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Case Report

Spindle cell tumor with thymus-like differentiation masquerading as a papillary carcinoma of thyroid gland

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Abstract Spindle epithelial tumor with thymus-like differentiation (SETTLE) is an indolent malignant thyroid tumor seen in children, adolescents, and young adults showing thymic or related branchial pouch differentiation. The tumors are composed of spindle cells along with focal epithelial component and glandular formations. We herein report a case of SETTLE with a follow-up period of 20 years.

Keywords: Papillary carcinoma of thyroid, SETTLE, thymus

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INTRODUCTION

Spindle cell tumor with thymus-like differentiation (SETTLE) is an extremely rare malignant thyroid tumor encountered in the children, adolescents, and young adults. We hereby present an unusual case of a 58-year-old male, who presented to our hospital for the treatment of a thyroid mass with nodal metastasis and recurrence involving the larynx, after undergoing a laryngectomy and being mistreated for papillary carcinoma of thyroid. On reviewing the slides, a diagnosis of SETTLE was offered. Although SETTLE is a very entity, it should always be considered as a differential in case of a thyroid mass in the young population.

CASE REPORT

A 58-year-old male presented to Gujarat cancer and research institute with a recent history of larygectomy and radical neck dissection for getting his slides reviewed. On tracing the history, the patient had a history of a midline neck mass 20 years back for which a right hemithyroidectomy was done. Ten years later, the patient presented with a nodal mass for which a wide local excision was done. Outside histopathological examination (HPE) revealed a papillary carcinoma of the thyroid gland. Hence, a completion thyroidectomy was done, and the patient was started on high dose radioiodine therapy. Further 9 years later, the patient developed a recurrent neck swelling for which a total laryngectomy and radical neck dissection was performed. Last computed tomography scan revealed, an soft-tissue density lesion in the right lower neck abutting thyroid and cricoid cartilage with erosion of right lamina of thyroid cartilage. On reviewing the slides, HPE showed a biphasic tumor, composed of fascicles of spindle cells admixed with epithelial cells forming glandular arrangement [Figures 1 and 2]. The differential diagnosis under consideration was settle and synovial sarcoma and an IHC panel was advised. IHC results showed that tumor cells were positive for AE1, Vimentin, CK 5/6, and CK7 [Figure 3]. Tumor cells were immunonegative for

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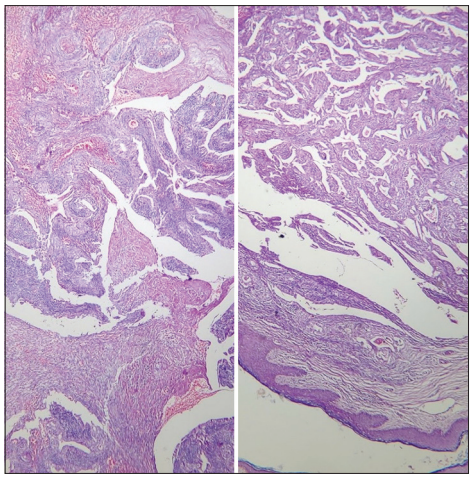

Figure 1: Scanner view shows a biphasic tumor showing the epithelial and spindle cell component. Spindle cells are arranged in fascicles and the epithelial cell cells show a focal ductular and glandular arrangement

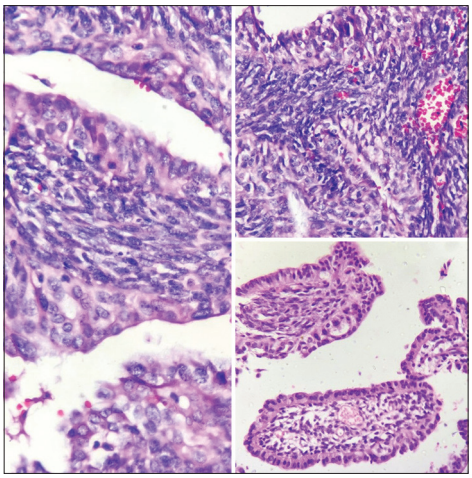

Figure 2: High-power view shows the spindle cells merging into the epithelial component. The epithelial cells are cuboidal to columnar forming ductular and glandular arrangements. The spindle cells are plump and elongated with blunted ends and bland hyperchromatic nucleus

CD 99, BCL2, Thyroglobulin, TTF-1, PAX8, C-kit, and CD 5. Hence, taking into consideration, the long clinical history, presentation, HPE and IHC results; a diagnosis of SETTLE was offered.

DISCUSSION

Chan and Rosai coined the term SETTLE in 1991 to define a group of tumors that originate in the neck with a thymic or related branchial pouch differentiation that included tumors such as carcinoma showing thymus-like differentiation, ectopic cervical thymomas, and ectopic hamartomatous thymomas.[1] During the embryological development, the foramen cecum gives rise to the thyroid gland. The ultimobranchial body gives rise to the lateral thyroid as well as thymic tissue, parathyroid, thyroid c cells, and salivary gland tissue. This process gives rise to ectopic tissue in the thyroid gland that undergoes neoplastic transformation.[2]

SETTLE is principally a tumor of children and young adults with and almost equal sex predilection.

It presents clinically as an asymtopmatic, slow growing, painless neck mass or an enlarged thyroid gland in most cases. However, in some case, it may grow rapidly or present with symptoms such as painful swelling or with obstructive symptoms. Commonly, the mass involves the right lobe of the thyroid gland.[3,4] Very rare cases may be observed arising from the adjacent soft tissue.

As the patient presents with a thyroid mass, the differentials considered by the physicians as per the usual young age are papillary and medullary carcinomas, immature teratomas, and synovial sarcoma. FNAB is the first investigation of choice for the preoperative diagnosis. Kaur *et al*. described that FNA showing a biphasic round and spindle cell population in an appropriate clinical setting may suggest the diagnosis of SETTLE, although it is a challenging diagnosis on FNA.[5]

The exact diagnosis is usually established after the surgical resection of the mass.

On gross examination, it is a smooth, white-tan mass that can be circumscribed or infiltrative. It can also show small cysts or mucin-containing areas.

On low power examination, the tumors were biphasic, cellular, showing admixture of fascicular and reticulated pattern, with areas of stromal hyalinization. The spindled areas merged into the epithelial areas forming glomeruloid glandular structures, tubules or small glands lined by cuboidal to columnar areas. On high-power examination, the spindled cells possessed elongated or oval nuclei with one or more pointed or blunted ends. The nuclei had a distinct nuclear membrane and finely dispersed chromatin with an inconspicuos nucleoli. Pleomorphism was minimal, and mitosis was occasional. The histology typically resembled a synovial sarcoma. Areas of sqaumous morules or mucinous cysts are often found. However, in our case, neither squamous morules nor cystic mucin-containing areas were noted. This can be attributed to inadequate sampling or recurrent nature of the lesion. However, the indolent course suggested a diagnosis of SETTLE over a synovial sarcoma in our case.

The main differential diagnosis under consideration is biphasic synovial sarcoma.

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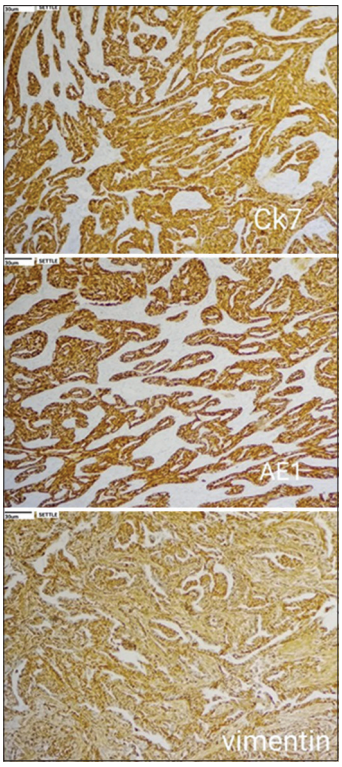

Figure 3: Shows immunopositivity for AE1, CK7, and Vimentin

On IHC, the spindle and glandular cells show positivity for high-molecular-weight cytokeratins. These cells lack positivity for EMA and low-molecular-weight cytokeratin. Strong membranous and cytoplasmic positivity for CD99 and BCL2 can also be seen in both the components. Membranous CKIT positivity can also be found.[6] These tumors are typically negative for TTF1.

Although molecular confirmation is considered the gold standard for distinguishing SETTLE from synovial sarcoma, we believe that histologic features, clinical history, and immunohistochemistry were sufficient to make a confident diagnosis of SETTLE in our case. Molecular studies would rather be really helpful in FNAC or biopsies where we have limited diagnostic material.

Surgery is the main stay of treatment for SETTLE and includes resection of primary and metastatic disease.[3] There is no proven role of adjuvant chemotherapy or radiotherapy.

The 5-year survival rate was approximately 83%. Although 50% of these patients had late metastasis,[3] the most common site of metastasis was cervical lymph nodes and lung.

CONCLUSION

SETTLE should be considered in the differential diagnosis of spindle cell tumor of thyroid origin as it often mistaken as papillary carcinoma of the thyroid gland. It has an indolent course and good prognosis if appropriately treated at presentation and monitored for a long period.

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Conflicts of interest
There are no conflicts of interest.

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