Case Report

Inflammatory Myofibroblastic Tumour of the Trachea in a Child with Asthmatic Symptoms

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Abstract

We here present a case of an 8-year-old girl with an inflammatory myofibroblastic tumour of the trachea. She was initially referred due to stridor, dyspnea, and low functional status. Diagnostic work-up revealed a cauliflower-like tumour in trachea, which was initially removed endoscopically and subsequently by tracheal resection. The patient was previously diagnosed with idiopathic thrombocytopenia, which resolved spontaneously postoperatively.

Inflammatory myofibroblastic tumours also known as inflammatory pseudo tumours are very rare primary tumours of the trachea and considered by the WHO to represent intermediate grade of malignancy. Any association between this condition and idiopathic thrombocytopenia has so far only been described sporadically.

INTRODUCTION

Primary tumours of the trachea are very rare and are usually malignant. Inflammatory myofibroblastic tumours (IMT) also known as inflammatory pseudo tumours (IPT) are benign tumours and morphologically characterised by proliferation of myofibroblasts and fibroblasts, mixed with variable number and type of inflammatory cells. It is an extremely rare disease with a frequency of 0.04-0.07% of all patients with tumours [1]. IMTs are primarily found in the respiratory tract, but may occur in virtually any tissue [2]. As the tumour is considered by the WHO to represent intermediate grade of malignancy radical surgery is recommended [3,4]. We here present a case of an 8-year old girl surgically treated for tracheal IMT. Additionally she was previously diagnosed with IPT which, interestingly resolved after removal of the tumour.

CASE REPORT

An 8-year-old girl, known with IPT was referred to the ENT department, Aarhus University Hospital due to increasing stridor, dyspnea, and low functional status. Haemoptysis was not reported. In the previous two years inhalation treatment had been attempted suspecting asthma, but without effect. Diagnostic work-up including fiber laryngoscopy, CT scan and subsequent tracheoscopy revealed a cauliflower-like tumour three centimetres below the vocal cords, causing a significant narrowing of the trachea.

The tumour was initially removed endoscopically with cold instruments, primarily to secure tissue for histological examination as well as restoring the airway. Macroscopically resection margins were narrow but considered significant. Histological examination revealed IMT; however margins could not be evaluated due to piecemeal resection. At initial follow-up the patient reported a significant improvement of the respiratory function. Since the tumour was considered to be intermediate malignant by WHO, a tracheal resection was initially scheduled. The following tracheoscopy and MRI indicated no signs of residual tumour and a tracheal resection was not performed. However, at subsequent follow-up, MRI and tracheoscopy indicated tumour recurrence. Hence, a tracheal resection was performed. Three tracheal rings i.e. 2 cm of the trachea was resected, with macroscopic clear and wide margins. There were no signs of deep infiltration. Trachea was sutured with end to end anastomosis and the patient left intubated until the following day. No perioperative complications were reported.

Histology confirmed IMT, however with some uncertainty regarding the superior and inferior margins. During the subsequent 8-year follow-up, which included regular tracheoscopy and initially MRI, there were no signs of recurrence and up to present time the patient is tumour free.

The course was complicated by the fact that the girl was known with IPT, necessitating thrombocyte infusion prior to surgery. Interestingly the thrombocytopenia resolved spontaneously after surgery. Thus thrombocyte antibody screening including bound antibodies turned negative and thrombocyte function normalized (Figure 3).

DISCUSSION

Inflammatory myofibroblast tumours of the trachea are rare neoplasms [1]. Due to the highly varied morphology IMT has many synonyms such as fibrosis histiocytophathia, fibroxanthoma...
The tumor was consistently increased in size and extent on the follow-up CT scans, most noticeably between the second and third scan. The tumor was resected endoscopically, with the aid of a surgical deformity in the posterior wall of the trachea, and a significant airway obstruction.

Figure 1: CT scan of trachea. A cauliflower-like tumor extends from the posterior wall of the trachea and into the lumen, causing a significant airway obstruction.

Figure 2: CT scan of trachea. A cauliflower-like tumor extends from the posterior wall of the trachea and into the lumen, causing a significant airway obstruction.

Figure 3: Circle 1 indicates the time when endoscopic removal of the tumor occurred (September 10th, 2007). Circle 2 indicates the time when the tracheal resection occurred (May 13th, 2008). In the period from 2007-2012 blood count tests were performed in variable intervals to follow the patient’s ITP, which is indicated by the numbers. The thrombocyte peaks around the operation points were due to thrombocyte transfusion.

CONCLUSION

In conclusion, we report a case of ITP in a child with tracheal IMT, and spontaneous disappearance after surgical removal of the tumor.

REFERENCES


Cite this article