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Case Report

A rare primary thyroid mixed germ cell tumour: a case report

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ABSTRACT

Germ cell tumours mostly occur in the gonads. Extragonadal site is rare and mainly located in the mediastinum, retroperitoneum and pineal gland. Germ cell tumour that occurs in the thyroid gland is extremely rare. We present our first description of a patient with a mixed germ cell tumour located primarily in the thyroid gland. A 35 years old gentleman presented with two months history of thyroid swelling associated with compressive symptoms. His neck ultrasound showed multinodular goiter with suspicious nodule (TR4) and FNAC revealed highly suggestive of a malignant lesion. Subsequently, the thyroid mass progressively enlarged in size causing acute upper airway obstruction that made him collapsed at Emergency Department. Core biopsy of TR4 thyroid nodule suggestive of malignant lesion which could be mixed germ cell tumour or poorly differentiated carcinoma. Abnormal laboratory tests showed a high concentration of LDH, Alpha fetoprotein and beta-HCG. He underwent emergency total thyroidectomy, debulking and tracheostomy. Histopathology reported the mass composed of mixed non-seminomatous germ cell tumour with area of necrosis (30%), primarily immature elements (80%), yolk sac tumour (15%) and embryonal carcinoma (5%). He was scheduled for chemotherapy post operatively however he succumbed to the illness due to advanced tumour and sepsis.

Keywords: Tumour, TR4, Germ cell tumour, Biopsy

INTRODUCTION

Germ cell tumours (GCT) mainly occur in the gonads. Extragonadal site is rare and mainly located in the mediastinum, retroperitoneum and pineal. GCT that occur in the thyroid gland are extremely rare but can occur in young adults. We present our first encounter of a patient with a mixed germ cells tumour located primarily in the thyroid gland.[1]

CASE REPORT

A 35 years old gentleman, an active smoker with no medical illness, presented with two months history of anterior neck swelling. It was associated with hoarseness of voice and dysphagia. His thyroid ultrasound revealed right multinodular goiter with spongiform appearance and some nodules have mix solid cystic, hypoechoic lesion and microcalcification described as TIRADS 4. There was no normal thyroid gland in the right lobe but left lobe was normal and no enlarged lymph node. Cytology aspiration of thyroid nodule was highly suggestive of malignant lesion. Subsequently, he developed difficulty in breathing due to progressively enlarged thyroid mass and collapsed at the Emergency Department because of acute upper airway obstruction. He was intubated for airway protection. Clinically the thyroid mass was 15×10 cm in size. Laryngoscope showed lateralization of right pharyngeal wall with right vocal cord palsy. Core biopsy of thyroid suggestive of malignant lesion could be mixed germ cell tumour or poorly differentiated carcinoma. However, ultrasound of testes was normal. CT Scan of neck and thorax showed hypodense thyroid mass with extension superiorly to parapharyngeal space and

inferiorly retrosternal space measuring 9×12.6×10 cm. The mass has a clear plane with other structures. The trachea was compressed with the narrowest luminal part was only 3 mm. There was multiple enlarged right cervical level II lymph nodes (Figure 1). Laboratory tests showed a high concentration of Lactate dehydrogenase (LDH) (1180 u/l), Alpha-fetoprotein (60981.2 IU/ml) and beta-HCG (56.5 IU/l) levels. After a multi-disciplinary team discussion (MDT), he was subjected to an emergency total thyroidectomy, debulking and tracheostomy (Figure 2). Macroscopically, part of the thyroid capsule was breached and ruptured with the gland measures 15×12×9 cm and weight 671 grams. Histopathology report disclosed it as mixed non-seminomatous germ cell tumour with area of necrosis (30%), primarily immature elements (80%), yolk sac tumour (15%) and embryonal carcinoma (5%). He was scheduled for chemotherapy BEP scheme (bleomycin, etoposide, cisplatin) but was postponed due to severe pneumonia postoperatively. Unfortunately, he developed recurrent neck swelling after 4 weeks and difficulty in breathing that required prolonged ventilatory support. Repeated CT thorax and neck showed recurrence tumour growth with multiple lungs and nodal metastases. His condition further deteriorated as complicated with hospital acquired infection and he succumbed to the illness because of advanced tumour and severe sepsis.

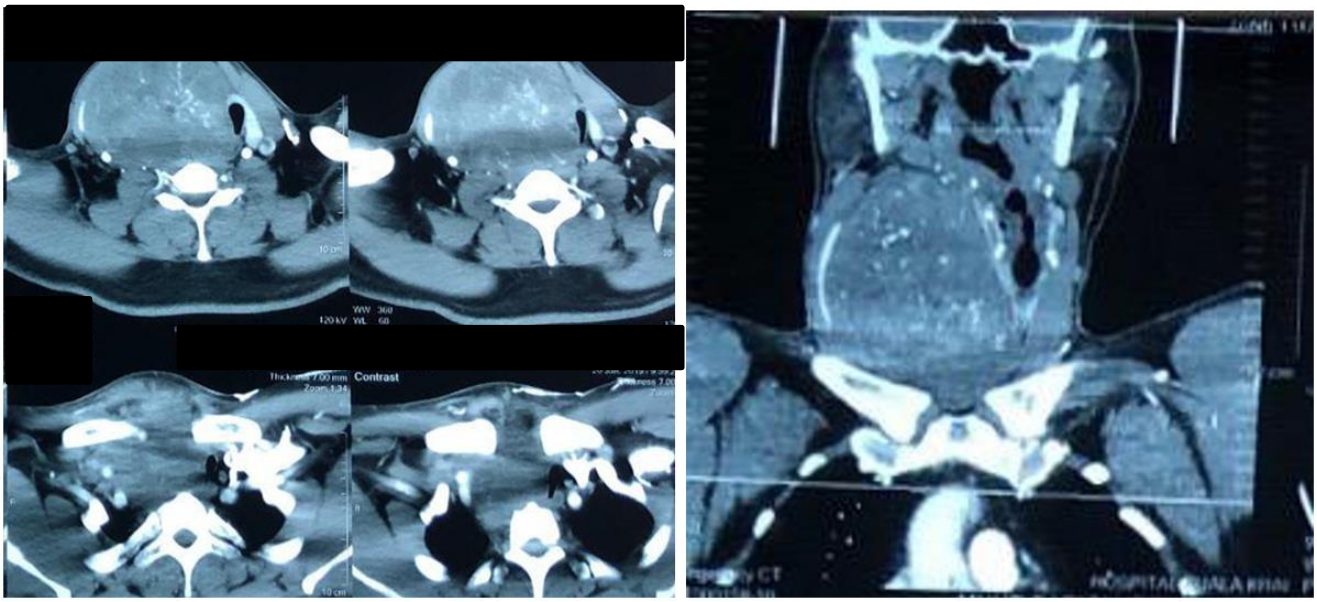

Figure 1: CT neck showed enlarged of thyroid of right lobe size 9×12.6×10 cm, superiorly extend till parapharyngeal space, inferiorly till retrosternal level, medially displaced larynx and trachea to the left. Trachea compression width 0.3 cm, and AP 1.8 cm. Left thyroid normal.

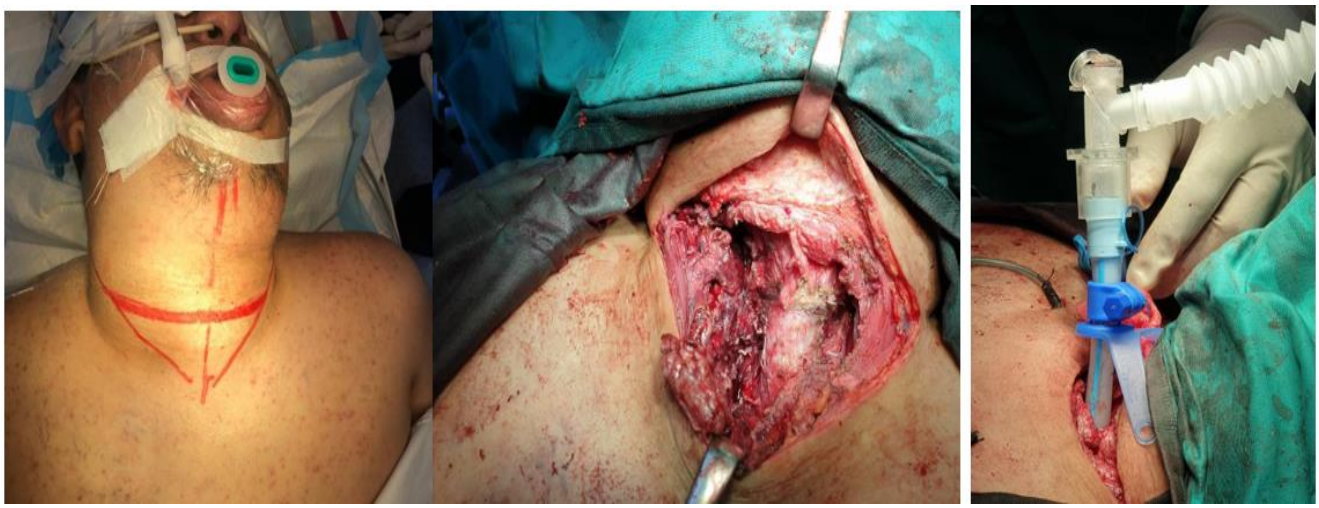

Figure 2: Post-operative images showed the wound after total thyroidectomy and trachestomy.

DISCUSSION

A germ cell tumour is (GCT) a tumour that occurs inside gonad organs (testis and ovary). Extragonadal germ cell tumour accounts for 1-5 % of all GCT.[8] In current practice, there is more acceptance that an extragonadal germ cell tumour can occur without a testicular focus.[7] However, extragonadal germ cell tumour rarely occurs primarily in the thyroid gland. The most common sites are tumours that arise from the midline: mediastinum, retroperitoneum, and cranial.[7-11] Extragonadal GCT can be divided into seminoma (30-40%) and non-seminomatous germ cell tumour (NSGCT) which accounts 60-70% of extragonadal GCT. NSGCT includes subtype histology such as teratoma, embryonal carcinoma, yolk sac tumour, choriocarcinoma and mixed. NSGCT is more aggressive compared to seminoma. In literature, most primary thyroid GCT cases were teratomas but occur more in children and young adult.[2-4,6,12-14] Most thyroid teratomas are benign. Malignant thyroid teratoma accounts for 1.3%, while only 19% of them occur in adults.[12] A case of primary mixed germ cell tumour of thyroid has been described by Joanna et al.[1] Diagnosing a primary thyroid germ cell tumour, metastatic from testis must be ruled out as it is more common. Ultrasound of testis should be performed in all cases to look for primary testicular GCT[8]. Most of the patients presented with rapidly enlarged thyroid nodules with some associated with compressive symptoms.[1-3,5,12-14] Diagnosis from fine needle aspiration cytology (FNAC) can be difficult. Some reported cases able to diagnosed from cytology while most of them mistakenly diagnosed as papillary thyroid carcinoma, follicular neoplasm and metastatic tumour.[1,3,5,13,14.] Conformation of diagnosis is from surgical specimen. There are no specific features of this tumor in ultrasound or CT scans. Most literature reported as ultrasound findings of hypoechoic lesion with area solid and cystic thyroid nodule. Patients may also have enlarged lymph nodes as this tumour may metastasize to lymph nodes. GCT of the thyroid may present with infiltration and compression such as hoarseness of voice and difficulty in breathing. Tumour marker will be raised in non-seminomatous germ cell tumour. It is very important to take tumour markes such as alpha feto-protein and ß-HCG as it helps in diagnosis of NSGCT. LDH is raised in high tumour burden. These tumour markers also can be use as monitoring during follow-up. Total thyroidectomy is surgery of choice followed by adjuvant chemotherapy. However, complete resection is difficult to achieve in locally advanced tumour. Based on management of testicular germ cell tumour, thyroid GCT also response well to chemotherapy using BEP regime (Bleomycin, Etiposide and cisplatin). Patients may develop recurrence after surgery, as an author reported as early as three weeks post total thyroidectomy.[2] Administration of chemotherapy early after surgery is mandatory to achieve complete response and prevent a recurrence. Even though surgery did not achieve complete tumour resection, with the adjunct of chemotherapy may prolong disease-free survival for more than four years[5]. Like in our case, the tumour recurs 4 weeks post thyroidectomy, indicative of an aggressive tumour. As our patient was in sepsis, chemotherapy was not able to be administered early. In managing this rare tumour, suspicion should be made in a rapidly growing thyroid mass with the aid of tumour markers. The initial biopsy may not be diagnostic but early total thyroidectomy is indicated as this tumour may cause airway obstruction. Confirmation from histopathology must be done urgently so that chemotherapy can be registered early. Delay in chemotherapy administration caused tumour to regrow and prognosis of the patient will be poor.

CONCLUSION

Primary thyroid mixed germ cell tumour is very rare. Diagnosing and managing this type of tumour is very challenging. High index of suspicion of a malignancy should prompt early thyroidectomy to gain definite diagnosis so that the patient would receive early chemotherapy for a better prognosis.

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