A rare case of anaplastic thyroid carcinoma in a young woman with prolonged survival

**INTRODUCTION**

Anaplastic thyroid cancer (ATC) is one of the most aggressive tumors in human medicine. Despite the multimodal therapy, the average survival of patients is just several months. Fortunately, it is an extremely rare tumor. Most frequently, ATC occurs in the elderly, with the average age of the patients being almost 70 years. It is extremely rare that ATC occurs in people under the age of 40. We report a case of a 33 year old woman with ATC, with the diameter of the tumor being 8x7cm, confirmed by histopathology and immunohistochemistry. In January 2001, a total thyroidectomy was performed, and the tumor was completely resected. Postoperatively, the patient received radiation therapy. Ten years after the surgery, on regular check-ups, the patient is still living and there is no evidence of a recurrent tumor or metastases. This case is instructive for two reasons, as it shows that ATC can occur in younger people and that there is a possibility of long term survival.

Key words: anaplastic thyroid cancer; thyroid gland; survival

CASE REPORT:

A 33 year old woman was admitted to the hospital for a scheduled surgery in our institution, in January 2001. One month prior to the hospital admission, the patient noticed non-painful, fast growing tenderness on the right side of the neck. The ultrasound of the neck revealed a solitary hypoechogenic nodule in the right thyroid lobe, with a diameter of 4x3cm. The rest of the thyroid tissue was homogenous. No enlarged lymph nodes were detectable in the neck by the ultrasound. The scintigraphy revealed a nonfunctional node. The patient was euthyroid, and the anti-thyreoglobuline and antiperoxidase antibodies were negative. The value of serum calcium was within the reference range. A fine needle aspiration biopsy revealed a malignant tumor.

At the time of hospital admission, the patient did not have any pain. Physical examination showed a non-tender node in right thyroid lobe, with a diameter of 7 cm (it doubled its size in less than a month). The node had an elastic consistency, it was painless, and less movable during swallowing. The physical examination did not reveal cervical lymphadenopathy. The laryngoscopy showed that both vocal cords were symmetrically movable. The chest X-ray presented a deviation of the trachea to the left. Biochemical analyses showed only a higher erythrocyte sedimentation rate (70 mm/h). The patient’s history was clear from other malignant diseases, previous neck radiation, other endocrine disorders or previous surgeries. The patient was free from other chronic illness and medical treatment. The family history was negative for thyroid disorders. She was not an inhabitant of an endemic goiter region. Intraoperatively, the tumor of the right thyroid...
lobe was identified, partially with capsule penetration and infiltration of the sub hyoid muscles. The total thyroidectomy en bloc with infiltrated muscle was performed. No thyroid remnant was seen macroscopically. Figure 1 shows a gross section of the tumor. Reliable identification and preservation of parathyroid glands and recurrent laryngeal nerve on the left side of the neck was achieved. On the right side of neck, the recurrent laryngeal nerve was carefully dissected from the tumor. The extempore biopsy pointed to ATC. The definitive histopathology (HE stain) showed gigantocellar type of ATC, composed of the giant cells, with the numerous atypical nuclei, mitotic figures, expressed pleomorphism and areas of the necrosis (Figure 2). This finding confirms the extempore biopsy. The immunohistochemistry was used to evaluate the tumor. It did not express the thyroglobulin and calcitonin stains, but it was positive for cytokeratin.

The postoperative course went well. The phonation was remarkable. The values of serum calcium were over 2.0 mmol/l. She was discharged from hospital on the fourth postoperative day. The substitution with L-thyroxine started immediately, first with 100 mcg per day, and after a month the dosage was increased to 125 mcg per day. Six weeks after the surgery, radiotherapy was applied (30Gy). Chemotherapy was not used. For the possibility that ATC originated from differentiated thyroid cancer, six months following the surgery the scintigraphy of the neck and whole body scan was performed. Examination was remarkable.

Ten years after the surgery patient is still living. She is coming to the regular six-month check-ups. At these check-ups, the patient was physically examined, ultrasound of the neck and abdomen, chest X rays were done. Based upon these results, no signs for recurrence of the disease nor metastasis were present. The TG values are not measurable, and the TSH values are ranging from 0.3 to 0.5 mU/L.

DISCUSSION

We have decided to report this case because of two reasons:

1. So far, this is our youngest ATC patient, among 150 ATC patients diagnosed in the past 15 years. It belongs to a younger group of ATC patients in literature published thus far. More than 90% of patients are older than 50 years, while the incidence rate under 40 years of age is extremely low. This case illustrates that, in differential diagnosis of thyroid alterations in younger patients, ATC should be kept in mind.

2. On the other hand, this is our patient with the longest survival, and it also shows that survival is possible in ATC patients, despite the fact that ATC is a tumor with one of the worse prognoses. It is ironic that one of the most indolent tumors in human medicine – papillary cancer, and also one the most fatal human cancers – ATC can originate from the thyroid gland. Besic et al., analyzed 188 ATC patients. They found that the median survival was 3 months, one-year survival 13%, two-year survival 6%. Their study showed that during the first month after the establishment of the diagnosis, 20% of patients died, and during first two months, 33% of ATC patients died. According to the results of Kihare et al., the average survival of ATC patients was 9 months, one-year survival was 21%, two-year survival 11% and five-year survival was 5%. Yau et al., analyzed 50 ATC patients, and the results showed that the median of survival was 3 months, one year survival 14%, and three-year survival was 8%. Median of survival in ATC patients was 4 months, found by Haigh et al. One year survival, which ranged from 10-15% of patients, was found by Sugitani et al., and Gilliland et all. Is it hard to find the ATC patients with five-year survival. This fact is opposite to the opinion of some authors, that patients, without signs of the disease five years after the surgery, are cured. Some authors think that long term survival in ATC patients is causing the doubt in the adequate diagnosis and that is
also necessary to reevaluate the diagnosis. In this case, we reevaluated the diagnosis. The revision of the specimen and additional immunohistochemistry test were ordered from the pathologist, but these tests also confirmed the diagnosis of ATC.

We can only make the assumption that the age of the patient had special influence over this long term survival, because the same model of treatment was applied in other patients. According to the literature, the prognosis of the ATC patients depends on the age of the patient. Longer survival can be expected in younger patients. Besic et al. claim that ATC patients over the age of 70 have a 1.5 times higher risk of shorter survival than patients under age of 70. According to a univariate regression analysis conducted in this study, comorbidity is significantly statistically correlated with survival, but it is not an independent factor analyzed by multivariate regression analysis. Yau et al. found that survival is statistically longer in patients under age of 65. The reason for that might be comorbidity, usually found in the older patients. Giuffrida et al. and Kebebew et al., found that survival is longer in ATC patients under age of 60. Gilliland et al. found that one year survival is decreasing in ATC patients as the age of the patient rises. In contrast with other authors, results of Haigh et al. and Sugitani et al., showed that the age of patients did not have any influence on survival in of the ATC patients.

Conclusions: This case is instructive for two reasons, as it shows that ATC can occur in younger people and that there is a possibility of long term survival.

SUMMARY


Ovaj slučaj je značajan iz dva razloga, prvo zato što pokazuje da se anaplastični karcinom može javiti i kod mladih osoba, a zatim i zato što pokazuje da je kod ovih bolesnika moguće dugoročno preživljanje.

Ključne reči: anaplastični karcinom štitaste žlezde; štitasta žležda; preživljavanje

REFERENCES: