CASE REPORT

Fatal outcome of an anaplastic thyroid tumour

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SUMMARY

Anaplastic thyroid cancer is a rare form of thyroid cancer, known for its very poor prognosis, even with the appropriate management. It is very aggressive, with a tendency to invade locally and lymph node metastasis is often present at the time of diagnosis. We present an elderly patient, who presented with rapid enlarging neck mass, which, within weeks, led to his death. His family gave their consent to perform an autopsy, and here we describe and illustrate the profound aggressiveness of this tumour.

BACKGROUND

Anaplastic thyroid tumours are extremely aggressive, and death often occurs within the first year after the diagnosis. This neoplasm is one of the deadliest tumours, where local growth leads to compromise of the structures of the neck, such as trachea and arteries.

Histologically, it is composed of cells, derived from thyroid gland, but showing very little or no evidence of differentiation. Usually, a small percentage of cells show differentiation in the form of papillary or follicular carcinomas, giving rise to the hypothesis of a dedifferentiation from those carcinomas. The very rapid growth of the tumour and the poor response after surgical treatment, chemotherapy or radiotherapy often leads to comfort care measures as the initial management plan.

CASE PRESENTATION

A 79-year-old patient was being monitored in an outpatient setting with check-ups every 6 months, for the last 4 years, as he had a 2 cm left thyroid nodule which was confirmed to be benign in a fine-needle aspiration (colloid nodule, Bethesda II¹). He came to the doctor's office because the nodule size had increased to 6 cm since the last check-up 2 months earlier (at that time, the nodule was 2 cm in size). Given the rapid growth of the thyroid nodule, a thyroidectomy was initially scheduled for 4 weeks later.

Three weeks later, however, the patient was admitted to the hospital presenting with fever, dysphagia and general malaise. On physical examination, the patient showed cachexia, dyspnoea and hyperextension of the neck. The thyroid mass had increased to 9 cm in diameter, it was non-tender, firm and hard in consistency.

A chest X-ray ([figure 1A, arrow](#)) showed tracheal displacement to the right. A few hours after

admission, the chest X-ray showed dense infiltrates in both lungs, suggestive of bronchopneumonia ([figure 1B](#)). A chest CT scan ([figure 1C,D](#)) showed a large cervical and mediastinal thyroid mass, pushing the trachea towards the right side. Less than 24 hours after admission, the patient's situation worsened and he passed away.

INVESTIGATIONS

We decided to ask the family for consent to perform an autopsy, as our primary intention was to determine the cause of death and identify the extent of the disease. The main finding was a large thyroid gland weighing 315 g (normal: 40 g). The right lobe measured 5.5×3.5×3 cm, while the left lobe measured 9.5×4.5×4 cm ([figure 2](#)). The lungs showed multiple nodules (1–2 cm in size), which were hard in consistency, suggesting metastases ([figure 3](#)).

Given the aggressive, quick and fatal course of the disease, the main suspicion had been anaplastic thyroid cancer, which was later confirmed by the aforementioned autopsy findings. Samples of the thyroid gland revealed large areas of necrosis and haemorrhage. Cells were highly pleomorphic, with large nuclei. Some areas were infiltrated by neutrophils, and some nested zones showed features of papillary thyroid carcinoma (images not available). Approximately 90% of the mass was considered anaplastic. Pulmonary and adrenal metastases were also sampled and showed the anaplastic involvement.

When immunohistochemistry techniques were performed, the samples were positive for AE1/AE3 (Pax-8 was not performed), indicating it was a carcinoma. Based on clinical presentation, radiological imaging, autopsy findings and histological samples, we established the final diagnosis of an anaplastic (undifferentiated) thyroid tumour with lymph node, pulmonary and adrenal metastases, that is, stage pT4bN1bM1 (grade IV), according to the seventh edition of the American Joint Committee on Cancer tumour staging system.²

DISCUSSION

The aim of this study is to give an illustrative clinical picture of the aggressive behaviour of anaplastic thyroid carcinoma (ATC), with a clear educational purpose. Since ATC is a rare thyroid tumour,³ it is not frequent to witness such a profuse presentation with plenty of symptoms and pathological features. This tumour can arise as a rapidly growing cervical mass, which can easily double in size within a week

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Figure 1 Chest X-ray (A; arrow) showing tracheal displacement to the right. A few hours after admission, the chest X-ray showed dense infiltrates in both lungs, suggestive of bronchopneumonia (B). Chest CT scan (C, D) showing a large cervical and mediastinal thyroid mass, pushing the trachea towards the right side.

and invade other neck structures. Progressive airway compromise is frequent, which can lead to dyspnoea and hyperextension of the neck. Dysphagia due to oesophageal invasion may lead to cachexia.⁴ Our patient showed all these symptoms.

Predisposing factors include a history of goitre, associated papillary thyroid carcinoma and exposure to radioactive iodine. Anaplastic thyroid carcinoma may arise from a well-differentiated thyroid cancer, that is, the occurrence of a dedifferentiation from a well-differentiated carcinoma, such as papillary thyroid carcinoma.⁴ Indeed, at least 20% have a history of a coexisting differentiated thyroid tumour.^{5 6} As mentioned before, our patient had a long-standing history of goitre, and

Figure 2 (A) Shows the rear view of a multiloculated cervical mass, with a left thyroid lobe measuring 9.5×4.5×4 cm, isthmus measuring 6×3.4×3 cm and right thyroid lobe measuring 5.5×3.5×3 cm. (B) Shows the gland split open, with cystic and solid zones and areas with haemorrhage.

Figure 3 The lungs showed multiple whitish nodules, often less than 1 cm in size, hard in consistency (A). (B) Shows a detailed view of the right lung. These nodules were confirmed to be metastases from the primary thyroid neoplasm.

the histopathological report described foci of papillary thyroid carcinoma within the undifferentiated neoplasm.

The disease is rarely confined to the thyroid gland, with local invasion and cervical and distant metastases, often identified at the time of diagnosis.⁷ ATC has an early dissemination capability. Regardless of local invasion, distant metastases can be found in lymph nodes, lungs, liver, adrenal glands, bones and brain.

Anaplastic thyroid carcinoma has very poor prognosis (median survival: 3 months), often despite aggressive treatment,⁸ since most patients show metastases at the time of diagnosis, mainly in the lungs. Although a management plan should be promptly formulated, the poor outcome often leads to palliative care medicine. Despite there being no standardised treatment approach, a multidisciplinary, aggressive approach, including surgery, chemotherapy and radiotherapy, may improve overall survival.⁹ In any case, surgical resection is often performed to provide palliative, symptomatic relief.¹⁰

It is worth noting that our patient was scheduled for a thyroidectomy 4 weeks after the mass was 6 cm in size. We believe there was no justification for having delayed a fine-needle aspiration technique. It is likely that both the diagnosis and the outcome would have been the same, but the patient would have benefited from palliative care medicine.

Learning points

- We would like to emphasise the fast-growing, aggressive, fatal course of this rare but highly lethal form of thyroid malignancy. We have depicted an illustrative example of the course of anaplastic thyroid carcinoma.
- After the diagnosis and the tumour, node, metastases classification stage are determined, both rapid assessment and a multidisciplinary management plan should be formulated. Realistic expectations should be given to the patient and the family.
- Given the poor prognosis, option of palliation should be given to patients with unresectable anaplastic thyroid carcinoma, instead of a surgical intervention that can significantly impair the patient's quality of life.
- The overall mortality remains high, even after aggressive surgical and chemotherapy treatment.

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Competing interests None declared.

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