CASE REPORT

RARE SURGICAL TREATMENT OF "ASTHMA" WITH POST-PARTUM CARDIOMYOPATHY: CASE REPORT AND REVIEW OF LITERATURE

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Abstract

We report the case of a 37-year-old woman who presented with severe persistent "asthma" requiring repeated courses of steroids and bronchodilators. Spirometry was suggestive of upper airway obstruction. Computerized tomography scan (CT) demonstrated a polypoid mass in the cervical trachea obstructing 90% of the lumen. Urgent tracheostomy was performed followed by bronchscopic excision of the mass. Histopathological diagnosis of schwannoma was confirmed using immunohistochemical markers. We found only 20 primary tracheal schwannoma cases previously reported in the English literature.

Keywords: ♦ Upper Airway obstruction ♦ Tracheal tumor ♦ Schwannoma, ♦ Neurilemoma.

Case presentation

A 37-year-old female presented to the emergency room with two days history of dry cough, shortness of breath, orthopnea and wheezing. The patient had similar symptoms for about one year earlier and was diagnosed to have bronchial asthma. She was started on inhaled bronchodilators and steroids. Her symptoms persisted despite being compliant with this therapy and required frequent emergency room visits for presumed exacerbations of asthma. She had hypertension and diabetes for six years. There was no history of childhood asthma, eczema, or hay fever and no history of smoking. She had no family history of asthma.

On physical examination she was tachypneic (30/min) and tachycardic (140/min). There was decreased air entry bilaterally with inspiratory and expiratory wheezing over both lung fields and trachea. Arterial blood gases (ABG) on room air showed: pH 7.36, pCO2 55mmHg, pO2 60 mm Hg, Bicarbonate 30 and O2 Saturation 89%. Echocardiography revealed moderate impairment of left ventricular systolic function (ejection fraction 43%), and right ventricular systolic pressure (RVSP) of 40mmHg. These changes were attributed to post-partum cardiomyopathy.

The patient was treated with IV steroids, antibiotics, inhaled salbutamol, ipratropium and oxygen. She did not improve. Spirometry suggested fixed upper airway obstruction. CT scan of the neck demonstrated a soft tissue mass within the trachea at the level of C6-C7 measuring about 1.5 x 1.5 cm, causing almost 90% obstruction of the tracheal lumen (Fig. 1).

The patient developed increasing respiratory distress and urgent tracheostomy was performed. Rigid bronchoscopy showed a large, pale polypoid mass just below the vocal cords, protruding into the trachea and almost completely obstructing it. Punch biopsy of the lesion revealed non-specific acute and chronic inflammation with no evidence of malignancy. The tracheal mass was removed piecemeal using biopsy forceps. Histopathology revealed

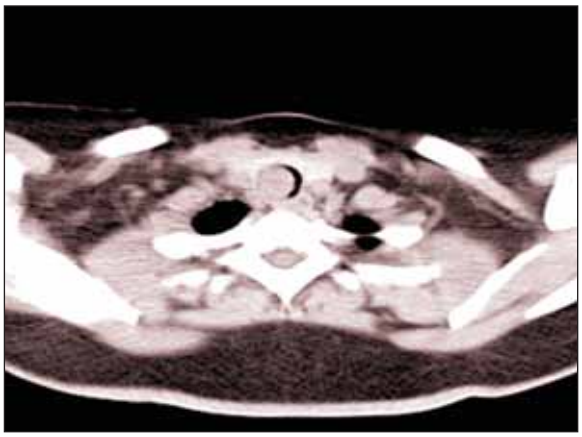

Fig. 1: *CT of the chest in cross section showing a large, rounded, smooth mass in the trachea attached to the lateral wall and severely narrowing the tracheal lumen.*

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spindle cells with wavy nuclei, verruca bodies and hyalinized blood vessels. Immunoperoxidase stains were positive with S100, and negative with SMA, AE1/AE3, Desmin, CD31, CD34 and Chromogranin, consistent with the diagnosis of schwannoma (Fig. 2A, 2B).

Repeated CT scan showed presence of residual tumor. Bronchoscopy was repeated to complete the excision of the mass. Her symptoms improved leading to rapid reduction of asthma medications. Tracheostomy tube was removed, and the patient was discharged. Four weeks post-operatively, her symptoms were completely resolved without any medications. On follow up visits up to 2 years, the patient is still asymptomatic, with no evidence of recurrence as confirmed by CT scan and spirometry. Echocardiography showed improvement in left ventricular function (50%) with resolution of tachycardia, but RVSP remained elevated (42 mmHg).

Discussion and review of literature

MEDLINE search for published reports in the English literature of primary tracheal schwannoma from 1950 to 2008 identified 20 such cases (Table-1)[1-19]. Including this case, the location of the tumor was: nine in lower, nine in upper, and three in middle trachea (Table 1). The age at presentation varied from 9 to 77 years. Twelve of these cases occurred in women (57%). The delay of diagnosis varied from 1 month to 7 years. There was no reported death. There was only one case of recurrence after 12 years after endoscopic resection[6].

Primary tumors of the trachea are rare, accounting for less than 0.1% of all tracheal tumors. Laryngeal schwannomas was described by Suchaneck in 1925[1]. Strauss and Guckien described tracheal schwannoma in 1951[1]. Strot described primary lung schwannoma in 1946 and named it schwannoma, attributing the histogenesis to schwann cells of the neural sheath[20].

Schwannomas are benign encapsulated neurogenic tumors, arising from schwann cells of the peripheral and cranial nerve sheaths. In about 50% of the times, this tumor occurs in the head and neck region[3]. It is almost always solitary and benign[1]. Malignant schwannoma usually shows histological infiltration, hypocellularity, high mitotic activity, tumor necrosis and focal or minimal expression of S-100 protein[21].

The presenting symptoms are usually chronic dry cough, and progressive exertional dyspnea that are not relieved with medication. Characteristic for this condition is dyspnea and cough that become worse on lying supine or on bending forward probably due to compression of the trachea by the tumor. Wheezing is usually generalized, and the presence of stridor may help in the diagnosis. Clinically it is often misdiagnosed as asthma, and the diagnosis is usually delayed for an average of 10 to 15 months.

Spirometry may show fixed obstructive airflow with no improvement after bronchodilators. Chest radiography is helpful especially if the tumor is in the upper trachea. CT scan is used to delineate tumor size, location, and extension. The use of multislice computerized tomography, Multiplanar and 3D images may be useful in the evaluation of the tracheobronchial tree. Histopathologial diagnosis is usually made after bronchoscopy and biopsy. Rigid bronchoscopy is

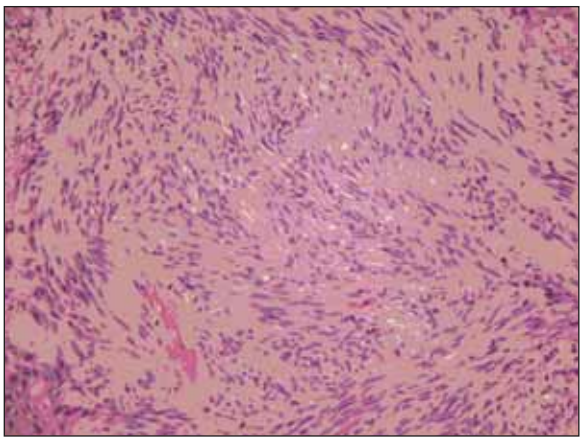

Fig.2A: *Microscopic view of the tumor showing fascicles of spindle cells with palisade of the nuclei (verruca bodies). (Hematoxylin & Eosin x 200).*

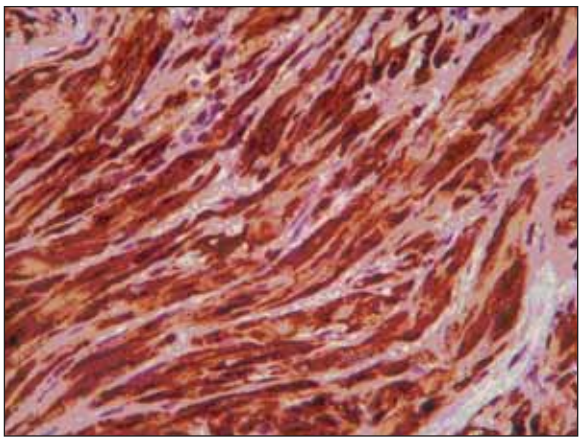

Fig.2B: *Immunoperoxidase stain with S-100 protein highlighting the neural sheath nature of the tumor (Immunoperoxidase x 400).*

Table 1: Reported cases of primary tracheal schwannoma[1-19]:

Ref no	Age	Sex	Site	Symptoms	Duration	Therapy	Follow up
1. Straus GD 1951	28	M	L	Cough, Fever, Pneumonia	12 M	Two stage endoscopy, silver nitrate cautery	Asymptomatic, 6 months
2. Karlan MS 1973	21	M	U	Difficult breathing, Cough, Pneumonia		Tracheal fissure resection	Asymptomatic, 1 year
3. Conley J 1975	28	M	U	Cough, Difficult breathing		Surgical resection	Lost Follow up
4. Nass RL 1979	36	M	L	Dyspnea, Hemoptysis, Wheezing	4 M	Endoscopy with electrocoagulation	Asymptomatic, 4 years
5. Ma CK 1981	23	F	U	Difficult breathing, Wheezing, Dyspnea		Tracheal Sleeve resection	Asymptomatic, 1 year
6. Horovitz AG 1983	38	F	L	Difficult breathing, Cough, Dyspnea	3 M	Endoscopic excision 1970, tracheal resection 1982	Tumor reoccur 12 years later, now asymptomatic
7. Ikezone J 1986	54	M	M	Mild Dyspnea on exertion	7 M	Tracheal sleeve resection	NA
8. Robin J 1988	35	F	U	Asthma, Dyspnea, Productive cough	2 Yr	Endoscopic resection	NA
9. Pang LC 1989	57	F	L	Dyspnea, Cough, Wheezing	3 M	Tracheal sleeve resection	Asymptomatic, 7 years
10. Pang LC 1989	34	F	L	Dyspnea,, Cough, Wheezing, Stridor	3 M	Tracheal resection	Asymptomatic, 6 months
11. Inoue H 1989	63	F	L	Dyspnea, Cough	8 M	Tracheal sleeve resection	Asymptomatic, 2 years
12. Stack PS 1990	23	F	U	Dyspnea	6 M	NA	NA
13. Rusch V W 1994	45	M	U	Dyspnea, Cough	6 M	Nd/YAG Laser	Asymptomatic, one year
14 Weiner DJ 1998	16	M	L	Asthma, Dyspnea	11 M	CO2 Laser	NA
15 Dorfman J 2000	33	M	L	Dyspnea	6 M	Surgical resection	Asymptomatic, 3 months
16 Erdogan S 2004	19	M	U	Dyspnea, Asthma	4 M	Tracheostomy then surgical resection	Asymptomatic, 1 year
17 Nio M 2005	9	F	M	Dyspnea, Cough,	1 M	Surgical resection	Asymptomatic, 4 months
18 Righini CA 2005	51	F	U	Dyspnea, Cough	12 M	Surgical resection	Asymptomatic, 3 years
19 Dincer SI 2006	49	F	L	Dyspnea	4 yr	Surgical resection	Asymptomatic, 1 month
20 Erol MM 2008	77	F	M	Dyspnea, Cough	7 yr	Polypectomy	Asymptomatic, 7 months
21 Current case 2008	37	F	U	Dyspnea, Asthma	3 M	Tracheostomy, then endoscopic resection	Asymptomatic, 21 months

preferred to obtain adequate biopsy, assess the extent of the tumor, secure the obstructed airways and control bleeding that may occur with biopsy. However flexible bronchoscopy can be safe and successful in some cases. Precise localization of the tumor and its relationship to the trachea and main bronchi are helpful in planning the surgical resection[21-23].

The management of schwannoma in published reports included endoscopic resection, laser endoscopic resection using Nd/YAG, or CO2 laser, and segmental tracheal resection (Table-1). Although initial endoscopic excision is successful, delayed local recurrence was reported up to 12 years after initial resection, suggesting that patients treated in this manner should be kept under long term surveillance[6].

Summary

Upper airway obstruction should be considered in all patients with chronic cough, dyspnea, or asthma, especially when the response to treatment is suboptimal. Schwannoma is one of the rare tumors which can present as upper airway obstruction. Spirometry, CT scan and bronchoscopy are helpful in the diagnosis. The therapeutic approach is influenced by the clinical presentation, the type of the tumor (pedunculated or sessile), the risk of tracheal resection, and the presence or absence of an extratracheal component. Trahceal schwannoma can have delayed recurrence after bronchoscopic resection, and continued long term follow-up is needed in such patients. ♦

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