

Management of anaplastic thyroid carcinoma spread over the trachea with mediastinal extension

C.E. FALCO, A.M. GRANDE¹, S. NICOLARDI², M. VIGANÒ¹, M. BENAZZO

SUMMARY: Management of anaplastic thyroid carcinoma spread over the trachea with mediastinal extension.

C.E. FALCO, A.M. GRANDE, S. NICOLARDI, M. VIGANÒ, M. BENAZZO

Introduction. We report a case of treatment of anaplastic thyroid carcinoma spread over the trachea with mediastinal extension.

Methods. Case report and review of the world literature concerning the treatment of anaplastic thyroid carcinoma are presented.

Discussion. The role of surgery in treatment of anaplastic carcinoma remains controversial. Our case we underlined two questions: the appropriateness of the surgery options with extra-thyroid spread and the better surgery approach to anaplastic thyroid carcinoma interesting the mediastinum controlling the great vessels of the neck. Even if curative resection cannot be achieved, surgical resection can immediately reduce the tumor bulk to facilitate the efficacy of post-operative radiotherapy and/or chemotherapy and to achieve a good local control to avoid the need of a subsequent palliative tracheostomy. Tumor upper mediastinal involvement made mandatory to open the sternum in order to allow a more complete resection of the macroscopic mass. The ministernotomy represents a valuable alternative that allows reduction in surgical trauma increasing patient's comfort.

Conclusion. The complete resection of the tumor mass without sacrificing vital structures can lead to some prolonged survival. Even if complete resection cannot be achieved, surgical resection can immediately reduce the tumour bulk and achieve good local control of the disease to avoid the palliative tracheotomy.

RIASSUNTO: Trattamento del carcinoma tiroideo anaplastico con infiltrazione della trachea e invasione mediastinica.

C.E. FALCO, A.M. GRANDE, S. NICOLARDI, M. VIGANÒ, M. BENAZZO

Introduzione. Descrivere il caso di un carcinoma anaplastico della tiroide con invasione tracheale ed estensione mediastinica.

Metodi. Vengono presentati il "case report" e la revisione della letteratura medica sul trattamento del carcinoma anaplastico tiroideo.

Risultati. Il ruolo della chirurgia nel trattamento del carcinoma anaplastico rimane controverso. Nel nostro caso sottolineiamo due questioni, ovvero l'importanza della corretta opzione chirurgica nel trattamento del carcinoma con invasione extra-tiroidea e di quello con invasione mediastinica, controllando i grossi vasi sanguigni del collo. Sebbene non sia curativa, la resezione chirurgica può immediatamente ridurre le dimensioni del tumore facilitando l'efficacia della radioterapia post-operatoria e/o della chemioterapia e consentendo quindi un buon controllo locale per evitare la necessità di una successiva tracheotomia palliativa. Quando il tumore coinvolge il mediastino superiore si deve obbligatoriamente procedere alla sternotomia per una più completa possibile resezione della massa tumorale. La ministernotomia mediana superiore rappresenta una preziosa alternativa che permette la riduzione del trauma chirurgico migliorando la qualità di vita dei pazienti.

Conclusioni. La resezione completa di tutta la massa tumorale, senza sacrificare le strutture vitali, può portare ad una maggiore sopravvivenza. Sebbene non si possa ottenere una exeresi completa del carcinoma anaplastico tiroideo con invasione mediastinica, la resezione chirurgica può ridurre immediatamente le dimensioni tumorali consentendo un controllo locale della malattia ad evitare la tracheotomia palliativa.

KEY WORDS: Anaplastic thyroid carcinoma - Mediastinal involvement - Ministernotomy.
Carcinoma anaplastico della tiroide - Estensione mediastinica - Ministernotomia.

Introduction

Anaplastic thyroid carcinoma is one of the most aggressive and lethal solid tumors. The mean age at diagnosis is 55 to 65 years. The peak incidence is in the 6th to 7th decade of life (1, 2). The disease has a preponderance in women, by a ratio of 3:1 to 1,2:1.

The mean size of the mass is 8 cm and ranges from 3 to 20 cm (3, 4). The cervical lymph nodes and the recurrent laryngeal nerve are involved between 40% and

University of Pavia, Foundation IRCCS Policlinico "S. Matteo", Pavia, Italy
Department of Otorhinolaryngology
¹ Department of Cardiac Surgery
² Department of Surgical Sciences

Authors' contributions

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30% of patients (5-7). The adjacent structures may be involved in up to 70% of the patients: muscles (65%), trachea (46%), esophagus (44%), and larynx (13%) (8).

The presence of metastatic disease is seen in 50% of the patients at the moment of the diagnosis and the lung is the organ more often involved (80%) (3-5, 9, 10). The mortality of anaplastic carcinoma thyroid carcinoma range between 95% to 98% (11).

We report a case of anaplastic carcinoma of the thyroid with massive invasion of the trachea and mediastinal extension.

Case report

A 64-years-old woman was admitted at our Department with a 2-month history of enlarged cervical mass and 9-year history of goiter diagnosed by sonography.

The patient, with type II diabetes, reported the following symptoms: cervical pain, dyspnea with initial dysphagia. Physical examination revealed a cervical mass (6x5 cm) covered by hyperaemic skin, fixed righ, hemilarynx, with the omolateral pyriform sinus bulged, and important reduction of airway space (Fig. 1). Endoscopically an ulcerated mass in the anterior wall of trachea, 1cm under the cricoid cartilage, was visualized. Neck and chest CT scans demonstrated a huge cervico-mediastinic tumor invading and infiltrating the trachea for 3,3 cm, the paraglottic spaces and crico-thyroid membrane; neck nodes at level III and V were present on the right side (Fig. 2). A suspected metastatic lesion was found in the terminal segment of inferior lobe of the left lung parenchyma. Findings of needle aspiration biopsy taken from the neck tumor showed a not defined carcinoma.

The patient underwent total thyroidectomy with ministernotomy combined with total laryngectomy and exeresis of first four cartilageneous tracheal rings; "en bloc" lymph node removal by bi-



Fig. 2 - CT scan showing tumor mass spread over the trachea (1) and to the mediastinum (2).

laterally modified radical neck dissection (type III) was performed. Total tiroidectomy with total laryngectomy extended to first four cartilageneous rings was achieved by a classical technique.

Ministernotomy was achieved prolonging the incision in the mid-line position toward the sternal notch and for about three cm over the manubrium. After careful dissection performed under the sternal notch, the manubrium was separated in the midline up to the angle of Louis using a sternal saw; a Finocchietto retractor was applied and the sternal edges spread. In this way it was possible to perform a careful dissection of the vascular structures, i.e. right jugular and subclavian veins, and brachiocephalic artery, from the neoplastic tissue that was removed (Fig. 3).

The minimally approach utilized should be considered a variant of the usual ministernotomy where sternal division is performed from the sternal notch to the third or fourth intercostal spaces forming a "J" incision (16,17). This approach was avoided in order to limit bleeding and mammary artery damage.

Hystological examination of the surgical specimen confirms the suspicious: anaplastic thyroid carcinoma pT4aN2. The patient recovered locally and had a regular immediate post-operative period; later, due to the expanding pulmonary mass, she developed a fast worsening respiratory failure and after three months she died, without signs of surgical wound infection or dehiscence.



Fig. 1 - Cervical mass covered by hyperaemic skin.

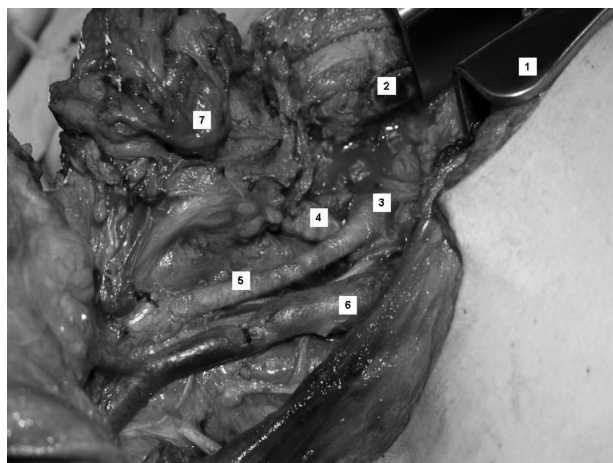


Fig. 3 - Total thyroidectomy with ministernotomy combined with total laryngectomy: 1) Finocchietto retractor, 2) sternal notch, 3) anonymous artery, 4) trachea, 5) common carotid artery, 6) internal jugular vein, 7) tumoral mass.

Discussion

The role of surgery in treatment of anaplastic carcinoma of the thyroid remains controversial. In fact, the majority of patients has a disease beyond the bounds of any meaningful resection.

In our case we underlined two questions. The first one is the appropriateness of the surgery option in the case of an aggressive disease and where the curative resection cannot be achieved. The second is the better surgery approach to anaplastic thyroid carcinoma to control the great vessels of the neck.

Even if curative resection cannot be obtained, surgical resection can immediately reduce the tumor bulk to facilitate the efficacy of post-operative radiotherapy and/or chemotherapy and to achieve a good local control to avoid the need of a subsequent palliative tracheostomy (12-15). Junor highlights prolonged survival in a series of 91 patients underwent total or partial thyroidectomy followed by radiotherapy, when compared with patients who had only biopsy (18). Kobayashi, in a series of 37 patients, noted an increase in survival from 2 to 6 months with complete macroscopic resection of tumor (19). Sugino et al. reported, in 40 cases, one year survival in patients who underwent surgical debulking followed by radiation (60%) while the same survival was reached only by 20% of patients who did not undergo surgery (23).

The management of patients who present mono or bilateral vocal cord paralysis or tracheal invasion can become extremely complex for cervico-mediastinal involvement with rapid evolution of symptoms. Airway obstruction is the cause of death in 50% of patients with thyroid carcinoma (20). Shin et al. have proposed a staging system based on the anatomic modality of tumour extension to the trachea. They report a poorer prognosis when the tumor extends through the entire thickness, expands into the tracheal mucosa and is visible at bronchoscopy as a nodule or ulcerated mass (21).

There are different opinions regarding airway management (11) ranging from elective tracheostomy, tracheal and laryngotracheal resection, laryngectomy with or without cervical exenteration, emergency tracheostomy. Only a few notes have been published. Gaissert et al. (22) in 82 patients (76% differentiated carcinoma of thyroid) considered tracheal resection when complete resection of gross airway disease appeared feasible. Symptoms caused by airway obstruction or hemoptysis were an indication for palliative resection even when grossly positive peritracheal margins or pulmonary metastasis were noted. Limited presumed or known pulmonary metastasis does not represent a real contraindication to resection. A tumor is considered unresectable

when advanced metastatic disease is present, the length of involved airway at bronchoscopy precluded primary anastomosis or mediastinal tracheostomy, or invasion of vital adjacent organs is found during operative exploration. Shaha et al. (11), in 30 patients with anaplastic thyroid cancer and acute airway problems, defined airway management depending on patient and family wishes, by the availability of definitive treatment and the disease extension. Further the patient with acute airway distress may benefit from airway bypass with tracheostomy or cricothyrotomy but this procedure can result arduous to perform. The trachea may be considerably deviated or surrounded by a large tumoral mass or, in case of extensive tumor, should be impossible to identify the trachea or tracheal lumen. Many patients with tracheostomy develop a carcinosis of the neck that worsens the local condition.

The involvement of the upper mediastinum in diseases of thyroid gland is not relatively frequent: the lesions originating from thyroid tissue are 5,8% of all masses of mediastinum (24). Instead of median sternotomy, the minimally invasive approach to the sternum represents a valuable alternative that allows reduction in surgical trauma increasing patient comfort. In cardiac surgery ministernotomy provided several benefits to the patient: Rodriguez (25) and coll. showed a significant earlier discharge from ICU and hospital. In addition, other advantages of this technique are: less haemorrhage and infection risk, minor ventilatory support time, less postoperative pain and better aesthetic results and less costs.

Tumour upper mediastinal involvement made mandatory to open the sternum in order to allow a more complete resection of the macroscopic mass. As stated by Are and Shaha (26), a neck dissection should be performed only in the setting of complete macroscopic resection. We do believe that this objective is obtained through a ministernotomy that allows to resect the upper mediastinal mass and, at the same time, to perform a save dissection of the neuro-vascular structures in the neck. In fact, the enlarging of the surgical field makes easier and more comfortable vital structures dissection.

The neoplastic mass in our patient involved the right internal jugular vein from the mandible angle to the confluence with the subclavian vein; dissection was accomplished and favoured by the adopted technique.

Conclusion

The complete resection of all gross disease without sacrificing vital structures can lead to some prolonged survival. Even if the resection cannot be achieved, curati-

ve surgical resection can immediately reduce the tumour bulk and achieve good control to avoid the palliative tracheotomy. It is important to remember that anaplastic

thyroid carcinoma is rare and it is hard to find a sufficient number of patients to study the natural history of the tumour and its response to treatment.

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