ADENOID CYSTIC CARCINOMA (CYLINDROMA)- A CASE REPORT

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Introduction

Adenoid cystic carcinoma (Cylindroma) is a histologically distinctive neoplasm characterised by local infiltrative growth and a prolonged clinical course. Although precise figures are difficult to obtain in western study, it is the most common sub-type of tracheobronchial gland tumour, probably accounting for atleast 75-80 percent of reported cases. In our country there is no statistics. Approximately 80 percent arise with about equal frequency in the trachea and mainstem bronchi[1,2]. Most of the remainder occur in lobar bronchi. The aetiology is unknown. In one series in which histories were adequate six of seven patient were non smokers[3]. Pathologically adenoid cystic carcinoma characteristically grows into the airway lumen forming a smooth surfaced, somewhat polypoid tumour, occasionally growth is circumferential and annular. Submucosal extension sometimes to a considerable distance from the main tumour is not uncommon[1,4,5]. The overlying tracheobronchial epithelium is usually intact although it may ulcerate. Histologically consists of uniform cells with relatively little cytoplasm arranged in well defined nests or trabeculae of moderate size. The nests frequently show a cribriform pattern in which the cells care separated by cystic spaces containing a mucinous substance that stains strongly with alcian blues. Occasionally the intereellular material has a homogeneous, eosinophilic (hyalinized) appearance. Mitotic activity & necrosis are seldom observed. The tumour may extend directly into and destroy cartilage & perineural invasion is frequent.

Conventional tomography in anteroposterior and lateral projections may be useful in the evaluation of this tumours (indeed for all forms of tracheobronchial gland tumours) although CT is clearly a more useful technique, particularly for assessing the presence or absence of mediastinal extension[6]. Fibreoptic bronchoscopy and biopsy is an easy and safe method for confirmatory diagnosis.

Clinical course is usually prolonged and the prognosis much better than that of the more common forms of pulmonary carcinoma[1,7]. Because of the rather slow tumour growth, patients must be followed for a considerable period of time before the final outcome is established; death from recurrent disease has been documented upto 30 years after the initial discovery of the tumour[7].

Case report

A 32-year-old physician who is a life long nonsmoker presented to us with chronic inter mittent cough for the past 2 years and occasional streaky hemoptysis for 2 months. During this period there was no loss of weight & no fever. Suddently he developed wheezing for a period of 10 days before presentation. On physical examination he looked well. There was diminished breath sound over the whole of left hemithorax. His breath sounds were harsh and suggestive of mild stridor during forced breathing. There was no other positive findings. Serial chest X-ray revealed no abnormalities. Complete blood picture revealed no abnormalities. Tuberculin test was 10mm induration. Sputum for AFB was negative. Fibreoptic bronchoscopy showed a vascular polypoidal mass arising from lower end of trachea and main carina, completely obstructing the left main bronchus. The right main bronchus was also partially obstructed but the scope could be passed through to visualise the right upper lobe, middle & lower lobe bronchi. These were clear of tumour. Biopsy was taken from the lesion. The report revealed adenoid cystic carcinoma.

CT thorax showed multiple soft tissue masses in the right & left main bronchi. There was also thickening of the lower trachea by a ring of soft tissue of similar aetiology. The masses were seen to extend extraluminally into the prevertebral region.

In view of the severe obstruction to the airway and the inoperability of the tumour, we decided on clearance of the endobronchial tumour with Nd-Yag laser. There is no arrangement for laser therapy in Bangladesh. This was done at TanTock Seng Hospital in Singapore. Extensive resection of the tumour masses were done with Nd-yag laser via a rigid bronchoscopy. The left main bronchus was clear after resection but in view of the extrinic compression from the external tumour and the likely recurrent of the endobronchial tumour, a

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stent (Duman Silicon Stent) was put into both the left and right main bronchi.

He has improved symptomatically and breath sounds can be clearly heard in the left lung. His lung function has improved significantly post-laser therapy. At the check bronchoscopy revealed that the stents are patient and the distal airway can be visualised

Discussion

Adenoid cystic carcinoma is a tracheobronchial gland tumour. This tumour are quite uncommon and represents only about 15% of bronchial adenoma8 although it is probably a commonest primary malignant tumour of the trachea[9]. Over 50 year period at the Mayo clinical 20 cases of adenoid cystic carcinoma were seen[10]. It is a slow growing locally invasive malignancy[9].

Cylindromata may occur at any age and cause hemoptysis and symptom of bronchial obstruction and is often mistaken for other common disorders such as asthma[11]. They should be suspected particularly if a young or middle aged adult presents with increasing stridor of a few weeks or months duration[10]. The chest radiograph is frequently normal, may show a hilar tumour or distal collapse or consolidation. If the tumour is arising in the trachea the film will usually be normal. Clinical suspicion may be heightened by the characteristic appearance of the flow volume loop[12]. CT is useful for assessing the presence or absence of mediastinal extension[6]. Confirmation is by bronchoscopy & biopsy but caution should be exercised in the choice of instrument as the fibroeptic bronchoscope may occlude an already narrowed opening & where the stenosis is critical, biopsy using either instrument may result in complete occlusion of a small orifice. Bronchography may be used to delineate the extent of a central tumour. The tumour may rarely metastasize to nodes & liver but more commonly invaes widely into local structures[10].

Ideally treatment is by excision, though due to the usually central situation of the tumour this may not be feasible. If however, the disease apparently confined to the trachea, complete excision has been achieved in some centre using tracheo-bronchoplastic procedures and this procedures have allowed complete removal[13].

If resection proves impossible radio therapy which may be administered externally or endotracheally will often produce prolonged remission & relief of symptoms where tracheal obstruction is threatening life[14]. The use of LASER therapy holds promise of controlling the obstruction if resection is impossible. One series described the use of a neodymium yag laser in 21 patients with tracheal tumours of which almost half were primary[15]. The majority were treated as emergencies for impending asphyxia with symptomatic benefits allowing time to be gained for full diagnostic evaluation & treatment planning which included the use of further laser treatments, surgery or radiotherapy. Sometimes patients with tracheo bronchial obstruction that is not amenable to curative surgery may benefit from the insertion of a prosthetic stent[6].

Despite the fact that adenoid cystic carcinoma is a malignant neoplasm capable of metastasizing and causing death, the clinical course is usually prolonged & the prognosis much better than that of the more common forms of pulmonary carcinoma[1,7]. Although approximately 50% of patients described in older literaure either died of the neoplasm or developed recurrent disease[2] the more sophisticated surgical and diagnostic techniques available now a days may be expected to result in cure in a high proportion of cases, especially when the tumour is relatively small. Metastasis are uncommon & usually appear late in the course of the disease, death is usually the result of local intrathoracic complication[3,17,18].

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