Inflammatory Myofibroblastic Tumour of the Trachea with Paratracheal Lymph Nodes and Mediastinal Invasion

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ABSTRACT

Objective: We describe a rare case of inflammatory myofibroblastic tumour of the trachea in a 47 years old female with paratracheal lymph nodes infiltration and mediastinal extension, who presented with progressive difficulty in breathing. (Rawal Med J 2009;34:115-116).

Key words: Inflammatory myofibroblastic tumor, trachea.

INTRODUCTION

Inflammatory myofibroblastic tumor has many synonyms including inflammatory pseudotumor, plasma cell granuloma, fibrous histiocytoma, fibromyxoma or inflammatory myofibrohistiocytic proliferation. These tumors can occur at any age, but they are more prevalent in children and young adults. Bumber et al1 found only 13 cases of inflammatory pseudotumor of the trachea. However, only one case involving the neck with tracheal invasion was found in the literature.2 We describe a rare case, an adult with inflammatory myofibroblastic tumor of the trachea with paratracheal lymph nodes infiltration and mediastinal extension.

CASE REPORT

A 47-year-old female presented with a history of progressive difficulty in breathing and stridor. Physical and laboratory examination were unremarkable. A CT scan (Fig. 1a) showed a focal mass of soft tissue density at the right lateral wall of the trachea with mediastinal extension. An MRI scan (Fig. 1b) revealed intraluminal mass of 5 cm in length causing narrowing of the lumen of the trachea with
mediastinal extension. Tracheoscopy revealed a pink polypoidal mass obstructing almost 80 per cent of the tracheal lumen (Fig. 1c). The tumor was excised piece-meal. The pathological diagnosis was inflammatory myofibroblastic tumor (Fig. 1d and 1e). She became asymptomatic after this initial procedure which was aimed at establishing a better airway and reaching a diagnosis.

One month later, anterior neck and mediastinal exploration were performed. The tumor was adherent to the trachea approximately 6 cm from the vocal cords extending 5 cm towards the carina inferiorly and deep to the innominate artery and vein posteriorly. There were multiple matted pretracheal and paratracheal lymph nodes. Complete excision of the tumor and trachea resection and anastomosis was not possible because the tumour involved 5 cm of the tracheal in length and it also extended deep to the innominate vessels. We debulked the tumor as much as possible.
Figure 1

Radiological, histological, pre-operative and post-operative findings of the case. A 47-year-old female with inflammatory myofibroblastic tumour of the trachea. a & b: Admission CT scan and MRI of the neck showed inflammatory myofibroblastic tumour of the trachea with paratracheal lymph nodes infiltration and mediastinal extension. c: Tracheoscopy revealed a firm pedunculated lesion located on the right posterolateral tracheal wall causing 80% luminal obstruction. d & e: Photomicrographs of trachea and neck specimens. Tumour composed of fibrous components, inflammatory cells and plasma cells intermingled with myofibroblastic cells. f: On CT scan obtained a year after surgery, there is no evidence of tumour recurrence.
She remains symptom-free at follow-up two year after surgery. A repeat CT scan of the neck and thorax also showed no evidence of tumor in the trachea and mediastinum (Fig 1f).

**DISCUSSION**

Inflammatory myofibroblastic tumour (IMT) was first described by Brunn in 1939. These tumors most commonly arise in the lungs and abdomen. They seldom occur in the trachea, accounting for 2.7% of all reported cases. In 1994, WHO defined IMT as an intermediate soft tissue tumor that is composed of myofibroblast-differentiated spindle cells and accompanied by numerous inflammatory cells, plasma cells, and/or lymphocytes. Meis et al designated it as malignant based on their clinical behavior (multifocality in 30%, recurrences in 27% and the presence of regional metastases in 8%) and cellular atypia. Su et al described the cytogenetic evidence of acquired clonal chromosomal abnormality within the tumor of a small number of cases concluding that an IMT may be a clonal neoplasm.

The treatment modalities that have been used are endoscopic removal with CO₂ laser or electrocautery or open surgical approach with segmental tracheal resection. Radiotherapy has been used in a small number of cases with only fair results. It may be the treatment of choice when surgery or corticosteroid therapy are contraindicated or unsuccessful. Corticosteroid therapy is the standard treatment for the IMT of the orbit. In our case, the patient showed improvement with steroid therapy before surgery. Post-operatively no steroids were given. There has been no recurrence to date. Careful long-term follow-up is essential, as in our patient complete surgical resection was impossible, as sarcomatous transformation and recurrences can occur.

**REFERENCES**


