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CASE REPORT

Spindle Epithelial Tumor with Thymus-like Element of the Thyroid Gland

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ABSTRACT

Spindle epithelial tumor with thymus-like different ion (SETTLE) is an extremely rare tumor arising from thyroid gland which was first time recognized and reported by Chan and Rosai. It is believed to be arising from the branchial pouch or the remnant of thymus within the thyroid gland as it reveals primitive thymic elements. It has been reported more than 20 times as an individual case reports in the searched English literature. It mostly occurs in the children and adolescent but cases of SETTLE are reported in adults and even in elderly patients. Age of patient is not diagnostic feature of the SETTLE tumor. According to the published data, SETTLE tumor has an indolent course but delayed blood-borne distant metastases is seen in the cases with long-term follow-up. The present case is 13-year-old boy who presented with solitary nodule of left lobe of the thyroid without any other symptoms. The patient was subjected to ipsilateral hemithyroidectomy. The histopathological report revealed the diagnosis of SETTLE tumor which was confirmed with the immunohistochemistry. The patient is followed-up regularly and there is no evidence of metastasis with more than 4 years follow-up. There are no existing guidelines regarding the optimum treatment and the duration of follow-up due to paucity of data about management of SETTLE tumor. The long-term follow-up is needed to detect recurrence or metastases.

Keywords: Thyroid tumor, SETTLE, Thymoma, Sarcoma.

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INTRODUCTION

Spindle epithelial tumor with thymus-like element (differentiation), i.e. (SETTLE), is very uncommon tumor arising from thyroid gland first reported by Chan and Rosai.[1] The tissue of origin of the SETTLE is either branchial pouch or remnant of thymus within the thyroid gland. It mostly occurs in the children and adolescent.[1-3] The tumor exhibits indolent course but delayed metastases are known to occur.[1-3] Herein, we report a case of SETTLE tumor with more than 4 years uneventful postoperative follow-up.

CASE REPORT

A 13-year-old boy presented with swelling in anterior midline of neck extending to left side for 2 months. The swelling was insidious in onset and progressive in growth without any other symptoms. No any other significant history was sought. General physical examination was within normal limit. Examination of the swelling revealed a solitary, oval swelling of 3 × 2 cm on just left side of midline of lower part of neck. Swelling revealed rounded borders, smooth surface with free upward movement with deglutition. It was nontender and firm to palpation. There was no enlargement of regional lymph nodes. It was diagnosed as a nontoxic solitary nodule of thyroid. Investigations revealed normal thyroid profile. Neck ultrasonography showed features suggestive of solitary thyroid nodule. Fine needle aspiration cytology (FNAC) from the nodule revealed findings in favor of medullary carcinoma or intrathyroidal thymoma. So, the possibility of MEN syndrome was considered which was ruled out with urinary VMA and catecholamine estimation, abdominal sonography and serum calcitonin estimation. Contrast-enhanced computerized tomography revealed the single nodule in the left lobe of the thyroid (Fig. 1). Patient was subjected for epsilateral lobectomy with written informed and valid consent from the patient and his mother. Intraoperatively, there was a single nodule in the left lobe of thyroid with no enlargement of lymph nodes. The gross specimen revealed a single-rounded nodule with homogenous cut surface (Fig. 2) in addition to the normal thyroid tissue. Histopathology of the tumor revealed capsulated lesion with biphasic cell pattern (Fig. 3). Cells showed vesicular to hyperchromatic nuclei and prominent nucleoli. The spindle cell component exhibited pseudosarcomatous trabecular pattern and areas of glandular differentiation surrounded by spindle tumor cells. Thyroid

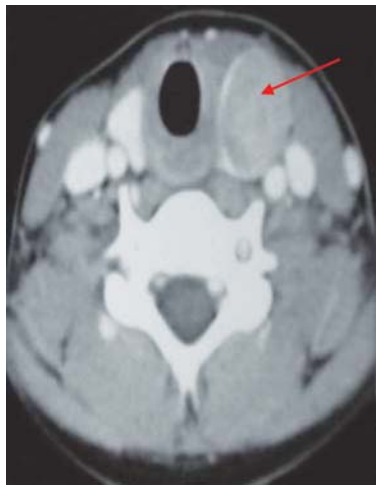

Fig. 1: Contrast-enhanced computerized tomography revealing the solitary nodule in left lobe of thyroid (arrow)

tissue outside the nodule showed normal morphology. Since the histopathology report was suggestive of SETTLE, the tumor tissue was subjected for immunohistochemistry (IHC) for confirmation of diagnosis. IHC demonstrated positivity for cytokeratin and thyroglobin and negativity for S-100, calcitonin and SMA (Figs 4 and 5). The patient had uneventful postoperative recovery and there was no evidence of distant metastases with more than 4 years follow-up.

DISCUSSION

Chang and Rosai first reported the unifying concept of SETTLE tumor of the thyroid.[1] They postulated that SETTLE tumor arises from the ectopic thymic tissue inside the thyroid gland or from the remnanent of branchial pouch.[1-5] Since then, about more than 20 cases of SETTLE tumor have been reported in the English literature.[2] Initially, SETTLE tumor was named as spindle cell tumor, thymoma or malignant teratoma of thyroid gland.[4] It mostly occurs in the children and adolescent and young adults. Youngest reported case was of 4 years and oldest reported case was of 59 years old with median age of 15 years.[1,2,5-9] Male to female ratio is 1.8:1. It is slow growing tumor, as in most of the reported cases, the mass was present for either entire adult life or more than 4 years. Sometimes sudden rapid growth is observed in the longstanding swellings. Despite of low malignant potential, it has tendency for delayed distant metastases. The incidence of metastases increases markedly as the period of follow-up is more than 5 years.[2,5] Two cases have exhibited distant metastases even after 20 years which signifies the long-term follow-up with special attention to lungs which is most common site of metastases.[5] The present case did not show local or distant metastases with more than 4 years postoperative follow-up but patient need to be kept under follow-up for many years to detect possible delayed metastases as early as possible.

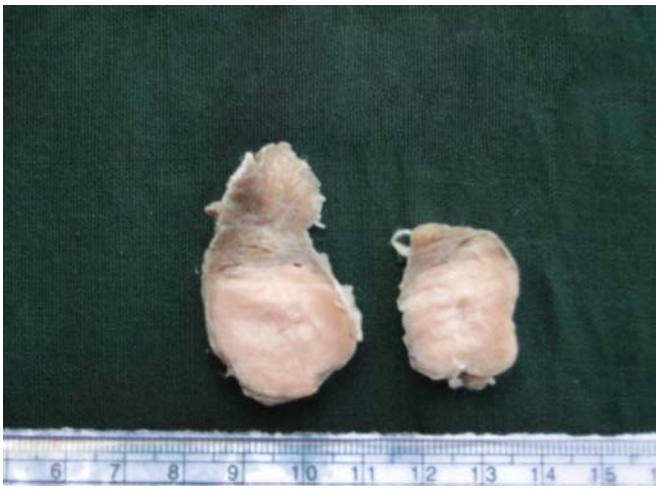

Fig. 2: Cut section of gross specimen of left lobectomy of thyroid showing the tumor and the normal thyroid tissue

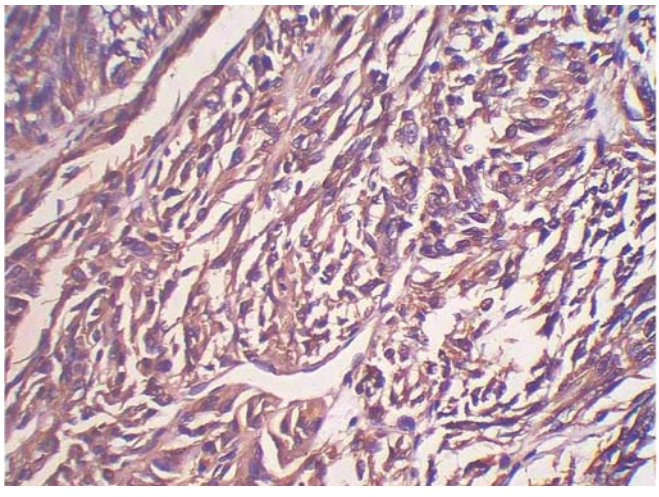

Fig. 4: Immunohistochemistry suggestive of positivity for cytokeratin, magnification 40×

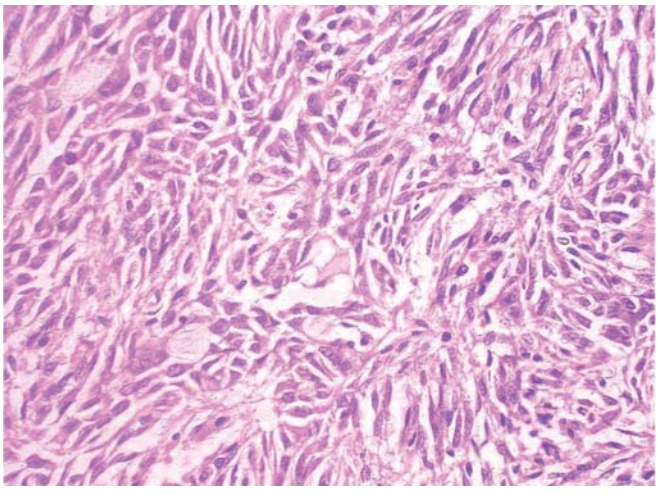

Fig. 3: Histopathology of the tumor revealed capsulated lesion with biphasic cell pattern

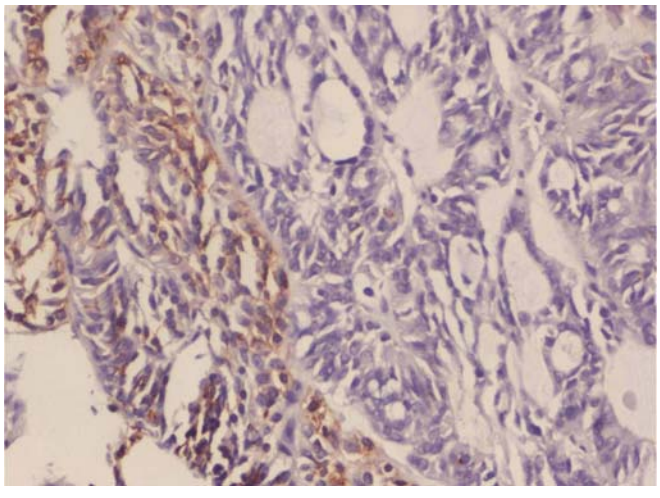

Fig. 5: Immunohistochemistry positivity for thyroglobulin, magnification 40×

SETTLE tumor is composed of spindle cells of epithelial origin forming fascicles leading to formation of papillae, tubule and cystic spaces. Normally, tumor displays biphasic pattern but occasionally monophasic pattern is also seen.[5,7] These spindle cells show positivity for cytokeratin (epithelial membrane antigen and vimentin and negativity for calcitonin, chromogranin, desmin, myoglobin and actin).[2,5,7] Bing Xu et al reported the molecular genetic analysis revealing Ki-ras gene mutation at codons 13 and 15 on the same allele which may be the cause for tumorogenesis.[2,4] However, there is no convincing evidence regarding the thymic origin of the tumor since SETTLE tumor is negative for CD20 and CD5 and TdT positive immature lymphocytes.[2] Cheuk et al reported that the histogenesis of SETTLE tumor still remains an enigma and they considered the possibility of thyroblastoma instead of thymoblastoma.[2,5] Medullary carcinoma of thyroid, sarcomatoid anaplastic CA, synovial sarcoma and malignant teratoma should be considered as differential diagnoses.[2,5,7] Medullary CA is positive for calcitonin which is negative in SETTLE.[2,5] Anaplastic CA reveals extrathyroidal invasion and extensive pleomorphism, mitoses and necrosis.[2,5,7] The uncommon monophasic variant can be differentiated from synovial sarcoma which shows monophasic spindle cells, hyperchromatic and more mitotic activity than SETTLE.[2,7] Malignant teratoma exhibits some mesenchymal tissues, like cartilage, bone and undifferentiated neural component.[2,7] There is no adequate data available to decide about the real malignant potential of the SETTLE tumor as most of the publications are about individual case reports with inadequate follow-up.[1-9] Aleksandr et al expressed the need to examine more cases to correlate between the morphological features of tumor with the metastatic potential.[2] There are no existing guidelines regarding the treatment and follow-up of cases due to paucity of data about management of SETTLE tumor. Hence, long-term follow-up is mandatory to detect possible recurrence or distant metastases.

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