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**Case Report**

# Carcinoma showing thymus-like differentiation (CASTLE) – A Rare Extra-Thyroid Disease

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**Abstract**
*Carcinoma showing thymus-like differentiation (CASTLE) is a rare malignant tumor occurs in soft tissues of neck or thyroid gland. It accounts for 0.1% to 0.15% of all thyroid cancers. Immuno-staining with CD5 is a useful marker for differentiating from other malignancy of thyroid.*
*Case report: A 72-year-old female presented with painless neck swelling which on examination was firm to hard mass at left level III region extending inferiorly upto medial supraclavicular fossa. Imaging showed a solid conglomerate lymph-nodal mass and biopsy reported as poorly differentiated neoplasm. IHC markers confirmed the diagnosis of CASTLE disease. She underwent left modified radical neck dissection along with left hemithyroidectomy and Adjuvant radiation therapy. Now at 8 years of follow-up, no evidence of disease recurrences.*
*Thus, being a rare entity, early diagnosis and prompt treatment plays an important role in the outcome.*
**Keywords:** *Castle disease, thymus like differentiation, thyroid neoplasm, long-term follow-up, extra-thyroidal disease.*

## Introduction

Carcinoma showing thymus-like differentiation (CASTLE) is a rare malignant disease that occurs in soft tissues of neck or thyroid gland[1]. These tumors were proposed an origin from ectopic thymus or along the remnant of branchial pouches[2]. Thus, the clinical presentation and radiological features can be similar to malignant lesion arising from either thyroid or thymus. Immuno-staining with CD5 is a useful marker for

confirming the diagnosis of CASTLE disease from other malignancy of thyroid. Although majority of patients undergo surgery, being a rare entity, the ideal treatment approach is still unknown. According to literature, it accounts for 0.1% to 0.15% of all thyroid cancers of which only 3 cases were extra-thyroid tumors[3-6]. Here, we report a case of extra-thyroid CASTLE, successfully treated and on follow-up for eight years after obtaining written and informed consent from the patient.

**Case Report**

A 72-year-old women noticed a painless swelling over left medial supraclavicular region in 2013, which was gradually increasing in size. On physical examination, 5 x 4.5 cm hard and fixed mass located at left level III region extending below till left medial supraclavicular fossa. No obvious lesion was found in the oral cavity, nasopharynx, oropharynx or hypopharynx. She was evaluated with ultrasonogram (USG) neck that showed a well-defined hypoechoic mass situated below the left lobe of thyroid. Computer Tomography (CT) showed 4.9x4.5 cm solid enhancing superior mediastinal conglomerate lymph-nodal mass. Fine-needle aspiration (FNA) biopsy of the mass was performed and only spindle cells were found. Following which Trucut biopsy reported as poorly differentiated neoplasm. Immunohistochemistry (IHC) was focally positive for CD5, Cytokeratin, HMWCK and HBME and negative for LCA, Vimentin, S100, CEA, synaptophysin and thyroid transcription factor-1. In-view of clinical, radiological and pathological correlation, features were suggestive of carcinoma with Thymus like Differentiation (CASTLE Disease). Being a rare entity, report was discussed in multi-disciplinary Tumour Board and decided for surgical intervention but was refused by the patient. She received 8 cycles of oral cyclophosphamide and was under close follow-up. She had a stable disease. In July 2015 she noticed an increase in size of the mass with no evidence of disease or metastasis elsewhere. Thus, underwent left modified radical neck dissection along with left hemithyroidectomy. Postoperative histopathology reconfirmed the diagnosis as CASTLE disease and all other lymph-nodes were reactive nodes. She received radiation therapy to the postoperative region of dose 6000cGy in 30 Fractions using 3-dimensional conformal radiotherapy (3DCRT) in 2015 [Figure No 1]. At the most recent follow-up in 2021, i.e., eight years since diagnosis, no evidence of tumour recurrence was present.

**Discussion**

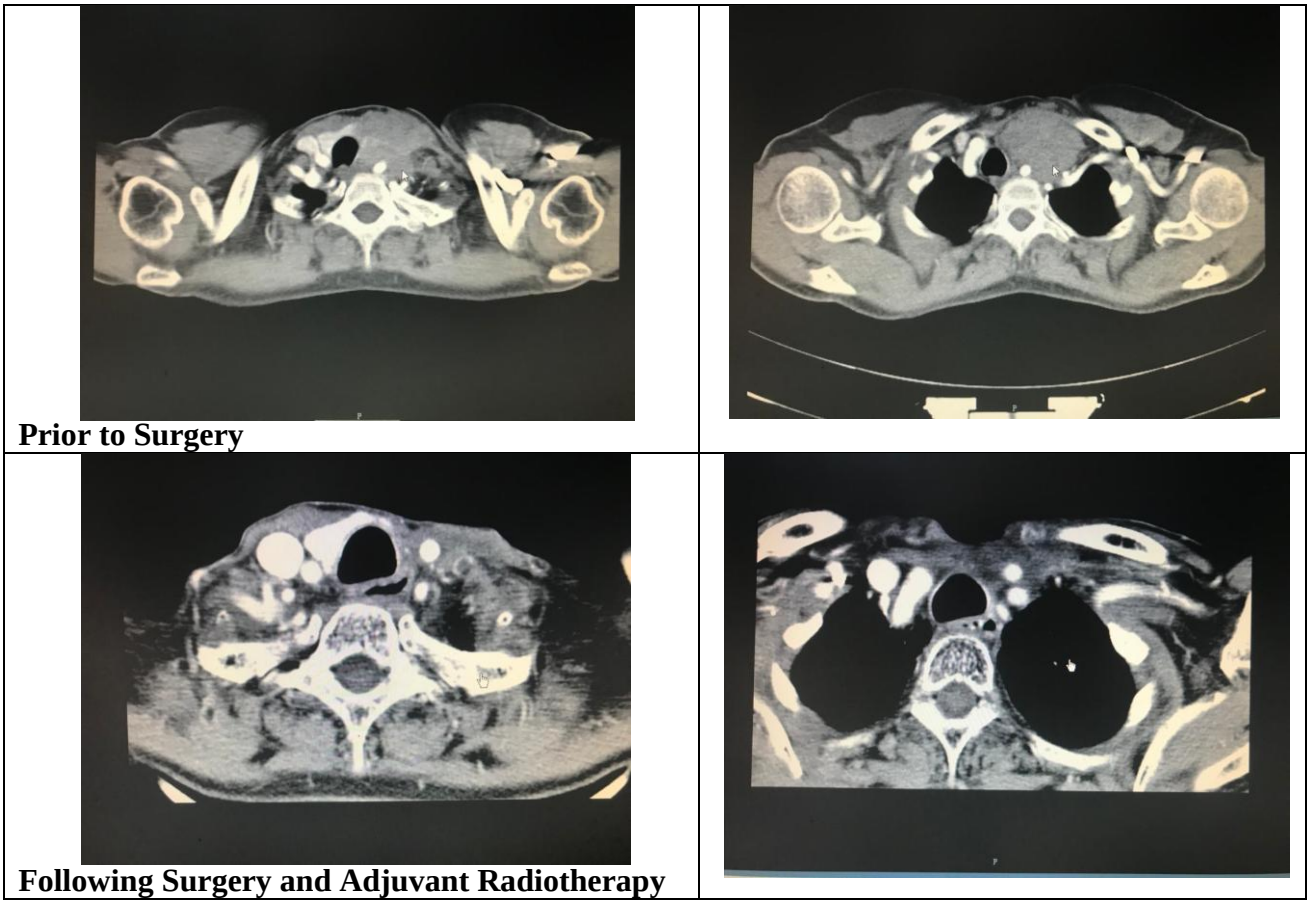
CASTLE tumour – Carcinoma showing thymic like differentiation is a rare malignant neoplasm occurring in thyroid gland or soft tissue of the neck. It is assumed to arrive from remnant of thyropharyngeal duct or from ultimobranchial body remnants within the thyroid gland[2,7]. According to the review of literature, only very few cases have been reported with extrathyroidal presentation[3-4]. Thus, we report a case with extrathyroidal presentation.

CASTLE tumours are generally an indolent tumour, occasionally pursue a more aggressive course[2]. A FNA biopsy seems to offer limited diagnostic value as majority of cases in literature being identified as malignant tumour without further definitive classification. Histologically CASTLE present with lobulated and expansile growth pattern with fibrous septa dividing the tumour, with prominent nucleoli and associated with lymphoplasmacytic infiltration[2-5,7-9].

There is no consensus on the management due to the rarity of the disease. Treatment consists of surgical excision with or without radiation treatment.

Youens et al. reported 27 cases of which 25 were intrathyroidal cases. Ten cases had recurrences of which five died either with local or distant recurrences (liver/lung)[1]. Only three extrathyroidal CASTLE reported till date, were managed by resection and adjuvant radiotherapy, neither showed loco-regional recurrences. Choi et al.[5] reported seven more new cases from the

literature regarding the treatment modalities and outcome. It is evident from all these cases that excision with lymph nodal dissection were the primary mode of treatment with adjuvant radiation showed a better outcome and local control. Though there no strong recommendation regarding chemotherapy, in our case, since the patient refused surgery and radiation primarily, our oncologist offered chemotherapy – Cyclophosamide. She received 8 cycles and was kept on close follow-up. After 2 years, increase in size of neck mass noted and thus underwent surgical resection and adjuvant radiotherapy. Almost 8years of follow-up since diagnosis and 6 years since resection and radiotherapy, no evidence of tumour recurrence noted.



**Prior to Surgery**

**Following Surgery and Adjuvant Radiotherapy**

**Figure No 1:** Above CT images showing the response following surgery and adjuvant radiotherapy

## Conclusion

This is an extrathyroidal CASTLE disease which was successfully treated and on follow-up. Being a rare entity, early diagnosis and prompt treatment modality plays an important role in the outcome.

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## References

1. Youens KE, Bean SM, Dodd LG, Jones CK: Thyroid carcinoma showing thymus-like differentiation (CASTLE): case report with cytomorphology and review of the literature. Diagn Cytopathol 2011, 39:204–20

2. Chan JK, Rosai J: Tumors of the neck showing thymic or related branchial pouch differentiation: a unifying concept. Hum Pathol 1991, 4:349–367
3. Luo CM, Hsueh C, Chen TM: Extrathyroid carcinoma showing thymus-like differentiation (CASTLE) tumor – a new case report and review of literature. Head Neck 2005, 27:927–933.
4. Ahuja AT, Chan ESY, Allen PW, Lau KY, King W, Metreweli C: Carcinoma showing thymic-like differentiation (CASTLE tumor). Am J Neuroradiol 1998, 19:1225–1228.
5. Choi et al. Extrathyroid carcinoma showing thymus-like differentiation (CASTLE): a new case report and review of the therapeutic role of neck dissection and radiotherapy World Journal of Surgical Oncology 2014, 12:247
6. Tsutsui H, Hoshi M, Kubota M, Suzuki A, Nakamura N, Usuda J, Shibuya H, Miyajima K, Ohira T, Ito K, Ikeda N. Management of thyroid carcinoma showing thymus-like differentiation (CASTLE) invading the trachea. Surgery today. 2013 Nov;43(11):1261-8.
7. Reimann JD, Dorfman DM, Nose V: Carcinoma showing thymus like differentiation of the thyroid (CASTLE): a comparative study: evidence of thymic differentiation and solid cell nest origin. Am J SurgPathol 2006, 8:994–1001
8. Miyauchi A, Kuma K, Matsuzuka F, Matsubayashi S, Kobayashi A, Tamai H, Katayama S: Intrathyroidal epithelial thymoma: an entity distinct from squamous cell carcinoma of the thyroid. World J Surg 1985, 1:128–135.
9. Mizukami Y, Kurumaya H, Yamada T, Minato H, Nonomura A, Noguchi M, Matsubara F: Thymic carcinoma involving the thyroid gland: report of two cases. Hum Pathol 1995, 26:576–579.