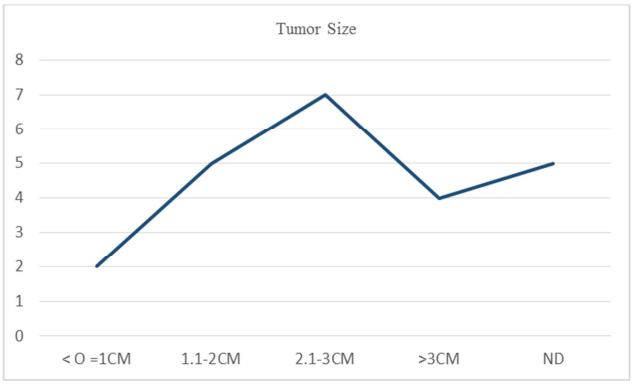

Figure 7. *Reported tumor size in centimeters. ND: not disclosed.*

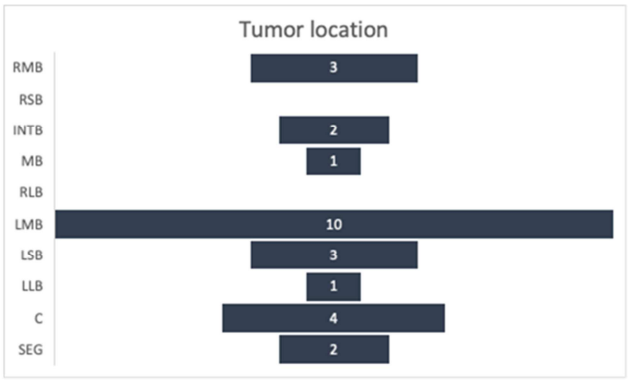

Figure 8. *Tumor location in reported cases.*

RMB: right main bronchus, RSB: right superior bronchus, INTB: intermediary bronchus, MB: middle bronchus, RLB: right lower bronchus, LMB: left main bronchus, LSB: left superior bronchus, LLB: left lower bronchus, C: carina, SEG: segmentary bronchus.

The most often performed resective procedure was flexible bronchoscopy in 10 cases (43.48%), often with the aid of electrocoagulation or argon-plasma coagulations. Including rigid bronchoscopy, endoscopic management of the tumor was performed in 16 (69.56%) patients. Of those, two procedures were used only for debulking and airway permeabilization prior to tracheal surgery. Simple lobectomy was performed in 5 (21.74%) patients, while one patient underwent a sleeve lobectomy, while another required pneumonectomy due to parenchymal destruction secondary to chronic obstruction.

In all 26 tumors of the 23 patients, histopathologic diagnosis of Schwannoma was confirmed. All cases had positivity for protein S100.

Postoperative follow-up was reported in only 9 (39.13%) of cases. Of those, 7 had a follow-up no longer than six months, while one patient was monitored for a year and another for 4 years. Only 3 recurrences were reported, however lack of recurrence was reported in only 8 patients, while in 12 patients there was no report of presence or absence of recurrence. Of the reported recurrences, all were in patients with history of endoscopic resection, 1 through rigid and 2 through flexible bronchoscopy. Of those 3, one patient required a lobectomy, while bronchoscopic resection was performed in the other two cases. It is worth noting that of these two patients, one was advised to undergo a lobectomy from start, but refused the procedure, so repeated bronchoscopic resections were performed. No malignant transformations were reported.

5. Conclusions

Endobronchial Schwannomas are an uncommon type of airway tumor, and are rarely thought of as an initial diagnosis. Total resection of the tumor is of upmost importance to avoid recurrence. Although bronchoscopic resections have proven to be effective, lack of follow-up and recurrence information prevents it from becoming the surgical standard. Surgical resections should be considered in all patients with high probability of residual tumor through endoscopic resection, most notably those with a known or suspected extraluminal component. More case reports are needed to develop a better understanding of the best diagnostic, treatment and follow-up options.

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