CAZURI CLINICE

Inflammatory myofibroblastic tumor of the trachea

Boloursaz MR[1]., Khalilzadeh S[1]., Abbasi Dezfoli A[2]., Kahkoee Sh[3]., Karimi Sh[4]., Abbaszadeh M[1]., Masjedi MR[5]., Velayati AA[1].*

[1]Pediatric Respiratory Disease Research Center,

[2]Tracheal Disease Research Center,

[3]NRITLD,

[4]Mycobacteriology Research Center,

[5]Chronic Respiratory Research Center,

National Research Institue of Tuberculosis and Lung Disease (NRITLD),

Masih Daneshvari Hospital, Shaheed Beheshti University of Medical Science, Tehran, Iran.

ABSTRACT

This report described a 2-year-old boy who presented with severe respiratory distress and stridor. Bronchoscopy and CT revealed a mass in the left anterolateral tracheal wall and histopathology showed a tracheal inflammatory myofibroblastic tumor. Initial removal by rigid bronchoscopy resulted in prompt recurrence of the tumor. Therefore he underwent tracheal surgical resection. A bronchoscopy at 12 months after surgery did not show any recurrence sign.

Keywords: *Inflammatory Myofibroblastic tumor, Trachea, Pediatric age.*

REZUMAT

Tumoră inflamatorie miofibroblastică traheală

Acest raport descrie cazul unui băiețel în vârstă de 2 ani ce s-a prezentat cu insuficiență respiratorie severă și stridor.

Bronhoscopia și examenul CT au dezvăluit o masă tumorală pe fața antero-laterală a peretelui stâng traheal ce examinată histopatologic s-a dovedit a fi tumoră traheală inflamatorie miofibroblastică.

Îndepărtarea inițială a tumorii prin bronhoscopie rigidă a dus la recurență tumorală rapidă. Așadar, pentru ablația tumorală s-a intervenit chirurgical. Bronhoscopia efectuată la 12 luni post chirurgical nu a decelat semne de recurență.

Cuvinte cheie: *trahee, tumoră inflamatorie miofibroblastică, vârstă pediatrică.*

Introduction

Inflammatory myofibroblastic tumor (IMT), commonly known as Inflammatory Pseudotumor (IPT) is a rare lesion with the frequency of 0.04%-0.07% of all respiratory tract tumors[1]. The etiology of IMT is still uncertain and it can occur at any age but it is more probable to be detected in pediatric and young children. IMTs are frequently found in the lung but similar lesions are also have been reported almost every site in the body[2,3]. To our knowledge reported cases of this tumor presenting in the trachea in children are very few. We report here a 2-year-old boy surgically treated for tracheal IMT.

Case report

A 2 year-old boy was presented to our pediatric respiratory unit in June 2009 with severe respiratory distress and stridor. He had a history of cough and wheeze for 5 months and was wrongly treated for presumed asthma. The patient underwent bronchoscopy three months ago for possibility of foreign body in his airways. As a result, soft tissue mass was removed and the pathology report showed infantile fibrosarcoma. He obtained prompt relief of his symptoms after bronchoscopy. One month prior to admission in our center his symptoms reccured. A physical examination showed a well-developed child with severe bilateral inspiratory and expiratory stridors. He was afebrile and his blood pressure was 85/60 mmHg and respiratory rate 34 per minute. A chest roentgenogram and routine laboratory studies were normal. Computed tomography showed soft tissue projection in the left anterolateral aspect of tracheal lumen in upper thoracic region (Figure 1). Direct laryngoscopy and bronchoscopy were performed. The bronchoscopy results showed a mass with 80% luminal obstruction. The mass was 27 mm in length at 15mm from main carina and 20 mm from the vocal cords with no extraluminal extension. The tumor was removed during rigid bronchoscopy and patient recoverd well from this procedure. Microscopic section of submitted tissue showed a cellular mass with diffuse proliferation of uniform spindle cells admixed with some plasma cells,small lymphocytes and rare PMNs and eosinophils (Figure 2A, 2B). There was no evidence of significant nuclear atypia or necrosis. The mitotic figures was about 1-2 per 10 high power field. Immunohistochemistry was positive for Muscle-Specific Actin and weakly to moderately positive for cytoplasmic Anaplastic Lymphoma Kinase (ALK-1), but negative for pancytokeratin, Desmin, Smooth Muscle Actin, CD31 and S100. These features were compatible with IMT.

The patient presented again 3 months after his first discharge with respiratory distress and stridor. The repeat bronchoscopy showed the recurrence of tumor at the previous mass site. Thus, about one month after second bronchoscopy we decided to perform a tracheal surgical resection. Through cer-

*Correspondence: M. Abbaszadeh, Associated Professor of Pediatrics, Tel: +982127122478, Fax: +982126109549
Adress: pediatric ward, Masih Daneshvari Hospital, Daar-abad, Niavaran, Tehran, Iran
email: marjanabbaszadeh@gmail.com

Figure 1.
A computed tomographic scan of the trachea demonstrating a circumscribed intraluminal tracheal mass (arrow) located at the left antero-lateral wall.

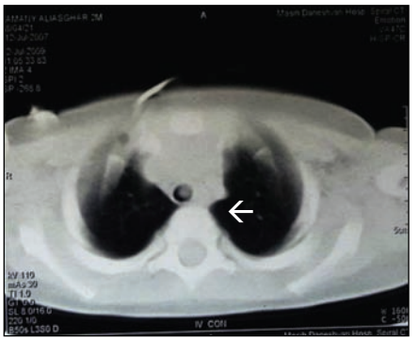
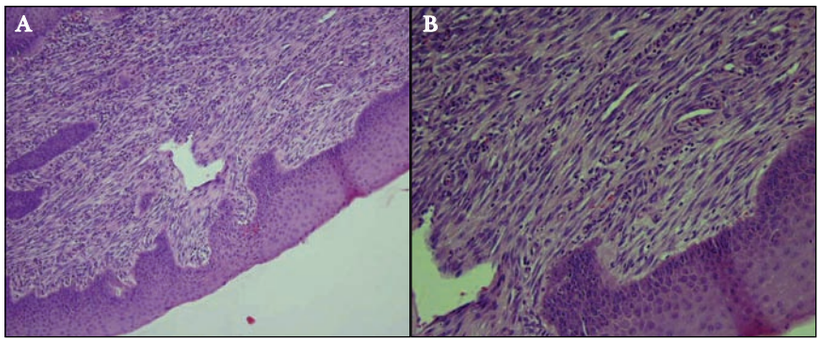


Figure 2.
(A) Hematoxylin-eosin (H& E) 10× (B) H &E 40× ; sections reveal metaplastic squamous epithelium of trachea, covering diffuse neoplastic proliferations of tumoral spindle cells admixed with inflammatory cells.



vicotomy the middle part of the trachea (abouth 27 mm, 5 rings) was resected. Releasing of anterior portion of cricoid to above the hyoid bone and anterior section of the trachea from carina to the first ring of the trachea were performed. End to end tracheal anastomosis was accomplished with continuous sutures (vicryl 5-0). A pathologic diagnosis again was consistent with IMT. The patient remained well, without evidence of recurrence, 1 year after resection.

Discussion

IMT is a proliferation of spindled myofibroblasts along with an inflammatory infiltrate including plasma cells, lymphocytes, histiocytes, eosinophils[2]. Given its various histologic characteristics, this tumor has been described in literature with many synonyms such as: inflammatory pseudotumor, fibrous histiocytoma, fibroxanthoma, xanthogranuloma etc[2,4,5]. Spindle cells are the principle cells in the histologic picture in 70% of the cases which was similar in our case[6,7]. Due to its wide variety, IMT might be confused even by experienced pathologists with neoplasms such as rhabdomysarcoma, infantile fibrosarcoma, infantile myofibromatosis and Hodgkins lymphoma[3,6]. Zapatero et al, reported a case of 6-year old boy whose biopsy sample was diagnosed as rhobdomyosarcoma but further histologic studies showed an IMT[8]. In our patient, initial biopsy sample was diagnosed as infantile fibrosarcoma but other biopsies indicated an IMT. Differential diagnosis can be excluded by using immunohistochemistry[5]. Probable predisposing factors include an inflammatory reaction secondary to trauma, an autoimmune reaction or infection[3,7]. In our case there was no history of trauma or infection which was akin to the majority of other reported cases[3]. All in all it is a benign lesion that most commonly occurs in children and adolescents. In 50% of cases at pediatric age it is found in the lungs and tracheobronchial tree[7,9]. Although variable clinical manifestations of airway inflammatory pseudotumors such as dyspnea, stridor, wheezing, hemoptysis, chronic cough, dysphagia, fever, pleutoric pain, right upper quadrant or epigastric pain and severe constitutional symptoms have all been described[1,10]. Diagnosis and treatment of pseudotumor are often delayed due to nonspecific symptoms. CT-scan may reveal a well-defined or circumscribed endotracheal mass[7,9]. Direct laryngoscopy and bronchoscopy with biopsy are essential for diagnosis and assessment of airways involvement[7]. After histopathological confirmation, the commonest modality is conservative endoscopical resection through ultimate Co2 laser[1]. In spite of the evidence of its histologically benign features, there have been documented reports of local recurrences, metastases, medical invasion and sarcomatous transformation. Recurrence after local resection is more frequent than distant metastases[9]. Venizelos I et al reported that extrapulmonary IMTs have a recurrence rate of ~25% and a metastasis rate of <50%[2]. The choice of treatment in recurrent tracheal IMTs is open surgical intervention with segmental tracheal resection[1,6]. Adjuvant therapy (radiotherapy and chemotherapy) are not indicated in most cases and should be reserved when surgery is not an option[3,10].

Conclusion

Inflammatory pseudotumor has to be considered among differential diagnosis for any pediatric patient presenting with asthma and tracheal mass. Thorough morphologic assessment along with Immunohistochemical stains is mandatory in order to avoid misdiagnosed IMT with other similar appearing lesions. In case of recurrence after endoscopical treatment, tracheal resection is mandatory. Recurrences after tracheal resection are not common but careful follow up is essential however complete resection of the lesion is preferable. The overall prognosis for IMT in children is excellent even after tracheal resection.

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