ELSEVIER

European Journal of Cardio-thoracic Surgery 16 (1999) 464–468

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

www.elsevier.com/locate/ejcts

Case report

Fibrohistiocytic tumor of the trachea in a child ☆

Sibel Öztürk[a], Hakan Kutlay[b], Serpil D. Sak[a,*], Şinasi Yavuzer[b]

[a]*Department of Pathology, Ankara University School of Medicine, Ankara, Turkey*
[b]*Department of Thoracic Surgery, Ankara University School of Medicine, Ankara, Turkey*

Received 26 January 1999; received in revised form 4 July 1999; accepted 28 July 1999

Abstract

A 10-year-old girl who had been treated for presumed asthma during the previous year, was admitted to our hospital with dyspnea and wheezing. An endotracheal tumor was detected and a tracheal resection was performed. The tumor was diagnosed as a fibrohistiocytic tumor. Biologic behavior of fibrohistiocytic tumors of the trachea is not clear and morphologically similar cases are interpreted as malignant or benign by different authors. However, an analysis of the present case and 11 well documented cases in the literature shows that this tumor must be evaluated as a low-grade malignancy and initial complete tumor resection is necessary. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Trachea; Fibrohistiocytic tumors

1. Introduction

Primary fibrohistiocytic tumors of the trachea are very rare neoplasms. We describe here a case diagnosed in a 10-year-old female, who presented with asthma-like symptoms. To our knowledge, the present case is the 16th reported case of fibrohistiocytic tumor (FHT) of the trachea [1–10]. Due to the limited number of cases in the literature, biological behavior of these tumors is not clear and histologically similar cases are diagnosed as benign [1–4] or malignant [1,7–9] by different authors. This paper presents the clinical and histopathological features of the present case and compares the characteristics of other cases in the literature in an attempt to clarify this issue.

2. Case report

A 10-year-old white female was admitted to our hospital in March, 1997 with progressive exertional dyspnea and wheezing unrelieved by outpatient management for presumed asthma. The patient did not have a history of asthma in childhood but she had been treated for asthma-like symptoms during the past year. Physical examination was remarkable for marked inspiratory and expiratory stridor, wheezing, tachypnea and orthopnea. A CT scan of the neck and chest showed an endotracheal mass of 2 cm in diameter. MRI showed that the tumor was 20 ± 2 mm, and it was occluding 80–90% of the lumen with a crescentic luminal space left, on the right lateral wall.

The patient underwent bronchoscopy that showed a polypoid mass almost completely obstructing the tracheal lumen. The bronchoscope could not be passed beyond the tumor and an adequate airway could not be maintained. In the mean time end tidal pCO_2 increased to 120 mmHg. A right posterolateral thoracotomy was performed without delay and the trachea was divided 1 cm distal to tumor. Ventilation was provided via an endotracheal tube placed in the left main bronchus. A tracheal segment of 3.6 cm was resected and tracheal reconstruction was performed by 3/0 vicryl using interrupted sutures. Prior to anastomosis, inferior pulmonary ligament, right hilum and left main bronchus were released to decrease anastomotic tension. The patient did well postoperatively, was discharged on the 12th postoperative day without any complications and received 5000 rads 1 month after the operation. In the last follow-up, which was 17 months after the operation, there was no stenosis or tumor recurrence.

Upon pathologic examination, the specimen was 3 cm long and consisted of five tracheal rings. There was an irre-

☆ Presented at the 13th Annual Meeting of the Turkish Congress of Pathology, September 6, Istanbul, Turkey, 1997.

* Corresponding author. Ankara Üniversitesi Tıp Fakültesi Patoloji ABD, Morfoloji Binası 06339 Sıhhiye, Ankara, Turkey. Fax: +90-312-310-6370.

E-mail address: sak@dialup.ankara.edu.tr (S.D. Sak)

1010-7940/99/$ - see front matter © 1999 Elsevier Science B.V. All rights reserved.
PII: S1010-7940(99)00288-2

Downloaded from https://academic.oup.com/ejcts/article/16/4/464/404189 by Ankara University Library (ANK) user on 11 December 2021

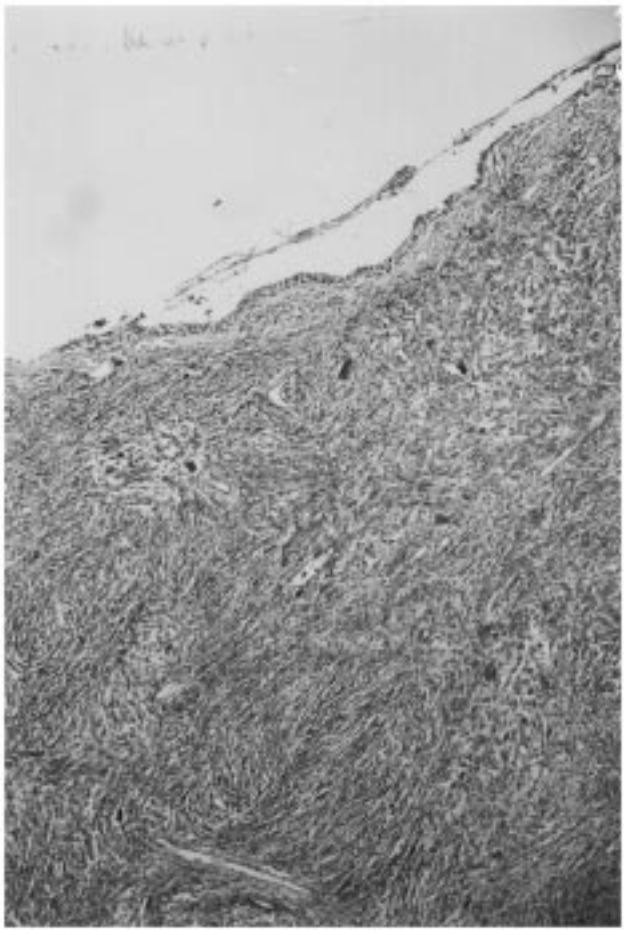


Fig. 1. Beneath the epithelium, spindle cells forming interlacing fascicles and storiform pattern with occasional multinucleated giant cells (hematoxylin and eosin × 40).

gularly surfaced, yellow-pink, focally hemorrhagic tumor projecting into the lumen which measured 2.0 × 1.5 × 1.4 cm. Microscopically a neoplasm continuous with proximal margin of excision was observed which invaded a tracheal segment of 2 cm in length and destroyed the tracheal cartilage but did not infiltrate the adventitia. The tumor projected in a polypoid fashion into the tracheal lumen and it showed acute and chronic inflammation with granulation tissue and squamous metaplasia. The neoplasm was composed of spindle or polygonal cells with mild to moderate degree of nuclear pleomorphism. Spindle cells formed interlacing fascicles which in some areas had a storiform arrangement and two mitotic figures were seen per 50 high power fields. There was also a population of multinucleated giant cells and lipid-laden macrophages (Fig. 1). A paratracheal lymph node was negative for tumor. Immunohistochemically, spindle cells were positive for vimentin and negative for desmin, S-100 and cytokeratin. Multinucleated giant cells and histiocytes were both vimentin and CD-68 positive. We interpreted the case as fibrohistiocytic tumor on the basis of the aforementioned findings. It was difficult to comment on the possible biological behavior of the tumor. There was no definite feature of evident malignancy but destruction of tracheal cartilage, presence of mild to moderate pleomorphism and mitotic activity, albeit rare, prevented us to diagnose the case as 'benign' fibrous histiocytoma.

3. Discussion

Primary tracheal tumors are very rare neoplasms, most of them being epithelial in origin. Primary fibrohistiocytic tumors of trachea are even rarer, only 15 cases have been reported in the literature [1–10] previously and only 11 of them are well documented with clinical and histological characteristics (Table 1).

As one examines the histological descriptions of the cases in the literature, it is striking to observe that apparently very similar tumors are defined as benign fibrous histiocytoma [1–4] or malignant fibrous histiocytoma [1,7–9] by different authors. One case was diagnosed as invasive fibrous tumor of the tracheobronchial tree [6]. In all cases, spindle cells forming fascicles were seen, usually with a focal or prominent storiform pattern. In half of the tumors, multinucleated giant cells were observed [1,2,4–7]. Although there is no numerical information about mitosis in the descriptions of the reported cases, nuclear pleomorphism and mitoses were not prominent except for the cases of Louie and Wang [5,8]. The histological criteria separating malignant from benign lesions are not well established and assessment of the behavior of FHTs of the trachea on the basis of their histology is difficult [1]. Although the follow-up of the tracheal FHTs in the literature is limited, there was only one death in the 3rd month. This tumor was an unusually large tumor with prominent pleomorphism, frequent mitoses and tissue invasion in an elderly female [5]. All the other patients were free of disease in their last follow-up and none of the patients had a history of metastasis. Although FHT of the trachea is usually a polypoid tumor, it shows a close relationship with tracheal cartilage and endoscopic or incomplete excision seems to be insufficient in treatment of these patients because of the high risk of recurrence which is the case in four tumors [1,2,4,6].

In conclusion, the distinction between benign and malignant fibrous histiocytoma is not always easy and depends as much on the size and location of the tumor as it does on histological features. The majority of FHTs of the trachea show similar clinical and morphologic features suggestive of a low grade malignancy, nevertheless they are diagnosed either as malignant or benign by different authors. Awareness of this fact and initial complete resection may avoid recurrences which may require a more extensive resection than would have been possible initially.

Acknowledgements

We wish to thank Dr S. Han for his help.

Downloaded from https://academic.oup.com/ejcts/article/16/4/464/404189 by Ankara University Library (ANK) user on 11 December 2021

Table 1
Well documented cases of fibrohistiocytic tumors of the trachea: clinical and histological characteristics

Author	Age–sex	Tumor size (cm)	Presenting symptoms	Histology and diagnosis	Treatment and follow-up
Karlan et al. [1]	57–F	2.5 × 2.5 × 0.8	Intermittent hemoptysis 15 years inspiratory wheezing 3 months	Spindle cells, foamy histiocytes, whorled/storiform pattern –fibrous histiocytoma of the sclerosing hemangioma type	Endoscopic excision, LR[b] at 1 year; endoscopic re-excision, NED[a] at 2 years
Hakimi et al. [1]	26–M	2.0 × 1.5 × 1.5	Cough, hemoptysis 1 week Weakness, weight loss	Spindle-polygonal cells, osteoclastic giant cells, mild pleomorphism, tumor cells in one vascular space, extension to the external surface of the cartilage, no mitoses –benign fibrous histiocytoma	Tracheal resection, NED[a] at 27 months
Sandstrom et al. [2]	15–F	0.9 × 0.8 × 0.8	Cough, progressive exertional dyspnea, stridor 6 months	Spindle cells forming fascicles and storifom pattern, multinucleated giant cells, occasional mitoses, vascular invasion, invasion through tracheal cartilage, extensive replacement and destruction of thyroid –fibrous histiocytoma	Endoscopic excision, LR[b] at 2 months; bronchoscopic resection, 2nd LR[b] at 5 months; Tracheal resection, NED[a] at 12 months
Cohen et al. [3]	2–F	0.8 × 0.5 × 0.5	Fever, tachypnea, cough, dyspnea 5 months	Spindle cells, foamy macrophages, rare mitoses –xanthofibroma (fibrous histiocytoma)	Endoscopic excision (twice), NED[a] at 10 years
Gonzalez-Campora et al. [4]	16–M	Grain of rice (first) 1.5 × 1.0 × 0.5 (recurrence)	Severe obstructive dyspnea, hemoptysis 6 weeks	Spindle and polygonal cells, multinucleated cells, storiform pattern, no atypia, scanty mitoses, no cartilagenous or vascular invasion –benign fibrous histiocytoma	Local resection, LR[b] at 6 months; widened local resection, NED[a] at 15 months
Louie et al. [5]	77–F	? (> 10)	Dyspnea, intermittent stridor, enlarging right neck mass – 11 years after radiation to right neck and thyroid for papillary carcinoma	Spindle cells with prominent pleomorphism, frequent mitoses, many multinucleated giant cells –postirradiation malignant fibrous histiocytoma	Bronchoscopic laser therapy, Adriamycin, expired at 3 months
Tan-Lui et al. [6]	8–F	3.0 × 3.0	Wheezing few years	Spindle cells with moderate atypia, rare mitoses, multinucleated giant cells –invasive fibrous tumor of the tracheobronchial tree	Tracheal resection, LR[b] at 1 year; Carinal resection, NED[a] at 4 years

Downloaded from https://academic.oup.com/ejcts/article/16/4/464/404189 by Ankara University Library (ANK) user on 11 December 2021

Table 1 (*continued*)

Author	Age–sex	Tumor size (cm)	Presenting symptoms	Histology and diagnosis	Treatment and follow-up
Sculerati et al. [1]	17–F	? (2–3)	Wheezing, dyspnea, hemoptysis 3 weeks	Spindle cells, areas of storiform arrangement, some pleomorphism, occasional mitoses, no multinucleated giant cells –low grade malignant fibrous histiocytoma	Endoscopic debulking, tracheal resection, radiotheraphy, NED[a] at 20 months
Randleman et al. [7]	17–F	1.5 × 1.5 × 1.3	Dyspnea, wheezing 1 year	Spindle cells, scattered Touton giant cells, moderate pleomorphism, rare mitoses –low grade malignant fibrous histiocytoma	Tracheal resection, NED[a] at 14 months
Wang et al. [8]	58–F	2.5 × 0.6	Cough with scanty sputum, progressive exertional dyspnea 3 months	Spindle cells with severe nuclear pleomorphism, frequent mitoses –malignant fibrous histiocytoma	Tracheal resection, NED[a] at 30 months
Sennaroğlu et al. [9]	32–M	1.5 × 1.0	Progressive dyspnea 1 year Hemoptysis 6 months	Fibrohistiocytes with storiform pattern –malignant fibrous histiocytoma	Tumor resection and radiotherapy, NED[a] at 3 years
Present case	10–F	2.0 × 1.5 × 1.4	Exertional dyspnea, wheezing 1 year	Spindle cells with moderate pleomorphism, focal storiform organization, multinucleated giant cells, occasional mitoses –fibrohistiocytic tumor	Tracheal resection and radiotherapy, NED[a] 17 months

[b] LR, local recurrence.
[a] NED, no evidence of disease.

Downloaded from https://academic.oup.com/ejcts/article/16/4/464/404189 by Ankara University Library (ANK) user on 11 December 2021

References

[1] Sculerati N, Mittal KR, Greco MA, Ambrosino MM. Fibrous histiocytoma of the trachea: management of a rare cause of upper airway obstruction. Int J Pediatr Otorhinolaryngol 1990;19:295–301.
[2] Sandstrom RE, Proppe KH, Trelstad RL. Fibrous histiocytoma of the trachea. Am J Clin Pathol 1978;70:429–433.
[3] Cohen SR, Landing BH, Isaacs H. Fibrous histiocytoma of the trachea. Ann Otol Rhinol Laryngol Suppl 1978;82:2–4.
[4] Gonzalez-Campora R, Matilla A, Sanchez-Carrillo JJ, Navarro A, Galera H. 'Benign' fibrous histiocytoma of the trachea. J Laryngol Otol 1981;95:1287–1292.
[5] Louie S, Cross CE, Amott TR, Cardiff R. Postirradiation malignant fibrous histiocytoma of the trachea. Am Rev Respir Dis 1987;135:761–762.
[6] Tan-Liu NS, Matsubara O, Grillo HC, Mark EJ. Invasive fibrous tumor of the tracheobronchial tree: clinical and pathologic study of seven cases. Hum Pathol 1989;20:180–184.
[7] Randleman CD, Unger ER, Mansour KA. Malignant fibrous histiocytoma of the trachea. Ann Thorac Surg 1990;50:458–459.
[8] Wang YH, Wang JL, Lin AS, Cheng L. Malignant fibrous histiocytoma of the trachea combined with thymic carcinoma – a case report. Chang Keng I Hsueh 1993;16:271–274.
[9] Sennaroğlu L, Sözeri B, Ataman M, Gököz Ö. Malignant fibrous histiocytoma of the trachea: case report. Acta Oto-rhino-laryngologica Belg 1996;50:147–149.
[10] Refaely Y, Weissberg D. Surgical management of tracheal tumors. Ann Thorac Surg 1997;64:1429–1432.

Downloaded from https://academic.oup.com/ejcts/article/16/4/464/404189 by Ankara University Library (ANK) user on 11 December 2021