CASE REPORT

Adenoid Cystic Carcinoma of the Trachea: A Case Report

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Summary

A case report of adenoid cystic carcinoma of the trachea is presented. A high index of suspicion is required to make the diagnosis of tracheal tumours early since the patients tend to have normal chest radiographs which on closer examination may show an abnormality of the tracheal column. Adenoid cystic carcinoma is the most common malignant tumour of the trachea. Locoregional control of this disease is achieved by a combined modality therapy of surgery and postoperative radiotherapy.

Despite this, it still has a prolonged clinical course and the tendency for delayed onset of distant metastases.

Key Words: Tracheal tumours, Tracheal resection and Reconstruction, Adenoid cystic carcinoma

Introduction

Primary neoplasms of the trachea are very rare, about 2.7 new cases per million persons[1]. Over 90% of these tumours are malignant with 2 main histological types: squamous cell carcinoma and adenoid cystic carcinoma. Adenoid cystic carcinoma is notorious for its insidious onset, perineural invasion of major nerves and submucosal spread. Only 10% of adenoid cystics will metastasize to regional lymph nodes. Combined treatment of surgical resection and radiotherapy has yielded the best results for adenoid cystic carcinomas of the head and neck with a disease specific survival at 5 years being 89%[2]. However, these tumours are known for local recurrence with a disease specific survival at 10 and 15 years dropping from 67.4% to 39.6% respectively[2]. To date there have only been 2 cases of primary tracheal tumours reported from this hospital over the last 10 years, with the latest case being the first managed here.

Case Report

A 35 year old Indian lady was referred from the outpatient clinic with a history of progressive dyspnoea, cough and dysphagia of 8 months duration. She had received treatment for a recent adult-onset bronchial asthma without improvement. Her dyspnoea was accompanied by paroxysmal exacerbation occurring frequently nocturnal. Clinically she had a wheeze best heard at the end of forced expiration. She was uncomfortable when lying prone and at the same time mild pressure applied to the suprasternal notch. Her head and neck examination was normal. Mirror examination of the larynx revealed normal, mobile vocal cords. Auscultation of the chest revealed occasional bilateral ronchi.

Flexible laryngoscopic examination of the upper airway was attempted but abandoned when she went into laryngospasm. She recovered with immediate

resuscitation. Later, a plain lateral neck radiograph revealed an abnormality of the tracheal column. A repeat of the lateral neck view, with the neck hyper-extended revealed a rounded intratracheal radioopaque lesion. This lesion was confirmed on magnetic resonance imaging (MRI). It showed a rounded isointense mass arising from the right postero-lateral aspect of the trachea, almost obliterating it at the level of C6-C7vertebral bodies (Figure 1).

On the 21st January 1999, she underwent tracheal resection and reconstruction with primary anastomosis. Anaesthesia was administered via an awake fibreoptic intubation with a small endotracheal tube insinuated past the obstructive tumour. Intraoperatively the solid tumour, which was arising from the right postero-lateral wall of the trachea and almost occluding it, had caused a fusiform dilatation of the trachea extending from the 1st tracheal ring to the 5th ring. A 3cm segment of the trachea starting from the lower level of the cricoid was resected under frozen section control.

Although macroscopically and frozen histological examination of the margins appeared clear, subsequent paraffin sectioning revealed microscopic involvement of the upper margin. There was no cervical lymphadenopathy.

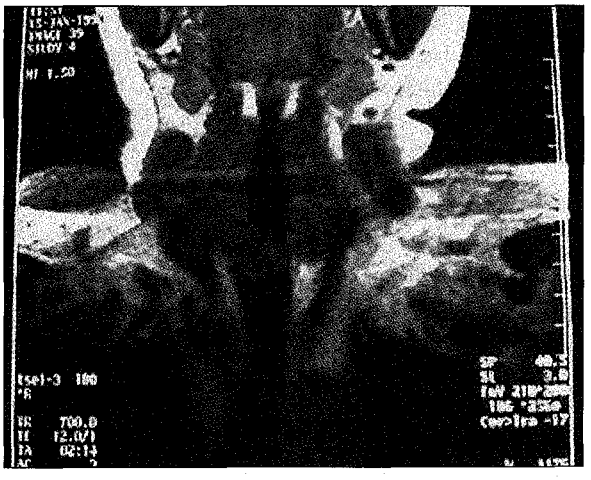

Fig 1: Coronal view of tracheal tumour as seen on MRI.

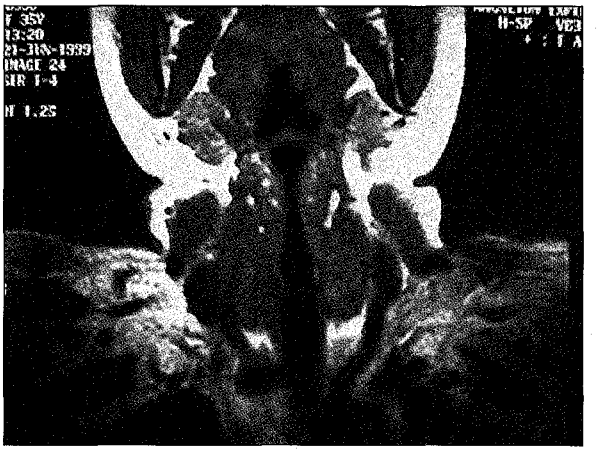

Fig 2: Coronal MRI 6 months after surgery.

The final histology was that of adenoid cystic carcinoma. At the conclusion of the procedure, the patient was extubated and breathing spontaneously.

Cervical flexion was maintained by incorporating steel sutures from the chin to the sternum. She had an uneventful postoperative period and was discharged well on the 4th week. A week later, at review, she was noted to be 8 weeks pregnant (her last menstrual period was on the 21st December 1998) but had to be terminated on medical grounds. She later underwent radical radiotherapy to the neck at the Hospital University Kebangsaan Malaysia, Kuala Lumpur which she completed on the 14th April 1999. At review, 6 months after the surgery, she was found to be well and free of disease (Figure 2).

Discussion

Neoplasms of the trachea are rare. Few institutions have been able to accumulate enough experience to allow definite conclusions to be made. It is estimated that 2.7 new cases of primary tracheal tumours per million persons occur annually[1].

There are two main histological types of primary tracheal carcinoma: squamous cell carcinoma (SCC) and adenoid cystic carcinoma.

The other primary malignant tumours are carcinoid tumours and mucoepidermoid carcinoma. Approximately 10% of primary tumours are benign. The most common benign tumour is squamous papilloma. Other types include pleomorphic adenoma, granular cell tumour and cartilaginous tumours. Most secondary tracheal tumours are caused by direct extension of a local tumour into the trachea, such as papillary or follicular thyroid carcinoma, lung cancer or oesophageal carcinoma. Distant metastatic disease to the trachea is rare[1].

Tracheal tumours frequently have an insidious onset, and early signs and symptoms may be disregarded or mistaken for a variety of other disorders. Shortness of breath on exertion, which may progress to dyspnoea at rest, a brassy cough, recurrent pneumonitis, wheezing, hoarseness and stridor may all be part of a clinical presentation. Many of these symptoms, especially dyspnoea on exertion and wheezing can be easily attributed to other respiratory disorders such as chronic bronchitis and asthma. A patient's past medical history is thus important. Occasionally the patient may have dysphagia as a result of pressure or invasion of the oesophagus. Haemoptysis occurs with some tumours.

The primary diagnostic technique for tracheal abnormalities is radiological. Chest radiograph and lateral neck view all too often appear to be normal which on closer inspection shows an abnormality of the tracheal column. The lateral neck view is usually taken with the neck hyperextended so as to bring the trachea above the clavicles. MRI has a very useful role to play. Sagittal and coronal views accurately define the tumour and provide additional information as to the presence or not of an extra tracheal component.

The office diagnosis by endoscopy with a flexible fibreoptic bronchoscopy, under local anaesthesia, is in general deferred until the time of correction in order not to precipitate an acute obstruction where subtotal obstruction is already present.

Airway management is crucial, especially if there is a high degree of obstruction distally. With this respect there should be a close liaison between the anaesthetist and surgeon since both share the same airway.

Primary tracheal tumours provide the second most common indication for tracheal resection and reconstruction with primary anastomosis[1]. However, the unique structure of the human trachea with its lateral rigidity, unpaired nature, segmental blood supply from branches of the inferior thyroid artery above and bronchial arteries below, and its relatively short length pose immense problems for direct reconstruction. The addition of cervical flexion allowed for direct reconstruction with a tension free primary anastomosis. Additional mobilization techniques may be required: suprahyoid laryngeal release and for the distal trachea mobilization of the right pulmonary hilum so as to provide a tension free anastomosis. Access to the upper trachea is via an anterior cervical approach and for the lower trachea and carina the transthoracic approach via a right postero-lateral thoracotomy.In case of unresectable disease, the airway may be maintained by repeated bronchoscopic debulkings, use of CO2 laser, the silicone Montgomery T-tube and tracheal stents. An example of such a stent is the Westaby T-Y tracheobronchial stent which is utilized in situations of complicated airway problems where endotracheal intubation may be impossible or undesirable, and a standard tracheostomy would be ineffective.

The two main histological types of primary tracheal malignancy have different natural histories. SCC may be ulcerative or exophytic in appearance. It tends to metastasize to regional lymphnodes and often directly invades mediastinal structures. Second primary squamous cell carcinomas are common. Locoregional recurrence is common and the disease is usually fatal within months. The 5 year survival after curative resection is 20 - 40%[1].

Adenoid cystic carcinomas have a much slower growth pattern. It is notorious for its perineural invasion of major nerves and submucosal spread. Only about 10% metastasize to regional lymphnodes. Although local recurrence is common, it may not present clinically for 5 or 10 or more years. Patients may survive for many years with microscopic or metastatic disease[1].

Both adenoid cystic and SCC are usually radiosensitive and radiotherapy has been utilized as both a primary and an adjunct treatment for primary tracheal carcinomas. Jim Fordice et al recently analyzed 140 patients from a

single institution for adenoid cystic carcinoma during 20 years between 1977 and 1966 who had a consistent treatment of surgery and postoperative radiation therapy[2]. This combination of surgery and ostoperative radiation therapy improved the locoregional control of adenoid cystic carcinoma of the head and neck with 85% freedom from relapse but the disease specific survival at 5, 10 and 15 years was 89%, 67.4% and 39.6% respectively. They also noted that the presence of four or more symptoms at diagnosis, positive lymphnodes, perineural invasion and a "solid" histology (a histopathological variant of adenoid cystic carcinoma that had the presence of solid histological pattern in 10% of tumour tissue examined or more, with the presence of anaplasia within this solid archictecture) were associated with increased mortality from disease[2,3].

Perineural invasion of major nerves, solid histological features and positive margins at surgery were associated with increased treatment failures[2].

In conclusion, this case highlights the need to constantly review the diagnosis especially if the patient does not respond to initial management.

Secondly, in the treatment of adenoid cystic carcinoma of the trachea, in particular and the head and neck, in general the best option available, where feasible is a combined modality of surgical excision and postoperative radiotherapy. If successful the patient can be expected to survive for many years. Even as we enter the new millennium, we are still without satisfactory answers as to why the disease recurs many years later after initial "curative" management. Hence, prolonged follow-up is essential for early detection and management of delayed complications or metastases.

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