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Case report

# Primary Squamous Cell Carcinoma of Thyroid – A Case Report with Emphasis on Treatment Options and Review of Literature

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**Introduction:** Primary squamous cell carcinoma of thyroid (PSCCT) is a rare entity with less than 100 cases reported in literature. It constitutes about 1% of all thyroid cancers. It has an aggressive behaviour and unfavourable prognosis. We report a rare case of PSCCT and provide a review of literature on PSCCT.

**Case Report:** A 45 year old gentleman with hypertension as co morbid condition reported to our hospital with complaints of swelling in anterior part of the neck. He was diagnosed as a case of carcinoma of thyroid. He underwent Total Thyroidectomy under general anaesthesia. With histopathology and IHC correlation, diagnosed as PSCCT. He received adjuvant RT to tumor bed to a total dose of 60 Gray (Gy) over a period of 6 weeks.

**Conclusion:** PSCCT is a very rare disease with very aggressive course. There is no consensus on the line of management. Lenvatinib can be tried in PSCCT as it resembles ATC.

## Introduction

Primary squamous cell carcinoma of thyroid (PSCCT) is a rare entity with less than 100 cases reported in literature. It constitutes about 1% of all thyroid cancers [1]. It has an aggressive behaviour and unfavourable prognosis. The diagnosis can be challenging because of the presence of squamous metaplasia in other thyroid malignancies and direct extension into thyroid from locally aggressive squamous cell carcinoma (SCC) [2]. PSCCT can be diagnosed only after ruling out metastasis from other sites, local part imaging, surgical excision of thyroid and immunohistochemistry (IHC) analysis [3]. We report a rare case of PSCCT and provide a review of literature on PSCCT.

### Case Report

A 45 year old gentleman with hypertension as co morbid condition reported to our hospital with complaints of swelling in anterior part of the neck. He had no other significant history like radiation exposure, family history of thyroid malignancies. Examination revealed a 3 x 3 cm hard swelling in left side of anterior neck. The swelling was moving with deglutition. Multiple small nodes were felt in left level III region. Baseline blood investigations were done and were normal. Ultrasound neck was done at an outside hospital which showed an irregular hypoechoic solid mass in left lobe of thyroid. FNAC done at an outside hospital was suggestive of malignant neoplasm. He was

diagnosed as a case of carcinoma of thyroid. He underwent Total Thyroidectomy under general anaesthesia. Post-operative period was uneventful. Immunohistochemistry (IHC) like keratin, CK 5/6, p40, CK 7, TTF -1, calcitonin, CEA, PAX 8, LMP, CD 21, LCA were requested on the surgical pathology specimen. IHC keratin, CK 5/6, p40, CK 7 showed positive reaction. IHC TTF -1, calcitonin, CEA, PAX 8, LMP, CD 21, LCA showed negative reaction. With histopathology and IHC correlation, diagnosed as PSCCT. Case was discussed in our hospital multidisciplinary board and he was planned for adjuvant radiotherapy (RT). He received adjuvant RT to tumor bed to a total dose of 60 Gray (Gy) using IMRT technique over a perior of 6 weeks. No RT related morbidity was noted. He was kept under follow up.

## Discussion and Review of Literature

PSCCT is rare entity which accounts only for about 0.2 to 1 % of all thyroid malignancies. Aetiology is unknown. There are 3 theories proposed. First, Embryonic origin involving branchial arch or thyroglossal duct remnant. Second is the metaplastic transformations secondary to chronic inflammatory thyroiditis. Last is the dedifferentiation of other thyroid carcinomas [1, 4, 5]. The last theory has led a few to consider PSCCT as a variant of anaplastic carcinoma, although the association remains unclear. Females are affected more, with a median age of presentation in $5^{th}$ and $6^{th}$ decade of life [6]. Patients usually present with a mass in neck associated with compressive symptoms. IHC plays a critical role in diagnosing PSCCT and distinguishing it from others. IHC can be positive for epithelial markers like CK 5/6, CK 7 which would confirm squamous cell of origin. TTF 1 and thyroglobulin negativity will rule out other common thyroid malignancies. PAX 8 positivity will confirm primary thyroid aetiology [7-9]. Management of PSCCT remains unclear due to the rarity of disease. Mostly they have been treated with surgical resection followed by adjuvant radiotherapy. If found to be unresectable, then concurrent chemoradiation has been tried, though response is poor [4]. Cook et al, who had published the largest review on PSCCT, showed that patients who underwent complete surgery followed by adjuvant radiotherapy had a longer survival when compared with others who underwent surgical resection alone or others who had incomplete surgical resection [10]. It is still unclear whether multimodality treatment will be beneficial in patients with PSCCT [11]. Some authors suggest surgical resection with or without adjuvant radiation as the main stay of treatment [12]. Our patient was treated with complete surgical resection followed by adjuvant radiotherapy. In regard to systemic therapy, chemotherapy has been used to little or no response. It was reported that almost half of the patients with PSCCT had distant metastasis at presentation. Thought conventional chemotherapy has not shown any survival benefit. Many published literature have shown better outcomes of anaplastic carcinoma of thyroid (ATC) when treated with lenvatinib. Lenvatinib is a tyrosine kinase inhibitor which selectively inhibits multiple angiogenic and oncogenic signal pathways. The evidence for lenvatinib comes from SELECT trial and phase II data on lenvatinib in ATC [13-15]. Though trials did not include PSCCT patients, lenvatinib can be used for PSCCT given the fact that it behaves like ATC. There is very limited literature on the use of lenvatinib in PSCCT.

In conclusion, PSCCT is a very rare disease with very aggressive course. Histopathology and IHC correlation help in differentiating PSCCT from other common thyroid malignancies. There is no consensus on the line of management, but given the aggressiveness of the disease, complete surgical resection followed by adjuvant radiotherapy should be given as it had improved survival based cook et al. Chemotherapy has limited role in PSCCT. Lenvatinib can be tried in PSCCT as it resembles ATC.

Larger prospective studies will be difficult given the rarity of the disease.

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*Statement of Transparency and Principals*

• Author declares no conflict of interest

• Study was approved by Research Ethic Committee of author affiliated Institute.

• Study's data is available upon a reasonable request.

• All authors have contributed to implementation of this research.

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