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Curative surgery for anaplastic thyroid carcinoma: A case report

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Abstract

Anaplastic thyroid carcinoma accounts for 3%–4% of thyroid malignancies worldwide and is aggressive in nature with a median survival of 6 months at diagnosis. A 67-year-old lady with a hard goitre presented with compressive symptoms and mild hypothyroidism. Ultrasound scan revealed a Thyroid Imaging Reporting and Data System 5 lesion with suspicious left-sided cervical lymphadenopathy. Anaplastic carcinoma was diagnosed by fine needle aspiration cytology. Left-sided thyroid tumour with possible carotid sheath infiltration and left-sided cervical lymphadenopathy was seen on contrast-enhanced computed tomography of the neck. She underwent total thyroidectomy with therapeutic bilateral selective central and lateral cervical lymphadenectomy. Involvement of the aero-digestive tract and carotid sheath was not observed intra-operatively. Histology reported anaplastic carcinoma with deposits of papillary carcinoma in affected lymph nodes. Oncological management was commenced thereafter. Anaplastic thyroid carcinoma usually presents as advanced disease. However, current guidelines suggest a multimodal approach comprising of curative surgery whenever feasible with adjuvant radiotherapy and chemotherapy. For patients with stage IV^{a}/IV^{b} loco-regional disease as in our patient, total thyroidectomy with therapeutic lymphadenectomy to achieve R0/R1 resection plus adjuvant therapy is the current accepted practice. For locally advanced disease, surgery maybe opted after down-staging. The aim is to resect tumour wholly and not merely de-bulking. The presence of papillary carcinoma in lymph nodes points towards anaplasia occurring in a background of differentiated thyroid carcinoma in our patient similar to what literature suggests. This has implications in post-operative thyroxine suppression and radioiodine ablative therapies.

Keywords

Anaplastic thyroid carcinoma, thyroidectomy, oncosurgery, Sri Lanka

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Introduction

Thyroid malignancy is the commonest endocrine-related malignancy globally and the second commonest in Sri Lanka.[1,2] The vast majority of which, however, fall into differentiated thyroid malignancy, which include papillary and follicular variants. Anaplastic thyroid carcinoma (ATC) is the rarest subtype accounting for 3%–4% of all thyroid cancers. The estimated number of new anaplastic cancer cases annually in the United Kingdom is about 70–90 whereas according to Sri Lankan cancer registry, 40 new cases were reported in 2015; 27 of whom were females.[2,3] As the name implies, this type is poorly differentiated and is often incurable at presentation. Median survival at diagnosis is about 6 months and it accounts for about 50% of all thyroid cancer–related deaths in the United States.[1] ATC occurs more in the elderly and only 10% occur in those under 50 years.

ATC can originate de novo or in the background of differentiated thyroid carcinoma. Sequence of events and precipitants leading to anaplasia is poorly understood, but mutations in BRAF, TP53, NRAS, PIK3CA, APC, PTEN, IDH1, ALK and TERT promoter genes have been linked to its origin.[3–7] Its gross appearance is usually a bulky tumour with homogeneous or variegated appearance with areas of necrosis and haemorrhage. Evidence of spread to surrounding tissues is often evident. Microscopic evaluation reveals three morphological forms: Squamoid, Spindle cell and Giant cell. A single tumour can occur singly or in a combination of

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the above forms.[3] Clinically, these tumours present as rapidly growing, painful anterior neck lumps with compressive symptoms involving the upper aero-digestive tract. This is often associated with regional lymphadenopathy and voice change. Ultrasound sonography (USS) findings include large solid masses with hyper-echogenicity, microcalcifications, irregular margins and cervical lymphadenopathy. Fine needle aspiration (FNA) of the nodule reveals marked atypia with bizarre mitoses and a biphasic cellular population.[4]

Case

A 67-year-old otherwise well lady presented to surgical clinic with a goitre spanning over a period of 1 year with recent onset rapid enlargement, difficulty in breathing and swallowing. There was no pain over the lump or overlying skin changes. She was clinically euthyroid and rest of her systemic enquiry did not reveal any abnormalities. The patient had not undergone previous surgeries and was not on any medication. Clinical examination of her neck revealed a large goitre with a left-sided hard nodule which was fixed to surrounding tissue. It was non-tender and was associated with left-sided deep cervical lymphadenopathy. Rest of her system examination was normal.

Her workup was initiated with a thyroid profile, neck USS and fine needle aspiration cytology (FNAC). Thyroid-stimulating hormone (TSH) was mildly elevated at 5.35 mIU/L and her thyroxine (T4) level was 7.55 pmol/L which meant she was hypothyroid. USS of the neck revealed Thyroid Imaging Reporting and Data System (TIRADS) 5 lesion with suspicious left-sided level 3 cervical lymphadenopathy. FNA was reported as anaplastic carcinoma of the thyroid. These initial tests were followed up by a contrast-enhanced computed tomography (CECT) of the neck and chest which was reported as anaplastic carcinoma of thyroid with left-sided level 3 cervical lymphadenopathy with possible local invasion of the left jugular vein, tracheal deviation without evidence of retrosternal extension.

Upper gastrointestinal (GI) endoscopy and laryngoscopy were normal and so was the erect postero-anterior (PA) chest X-ray. Routine blood investigations showed the following: white blood cell (WBC) – 8.47×10^9/L, Haemoglobin – 11.5 g/dL, platelet count – 248×10^9/L and prothrombin time international normalized ratio (PT INR) – 1.0. Renal and liver functions were insignificant and so was the electrocardiogram.

She then underwent total thyroidectomy with bilateral selective central and lateral cervical lymph node dissection. Intra-operatively, it was observed that the aero-digestive tract and carotid sheath were not infiltrated by the tumour and so were left alone. Histology of the specimen revealed anaplastic carcinoma in the left lobe, whereas the right showed Hashimoto thyroiditis. Well-formed papillary carcinoma deposits were observed in one and four cervical lymph nodes from left and right sides, respectively. Her post-operative stay was uneventful without evidence of nerve injury and hypocalcaemia. She was discharged and adjuvant chemo-radiation was commenced thereafter. We have followed up the patient monthly and after a year of surgery, she is healthy.

Discussion

Due to its aggressive nature, anaplastic carcinoma usually presents as advanced unresectable disease. However, current British Thyroid Association (BTA) and National Comprehensive Cancer Network (NCCN) guidelines suggest a multimodal approach comprising curative surgery whenever feasible with adjuvant external beam radiotherapy (EBRT) and chemotherapy.[3,8] In fact, a multi-institutional retrospective analysis of 114 patients have shown a significant improvement in outcome when surgery and radiotherapy was combined in combatting ATC.[9] For patients with stage IV[a] or IV[b] loco-regional disease as in our patient, total thyroidectomy plus therapeutic lymph node dissection to achieve R0/R1 resection is the current accepted practice.[3,8] Adjuvant EBRT and chemotherapeutic regimes are to be followed. Even for locally advanced disease, surgery is opted after down-staging with EBRT and chemotherapy because complete removal has shown to prolong survival.[3,8,9]

The American Thyroid Association (ATA) too suggests a similar approach.[1,10] Current evidence points towards an increased survival with total thyroidectomy with high-dose radiotherapy alone; however, targeted therapy is often used alongside. Due to its low incidence and high mortality at presentation, current guidelines on management are still based on low to moderate evidence even in the developed world.

Decision to undergo curative resection depends on the structures involved, possibility of R0/R1 resection and whether resection of involved structures causes morbidity/mortality. The aim of curative surgery is to resect tumour wholly and not merely de-bulking.[10] Routine preoperative imaging using ultrasonography and computed tomography (CT)/magnetic resonance imaging (MRI) is carried out to define the spread of disease prior to surgery. For systemic disease, resection of primary tumour, even if the resection is not R0/1, is considered to prevent upper aero-digestive tract obstruction.[10]

Ours is a rare case where the patient presented early and the workup was instantaneous which resulted in early diagnosis. We were then able to go ahead with the curative therapy (surgery followed by adjuvant chemo-radiotherapy) which is extremely rare for anaplastic carcinoma of the thyroid not only in the developing world but also in the west as discussed earlier. Multiple studies world-over suggest increased overall survival with this approach for stage Iv[a] and IV[b] with uncertain results for IV[c].[11,12]

The presence of papillary carcinoma deposits in the excised lymph node mass points towards anaplasia occurring in a background of differentiated thyroid carcinoma (DTC) in our patient. Indeed, many studies have shown most ATCs

to occur in a background of papillary carcinoma which later undergoes anaplasia due to mutations in multiple genes as mentioned earlier.[3,4] Decision to carry out en-block resection of lymph nodes is further backed by the fact that it enabled us to point the origin towards DTC which has implications in post-operative thyroxine suppression therapy and radioiodine ablative therapies even though de novo ATC has no place for this.

Conclusion

ATC is the rarest of the thyroid malignancies and is by far the most aggressive. Current treatment of choice involves a multimodal approach with surgery and adjuvant chemo-radiotherapy which has improved survival and overall patient outcome. Here, we present a rare case where curative surgery was performed for ATC following early detection and workup which already has prolonged patient survival considerably.

Author contributions

S.M.P.V. drafted the initial manuscript. M.D.P.G. and M.L.M.M. gathered further literature and added on to initial draft. E.A.D.U. oversaw the process and all confirmed the final draft.

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Ethical approval

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