Case report

Fibrohistiocytic tumor of the trachea in a child

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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.

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 pCO2 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-
gularly surfaced, yellow-pink, focally hemorrhagic tumor projecting into the lumen which measured 2.0 \times 1.5 \times 1.4 \text{ 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

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

<table>
<thead>
<tr>
<th>Author</th>
<th>Age-sex</th>
<th>Tumor size (cm)</th>
<th>Presenting symptoms</th>
<th>Histology and diagnosis</th>
<th>Treatment and follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Karlan et al. [1]</td>
<td>57±F</td>
<td>2.5 × 2.5 × 0.8</td>
<td>Intermittent hemoptysis 15 years inspiratory wheezing 3 months</td>
<td>Spindle cells, foamy histiocytes, whorled/storiform pattern fibrous histiocytoma of the sclerosing hemangioma type</td>
<td></td>
</tr>
<tr>
<td>Hakimi et al. [1]</td>
<td>26±M</td>
<td>2.0 × 1.5 × 1.5</td>
<td>Cough, hemoptysis 1 week Weakness, weight loss</td>
<td>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</td>
<td></td>
</tr>
<tr>
<td>Sandstrom et al. [2]</td>
<td>15±F</td>
<td>0.9 × 0.8 × 0.8</td>
<td>Cough, progressive exertional dyspnea, stridor 6 months</td>
<td>Spindle cells forming fascicles and storiform pattern, multinucleated giant cells, occasional mitoses, vascular invasion, invasion through tracheal cartilage, extensive replacement and destruction of thyroid benign fibrous histiocytoma</td>
<td></td>
</tr>
<tr>
<td>Cohen et al. [3]</td>
<td>2±F</td>
<td>0.8 × 0.5 × 0.5</td>
<td>Fever, tachypnea, cough, dyspnea 5 months</td>
<td>Spindle cells, foamy macrophages, rare mitoses xantho®broma (fibrous histiocytoma)</td>
<td></td>
</tr>
<tr>
<td>Gonzalez-Campora et al. [4]</td>
<td>16±M</td>
<td>Grain of rice (first) 1.5 × 1.0 × 0.5 (recurrence)</td>
<td>Severe obstructive dyspnea, hemoptysis 6 weeks</td>
<td>Spindle and polygonal cells, multinucleated cells, storiform pattern, no atypia, scanty mitoses, no cartilagenous or vascular invasion benign fibrous histiocytoma</td>
<td></td>
</tr>
<tr>
<td>Louie et al. [5]</td>
<td>77±F</td>
<td>? (&gt; 10)</td>
<td>Dyspnea, intermittent stridor, enlarging right neck mass – 11 years after radiation to right neck and thyroid for papillary carcinoma</td>
<td>Spindle cells with prominent pleomorphism, frequent mitoses, many multinucleated giant cells postirradiation malignant fibrous histiocytoma</td>
<td></td>
</tr>
<tr>
<td>Tan-Lui et al. [6]</td>
<td>8±F</td>
<td>3.0 × 3.0</td>
<td>Wheezing few years</td>
<td>Spindle cells with moderate atypia, rare mitoses, multinucleated giant cells invasive fibrous tumor of the tracheobronchial tree</td>
<td></td>
</tr>
</tbody>
</table>

Endoscopic excision, LRb at 1 year; endoscopic re-excision, NEDa at 2 years

Tracheal resection, NEDa at 27 months

Endoscopic excision, LRb at 2 months; bronchoscopic resection, 2nd LRb at 5 months; Tracheal resection, NEDa at 12 months

Endoscopic excision (twice), NEDa at 10 years

Local resection, LRb at 6 months; widened local resection, NEDa at 15 months

Bronchoscopic laser therapy, Adriamycin, expired at 3 months

Tracheal resection, LRb at 1 year; Carinal resection, NEDa at 4 years
<table>
<thead>
<tr>
<th>Author</th>
<th>Age-sex</th>
<th>Tumor size (cm)</th>
<th>Presenting symptoms</th>
<th>Histology and diagnosis</th>
<th>Treatment and follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sculerati et al. [1]</td>
<td>17–F</td>
<td>? (2–3)</td>
<td>Wheezing, dyspnea, hemoptysis 3 weeks</td>
<td>Spindle cells, areas of storiform arrangement, some pleomorphism, occasional mitoses, no multinucleated giant cells – low grade malignant fibrous histiocytoma</td>
<td>Endoscopic debulking, tracheal resection, radiotherapy, NED³ at 20 months</td>
</tr>
<tr>
<td>Randleman et al. [7]</td>
<td>17–F</td>
<td>1.5 × 1.5 × 1.3</td>
<td>Dyspnea, wheezing 1 year</td>
<td>Spindle cells, scattered Touton giant cells, moderate pleomorphism, rare mitoses – low grade malignant fibrous histiocytoma</td>
<td>Tracheal resection, NED³ at 14 months</td>
</tr>
<tr>
<td>Wang et al. [8]</td>
<td>58–F</td>
<td>2.5 × 0.6</td>
<td>Cough with scanty sputum, progressive exertional dyspnea 3 months</td>
<td>Spindle cells with severe nuclear pleomorphism, frequent mitoses – malignant fibrous histiocytoma</td>
<td>Tracheal resection, NED³ at 30 months</td>
</tr>
<tr>
<td>Sennaroglu et al. [9]</td>
<td>32–M</td>
<td>1.5 × 1.0</td>
<td>Progressive dyspnea 1 year</td>
<td>Fibrohistiocytes with storiform pattern – malignant fibrous histiocytoma</td>
<td>Tumor resection and radiotherapy, NED³ at 3 years</td>
</tr>
<tr>
<td>Present case</td>
<td>10–F</td>
<td>2.0 × 1.5 × 1.4</td>
<td>Exertional dyspnea, wheezing 1 year</td>
<td>Spindle cells with moderate pleomorphism, focal storiform organization, multinucleated giant cells, occasional mitoses – fibrohistiocytic tumor</td>
<td>Tracheal resection and radiotherapy, NED³ 17 months</td>
</tr>
</tbody>
</table>

³LR, local recurrence.

⁴NED, no evidence of disease.
References