Spindle Epithelial Tumor of Thymus-like Differentiation (SETTLE) in a 3-year-old African girl

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Key Clinical Message
Spindle epithelial tumor with thymus-like differentiation (SETTLE) is a rare tumor of the thyroid gland occurring in children and young adults. This report presents SETTLE as a tumor that can also affect young children (under fives). Although majority of the reports were in Caucasians, the tumor can affect all races.

KEYWORDS
ectopic, spindle epithelial tumor with thymus-like differentiation, thymus, thyroid, tumor

1 | INTRODUCTION

Spindle epithelial tumor with thymus-like differentiation (SETTLE) is an uncommon thyroid tumor of pediatric age group and young adults.1 The tumors often develop from ectopic thymus tissue and sometimes from remnants of branchial pouches that has the ability to develop along the thymic line.2 Histologically, the tumor is lobulated with biphasic cells consisting of spindle-shaped epithelial cells that merge into glandular cells.3 It is often malignant but the tumors occurring in children may be benign.4 The youngest patient reported in the literature was 2 years old,1 we present the first case of SETTLE in a 3-year-old African girl who has had six years of follow-up with no recurrence.

2 | CASE REPORT

A 3-year-old girl presented with a painless and progressively increasing lateral neck mass since birth. Examination revealed a nontender, mobile, and multinodular left-sided neck mass measuring 8 cm × 6 cm in dimension and there were no cervical or supraclavicular lymphadenopathies.

Thyroid function test was normal while ultrasound of the neck revealed a solid lesion mainly on the upper pole of the left lobe of the thyroid gland. Fine needle aspiration cytology was suspicious of malignancy. Plain radiograph of the neck showed deviation of the trachea to the right side. At surgery, the left lobe of the gland was involved and a left lobectomy was done with no adjuvant therapy. Gross (macroscopic) examination of the tumor showed a nodular mass measuring 6 cm × 4 cm × 4 cm and weighing 35 g. Cut sections revealed a tan colored lobulated tumor, firm in consistency, and disposed in whorled appearance. Focal areas of cystic spaces were seen. The tumor was highly cellular with proliferating spindle and polygonal (epithelial) cells occurring predominantly in lobulated and fasciculated patterns (Figure 1). The spindle cell component had hyperchromatic oval nuclei with scanty to moderate eosinophilic cytoplasm (Figure 2A-C), whereas the polygonal cells exhibited large vesicular nuclei. (Figure 2D) Foci of cystic spaces lined by epithelia cells that were disposed in irregular papillary patterns were also seen.
**FIGURE 1** A-D. Highly cellular tumor with proliferating spindle and polygonal (epithelial) cells occurring predominantly in lobulated and fasciculated patterns (H&E ×40)

**FIGURE 2** A-C. The spindle cell component had hyperchromatic spindle nuclei with scanty to moderate eosinophilic cytoplasm (A&B–H&E ×200). D: The polygonal cells exhibited large vesicular nuclei (C&D–H&E ×400)

**FIGURE 3** Cystic spaces lined by epithelial cells and containing irregular papillary structures (A–H&E×100, B–H&E ×200)
SETTLE has atypical clinical symptoms and signs and this makes preoperative diagnosis difficult as depicted from the review of previous cases. A review of the literature showed 41 reported cases of SETTLE with the usual presentation of a painless thyroid or neck mass of either a short or long duration depicting the generally slow growth of the tumor.

There is wide variation in the duration of clinical symptoms before the diagnosis is made with the range being 3 weeks to more than 10 years. The tumor is located in the right lobe of the thyroid gland in most patients, occasionally the left lobe may be involved and sometimes both lobes of the gland. Rapid growth of the tumor leading to tracheal compression has been reported. The patient, however, presented with deviation of the tracheal to the right side with no demonstrable lymphadenopathy. Preoperative diagnosis could not be made in most of the previously reported cases and the diagnosis was delayed in three cases where autoimmune thyroiditis was suspected because of a confusing clinical and ultrasonographic picture. Fine needle aspiration is often inconclusive but may arouse a suspicion of malignancy as in our patient. The relatively slow growth of the tumor makes the majority of reported cases amenable to surgical excision alone. Surgical excision may range from nodulectomy, lobectomy to subtotal thyroidectomy. Adjuvant chemotherapy and radiotherapy have been used in cases of widespread metastasis.

Although SETTLE exhibits delayed metastasis, it must be treated as a potentially malignant tumor as it can metastasize several years after surgical resection with the lungs as one of the commonest sites of metastasis.

In conclusion, we report here the first case of SETTLE in Nigeria and most probably from sub-Saharan Africa. The clinical presentation and behavior were typical and similar to the previously reported cases. Early diagnosis and surgical resection of the tumor with meticulous long-term follow-up of the patients is key to ensuring a good prognosis.

ACKNOWLEDGMENTS

The authors want to acknowledge Dr Mustapha Ajani for accepting to read through and correct the manuscript.

CONFLICT OF INTEREST

We also want to declare that there are no conflicts of interest in the production of this manuscript and the work was solely funded by the authors.

AUTHOR CONTRIBUTION

OOO: Has made substantial contributions to conception and design and acquisition of data; has been involved in...
drafting the manuscript and revising it critically for important intellectual content. Has given final approval of the version to be published and has agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. GOO: Has been involved in analysis and interpretation of data; drafting the manuscript and revising it critically for important intellectual content. Has given final approval of the version to be published and has agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. AOO: Has made substantial contributions to conception and design; acquisition of data; analysis and interpretation of data. Has been involved in drafting the manuscript and revising it critically for important intellectual content and has given final approval of the version to be published. TAJ: Has been involved in analysis and interpretation of data; drafting the manuscript and revising it critically for important intellectual content. Has given final approval of the version to be published and has agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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How to cite this article: Ogundoyin OO, Ogun G, Oluwasola A, Junaid TA. Spindle Epithelial Tumor of Thymus-like Differentiation (SETTLE) in a 3-year-old African girl. Clin Case Rep. 2019;7:1119–1122. https://doi.org/10.1002/ccr3.2163