Case report

A primary squamous cell carcinoma of the trachea: Case report and review of the literature

Carcinoma squamocellulare primitivo della trachea: Caso clinico e revisione della letteratura

G. Abbate, A. Lancella, R. Contini, A. Scotti
Division of Otorhinolaryngology and Head and Neck Diseases, “S. Biagio” Hospital, Domodossola, Italy

Summary

Primary windpipe tumour is extremely rare. Squamous cell carcinoma is the most common malignant form, especially in smokers. The radiological appearance of these neoplasms can be classified as intra-luminal, wall-thickening, exophytic form; the majority of the lesions are obstructive in nature and then tend to extra-luminal invasion. This tumour is usually diagnosed late on account of delayed specific symptoms: haemoptysis, dyspnoea, coughing, hoarseness, stridor. Surgery, followed by adjuvant radiotherapy, is the treatment of choice; primary radiotherapy, in inoperable cases, can represent a curative management option. A case of primary tracheal cancer is described and a review of the literature is presented.

Key words: Trachea • Malignant tumours • Squamous cell carcinoma • Dyspnoea

Introduction

Primary carcinoma of the trachea is not common. This rarity makes research into the natural history and treatment very difficult. Furthermore, data related to these tumours are limited. These tumours although extremely rare present extremely variable clinical and histological features.

Most primary cervical tracheal tumours are malignant: adenoid cystic carcinoma (ACC), squamous cell carcinoma (SCC), adenocarcinoma, mucocoeidermoid carcinoma, carcinoid tumour, oat cell carcinoma.

SCC is the most common pathology in smokers, ACC is more prevalent among non-smokers.

Benign tumours are xanthogranuloma and pleomorphic adenoma.

A carcinoma arising in the thyroid or oesophagus can spread to the trachea; moreover, the trachea can be the site of a metastasis from recurrent carcinoid tumour in the left main bronchus, larynx, lung, colon.

The radiological appearance of the tumours can be classified as: intra-luminal, wall-thickening, exophytic form. Endoscopic evaluation reveals that the majority of the lesions are bulky and obstructive in nature.

Malignant tumours tend to spread to extra-luminal invasion, for example, into the thyroid gland.

These tumours also tend to be diagnosed late on account of delayed specific symptoms; haemoptysis, dyspnoea, cough, hoarseness, stridor being the most common; when patients do not present blood in the sputum, the initial diagnosis is thought to be bronchial asthma; in many cases, therefore, suffocation almost occurs before surgical treatment.

Computed tomography (CT), in coronal projection, is the most useful radiological examination for tracheal tumours. Bronchoscopy and radiological examination are
complementary procedures. The main advantage of imaging is the demonstration of tracheal wall thickening and extra-luminal changes. Persistent or progressive local disease can cause complications: fatal haemorrhage, oesophago-tracheal fistula, tracheal necrosis, tracheal stenosis. Management of tracheal tumours includes interventional endoscopy, surgery, radiotherapy, endoluminal brachytherapy.

Extensive segmental resection of the trachea is the treatment of choice for primary malignant, and, occasionally, for benign tracheal tumours. Intervventional endoscopy is part of modern tracheal surgery. The resulting tracheal clearance has both palliative and curative purposes. Post-operative complications are mediastinitis, bilateral pneumonia, wound-healing disorders. Radiation therapy is effective treatment for primary tracheal neoplasm. Surgery, followed by adjuvant radiotherapy and primary radiotherapy in inoperable cases represent potentially curative treatment options. Positive lymph nodes or invasive disease at resection margins appear to have an adverse effect on the management of SCC; such an effect is not demonstrable with adenoid cystic carcinoma.

Surgery is the treatment of choice and up to 50% of the trachea can be resected with modern techniques. With tracheostomy a curative excision or a palliative excision is possible. The sleeve trachea resection is one of the optimal surgical modalities, the other options are: partial tracheal wall resection, immediate tracheal reconstruction, total laryngectomy + partial resection of trachea and thyroid lobectomy, resection and primary reconstruction, laryngotraheal resection, cervico-mediastinal exenteration, carinal resection and reconstruction. Trachea anastomosis is suitable for small defects. The platysma myocutaneous flap combined with the facial flap of the sternohyoid muscle, sternocleidomastoid myoperiosteal flap and the pectoralis major musculocutaneous flap are applied to reconstruct the defects of cervical trachea. Resection and post-operative radiation therapy are the treatment of choice, complete resection is the desired goal and demands knowledge of the principles of tracheal surgery. Debulking surgery, followed by radiotherapy, may provide effective and permanent control in ACC and can be effective in obviating local recurrence. Endo-bronchial high dose-rate brachytherapy may be used for tracheal tumours, even as a boost for external beam irradiation or in recurrences. Long-term survival may also be expected, particularly for tumours with adenoid cystic histology.

Palliation has improved with the introduction of laser resection, brachytherapy and stents. SCC may have a better prognosis, in the trachea, than in the lung. According to some Authors, patients who undergo primary surgery with adjuvant radiotherapy appear to have better disease-specific and overall survival rates compared with patients undergoing primary radiotherapy with or without chemotherapy.

After surgical management, the 3- and 5-year survival rates are 79.80%, 48.36% for ACC, 80% and 20% for SCC; in 1, 3, 5, 10 years are 82.3%, 75%, 75%, 50%.

Carvalho Hde, et al. presented their experience with high dose-rate endobronchial brachytherapy; they reported a good local control, at the time of the first bronchoscopic control. They treated 4 patients with non-resectable tracheal tumour: two patients with SCC died at the 6th and 33rd month, respectively, after treatment, only the second presented local recurrence. The other two patients were alive after 64 and 110 months of follow-up.

Some Authors discussed the role of radiation therapy alone: the median survival for SCC was 33 months, for ACC 94.2. The 1-, 3-, 5-year survival rates were 64.7%, 64.7%, 26% for SCC, 85.7%, 85.7%, 85.7% for ACC. Patients with SCC and patients with complete remission following treatment had a significantly better survival probability.

Other Authors reported that of 14 resected patients suffering from primary tracheal tumours, at the last follow-up (10 years), 9 are still alive. These Authors observed 5 long-term survivors > 6 years with adenoid cystic carcinoma or muco-epidermoid carcinoma. The 5-year survival rate for low grade malignant tumours arising in the trachea (carcinoid, muco-epidermoid carcinoma, adenoid cystic carcinoma) is 78.8%.

Case report
R.V. a 54-year-old male, heavy drinker and smoker, had been suffering from dyspnoea, sometimes haemoptysis, cough, weight loss ~10 Kg for approximately 2 months. The thorax studied by standard radiology showed a left pneumonia, routine blood tests revealed leukocytosis, neutrophilia, high levels of transaminases. Nevertheless, the patient refused admission to hospital and, therefore, underwent treatment with ceftriaxone, moxifloxacin, acetylcysteine, prednisone at home. After 9 days, X-rays and blood tests had improved. After 15 days dyspnoea had become more severe and was treated as asthma with beclomethasone aerosol and oral prednisone.

The rhino-pharyngolaryngeal region, explored by flexible fiberoptic examination, showed evidence of right laryngeal palsy. Radiographic examination of the oesophagus was normal (Fig. 1).

A tracheobronchososcopic examination demonstrated evidence of a voluminous tracheal mass; during performance...
of this test, heart arrest with respiratory failure occurred and the patient was reanimated with heart massage and mechanical respiration. Three hours later the patient underwent tracheostomy (IV – V tracheal ring). CT showed a voluminous wall-thickening solid tracheal mass, extending from the subglottic space to the upper mediastinic space, about 6-7 cm in length (Figs. 2, 3, 4). Due to the size of the lesion, the patient was submitted to palliative radiotherapy on T and n (trachea and neck), total dose 50.4 Gy, fractionated 1.8 Gy/die, with boosts on the trachea reaching 61.2 Gy. During follow-up, a relapse was observed on the posterolateral wall of the trachea, about 4 months after radiotherapy. R.V. died one year after diagnosis.

Discussion and conclusions
Primary cancer of the trachea is extremely rare and, therefore, pertinent data are limited. Most primary cervical tracheal tumours are malignant, for example, SCC in smokers, ACC in non-smokers, carcinoid tumour, mucoepidermoid carcinoma, oat cell carcinoma; benign neoplasms can also be observed, for example, xanthogranuloma and pleomorphic adenoma. We can diagnose tumours localized in the trachea as metastasis from tumours of thyroid gland, bronchus, oesophagus, larynx, lung, colon. Tracheal neoplasms, benign and malignant, are usually bulky and obstructive; they increase in size and tend eventually to extra-luminal invasion into the thyroid or upper mediastinic space. Unfortunately, these neoplasms are often diagnosed late on account of delayed specific symptoms (blood sputum, coughing, dyspnoea). Very often, the patient undergoes treatment for bronchial asthma, this mistake thus delays diagnosis and the tracheal tumour becomes inoperable. Tracheo-bronchoscopy and CT, in coronal projection, are basic examinations for these problems. Surgery (segmental resection, sleeve resection, immediate or secondary reconstruction using flaps) followed by radiotherapy is the treatment of choice. Radiotherapy alone is a possible treatment option in inoperable cases. Endo-bronchial brachytherapy may be used for tracheal tumours, especially if the tumour is small in size. Palliation has improved with the introduction of laser resection and stents.
The Authors chose to publish this report, given the rare occurrence of this form of cancer. A smoker presenting dyspnoea could be carrier of a tracheal neoplasm and early diagnosis is very important for the management.

References


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