Spindle Epithelial Tumor with Thymus - Like Differentiation (SETTLE): Case Report

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Summary

Spindle epithelial tumor with thymus-like differentiation (SETTLE) is a rare tumor with slowly progressive disease course with late metastases. Metastases develops in 20% patients but most patients are diagnosed when tumor is still localised. Surgery is the gold standard treatment for SETTLE. Limited data is available regarding efficacy of chemotherapy and radiotherapy. There have been less than 50 cases published in the English literature. We report a case of a 13-year-old male patient with neck metastasis. The patient presented asymptomatic right neck mass. He gave history of midline neck swelling since birth for which he underwent left hemithyroidectomy elsewhere and later on developed right neck swelling. He underwent right neck dissection at our institute and was followed up monthly thereafter.

Keywords: Neck swelling, spindle epithelial tumor, thymus like differentiation

Introduction

Some of the rare tumors occurring in the neck and the thyroid gland, show histologic resemblance to thymus and mediastinal thymomas.[1]

This type of tumors were classified into following groups by Chan and Rosai: ectopic hamartomatous thymoma, ectopic cervical thymoma, spindle epithelial tumors with thymic-like differentiation (SETTLE), and carcinoma showing thymus-like elements (CASTLE). The former two are benign tumors while the latter two behave like malignant tumors.[2]

Spindle epithelial tumor with thymus-like differentiation (SETTLE) is a slowly progressive tumor with metastasis occurring later in the course of the disease. It occurs in the younger age group, mainly children and adolescents, although it may present in older age group too.[3] They are highly cellular tumors comprised of compact bundles of long spindle epithelial cells which merge with tubulopapillary structures and/or mucinous glands.[1]

A study of eight cases done by Chan and Rosai in 1991, where they reported unusual spindle cell tumours of thyroid gland. These cases were earlier reported as thyroid spindle cell tumor with mucinous cysts and malignant teratoma of thyroid gland. SETTLE probably arises from ectopic thymus, vestiges of the thymopharyngeal duct, or branchial pouch. The term SETTLE was used to suggest its probable thymic origin and highlight its epithelial nature.[4]

There have been less than 50 cases published in the English literature. We report a case of a 13-year-old male patient with neck metastasis.

Case report

A 13-year-old male patient presented to surgical oncology OPD with complaint of asymptomatic right neck mass since 2 months. Patient gave history of midline neck swelling since birth which on Ultrasonography and Fine Needle Aspiration Cytology showed Thyroid swelling (?Adenomatous goitre/?Follicular Adenoma). He underwent left hemi thyroidectomy outside for the same in February 2020. The specimen biopsy reported mixed germ cell tumor – mature teratoma with yolk sac component. Slide review done at our institute showed SETTLE.

Six months after thyroid surgery, the patient developed right neck swelling and presented to our OPD in October 2021. The swelling was firm, non-tender located in the right supraclavicular region. Thorough examination of head and neck region was done. Direct laryngoscopic examination showed bulky tonsils with right tonsil larger than the left one. CECT of neck showed 25x22x25 mm lesion in right oropharyngeal wall. 18x21 mm right neck node in level IV. Incisional biopsy from the swelling done outside diagnosed it as extragonadal yolk sac tumor. Slide review of this biopsy done at our Institute showed SETTLE. Immunohistochemistry was done and yolk sac tumor was ruled out after discussion with pathologist.

Thereafter patient underwent PET CT which showed uptake in right supraclavicular lymph node of size 21x19x52 and SUV max=7 [Figure 1]. Patient underwent right modified neck dissection type III in December 2021. Specimen biopsy report showed single lymph node involved, largest lymph node being 6.5 cm without any extranodal extension. The diagnosis was metastasis from SETTLE in a known case of the same.

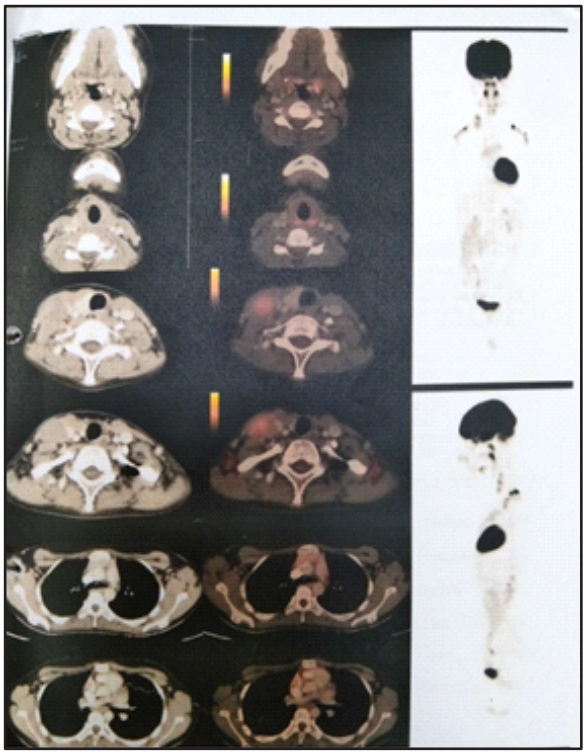

Figure 1: PET CT Images of patient

Patient was followed up monthly.

Discussion

SETTLE is a rare low grade slow growing tumors. It is a malignant tumor of the thyroid gland probably arising from thymic tissue or branchial pouch remnants.

"It is composed of spindle cells epithelial nature forming fascicles, merging into glandular structures taking the form of tubules, papillae, and cystic spaces. In some cases, cysts or glands lined by mucinous or respiratory epithelium may be present. Rare cases may be predominantly monophasic, with spindle cell predominance".[6]

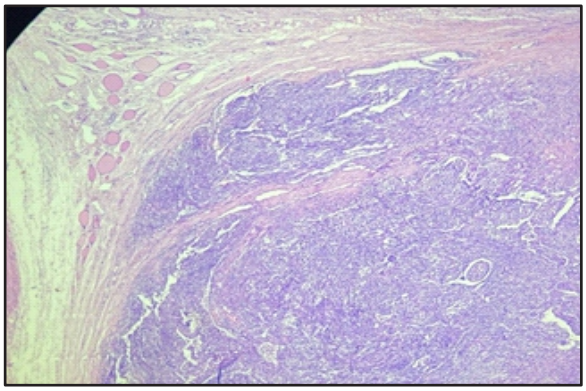

Figure 2: Histopathology image

Section shows circumscribed tumor with intervening fibrotic bands. Tumor shows biphasic component composed of epithelial and spindle cells [Figure 2].

The tumor cells test positive for epithelial markers on immunohistochemical staining which is consistent with tonofilaments and desmosomes observed in the ultrastructure. Also, the spindle cells stain strongly with 34bE12 suggesting high molecular weight cytokeratin expression which is seen in squamous cells.[6] Our patient showed positive staining for CK5/6, CK7, Vimentin and TLE1.

Metastases is seen in about 20% of the patients but most of the times it is diagnosed when the tumor is still localized. Relapse of metastases has been seen to occur many years after first diagnosis, with mean time to relapse being 11 years. Due to its rare nature, no standard treatment has been established for SETTLE with metastases. Limited data is available regarding chemotherapy or radiotherapy, their efficacy and tumor response to them.[7]

Ultrasonography may be useful in evaluating thyroid gland and localizing suspicious lymph nodes including their size and number. The gold standard for treatment is surgery. Advanced stages may be treated by chemotherapy or radiotherapy to control tumor growth as well as for metastases, vascular invasion or locally infiltrative disease. Amongst chemotherapeutic drugs, cisplatin, etoposide and cyclofosfamide have been found effective. In patients with inoperable disease, bone metastases or painful masses, radiation may be used as a neoadjuvant therapy.[8]

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